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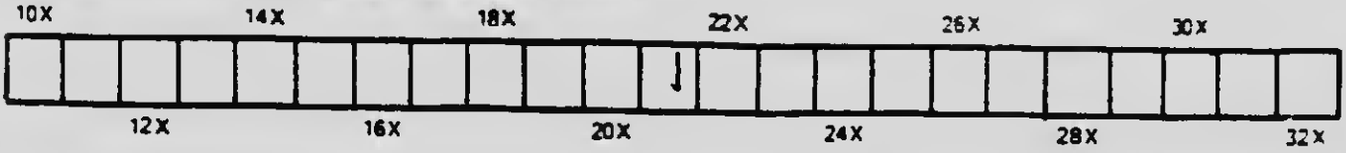
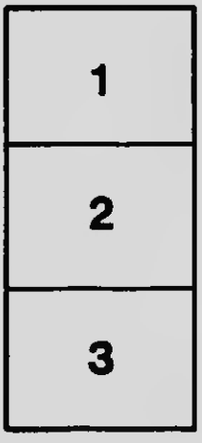
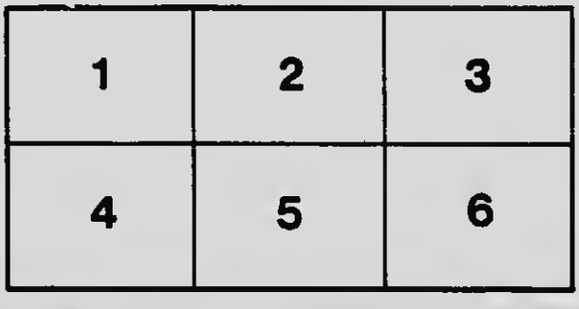
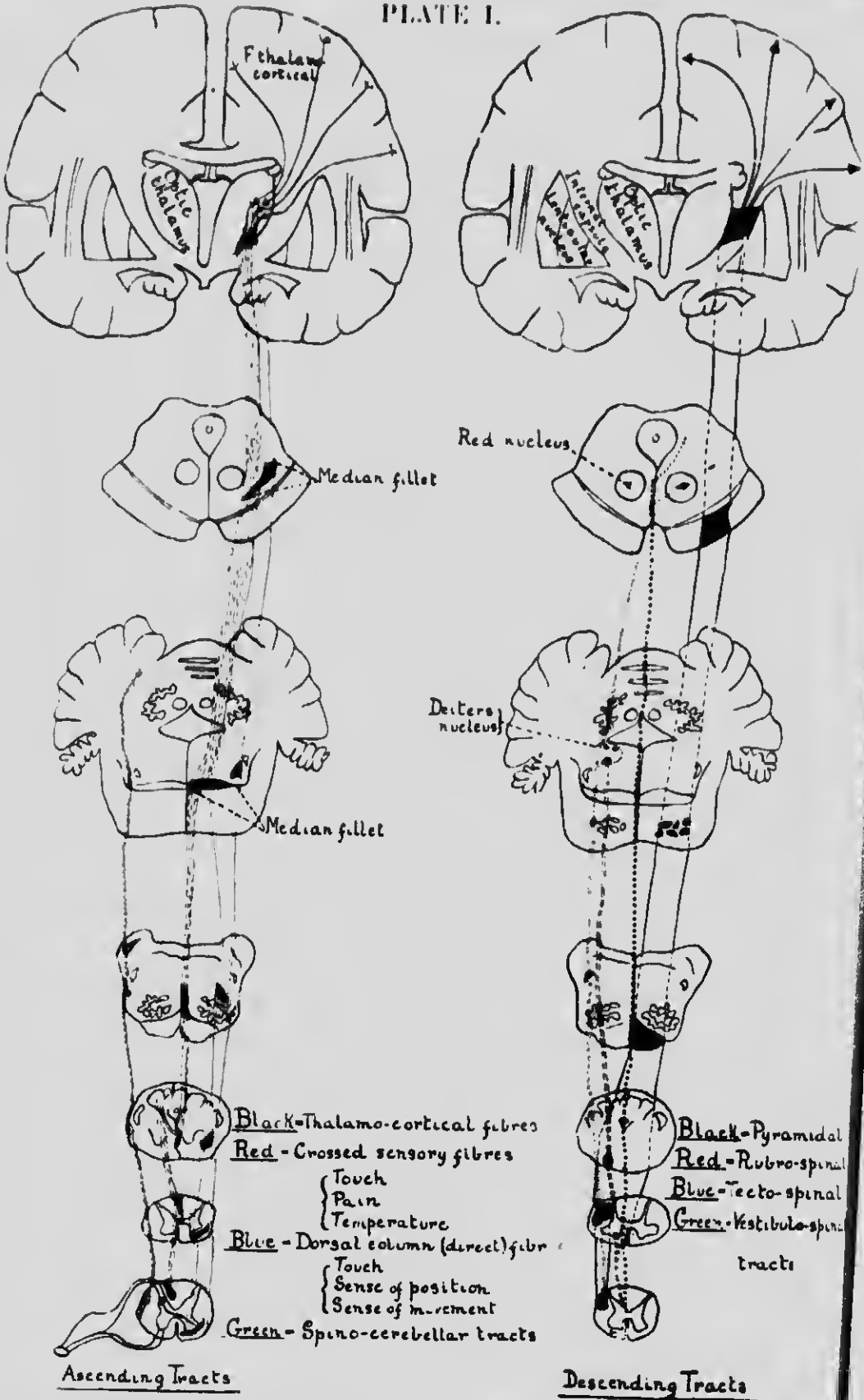






Diseases of the Nervous System

PLATE I.



The above drawing by my friend, Miss Karlowa, was made from a diagram on a lantern slide, kindly lent to me by Henry Head and Gordon Holmes.

DISEASES OF THE NERVOUS SYSTEM

BY

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Dedicated to
the Past and Present Students
of the
Manchester Medical School.

PREFACE.

In the present volume I have adopted, as far as is practicable, a clinical classification of nervous diseases, an arrangement which I have found to be the most useful to the student. In a class which I have conducted at the Manchester Royal Infirmary for the last twenty years, a description of the chief neuronic systems has been followed by the bringing forward of cases illustrating the various forms of paralysis and of other symptoms in relation to lesions of the corresponding neurons. In this way the student obtains a grasp of the principles of anatomical diagnosis, and is soon willing to admit that the investigation of diseases of the nervous system is less terrible than he previously supposed.

A student trained by this method, when shown a patient with atrophy of some of the muscles of the upper limb is able to say at once that there must be a lesion implicating either some part of the lower neurons of the affected limb or the muscles themselves. He then proceeds, by an examination of the sensation of the limb, to determine whether the lesion is limited to purely motor elements, namely, the spinal anterior horns or the muscles; or whether it involves also sensory elements, namely, the portions of the spinal cord which transmit sensation, or the sensory fibres of the posterior roots or of the peripheral nerves. Similarly, in a case of spastic paraplegia, the student recognises that the lesion must involve either the adjacent cortical centres for the legs in the uppermost portions of the ascending frontal convolutions, or the pyramidal fibres which proceed from these centres to the lumbar enlargement in the spinal cord. The student, having examined a number of cases mainly with the object of becoming efficient in the anatomical or regional diagnosis of nervous diseases, is then prepared to grasp the significance of the grouping of symptoms, and to investigate the history

of the case, especially as regards the influences of heredity and environment, and the order in which the symptoms developed; he thus obtains information which enables him to form an opinion as to the nature of the lesion, and gradually acquires skill in solving the frequently complicated problem of pathological diagnosis.

In teaching I have always avoided the use of the terms organic and functional, and these words are rarely mentioned in this book. Whether it is true, as I believe, or not, that definite, though to our present methods invisible, lesions underlie all "functional disorders," it is certainly important that the student should be instructed to investigate such disorders in precisely the same way as he proceeds to investigate the "organic diseases." The anæsthesia, the spasm, the paralysis or the contracted visual field which may be met with in a case of hysteria must be allocated, so far as he is able, to a particular part of the nervous system before he proceeds to consider the nature of the disturbance, whether chemical, vascular or other, which is interfering with the functions of the affected part.

The adoption of such a method is not only valuable as regards the attainment of accurate knowledge, but it lessens the likelihood of overlooking the beginnings of serious changes in the nervous system, which may have led to the manifestation of some of the phenomena known as hysterical.

A large number of illustrations in the book are from photographs of patients who have been under my care. For permission to reproduce some of the other illustrations I am indebted to the kindness of many friends, among whom I would specially mention Drs. Anderton, Batten, Farquhar Buzzard, Wardrop Griffith, Henry Head, Gordon Holmes, Mott, André Moussous, Poynton, Reynolds, Purves Stewart, Stoddart, St. Clair Thompson, Kinnier Wilson, Williamson, Professor Lorrain Smith, and Sir William Gowers. I would also express my acknow-

ledgments to Messrs. Longmans, Green & Co., and to Messrs. Frowde and Hodder & Stoughton for placing at my disposal a few of the diagrams contained in the anatomical works by Quain, Gray and Cunningham.

The unfailing courtesy of my publishers, Messrs. Sherratt and Hughes, and the care and attention they have given to the work have been of the greatest help to me.

I am much indebted to Dr. Tylecote for the preparation of the index.

In conclusion, I wish to offer my warmest thanks to my friend Dr. Dixon Mann, and to express my sincere appreciation of the invaluable help he has given me in the progress of this book through the press. He has read and criticised every proof-sheet, and it is due to his literary ability and his critical acumen that many errors have been rectified and many inaccuracies of statement avoided.

JUDSON S. BURY.

ST. JOHN STREET,
MANCHESTER,
March 30, 1912.

It is only four days ago that I received the proofs of the last section of this book, with suggestive notes from Dr. Dixon Mann, and to-day he has passed away. The blow is so sudden and crushing that I feel quite unable to realise its effects; the overwhelming feeling is that I have lost one of my oldest and best friends, a friend full of generous thoughts, and one ever ready to help. Our friendship of thirty years' standing, and our close association of the last few months are memories which I shall always cherish.

April 6, 1912. JUDSON S. BURY.

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LIST OF ILLUSTRATIONS IN THE TEXT.

SECTION I.

Anatomical and Physiological Introduction.

The elements or units of which the nervous system is constituted are called neurons. Each neuron is composed of a nerve cell with its protoplasmic processes called dendrites or dendrons and its axis cylinder process called the neuraxon or axon. Formerly it was supposed that the terminal branching processes of the

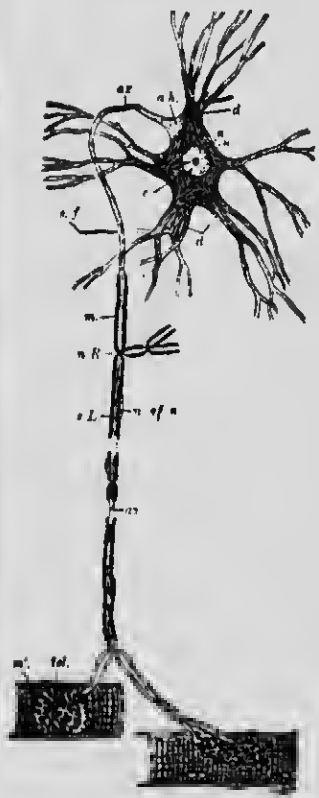

Fig. 1.—The Motor Cell Body, with its Protoplasmic Processes. a.h., axone-hillock devoid of Nissl bodies, and showing fibrillation; ax, axis cylinder or axone. This process, near the cell body, becomes surrounded by myelin, m., and a cellular sheath, the neurilemma, the latter not being an integral part of the neurone; c., cytoplasm showing Nissl bodies and lighter ground substance; d., protoplasmic processes (dendrites) containing Nissl bodies; n., nucleus; n′., nucleolus; n.R., node of Ranvier; s.f., side fibril; n. of n., nucleus of neurilemma sheath; tel., motor end plate; m′., striped muscle fibre; s.L., segmentation of Lantermann. (Barker.)

dendrites and axons of one neuron joined those of adjacent neurons. Recent investigations, however, are opposed to this view and now it is believed that the processes although contiguous, are not actually continuous with one another.

An axis cylinder is composed of a large number of conducting fibrils which separate in the nerve cell and pass to different dendrites; the dendrites themselves too are connected by fibrils which pass without interruption through the cell body. Hence it seems probable that the cell although able to maintain the nutrition of its various processes is unable to generate nerve force, any impulse arising within it being the result of a transmitted impulse; in other words impressions derived from the nutritional plasma surrounding the dendrites are transmitted by them to the fibrils which pass through the cell and converge together to form the axis cylinder. It is thus obvious that the cell body itself may be affected by impulses from many sources.

According to the direction of conduction in relation to the cerebral cortex, neurons may be classed as *afferent* and *efferent;* these form the main channels for sensory and motor conduction. A third class is constituted by a large number of neurons which act as connecting links between different parts of the nervous system. For example:—the fibres of the corpus callosum, formed of the axons of neurons that connect similar regions in the two hemispheres, and the neurons that connect each frontal lobe with the opposite half of the cerebellum. Such neurons are called *association* neurons.

EFFERENT PATH.

The efferent neurons constitute a path along which motor impulses are conducted, the exact distribution of which may be stated as follows: In the first place the path consists of two parts, namely, an upper or cerebro-bulbar and cerebro-spinal segment and a lower or bulbo-peripheral and spino-peripheral segment. The cell bodies of the neurons composing the **upper segment** of the motor path are situated in the Rolandic area of the cerebral cortex, and according to the experiments made by Sherrington and Grünbaum they have a narrower limitation than was formerly supposed. These experiments indicate that the motor functions are

mainly related to th. ascending frontal convolution and that the ascending parietal convolution does not

DIAGRAMS ILLUSTRATING POSITION OF MOTOR CENTRES IN CHIMPANZEE (AFTER SHERRINGTON AND GRÜNBAUM).

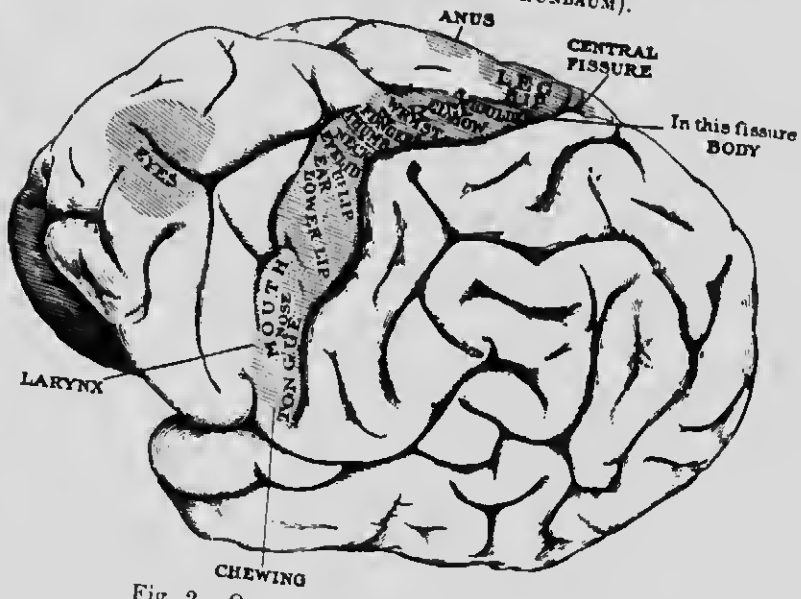

Fig. 2.—Outer surface of left hemisphere.

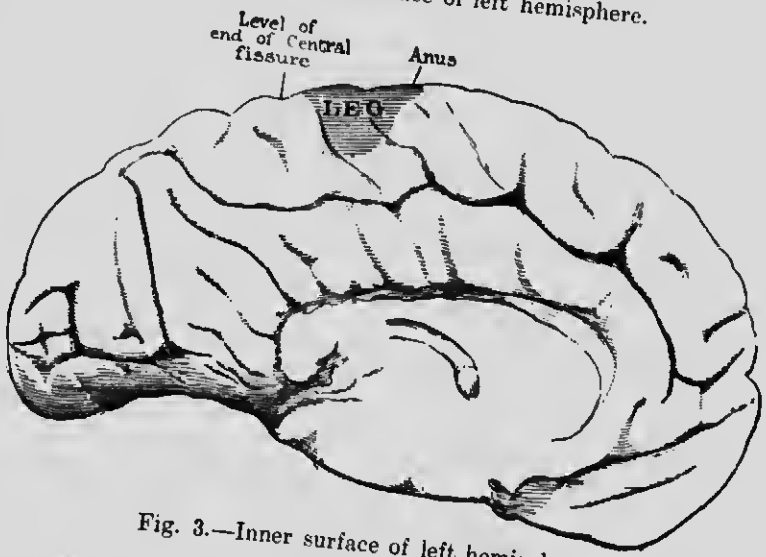

Fig. 3.—Inner surface of left hemisphere.

form any part of the motor area. In this connection it is interesting to note that the large pyramidal

nerve cells which are so abundant in the ascending frontal are not found in the ascending parietal convolution. The situation of the various motor centres is indicated in the accompanying diagrams. The leg centre occupies chiefly the upper fifth, the arm the middle two fifths, while the face, lips and tongue are represented mainly in the low two-fifths of the ascending frontal convolution. The motor centre for speech occupies the posterior end of the third frontal convolution together with the lower portion of the ascending frontal in the left hemisphere, and also in all probability the underlying island of Reil.

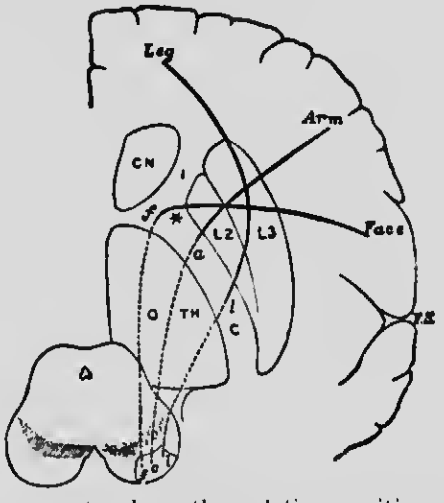

Fig. 4. Diagram to show the relative position of the several motor tracts in their course from the cortex to the crus. CN, caudate nucleus; O TH optic thalamus; L_2 and L_3, the middle and outer parts of the lenticular nucleus; f, a, l, face, arm, and leg fibres. (Gowers.)

The exact order of special movements from below upwards is stated by Sherrington and Grünbaum to be as follows:—Tongue, mouth, nose, ear, eyelids, neck, hand, wrist, elbow, shoulder, chest, abdomen, hip, knee, ankle, toes, perineal muscles, anus and vagina.

These areas are not so abruptly separated as the diagram might indicate; they overlap and the centre of each area must be stimulated in order to obtain the maximum of movement of the part it represents.

The axis cylinders of these cortical cells are prolonged into fibrils which collectively constitute the motor path, the situation of which is seen in the diagram. The fibres pass through the white substance of the hemisphere converging to the internal capsule where they occupy its "knee" and the anterior two-thirds of its posterior segment. The leg fibres are most posterior, and are next to the sensory tract, whilst still further backwards is the visual path. In the crus the motor

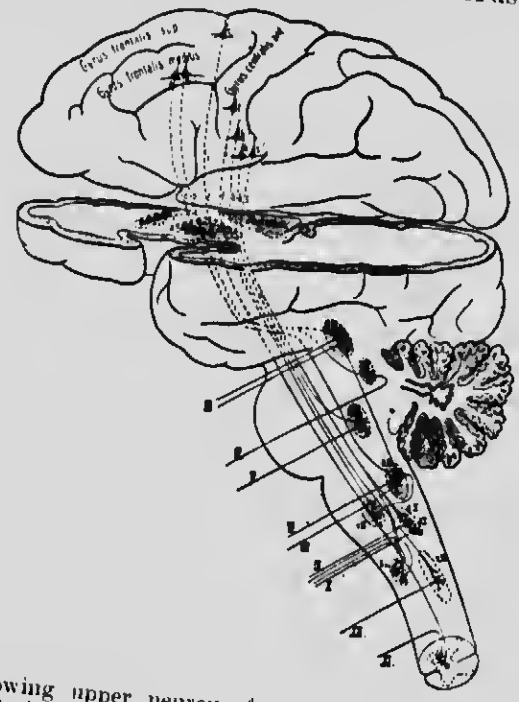

Fig. 5.—Showing upper neurons for the motor cranial nerves, according to Bechterew.

fibres occupy the middle two-fifths of the crusta. Entering the pons the fibres for the face and lower down those destined for the tongue leave the other motor fibres and cross the middle line to terminate in association with the facial and hypoglossal nuclei of the opposite side.[1] In the pons the limb and trunk fibres

1. With regard to the other motor nerves, namely, the 3rd, 4th, 5th, 6th, 10th and 11th the exact situation of the path of their upper neurons is not yet definitely known.

lie between the superficial and deep layers of transverse fibres; in the medulla they constitute the anterior pyramid, the larger proportion of its fibres crossing over at the decussation to run in the posterior half of the lateral column; this is the crossed pyramidal tract which, gradually diminishing in size, reaches to the lower end of the cord. A small variable proportion of fibres is continued into the anterior column of the same side of the spinal cord; many of these ultimately cross over within the cord to the opposite side. This direct pyramidal tract usually ceases about the middle of the dorsal region. Another small set of fibres (the homolateral pyramidal tract) runs down in the pyramidal tract of the same side, and conveys motor impulses to the homolateral leg. The fibres of these tracts end in arborisations in the grey matter on the floor of the fourth ventricle and in that of the posterior cornu of the spinal cord. Their terminal branches are in relation with the dendrites of the large cells which constitute in the one case the nuclei of the motor cranial nerves and in the other the nuclei of origin of the spinal nerves. These bulbar and spinal nuclei are composed of the cell bodies of the neurons of the lower segment of the motor path. The axons are prolonged into the fibres of the anterior roots and the peripheral nerves that are destined for the muscles.

The cortical centres preside over the nutrition of the pyramidal tract, and the bulbar and spinal nuclei over that of the motor fibres in the peripheral nerves. Thus there is a natural division of the motor path into two parts, an upper segment which extends from the cortical centres along the pyramidal tract to the bulbar nuclei and to the anterior horns of the spinal cord respectively, and a lower segment including the bulbar and the spinal nuclei together with the motor fibres which extend from them to the muscles. This division of the motor path into two parts corresponds to a convenient clinical classification of cases of paralysis, for we find that when the upper neurons are affected paralysis is

associated with spasm or rigidity of muscular tissue—*spastic paralysis*—whereas when the lower neurons are affected paralysis is associated with flaccidity and wasting of the muscles—flaccid or *atrophic paralysis*. The explanation of these differences is to be found in the trophic influence and the reflex functions of the nerve cells in the anterior cornua.

Fig. 6.—Diagram of upper and lower motor neurons; showing cortical cell, pyramidal fibre, anterior horn cell, nerve fibre, and muscle.

So long as the lesion is above the physiological level of these cells the nutrition of the muscles remains satisfactory, and their tone is increased, whilst since voluntary control over the reflex arcs is removed the tendon jerks become exaggerated. It is, however, to be noted that a *sudden* destruction of the cortical motor centres may produce at first a flaccid paralysis and a loss of the knee-jerk; this suggests the supervention of a dynamic factor which depresses the activity of the spinal centres. In lesions of the lower segment trophic

control over the muscles is removed and the reflex arc is hrokon, hence the paralysed muscles undergo atrophy and the tendon jerks cannot be evoked.

Our anatomical knowledge of the motor path shows the close correspondenco that exists hotween the distribution of paralysis and the site of a lesion. Thus in the case of the upper segment:—

1. If the lesion is above the decussation of the pyramids, the limbs and trunk muscles are paralysed on the opposite side of the hody.

(*a*) If it is situated above the middle of the pons, the opposite sidc of the face may also he paralysed.

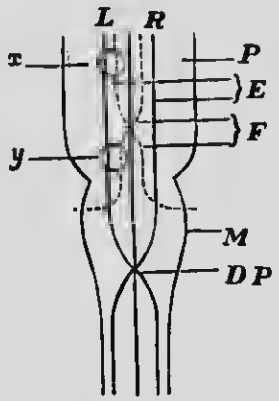

Fig. 7.—L, left; R, right; P, pons; M, medulla oblongata; DP, decussatio pyramidum; E, nerve fibres for the extremities; F, fibres destined for the facial nerve; x, lesion in the upper part of the pons; y, lesion in the lower part of the pons. (From Nothnagel.)

(*b*) If in the lower half of one side of the pons, the face is paralysed on thc same side as, hut the limbs on the opposite sidc to, thc lesion; this is callec "crossed paralysis." Such a lesion often involves the nucleus of the sixth nerve (around which the facial fibres in the pons form a loop) when the external rectus will be paralysed on the same side as the facial muscles.

(*c*) Another example of "crossed paralysis" is afforded by a lesion of the crus for in this situation the third nerve is usually implicated and then in addition to paralysis of the face, arm and leg on the opposite side of the body, there is paralysis of the ocular muscles supplied by the third nerve on the same side as the lesion.

(*d*) When a lesion involves the anterior pyramid of the medulla, a rare event, the face remains unaffected.

(*e*) Irritative lesions of the cortical centres cause convulsions, destructive lesions paralysis on the opposite side; and owing to the divergence of the motor fibres as they approach the cortex, and the consequent separation of those belonging to the face, arm, and leg respectively, the paralysis is often limited to the face or to one limb, when it is called *monoplegia*. The convulsions too of an irritative lesion are usually at first limited to a few muscles of the opposite side of the face or the opposite limb.

2. If the lesion is **below the decussation** of the pyramids, that is, is situated in some part of the pyramidal tract in the spinal cord, the limb and trunk muscles are paralysed on the same side. In the cord as a rule both tracts are affected, and hence both sides of the body are paralysed; in such cases the grey matter rarely escapes injury, and so modifications of sensation or of the reflexes usually accompany a spastic paralysis of spinal origin.

In the case of the **lower neurons** anatomical knowledge is also useful in determining the site of the lesion. Thus the anatomical relations of the bulbo-peripheral neurons indicate that the grouping of symptoms will be different when the nuclei or the nerve fibres are affected. For example it is common for the nuclei of the third, fourth and sixth nerves, which are close together on each

side of the fourth ventricle, to degenerate together, the result being paralysis of all the muscles of both eyes; whereas a lesion about the sphenoidal fissure may lead to paralysis of all the muscles of one eye.

Again in disease at the **base of the brain**, the fifth and sixth nerves may suffer together, or the seventh and eighth, or the spinal accessory and the hypoglossal. But in disease within the pons the sixth rather than the eighth would suffer with the seventh. It is interesting to contrast the successive involvement of the sixth, seventh and eighth nerves from a growth beginning in the centre of the pons with one beginning in

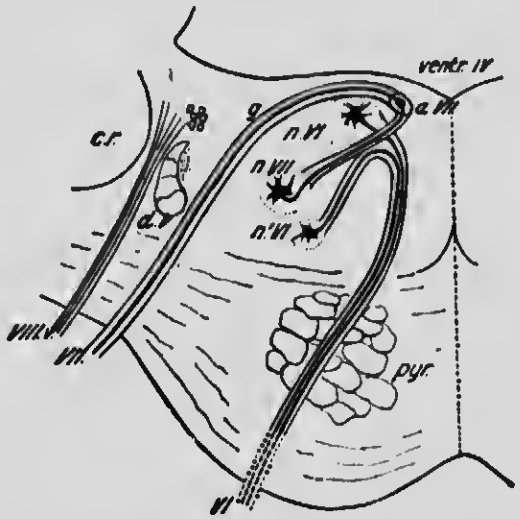

Fig. 8.—Showing the origin of the sixth and of the motor part of the seventh nerve. (E. A. Schäfer.) VI., sixth nerve; VII., seventh nerve; a.VII., ascending part of root of seventh, cut across near the floor of the fourth ventricle; g., genu of seventh nerve-root; n.VI., chief nucleus of the sixth nerve; n'.VI., accessory nucleus of seventh; d.V., descending root of fifth; pyr., pyramid-bundles; VIII.v., vestibular root of eighth nerve.

the cerebello-pontine angle and gradually invading the pons. In the former case both sixths are first implicated then as the tumour grows outwards to one side, the seventh and finally the eighth nerve becomes involved. In the latter case the eighth nerve is first caught, then the seventh and lastly the sixth.


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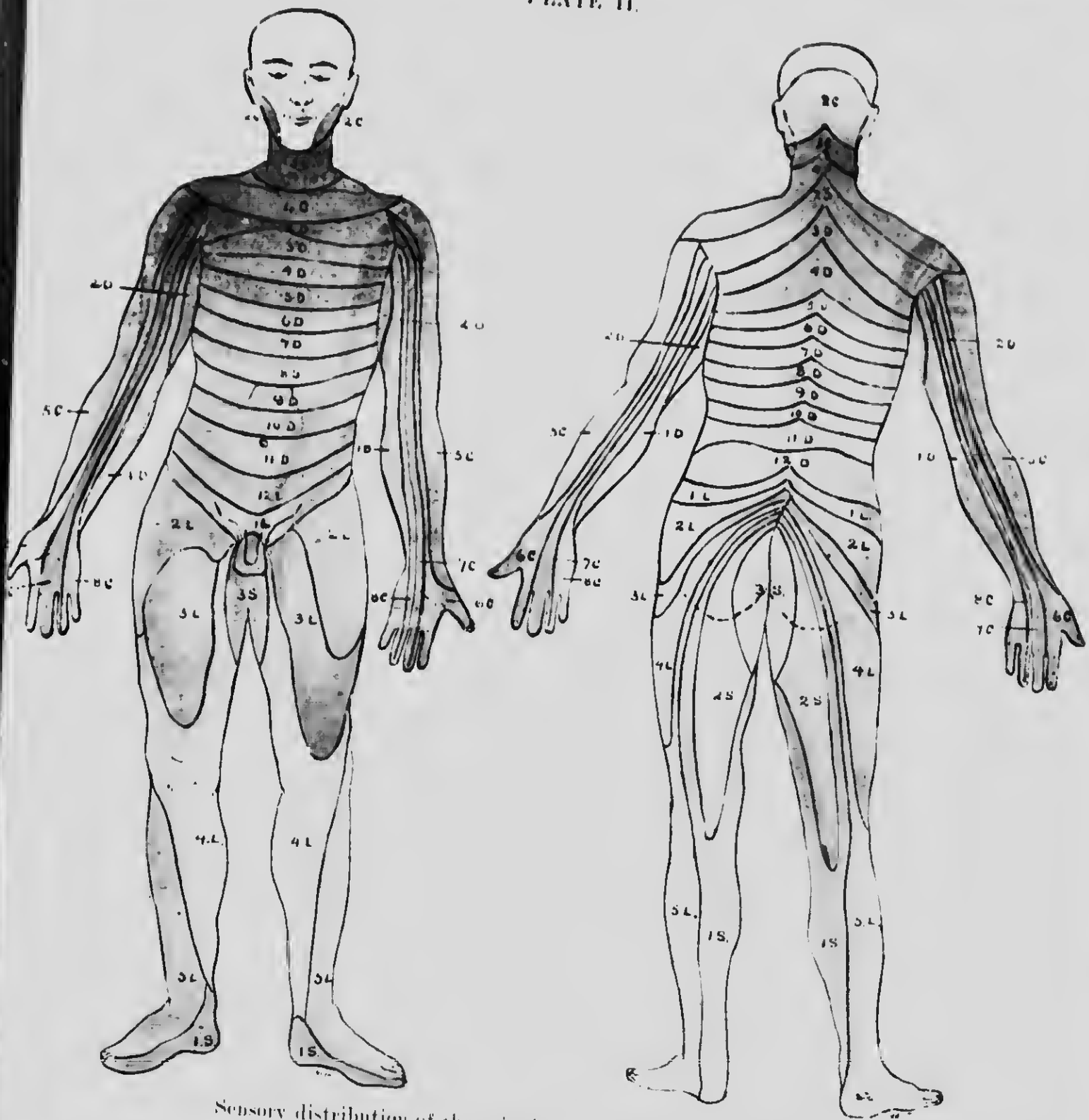
PLATE II.



Sensory distribution of the spinal nerve-roots (after Thorburn).

With regard to the **spino-peripheral neurons** a lesion limited to the cells of the anterior horns or to their axons in the roots and peripheral nerves will lead to an uncomplicated atrophic paralysis, but if the lesion is more extensive, sensory and other symptoms may accompany the paralysis. When the mixed peripheral neurons are diseased as in multiple neuritis sensory symptoms are often as prominent as, or more so than, the wasting of the paralysed muscles. Similarly in a myelitis in the lumbar part of the cord when the posterior as well as the anterior parts of the grey matter are diseased we find anæsthesia as well as atrophic paralysis in the legs. When disease is limited to one nerve the muscles supplied by it are alone affected, but when a spinal root or the group of cells from which it is derived is diseased then a group of muscles which act in functional association, but which are supplied by different nerves, become paralysed. Thus a lesion of the fifth root leads to atrophic paralysis of the deltoid, biceps and supinator longus and inasmuch as such a lesion is usually due to disease around the cord the sensory fibres of the root are also apt to be involved when a band of anæsthesia may be found down the outer side of the arm. The anæsthesia distinguishes a root lesion from an anterior horn lesion which leads only to an atrophic paralysis.

It is important to remember that the seat of a lesion which affects either a segment of the spinal cord or the superficial origin of its roots is at a higher level in the vertebral canal than that indicated by the paralysis or anæsthesia produced by the lesion. It is therefore necessary to be acquainted not only with the distribution of the nerve-roots but also with their relations to the vertebral spines. These facts are indicated in the following table, whilst the areas of skin supplied by the various spinal roots are mapped out in plate II.

Table showing the relation of the chief muscles to the spinal roots, and of the roots to the vertebral spines.

Apices of Spinous Processes.	Superficial Origin of Nerve-roots.	Muscles Supplied
C 1.....C 2	C 1, C 2, C 3	Deep muscles of the neck. Sterno-mastoid. Trapezius. Scaleni.
C 2.....	C 4	Diaphragm. Supra- and infra-spinatus.
C 3.....	C 5	Deltoid. Biceps. Brachialis anticus. Supinator longus. Pectoralis major.
C 4..... C 5.....C 6	C 6	Subscapularis. Pectoralis major. Serratus magnus. Latissimus dorsi. Pronators.
	C 7	Triceps. Extensors of wrist and fingers.
C 6.....	C 8	Flexors of wrist and fingers.
C 7.....D 1	D 1	Interossei and small muscles of the hand.
D 1.....	D 2, D 3	
D 2.....D 4		
D 3.....D 5		
D 4.....D 6		
D 5..... D 6.....D 9 D 7.....D10 D 8.....D11	D 7	Intercostals 7, 8, 9 — Rectus abdominis.
	D 8	Intercostals 10, 11, 12 — Internal and external obliques.
	D 9	
	D 10	
	D 11	
D 9.....	D 12	

Apices of Spinous Processes.	Superficial Origin of Nerve-roots.	Muscles Supplied.
D10.....	L 1	Quadratus lumborum.
	L 2	Cremaster.
D11.....	L 3	Sartorius. Adductors and flexors of hip.
	L 4	Quadriceps cruris. Abductors of hip.
	L 5	Flexors of knee.
	S 1	Calf muscles.
	S 2	Glutei. Peronei. Anterior tibial muscles. Intrinsic muscles of foot.
D12.....		
	S 3, S 4	Perineal muscles connected with micturition, defæcation, erection and ejaculation.
	S 5	
L 1.....		

AFFERENT PATHS.

Sensory Path. According to the recent researches of Head there are three sets of sensory fibres conveying different sensations, namely: (1) Fibres which subserve *deep sensibility*; these respond to pressure and conduct impulses produced by movements of tendons, joints and muscles and thus give information regarding the degree of movement and the position of a part. Such fibres are believed by Head to run with the motor nerves, for they are not destroyed by the division of all the sensory nerves to the skin. (2) Fibres which subserve *protopathic sensibility*; these respond to painful cutaneous stimuli and to the extremes of heat and cold. They regenerate rapidly when the ends of a divided nerve have been united. (3) Fibres which subserve *epicritic sensibility*; these fibres convey stimuli due to the lightest touch and enable the individual to discriminate between two points, and to recognise the finer grades of temperature. They are more readily injured and regenerate more slowly than those of the protopathic system.

The two latter forms of sensibility are conveyed by sensory fibres to the posterior roots. But "as soon as a sensory impulse reaches its first junction in the spinal cord, it becomes shunted into tracts devoted to the conduction of impulses, grouped in a way different from that found in the peripheral nerves." "It is no longer a question of protopathic, epicritic, or deep sensibility; the tracts in the central nervous system are devoted to the conduction of impulses concerned with pain, heat, cold and touch."

The exact position of these tracts in the spinal cord is not yet accurately determined. It is probable that all forms of sensibility with the exception of those concerned with passive position and movement, and with tactile discrimination cross to the opposite side of the cord, but there is reason to believe that the crossing is less sudden and uniform than was formerly supposed. The path for tactile sensations probably runs up the posterior column of the same side for a short distance and then crosses into the ventro-lateral tracts of the opposite side. The paths for painful and thermal sensibilities cross soon after their entrance into the cord; they run close together but are not intermingled, and impulses of cold are separated from those of heat. These paths run up the opposite side of the cord, either in the mesial part of the lateral column or in the posterior grey horn. That they are more centrally situated than the path for touch is indicated by the condition of cutaneous sensibility met with in syringomyelia. The impulses concerned with passive position and movement and with tactile discrimination, that is, recognition of two points of a pair of compasses, are conducted by the long intrinsic fibres of the posterior column up the same side of the cord to the nucleus gracilis and the nucleus cuneatus, and thence by fibres which cross to the opposite side. From the cord the sensory path passes into the formatio reticularis in the posterior half of the medulla and pons where the path from the fifth nerve joins it: then it ascends through the crus cerebri

to the optic thalamus. In this body the various sensory impulses from the opposite side of the body are collected together. Higher they are conducted by fibres which pass through the posterior limb of the internal capsule, and then diverge in their upward course through the white substance of the hemisphere to reach the post central convolution and the adjacent portion of the parietal lobe, for it is in those portions of the cortex that sensory representation is believed to be situated.

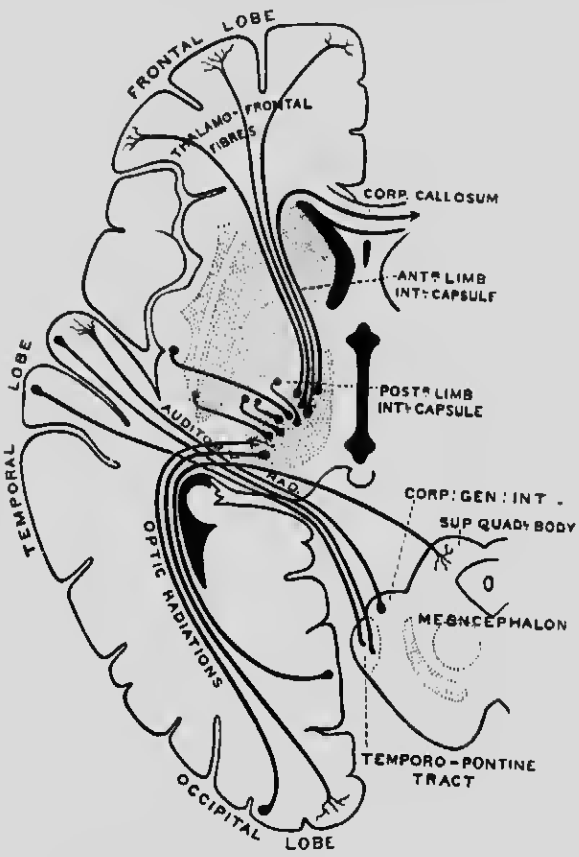


Fig. 9 (from Cunningham's 'Anatomy').—Shows the thalamo-ortical projection system of fibres.

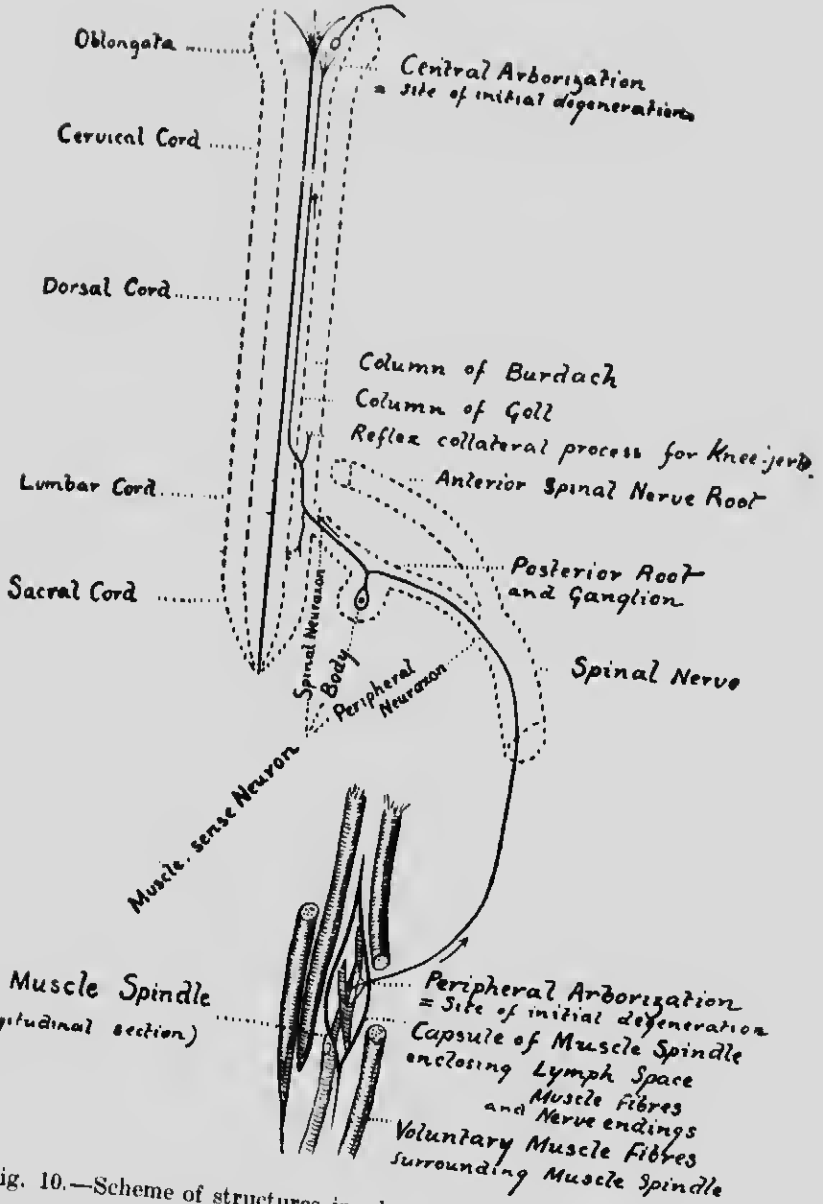
The sensory path as above described is not composed continuous fibres as in the case of the motor path, t is constituted by several systems of neurons. Thus

the ganglia in the posterior roots are connected by one set of neurons with cells in the grey matter of the cord and by another set with the cells of the nucleus gracilis and the nucleus cuneatus. From these different groups of cells axons arise which terminate in the thalamic region. Lastly sensory neurons connect the optic thalamus with the cerebral cortex.

A lesion of the pons involving the formatio reticularis near the superficial origin of the fifth nerve causes loss of sensation on the same side of the face and on the opposite side of the body; whilst a lesion of the lower part of the pons may produce loss of sensation on the opposite side of the body up to, but not above the level of the lower jaw. In the uppermost portion of the pons the sensory path for the fifth nerve crosses to the opposite side, hence a lesion at a higher level, namely at the crus, the hindmost part of the internal capsule or the parietal portion of the cortex will tend to produce loss of sensation in the skin and mucous membranes of the whole of the opposite half of the body. Lesions also of the so-called motor area are sometimes attended by blunting or perversion of sensibility, in the skin of the paralysed limb; moreover, motor spasms from irritation of the cortex are often preceded by a sensory aura.

A complete unilateral lesion of the cord produces what is known as Brown-Séquard's paralysis. On the opposite side of the body below the level of the lesion there is loss of sensibility to painful and thermal stimuli, whilst tactile and light pressure sensibilities may or may not be impaired. On the side of the lesion, in addition to loss of power, there is loss of the sense of position as well as of tactile discrimination, and of the vibrating sensation.

In cases of brain disease when anæsthesia attends paralysis the relative sequence of these symptoms would indicate the direction in which the lesion was advancing, thus the sensory being posterior to the motor tract, a lesion advancing from behind forwards would produce sensory before motor symptoms.



ig. 10.—Scheme of structures involved in tabes. (Langdon.)

OTHER AFFERENT PATHS. Of these, those which
rvey impressions from the periphery to the cerebellum,
ing it information regarding the position of the
ly in space, require a brief notice. They are—
(1) the muscular sense or equilibrium path;

(2) the vestibular path; and
(3) the path connecting the cerebellum with the centres for ocular movements.

(1) *The equilibrium path.* This is mainly constituted by the direct or dorsal cerebellar tract. The cell body of the muscle-sense neuron is in the posterior ganglion and has two branches. One branch, which may be regarded as a dendrite, receives impulses from the "muscle spindles" which are believed to be the end organs of muscle-sense fibres. The other branch or axon conveys these impulses to the cells of Clarke's vesicular column from which the direct cerebellar tract originates. Impulses along this tract as well as along the posterior median column, which may also convey

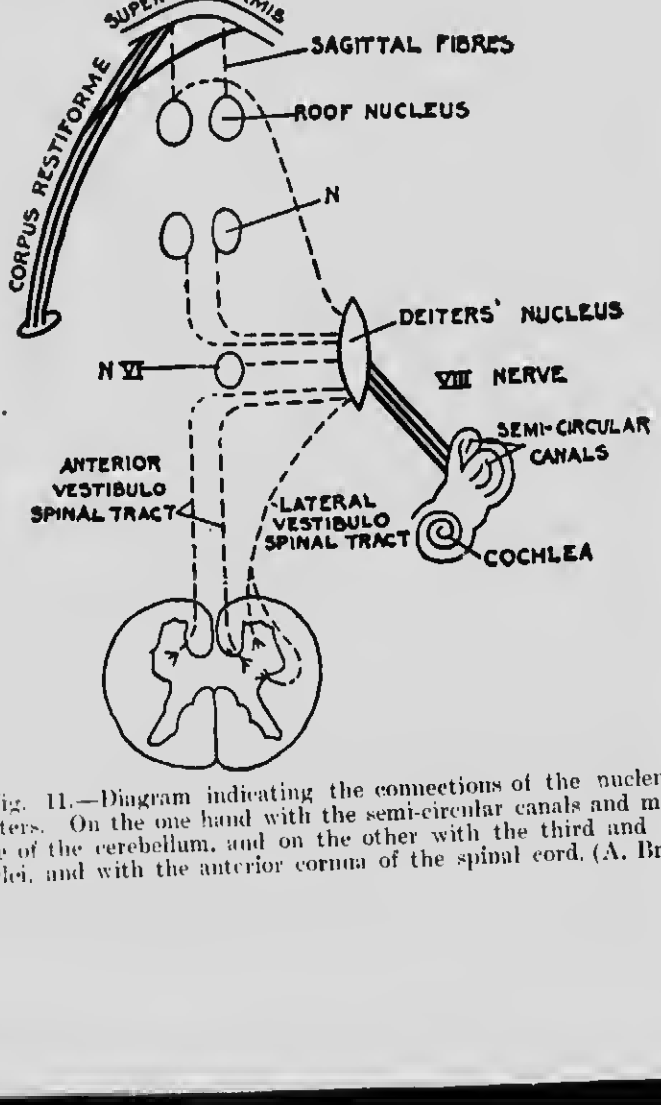


Fig. 11.—Diagram indicating the connections of the nucleus of Deiters. On the one hand with the semi-circular canals and middle lobe of the cerebellum, and on the other with the third and sixth nuclei, and with the anterior cornua of the spinal cord. (A. Bruce.)

sensations of position from the muscles are eventually conveyed to the cerebellum by means of its inferior peduncle.

A lesion of any part of this path whether it be in its peripheral or in its central axons will tend to produce inco-ordination of movement, the chief reason probably being that afferent impulses from the muscles are prevented from reaching the cerebellum. The cerebellum may indeed be regarded as the centre for the co-ordination of muscular contractions which maintain the body in a position of equilibrium. It acts, however, in strict subordination to the cerebrum, with which it has a crossed connection by means of the superior peduncles. It appears to regulate muscular contractions which are initiated and are subject to constant changes by the action of the motor centres in the cerebral cortex. Put in another way we may say that the cerebrum controls the finer, more conscious movements of the body whilst the cerebellum regulates the less conscious movements of locomotion and equilibrium. The cerebellum exerts an important influence upon the spinal cord by means of an efferent tract, which passes through the nucleus of Deiters and thence along the antero-lateral tract to the anterior horns of the same side of the cord. A cutting off of this tract may possibly explain the paresis occasionally met with in the limbs on the same side as a lesion in the cerebellum. It seems probable that each lateral lobe exercises a tonic influence on the muscles of the homolateral limb. Deiters's nucleus also receives fibres from the vestibular nerve and sends fibres into the posterior longitudinal bundle, through which probably it influences the nuclei of the oculo-motor nerves.

(2) *The vestibular path.* The fibres of the auditory nerve which receive impressions from the semi-circular canals unite to form its vestibular root which passes backwards through the pons on the inner side of the restiform body to end in nuclei, which are connected by neurons with the cerebellum, the dorsal longitudinal fasciculus and the sixth nucleus.

A lesion of the semi-circular canals or of the vestibular nerve tends to produce disturbance of equilibrium: the patient feels giddy and may find it impossible to stand. In Menière's disease such disturbance is frequently associated with tinnitus and deafness, and then the cochlear is affected as well as the vestibular nerve.

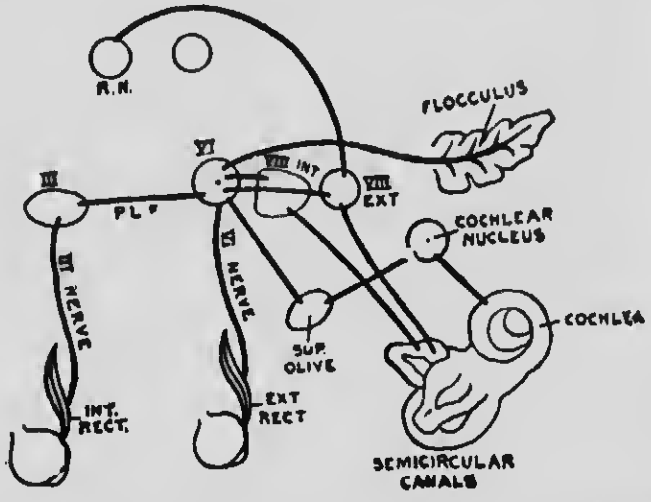


Fig. 12.—To show how the nucleus of the sixth nerve acts as the centre for conjugate deviation of both eyes to the same side, and also its connection with the auditory nuclei. (A. Bruce.)

(3) *The ocular path.* In all probability there is a connection between the centres for ocular movements and the cerebellum, for it is by the position of our eyes that we unconsciously estimate our relation to objects seen.

It is thus seen that the cerebellum receives impulses from the muscles through its inferior peduncles, from the semi-circular canals and the nuclei of the oculo-motor nerves through its middle peduncles, and from the cerebrum through its superior peduncles. According to Sherrington, the cerebellum is the "head ganglion of the proprio-ceptive system," that is of afferent impulses chiefly from the labyrinth and the muscles of the trunk and limbs. These impulses reach

the cortex of the cerebellum by way of the inferior and middle peduncles. From the cortex impulses pass to central nuclei, and thence along efferent tracts through the superior peduncles and red nuclei and the vestibulo-spinal tracts to exert an influence on the regulation and co-ordination of muscular movements.

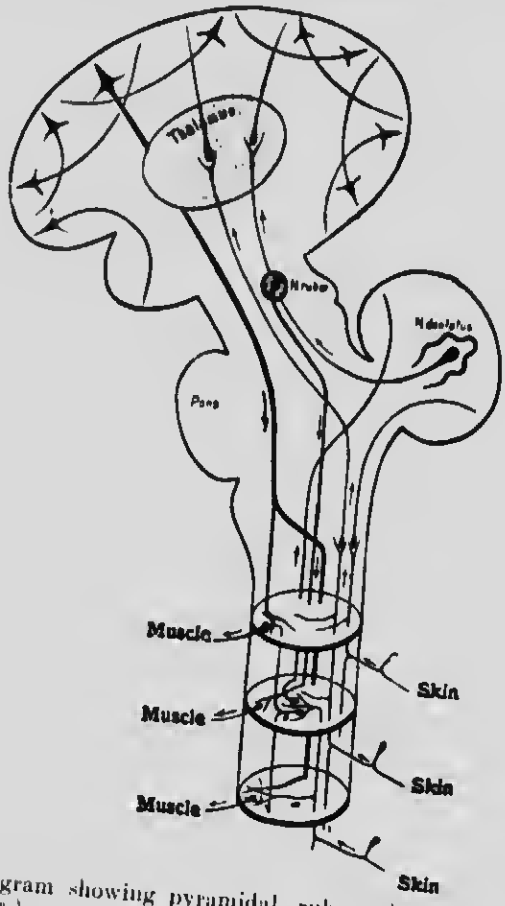


Fig. 13.—Diagram showing pyramidal, rubro-spinal and afferent racts. (Villiger.)

THE AUDITORY PATH is formed by a succession of eurons which link the fibres of the cochlear nerve with e superior convolution of the temporal lobe on the posite side. These neurons are (1) the bipolar cells of e organ of Corti, the axons of which constitute the hlear root; this enters the medulla on the outer side

of the restiform body and proceeds to (2) the terminal cochlear nuclei in the floor of the fourth ventricle from which (3) fibres go to the superior olive and the nucleus of the lateral fillet and thence to (4) those of the corpus geniculatum internum, the axons of which pass through

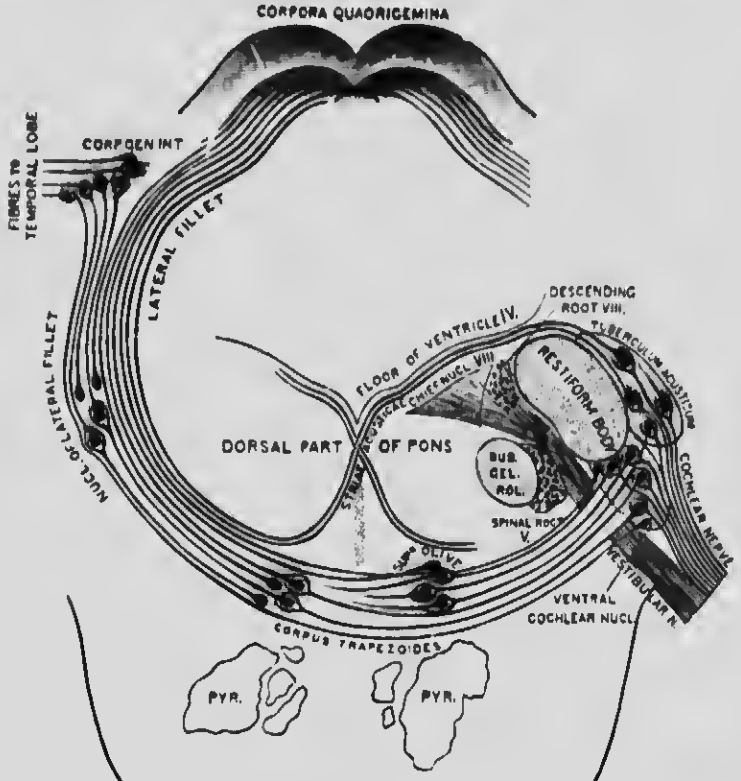


Fig. 14 (from Cunningham's 'Anatomy').—Showing the central connexions of the cochlear and vestibular divisions of the eighth nerve.

the posterior part of the internal capsule to the temporal lobe. The crossing of the auditory path takes place in the medulla and pons by way of the corpus trapezoideum and the lateral fillet; a few of its fibres, however, are transmitted by the lateral fillet of the same

side. The inferior quadrigeminal body, which is to be regarded as a ganglion accessory to the central auditory tract, is connected by fibres of the lateral fillet derived from the acoustic tubercle.

The Visual Path. The fibres of the optic nerves derived from the cells of the retinæ undergo a partial decussation in the chiasma, those from the inner half of each retina crossing to enter the optic tract of the opposite side, while those from the outer half of each retina enter the optic tract of their own side. Each tract passes round the crus cerebri to end in the pulvinar of the optic thalamus, the external geniculate body and the superior (anterior) quadrigeminal body. Fibres from these ganglia and particularly those from the external geniculate body enter the posterior segment of the internal capsule and pass by the "optic radiation" to end in the cuneus, being especially related to the sides of the calcarine fissure.

It is usually stated that the light reflex is subserved by the fibres in the optic nerve which go to the superior corpus quadrigeminum, some of them terminating in the cells of this ganglion, while others are related to the cells of the third nucleus for the sphincter of the iris. [illegible] evidence, however, seems to indicate that the ciliary ganglion is the peripheral motor nucleus which controls the sphincter pupillæ. The cortical half vision centres of the cuneus are connected with higher centres for visual memories which are situated in and about the angular gyri. In each angular gyrus the whole of the opposite field of vision is represented and to a less degree the whole field of the eye of the same side.

Any lesion that destroys the visual centre in the cuneus or interrupts the visual path between this centre and the chiasma impairs the functions of the outer half of the retina on the same side, and of the inner half of the opposite retina, and thus produces blindness on the side opposite to that of the lesion; this is called *lateral homonymous hemianopsia*. Thus if the right optic

tract is interrupted the patient when looking straight before him cannot see objects situated to his left. A lesion in the middle of the chiasma damaging the

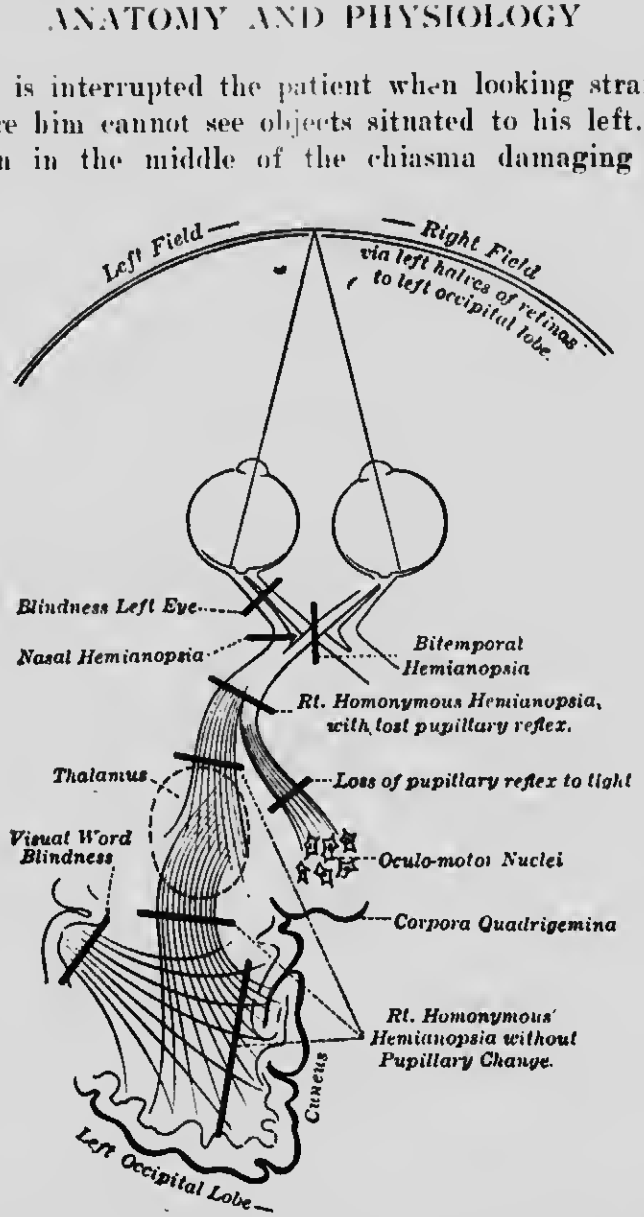


Fig. 15.—Diagram to show various forms of visual disturbance following lesions in different portions of the left visual pathway. (After H. Cushing.)

crossed fibres impairs the functions of the nasal half of each retina causing *temporal hemianopsia;* similarly injury to the uncrossed fibres by a lesion on each side of the chiasma leads to *nasal hemianopsia.*

In all probability a lesion of the angular gyrus would lead to concentric limitation of both fields of vision, the visual impairment being greatest on the side of the lesion. If the left angular gyrus is affected the variety of aphasia known as word blindness may be present.

THE OLFACTORY PATH. The olfactory bulb which lies on the under surface of the frontal lobe divides near the fissure of Sylvius into two roots; the outer root ends in the tip of the uncinate gyrus on the same side while the inner root crosses by way of the anterior commissure to the opposite hemisphere and reaches the cortex through the posterior limb of the internal capsule. Loss of smell, when not due to nasal disease, is usually caused by a lesion at the base of the brain, involving the olfactory bulb or tract: it also occurs in association with hemianæsthesia from disease of the posterior limb of the internal capsule on the opposite side.

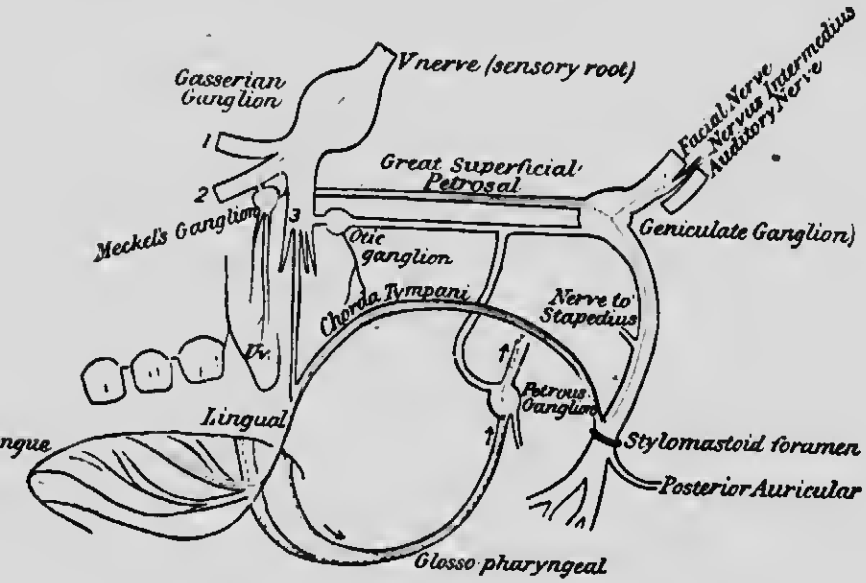


Fig. 16.—Diagram of trigeminal, facial, and glosso-pharyngeal nerves, showing course of taste fibres. (Purves Stewart.)

THE PATH FOR TASTE. The fibres for the anterior two-thirds of the tongue are derived from the lingual

nerve, those for the posterior third from the glosso-pharyngeal nerve. Between the tongue and the brain the course of the taste fibres is a complicated one, and may vary in different individuals. Some authorities believe that the sense of taste is conveyed to the brain solely by the roots of the fifth nerve, others by the sensory root of the facial and the roots of the glosso-pharyngeal nerve. The following appears to be the most probable route. The taste fibres in the lingual nerve pass into the chorda tympani and run along with the facial nerve to the geniculate ganglion. From this some fibres run in the great superficial petrosal nerve to Meckel's ganglion and ultimately join the second division of the fifth; other taste fibres leave the geniculate ganglion by the sensory root of the seventh (nerve of Wrisberg) and enter the medulla where in all probability they are connected with the glosso-pharyngeal nucleus.

The taste fibres from the posterior third of the tongue may be transmitted to the brain entirely by the glosso-pharyngeal nerve; some, however, may only run in this nerve as far as the petrous ganglion, their subsequent course being by Jacobson's nerve, the small superficial petrosal nerve to the otic ganglion from which they run in the third division of the fifth nerve to the brain.

The central route for taste is unknown; it may or may not pass through the internal capsule. The cortical centre for taste is in the anterior end of the temporo-sphenoidal lobe where it is in close relation with the olfactory centre.

For the anatomical relations of the other cranial nerves and their nuclei of origin the student is referred to works on anatomy. Here it must suffice to indicate in the accompanying diagrams some of the main features of their distribution and to make the following observations:—

(1) That there is a close connexion by means of fibres in the posterior longitudinal bundle between the nuclei of the third, fourth and sixth nerves which

govern the complex movements of the eyeballs. In this bundle there are fibres which connect the third nucleus of one side with the sixth nucleus of the opposite side: this connexion explains the associated action

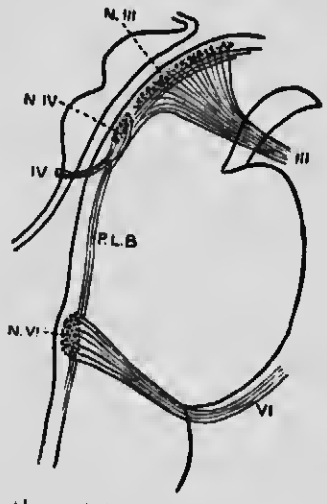


Fig. 17.—Plan of the origins of the third, fourth, and sixth nerves. (Quain.) III, third nerve; N.III, its nucleus; IV, fourth nerve; N.IV, its nucleus; P.L.B., posterior longitudinal bundle; V, sixth nerve; N.VI, its nucleus.

of the internal and external recti of opposite sides, which occurs in conjugate movements of the eyeballs.

(2) That the nucleus of the third nerve is made up of several groups which probably have distinct functions. These are classified by A. Bruce as follows: A median group associated with the movement of convergence, a postero-internal which subserves accommodation and pupil contraction, a postero-external which supplies the elevators of the lid and the eyeball, and an anterior group which sends fibres to the internal and inferior recti. Bruce points out that the first two groups are supplied from a set of vessels which do not enter the last two groups.

(3) That the sixth nerves have a long course before they enter the dura mater, and therefore are much exposed to injury and readily suffer from pressure as they pass beneath the pons.

(4) That the nuclear origin of the fifth nerve is a

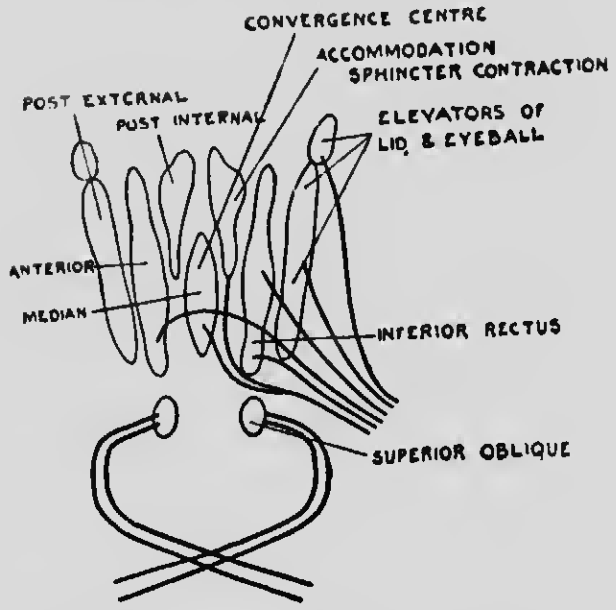


Fig. 18.—Diagram of the segments of the third nerve and their functions. The diagram shows also the position of the fourth nucleus and the course of its nerve. (A. Bruce.)

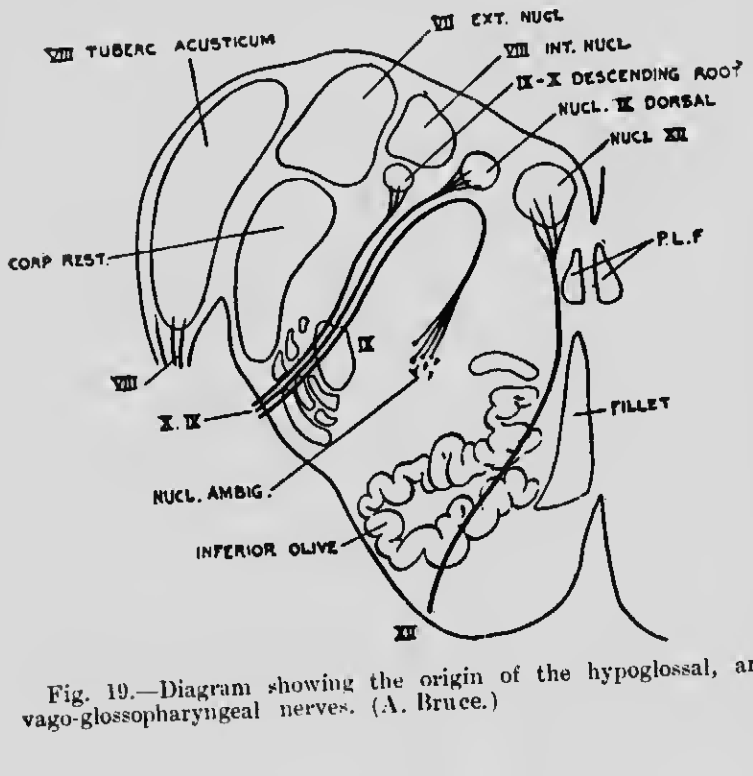


Fig. 19.—Diagram showing the origin of the hypoglossal, and vago-glossopharyngeal nerves. (A. Bruce.)

very extensive one; there is an upward extension of the motor nucleus as high as the corpora quadrigemina, while the larger sensory nucleus is connected with grey matter down to the level of the second cervical root.

(5) That the tenth nerve or vagus is now believed to include those roots which formerly were held to belong to the bulbar portion of the "spinal accessory." The latter term is now limited to the spinal part of the accessorius which comes from a separate nucleus. It is a purely motor nerve and is distributed to the sterno-mastoid and to part of the trapezius. A lesion of the roots of the hypoglossal and vagus nerves will cause paralysis of the tongue, palate and vocal cord on the same side while degeneration of the nuclei of these nerves will also often produce weakness of the lips, the probable explanation being that the fibres from the facial nerve to the orbicular oris are derived from the hypoglossal nucleus.

BLOOD VESSELS OF THE BRAIN.

The arterial supply is derived from the internal carotids and the vertebrals. The internal carotid artery after giving off the anterior cerebral is continued into the middle cerebral artery. The vertebrals unite to form the basilar artery which runs forward in the median groove of the pons and at its anterior border divides into the posterior cerebrals. The anterior cerebrals are united by the anterior communicating artery while the posterior cerebrals are connected with the internal carotids by the posterior communicating arteries and thus the anastomosis known as the circle of Willis is completed. The arterial supply to the brain may be divided into two systems of vessels, cortical and central. The cortical arteries anastomose freely with one another. But the central are terminal arteries and neither anastomose with each other nor with the cortical vessels.

Cortical Branches. The anterior cerebral supplies the first, a portion of the second frontal and the upper

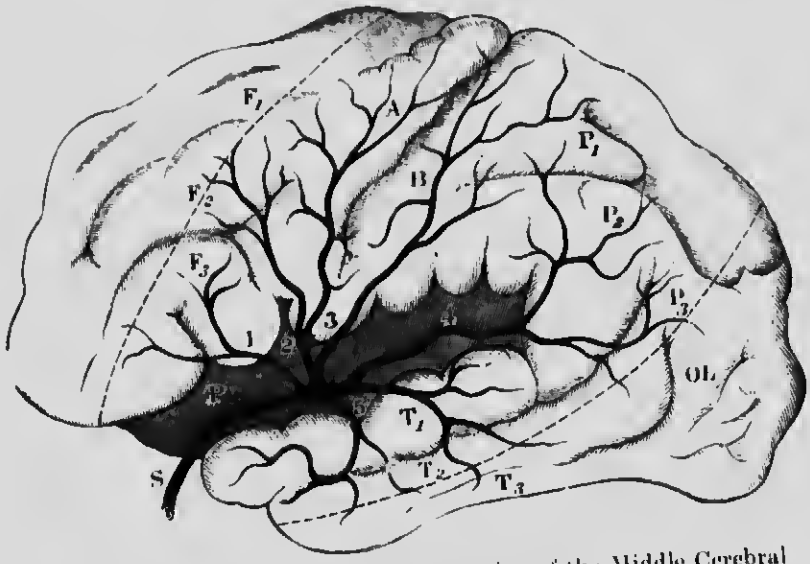


Fig. 20.—Showing the Area of Distribution of the Middle Cerebral Artery. (Duret.) S, Sylvian or middle cerebral artery; P, perforating branches; 1, inferior frontal branch; 2, ascending frontal branch; 3, ascending parietal branch; 4 and 5, parieto-sphenoidal and sphenoidal branches; A, ascending frontal convolution; B, ascending parietal convolution; F_1, F_2, F_3, first, second and third frontal convolutions; P_1, P_2, P_3, first, second and third parietal convolutions; T_1, T_2, T_3, first, second and third temporo-sphenoidal convolutions; OL, occipital lobe.

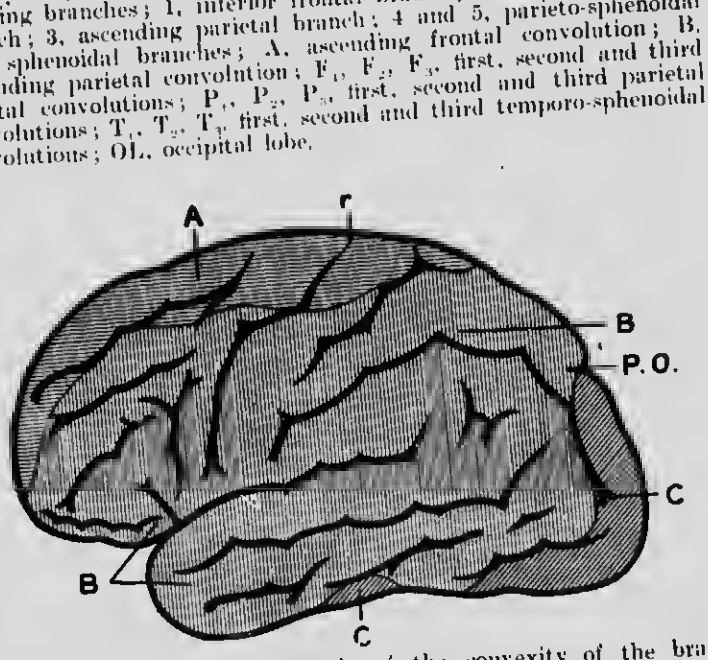


Fig. 21.—The vascular supply of the convexity of the brain. (Turner and Stewart.) A (horizontal lines), anterior cerebral artery; B (vertical lines), middle cerebral artery; C (oblique lines), posterior cerebral artery; P.O., external parietal-occipital fissure; r, fissure of Rolando.

parts of the ascending frontal and the ascending parietal convolutions, also the medium cortical surface of the hemisphere as far back as the internal parieto-occipital fissure. The posterior cerebral supplies the ventro-mesial portion of the temporal and occipital lobes with the exception of the anterior end of the temporal lobe and of the uncinate gyrus, the former being supplied by the middle cerebral and the latter by the anterior choroidal artery. The rest of the cortex is supplied by the middle cerebral artery which is distributed to the motor and sensory regions, to the auditory and speech centres and to the higher visual centre.

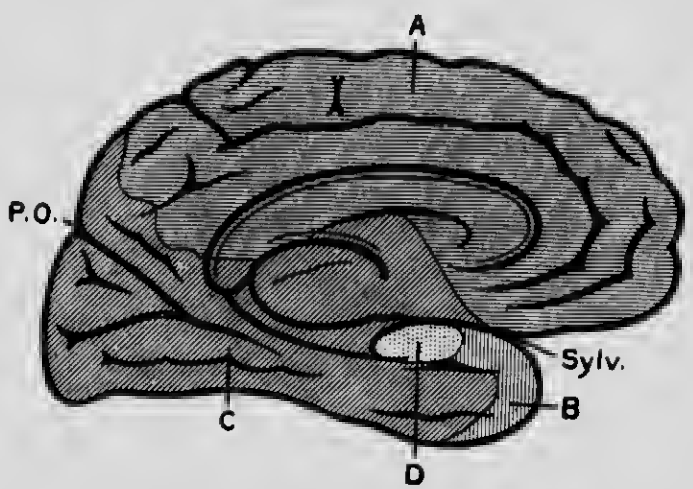


Fig. 22.—The vascular supply of the mesial surface of the hemisphere. (Turner and Stewart.) The lettering is the same as in fig. 21. D (dotted area), anterior choroidal artery.

Central Branches are derived from the circle of Willis and from the three cerebral arteries. The anterior cerebral supplies the floor of the third ventricle, the head of the caudate nucleus, the anterior limb of the internal capsule, portions of the lenticular nucleus, and the mesial half of the central ovale of the frontal lobe. The posterior cerebral gives branches to the crus, the red nucleus, the optic thalamus, the corpora quadrigemina, and the optic radiations.

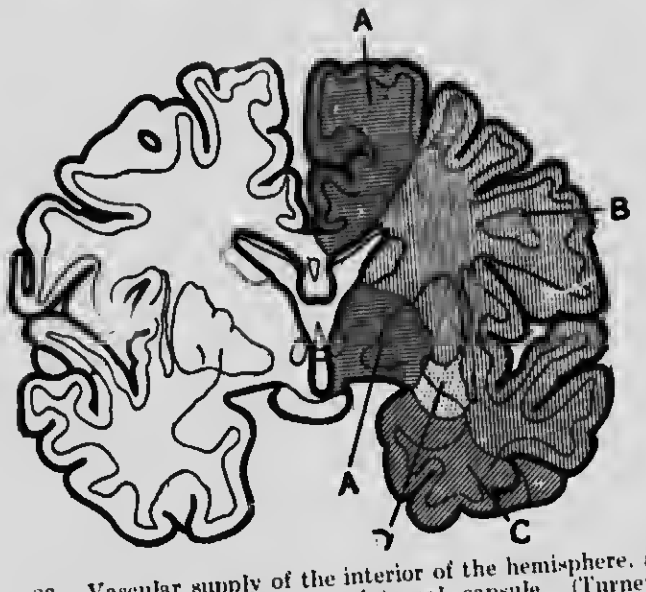


Fig. 23.—Vascular supply of the interior of the hemisphere, at the level of the anterior limb of the internal capsule. (Turner and Stewart.) The lettering is the same as in figs. 21 and 22.

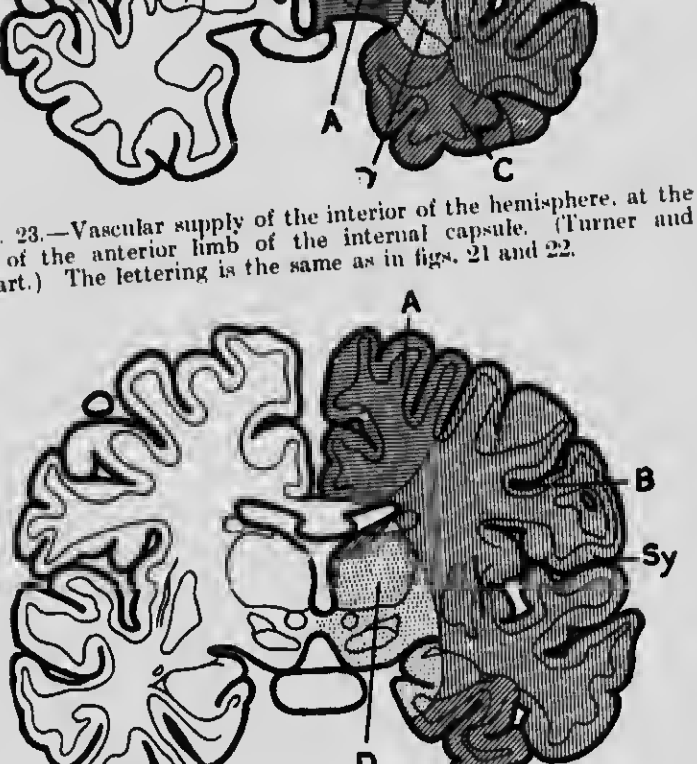


Fig. 24.—Vascular supply of the interior of the hemisphere, at the level of the optic thalamus and posterior limb of the internal capsule. (Turner and Stewart.) The lettering is the same as in the preceding figure.

Soon after its origin the middle cerebral artery sends branches to the basal ganglia and the posterior limb of the internal capsule. They go to the internal capsule

partly through the lenticular nucleus and partly outside it. The external branches are the largest and consist of two sets—an anterior set called lenticulo-striate branches which end in the caudate nucleus, and a posterior set called lenticulo-optic branches which run backwards to end in the optic thalamus. Both these sets but especially branches of the former set are liable to rupture. A third branch called the choroidal artery which runs backwards along the optic tract to enter the lateral ventricle is also derived from the middle cerebral artery.

As already stated branches of the central arteries do not communicate, hence thrombosis or embolism of one of them leads to permanent necrosis of brain tissue, whereas when a cortical artery is blocked the development of a collateral circulation may arrest the necrotic process. Senile softening is prone to occur in the ill-nourished zone of brain tissue between the terminal branches of the cortical and central arterial territories. Softening, too, is not uncommon in the pons and medulla, for the branches of the vertebral, basilar and cerebellar arteries which supply these parts do not communicate. It is rare, however, in the cerebellum owing to the free communication between its arteries.

The Veins. The blood after irrigating the brain is conveyed by veins into the various sinuses, the position of which is shown in figs. 180 and 181. It takes its exit from the cranium mainly by means of the internal jugular veins.

The chief points to be remembered about the venous circulation are (1) That the cortical veins enter the superior longitudinal sinus in a direction opposite to that of its blood stream. This together with the absence of valves in the cerebral veins and sinuses offers an explanation for the readiness with which clots are formed in them. (2) That the veins from the choroid plexus return blood from the lateral ventricles and end in the veins of Galen which discharge into the straight sinus. Any obstruction of the veins of Galen causes

effusion into the ventricles, but if there is much hydrocephalus it is probable that the foramen of Magendie, which maintains a communication between the ventricular cavity and the subarachnoid space, is completely occluded. (3) That in addition to the continuation of the lateral sinus into the jugular vein there are several other communications between the intra- and extracranial veins. Thus the veins of the nose and most of those of the scalp are connected with the superior longitudinal sinus; the occipital veins communicate with the lateral sinus by means of the mastoid veins, the deep cervical veins with the inferior petrosal sinus, while the ophthalmic veins connect the facial vein with the cavernous sinus.

BLOOD VESSELS OF THE SPINAL CORD.

It is probable that many diseases of the spinal cord are more closely related to vascular lesions than hitherto has been demonstrated, hence it is desirable to indicate the chief facts regarding the distribution of the vessels and to suggest the necessity of making a close study of the relations of lesions to arterial territories.

The spinal cord derives its arterial blood from an anterior and a posterior system of vessels

The anterior arterial system consists of the anterior spinal artery reinforced by branches from the intercostal, lumbar and sacral arteries. This artery which arises by two roots from the vertebrals runs downward in the region of the anterior fissure from the upper end of the cervical cord to the conus medullaris. In its course it gives off from 250 to 300 branches which are called anterior median arteries. Each of these branches passes horizontally into the anterior fissure, then turns to the right or the left, and passing through the anterior commissure enters the grey matter. There it divides into an anterior central artery, which supplies the anterior horn, and a posterior central which is distributed to the intermediate grey matter, to a portion of the posterior horn and to the vesicular column of

Clarke. The anterior central artery before dividing gives off an anastomatic artery which by means of its two branches is connected with the anastomatic arteries above and below it and thus a longitudinal chain of vessels is formed in the grey matter.

The posterior arterial system consists of the two posterior spinal arteries which arise separately from the

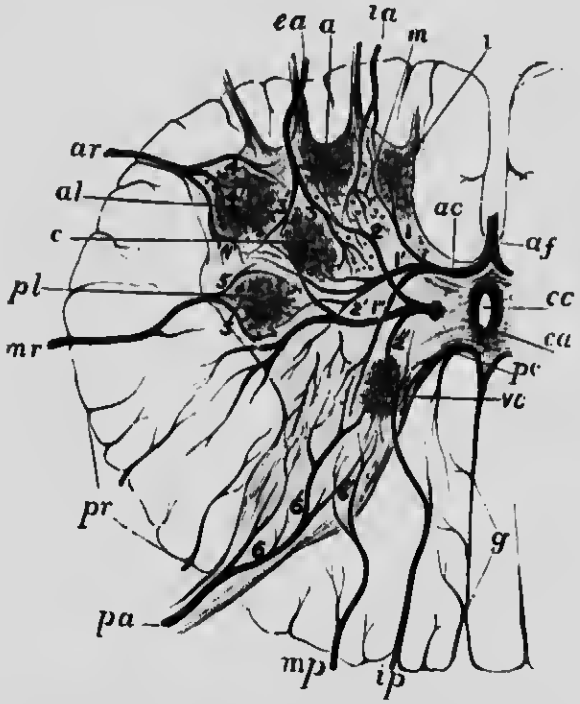


Fig. 25.—Diagram of the Distribution of the Blood-vessels, and Grouping of Ganglion Cells in the Spinal Cord. (Young.) Anterior median artery; af, arteries of the anterior median fissure; ac, artery of the anterior commissure; 1, anterior branch; 1′ median branch; 1″, posterior branch; ca, central artery; 2, anterior branch; 2′, median branch; 2″, posterior branch; pa, posterior root arteries; 6 6′ 6″, arteries of posterior horns; ia, internal anterior root artery; ea, external anterior root artery; 3 3′, internal and external branch; ar, antero-lateral branch; 4, anterior branch; 4′, median branch; 4″, posterior branch; mr, median lateral artery; 5 5′, anterior and posterior branches; pr, posterior lateral arteries; ip, internal posterior artery; mp, external posterior artery; g, arteries of the column of Goll; pc, artery of the posterior commissure; vc, vesicular column of Clarke; i, internal group of cells; a, anterior group; al, antero-lateral group; pl, postero-lateral group; c, central group; m, median area.

vertebral arteries and extend the whole length of the cord just in front of the posterior nerve roots, together with their communicating branches with the intercostal

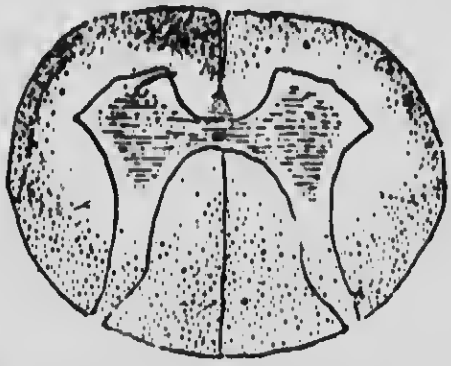


Fig. 26.—Section of the Cord showing division into three arterial districts (diagrammatic). Part supplied only by the anterior median and its branches is shaded with parallel lines. Part supplied only by the peripheral arteries is shaded with dots. Part supplied by both systems of arteries is unshaded. (Williamson.)

and lumbar arteries. Transverse anastomoses between the anterior and posterior systems are formed on the surface of the cord; there are also anastomatic branches

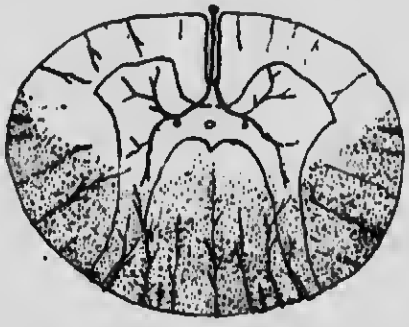


Fig. 27.—Transverse Section of Cord. Area supplied by posterior arterial system shaded with dots. Area supplied by anterior arterial system is not shaded. (Williamson.)

on the posterior aspect of the cord. From these branches small arterial twigs penetrate the cord and supply the posterior portions of the white and grey matter. The territories of the two systems are shewn

in the diagrams; their irrigation fields overlap and thus three arterial districts may be distinguished:—

(1) The district supplied only by the anterior system;
(2) That supplied only by the posterior system, and
(3) That irrigated by both systems.

Owing to the long and tortuous course of the spinal arteries, they are not subject to high pressure, which is such an important element in the production of degeneration and rupture of the arteries of the brain.

The arterial supply to the lower end of the cord is made with difficulty owing to the distance from the vertebral of the anterior and posterior spinal arteries. The reinforcing arteries too which accompany the nerves of the cauda equina have a long ascending course before they reach the cord: they are also very slender and tortuous. Hence the blood stream is sluggish and for this reason the lowest part of the cord has been said to be more prone to disease than its higher portions. But this is scarcely true, for certainly the mid-dorsal region of the cord is a commoner site for myelitis than the lumbar region.

Veins. There are three longitudinal veins, one vein follows the course of the anterior median fissure, another that of the posterior median fissure and the third lies near the central canal of the cord. The central vein receives blood from the grey matter and sends branches forwards to join the anterior spinal vein. The posterior spinal vein receives blood from veins on the lateral and posterior surfaces of the cord. The blood in the anterior and posterior veins is delivered by means of communicating veins into large venous plexuses which lie outside the dura mater. From the plexuses blood passes to the cervical and intercostal veins. At the upper part of the cord the plexuses communicate with the vertebral veins. The veins of the cord cannot be injected from the extra-dural plexuses, hence a hindrance to the return of blood from the plexuses does not produce over-distension of the veins of the cord.

Lymphatic Spaces ensheath the adventitia of the spinal blood vessels; there are also lymphatic clefts in the adventitia but there are no true lymphatic vessels in the cord itself.

According to Orr and Rows there is a constant flow of lymph upwards along the peripheral nerves, both cranial and spinal, towards the central nervous system, the main current of which lies in the inner meshes or lymph spaces of the fibrous perineural sheath.

SECTION II.

Etiology and General Pathology.

It is beyond the scope of this manual to give a detailed account of the etiology and pathology of nervous diseases. In the account of each disease a description will be given of its etiology together with the chief facts relating to its morbid anatomy. It seems desirable to make a few introductory remarks also on the various ways in which the functions of nerve-tissue may be deranged and on the resulting visible alterations in its structure.

In the main the causes are the same as those which produce disease in other parts of the body and similarly may be classified under the headings of *heredity* and *environment*. For the sake of simplicity we may restrict our attention to a single neuron and consider any morbid influences in its neighbourhood which may adversely influence it. The neuron is bathed in lymph and is surrounded by vessels and a fine network of connective tissue called *neuroglia*. It is also to be noted that its branches or dendrites are contiguous to those of other neurons. A little consideration of these facts makes it obvious that the ways in which a neuron may be injuriously affected are numerous. Thus its resistance may be lowered by inherited feebleness, by poisons in the lymph which surrounds it, or by changes in adjacent neurons. Again, a neuron may be lacerated or actually destroyed by external violence, or its structure may be seriously damaged in consequence of concussion, or of compression by a tumour. It may be torn or compressed by the sudden impact of blood which has escaped from a ruptured artery. In such a case hereditary tendencies so premature atheroma may be traced; this shows that the influence of heredity in the production of nervous disease may be due to early degeneration of extra-

neuronic as well as of neuronic tissue itself. Obviously the pathological problem in any given case may be a difficult one; we have to consider the influence of heredity on many tissues, the influence of impressions from other neurons, whether they be near the neuron under consideration, or situated at some distance from it, and finally the influences depending on changes in the surrounding vascular tissue and in the blood itself.

Our knowledge of the etiology of disease is in its infancy; hence it is incumbent on every student to record with care every fact that can be obtained from the patient or his friends regarding his medical history along with that of his parents, and other relations.

Heredity.

Referring to the subject a little more in detail, it may be pointed out that under the term heredity it is necessary to include all tendencies derived from the parents which may affect the reproductive cells before conjugation, or the organism itself during its intra-uterine development or at any period after birth. An inherited predisposition is one of the most important factors leading to nervous disease, and is no doubt frequently active even when its influence cannot be directly traced. Thus, in the case of such typically hereditary disorders as insanity and epilepsy, we sometimes meet with isolated cases in families in which no neuropathic tendencies can be discovered; yet it is easier to assume that some instability of cortex has been inherited than that a normal cortex has become deranged by an agent outside it. Such instability of nerve-tissue or congenital deficiency in resisting power to changes in its environment is seen, not only in epilepsy and insanity, but in migraine, hysteria, and other neuroses, and is particularly apt to show itself in some form or other in the offspring of alcoholic or syphilitic parents. Sometimes the vulnerability of the nervous system is shown in families by the appearance of different neuroses in different members; thus one member may suffer from

neuralgia, another from hysteria, epilepsy, or insanity, whilst a third may manifest a tendency to uncontrollable alcoholic excesses.

In other cases heredity is shown by the transmission of defective vitality in certain groups of neurons, or in muscular tissue, leading to early degeneration of the particular tissue involved. A good example of the former is Friedreich's disease, of the latter idiopathic muscular atrophy. These are family diseases, and may manifest themselves in several generations, and at about the same age in each case.

The influence of heredity in the production of nervous disease may also be due to its effects on extra-neuronic tissues, namely the neuroglia and the vessels. This is especially noticeable as regards a tendency in certain families to early vascular degeneration leading to apoplexy; it is also probably a factor in the development of new growths. Another example of indirect inheritance is tuberculous meningitis, as when several children in one family die in succession from convulsions and coma, not so much from inherent weakness of the nervous system as from a strong tendency to tuberculous disease.

Environment.

This term includes all the external influences that may be brought to bear on nervous tissue during any period of its development, growth, or subsequent life. Such agencies may be considered under the headings of injury or trauma, abnormal conditions of the blood supply and abnormal stimulation.

Injury.—Traumatism is an important factor in the production of nervous disease, quite apart from the results of fracture or disease of the skull or vertebral column, or of laceration of the peripheral nerves. A violent shaking or concussion of the intra-cranial contents may lead to meningeal hæmorrhage and even to rupture of a cerebral vessel; in the latter case there has usually been pre-existing disease of the vessel-wall, but

occasionally apoplexy, occurring some weeks after a blow on the head, appears to be due to changes started in the artery at the time of the injury.

Neurasthenia often owes its origin to cerebral concussion, and may depend on molecular changes in the cells of the cortex associated with overgrowth of neuroglial tissue, or on more obvious lesions, as minute lacerations, punctiform hæmorrhages, and other changes which collectively constitute a contused or bruised brain. The prolonged depression and irritability so often associated with the latter condition may even lead to insanity. There is evidence, too, that a tumour may owe its development to the nutritive changes produced by concussion.

Similarly in the case of the spinal cord molecular changes have been started by a simple concussion of the body, and have ultimately led to manifestations of cord disease; indeed, there is reason to believe that occasionally the degenerative changes which underlie the symptoms of tabes or of disseminated sclerosis are started by severe concussion, although no doubt in most cases the injury simply served to light up disease which was pre-existent, or for which the ground was duly prepared.

Abnormal conditions of the blood supply.—It is stated that the total quantity of blood within the brain undergoes few if any variations, and that owing to the absence of vaso-motor nerves [1] the size of the arteries does not vary. According to this view changes in the cerebral circulation will depend on changes in the general arterial and venous pressure; and hence it is incorrect to invoke spasm of a cerebral artery as an explanation of transient symptoms such as a hemiplegia lasting a few days or of more prolonged symptoms as in the case of many hysterical anæsthesiæ. On these points it is impossible to speak positively, and our knowledge is limited mainly to local alterations in the

1. "There is some, but not very definite, evidence pointing to the existence of weak cerebral vaso-motor nerves."—LEONARD HILL.

quantity of blood, of which a marked instance is the anæmia produced by the narrowing of an artery due to disease in its walls, or to its being blocked by a thrombus or an embolus.

A more striking cause of nervous disease is an alteration in the *quality* of the blood. This may depend on:

(1) A diminution of an excess of a normal constituent of the blood. The most noteworthy example of diminution or absence of a normal constituent of the blood giving rise to impaired brain functions is afforded by myxœdema and cretinism, the symptoms of which depend on absence of the thyroid secretion. Then the nervous phenomena of Graves's disease are ascribed either to an excess of thyroid secretion entering the blood, or to an insufficient quantity of the secretion of the parathyroid gland. Here, too, may be mentioned the lassitude and mental fatigue which result either from an insufficiency of oxygen, or from an excess of carbonic acid and nitrogenous waste products, as well as the stupor, delirium and psychical symptoms which occur in cases of heart failure owing to engorgement of the brain with venous blood.

(2) The presence of *abnormal constituents in the blood*. Under this heading are poisons which are introduced into the body, as alcohol, lead and arsenic, and poisons which are produced within the body, as the toxins of syphilis, tubercle and the specific infectious diseases. These toxins to which the nervous phenomena may be ascribed are the products of micro-organisms, many of which have been distinguished and are well recognised. In the case of chronic degenerative diseases disseminated sclerosis and amyotrophic lateral sclero-, and also of some acute nervous affections as acute terior polio-myelitis and Landry's disease, it is sumed that they also are set up by poisons, some of ich may be the products of bacteria, whilst others are ived from the products of over-fatigue or of abnormal estion in the alimentary canal, or from the perverted ction of some other organ or tissue.

The selective action of some poisons is very striking. Thus lead selects the nerves which supply the extensor muscles of the wrists and fingers, whilst alcohol picks out the higher centres of the brain as well as the peripheral nerves, especially those of the lower limbs. The poison of syphilis shows a preference for the vascular structures at the base of the brain, for branches of the cerebral and spinal vessels, for the afferent conducting paths of the spinal cord, and for the cortical cells in the anterior part of the brain. The early symptoms of diphtheritic paralysis, namely, paralysis of the soft palate and the ciliary muscles, indicate that the neurotoxin of this disease picks out special parts of the nervous system. Again, in rabies the medulla oblongata is chiefly affected, whilst in tetanus the virulent poison elaborated by the bacilli first affects the motor nucleus of the fifth nerve. Of the curiously selective properties of many poisons there appears to be no end, and indeed, it seems as if any variety of cell or fibre may be selected by a poison as its seat of attack. It is still uncertain whether toxic matter first attacks the neuron or the endothelial cells lining the vessel which supplies it. Possibly some toxins have their action limited to the nerve-cells, but the histological evidence in many cases is strongly in favour of a previous, or at any rate, of an associated vascular lesion. Certainly the prevalence of vascular lesions is a striking feature in the morbid anatomy of cord and brain diseases; in myelitis, in disseminated sclerosis, and in polio-myelitis the spots of disease are closely related to the distribution of blood-vessels, which may contain thrombi, or may have their coats thickened and their peri-vascular lymph-sheaths crowded with round cells. Such changes strongly suggest that the effects have been produced by the action of some irritant on the endothelial lining of the vessel, which leads to an increased flow of lymph and leucocytes into the peri-vascular tissues and thus initiates the earliest nerve lesions. Orr and Rows state that toxins reach the brain and spinal cord by the

current of lymph which ascends along the peripheral nerves, and although toxins spread to some extent in the lymph spaces of the piu-arachnoid, and so affect structures at a distance, they pass for the most part into the substance of the central nervous system. These observers believe that the vascular changes found in the cord in acute lesions are not of primary hæmatogenous origin, but are reactive changes due to a primary lymphogenous infection of the adventitia of the vessel walls.

Abnormal stimulation.—The effects of deficient or of excessive stimulation of neurons sometimes appear to be limited to the particular neurons which are subjected to the stimulation; at other times they are expressed by disturbance of the functions of neurons at a distance from those stimulated.

This statement is best illustrated by a series of examples. Prolonged mental worry or anxiety may, in those predisposed to neuroses, lead to an outbreak of hysteria or to melancholia or other forms of insanity. In these cases the higher cortical centres which have undergone the strain are those which break down, although it cannot be denied that other neuronic systems may be secondarily affected. In writer's cramp and in similar motor derangements associated with the excessive use of a particular group of muscles, the effects of over-stimulation appear to be limited to the lower neurons, although it is possible that irritation of sensory nerve-fibres has its first effect on cortical motor cells.

Arthritic muscular atrophy is a more obvious example f the spread of over-stimulation from sensory to motor-eurons—for the muscular atrophy of the affected limb ppears to depend on damaged nutrition of the motor lls of the cord as a result of morbid impulses from e irritated nerves of the affected joint. Indeed, periments have shown that lesions of the posterior ts produce visible changes in the spinal motor cells. doubtedly healthy sensory impulses are essential for proper nutrition of motor neurons, whilst morbid

sensory impulses will lead to detectable disease. A still wider diffusion of irritation occurs when convulsions are excited by over-stimulation of a child's intestine by the presence of a worm, or a mass of hard curd; although no doubt there is usually pre-existent irritability of the cortex owing to such malnutrition as is associated with rickets. In some cases the transference of stimulation is from the sympathetic to the cerebro-spinal system; thus, in angina pectoris the pain radiates along the cardiac branches of the sympathetic to the spinal cord, and thence is referred to the sensory nerves supplying the inner side of the left arm. Similar referred pains occur in many visceral diseases.

On the other hand *defective stimulation* will also lead to disease; a familiar example is the atrophy of both grey and white matter in the spinal cord, which gradually comes on after amputation of a limb. The nervous structures which thus suffer are those which have ceased to be used.

This principle of loss of function leading to permanent defect is also illustrated by many neuroses and psychoses; thus the impaired function of cortical cells associated with prolonged mental depression is apt to lead to actual melancholia, in which condition the functions of certain cells may be presumed to be abolished.

In many cases there is a co-operation of the agencies I have mentioned. For example, the effects on nerve-tissue during the growth of a tumour are the results partly of injury by compression, partly of irritation, and partly of interference with its blood-supply.

The Nerve Cell and its Degeneration.

A nerve cell is made up of protoplasmic material with a large oval nucleus and nucleolus situated in its centre. By the aid of suitable staining re-agents the protoplasmic material is seen to consist of two parts, an achromatic part which hardly stains and is called cytoplasm or trophoplasm, and a chromatic part which stains

deeply and is called kinetoplasm. The former forms the body of the cell and is continued into the axis cylinder; it is fibrillated, the fibrillæ passing into the processes. The latter is composed of a number of granular bodies called Nissl or chromophile bodies: it is continued into the dendrites but not into the axis cylinder.

In the brain and spinal cord the axis cylinder is covered by a myelin sheath; outside the central nervous system it has another covering, namely the primitive sheath or sheath of Schwann which is continued in the peripheral nerves (see figs. 1, and 28).

The earliest visible degenerative changes in a nerve cell that has been exposed to the action of a poison or other morbid influence is swelling and gradual disappearance of the Nissl or chromatic bodies. This is spoken of as *chromatolysis* and at this stage the degenerative process is reparable. A further change is represented by swelling of the nucleus which takes up a peripheral position; the chromatic bodies are replaced by fine dust, and later the cytoplasm becomes vacuolated and the whole cell with its processes becomes shrunken and disturbed. Such a condition is said to exclude the possibility of repair.

Degeneration of Nerve Fibres.

When a mixed nerve is divided changes occur on both sides of the lesion. Below the lesion, degeneration extends down to the ultimate nerve endings: this, called Wallerian degeneration, runs a rapid course. Above the lesion, the fibre degenerates more slowly and only for a short distance, usually as far as the next node; this is called *disuse atrophy*. The Wallerian degeneration of the distal portion of the nerve fibre is characterised by a breaking up of the medullary sheath into droplets of myelin: the axis cylinder succumbs to a granular degeneration and finally disappears. It is to be noted that the cell also suffers and presents the condition

of chromatolysis with displacement of its nucleus, and if the injury to the axis cylinder is a severe one, its cell of origin gradually atrophies. A similar result follows section of an anterior spinal root.

When a posterior spinal root is divided Wallerian degeneration progresses upwards in the part of the root attached to the cord and in the cord along its afferent tracts, namely the columns of Goll and Burdach and Lissauer, and downwards along the afferent comma tract. In the first mentioned tracts the degeneration

Fig. 28.—Wallerian Degeneration of Nerve Fibre when separated from its cell of origin (diagrammatic). Upper fibre and cell normal. Nissl's granules seen in the cell. Myelin sheath of fibre, deep black. Lower cell and fibre show changes following division of fibre. Myelin is broken up into globules (deep black). Lowest fibre shows final change—collapsed nerve sheath, myelin absorbed. Note spindle-shaped masses of proliferated cells of external sheath (faint black). (Williamson.)

extends upwards gradually diminishing in amount as far as the nucleus gracilis and nucleus cuneatus in the medulla.

Transverse lesions of the spinal cord whether produced by injury, pressure or disease, lead to secondary degenerations above and below the lesion, for both afferent and efferent axons are separated from their cells of origin. Descending degeneration is seen in the crossed and direct pyramidal tracts, in the comma tract, in the rubro-spinal tract which comes from the opposite red nucleus, and in the antero-lateral descending (vestibulo-spinal) tract from the cerebellum. Ascending

degeneration is seen in the direct cerebellar tract, the antero-lateral ascending tract of Gowers and in those tracts which as already mentioned degenerate as a result of division of the posterior roots. Lesions of the upper

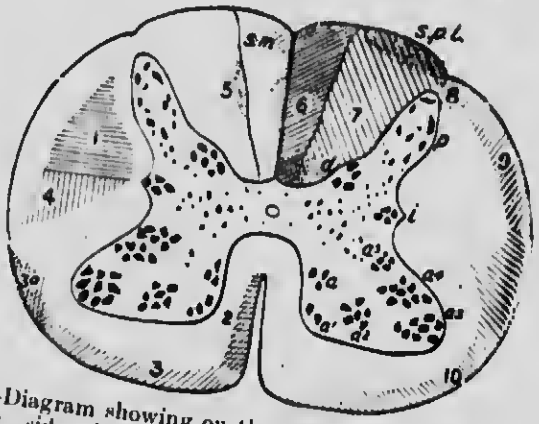


Fig. 29.—Diagram showing on the right side the 'ascending' and on the left side the 'descending' tracts in the spinal cord. 1, crossed pyramidal; 2, direct pyramidal; 3, antero-lateral descending; 3a, bundle of Helweg; 4, pre-pyramidal; 5, comma; 6, postero-mesial; 7, postero-lateral; 8, marginal; 9, dorsal cerebellar; 10, antero-lateral ascending or ventral cerebellar; s.m, septo-marginal; s.p.l., superficial postero-lateral fibres (dorsal root-zone of Flechsig); a to a^5, groups of cells in the anterior horn; i, intermedio-lateral group or cell-column in the lateral part of the grey matter; p, cells of posterior horn; d, dorsal nucleus or cell-column of Clarke. The dots represent 'endogenous' fibres (arising in grey matter of cord) having for the most part a short course. (Quain.)

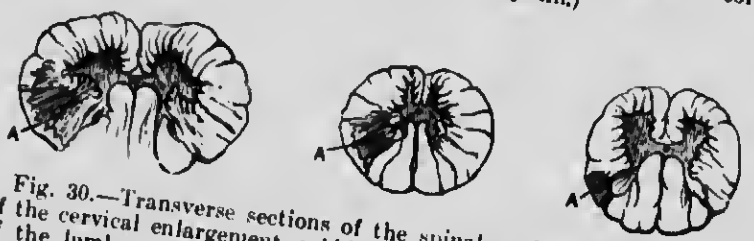


Fig. 30.—Transverse sections of the spinal cord, from the middle of the cervical enlargement, middle of the dorsal region, and middle of the lumbar region respectively, showing descending sclerosis of the pyramidal tract in the lateral column (A, A, A), secondary to a cerebral lesion. (Charcot.)

motor neurons in the brain lead to a secondary degenera-
ion of the crossed pyramidal fibres alone; this may be
raced down the cord to their terminal arborisations.
nother secondary degeneration may be observed at the

circumference of the antero-lateral column, that is in the vestibulo-spinal tract after a destructive lesion involving Deiters's nucleus; this degeneration is on the same side as the lesion.

In addition to these secondary degenerations, which are due to the cutting off of the axon from its cell of origin, there are *primary* degenerations which are set up by poisons circulating in the blood and lymph. The seat of the primary degeneration, that is the system of neurons affected, varies not only with the elective affinity of the particular poison, but also according to tendencies either inherited or acquired as a result of occupation, habits or other causes of stress, or of circulatory disturbances. As examples of such primary degeneration may be mentioned multiple neuritis, amyotrophic lateral sclerosis and tabes dorsalis.

Symptoms in Relation to Morbid Anatomy.

It is important for the student to recognise that no constant relation exists between the clinical phenomena and course of a disease and the changes found post-mortem. In many cases, indeed, a group of symptoms is the only part of the disease known to us. To this we give a name, and are said to have made a diagnosis. Thus to one group of symptoms we give the name of "paralysis agitans," to another "hysteria," to a third "spasmodic torticollis." The morbid anatomy of these conditions is unknown to us; we cannot speak with certainty in regard either to the site or the nature of the lesion. To such diseases, of which there are many, the term *functional* has been applied, whilst the term *organic* is applied to diseases in which morbid changes can be detected in the nervous system. But definite changes must underlie a so-called functional affection, and no doubt, with improved methods of investigating nerve-tissue, will ultimately be discovered. In the meantime it is necessary to recognise—(1) that the term "functional" must be taken to imply, not the absence of morbid changes, but only the absence of any which are

detectable; and (2) that symptoms run a course of their own which, even in the case of organic disease, cannot be adequately explained by an appeal to morbid anatomy. For example, the clinical progress of a brain tumour does not necessarily pursue a steady downward course. The tumour itself may be slowly and steadily enlarging, but the severity of the symptoms may vary from day to day, such variations no doubt partly depending on changes in the quantity or quality of the blood supplied to more or less healthy neurons in the vicinity of the growth. Indeed, symptoms almost identical with those occurring in organic disease are occasionally produced by poisons circulating in the system. Thus, to give only one instance, a boy under my care presented typical symptoms of tuberculous meningitis which were undoubtedly due to food intoxication.

Morbid anatomy, then, does not always throw light on the interpretation of the character and intensity of the symptoms observed during life, and sometimes the want of correlation between the two is striking. Hence the student is urged to make a *daily* study of symptoms even in chronic cases of apparently hopeless disease, for if he does so he will be astonished to find how frequently slight variations occur. Such a study will tend to convince him that the distinction between functional and organic disease is an artificial one, and will lead him to expect that in time some remedy may be discovered which will tend to accentuate the improvement that he observes from time to time in his study of the natural course of serious organic disease, a remedy which possibly may lead to actual recovery.

SECTION III.

The Clinical Manifestations of Nervous Diseases.

These may be conveniently grouped under the following headings:—

Motor symptoms—Electrical examination.
Sensory symptoms.
The reflexes.
Vaso-motor and Trophic changes.
The Special senses.
Speech and Articulation.
Mental symptoms and Disorders of consciousness.

In the investigation of a nervous case it is important to keep the above headings in mind otherwise striking symptoms may easily be overlooked. For example, a case of uncomplicated aphasia, moderate in degree, is shown to a student. He is told that the case is a nervous one and he is asked for a diagnosis. He looks for paralysis, examines the reflexes and tests the cutaneous sensibility and finds nothing abnormal. If now he remembers that there are other groups of symptoms to be considered he will naturally come to the investigation of speech. But a student imperfectly trained in the use of method might easily fail to make a diagnosis. It is also necessary to remind the student that his investigations must not be limited to the nervous system, but that he must also examine the heart, the kidneys and other organs of the body.

In taking the history of the case he should take pains to ascertain the initial symptoms, for these indicate the part of the nervous system which is first attacked. Thus if pain or numbness was first noticed by the patient we think of the sensory tract, if weakness of a limb we think of the motor path and so on. The exact sequence of symptoms should also be noted. A study of the

character, distribution, and grouping of the symptoms gives us information regarding the position of the lesion; this is the regional or *anatomical* diagnosis. A study of the mode of onset, the previous health and the family history of the patient furnishes evidence regarding the nature of the lesion; this is the *pathological* diagnosis. Thus a sudden onset of symptoms indicates a vascular lesion—hæmorrhage, thrombosis or embolism; an acute onset, the symptoms being developed in a few days, indicates some inflammatory condition; whilst a chronic onset, the full intensity of the symptoms not being reached for many weeks, suggests a slowly progressive lesion, such as a degeneration, or a new growth.

Examples. (1) Hemiplegia of the ordinary type indicates a lesion of the internal capsule. If the onset is sudden the lesion is a vascular one. If the patient is under forty and has had syphilis it is probable that there is thrombosis from thickening of the wall of the vessel. But if syphilis can be excluded and the patient has valvular disease of the heart it is probable that the vascular obstruction is due to embolism. If the patient is over forty and is the subject of chronic nephritis the lesion is probably hæmorrhage.

(2) The anatomical diagnosis of an *uncomplicated* paralysis, that is a paralysis unattended by sensory or other symptoms, with wasting of the muscles of both arms, is disease of the anterior horns in the cervical enlargement. The pathological diagnosis is based on the mode of onset and rate of development. If the onset is very gradual, the wasting palsy spreading slowly from one group of muscles to another, the lesion is progressive degeneration of the anterior horn-cells. But if the onset is sudden and the paralysis becomes intense within a few minutes, hæmorrhage into the anterior horns is the probable lesion. If the onset is acute, though not actually sudden, paralysis setting in within the first day or two of a feverish illness, the lesion is either inflammatory or is acutely degenerative, as for example anterior poliomyelitis.

In the two latter classes of cases the affected muscles are merely flaccid at first, atrophy not being conspicuous for some time.

MOTOR SYMPTOMS.

Many disorders of muscular action can be detected by a simple inspection of the patient. Thus when his body is at rest any decided alteration in the size or shape of a limb, the presence of tremor, of spasm, or of any abnormal attitude will be at once noticed, while attempts at active movements will reveal any decided paralysis, rigidity or inco-ordination.

Bulk. Considerable enlargement or diminution in the volume of a muscle or a group of muscles can be recognised at a glance. In order to detect slight alterations in size the limbs should be placed in symmetrical positions with their muscles as relaxed as possible, and a careful comparison be made between the two sides of the body. To compare the size of the arms, forearms and legs a measurement should be made of their greatest girths; in the case of the thighs a measurement should be taken about four or five inches above the patella.

Degenerative changes in a muscle do not always lead to a diminution in its size, for a muscle of normal, or even of increased bulk may be seriously diseased.

Spasm, is a term applied to increased muscular contractions which occur independently of any voluntary stimulus. It may be of the tonic or the clonic variety. *Tonic spasm* implies that the affected muscles are in a state of uninterrupted contraction, in other words that their tonus is excessive. When tonic spasm is long continued it leads to persistent rigidity of the affected limb: when it is limited to a particular group of muscles it is often termed *contracture*. Cramp implies the association of pain with spasm. Tonic spasm may occur apart from paralysis as in tetany and tetanus, or in combination with paralysis, a condition which is designated spastic paralysis.

In *clonic spasm* muscular contractions and relaxa-

tions quickly alternate; as a rule the movements are inco-ordinated and uninfluenced by volition, but in the variety known as *tic* they are co-ordinated and can be temporarily controlled by the exercise of the will. Clonic may pass into tonic spasms, or both may occur in the same case.

Tremor represents the most delicate, and convulsions the most severe and coarsest form of clonic spasm; between the two varieties every possible gradation is met with. When the tremor is limited to individual bundles of muscular fibres giving rise to wavy oscillations under the surface, it is called *fibrillary contraction;* such contractions are conspicuous in many cases of muscular atrophy. When tremor is more extensive in distribution it tends to cause manifest trembling of the limbs or trunk.

The term *convulsion* is applied (1) to cases in which a large number of the muscles of the body are simultaneously affected with spasms as in epilepsy, and (2) to severe forms of local spasm, such as the clonic spasms of a limb produced by irritative lesions of the cortical centres. There may be a single fit or a succession of fits. The term *eclampsia*, which is never applied to a single fit, is sometimes used to designate the recurrent convulsions which occur in uræmia, in the puerperal state and in young children apart from detectable disease of the brain.

Athetosis, a term given to slow irregular muscular contractions of a limb, and other disorders of movements which are occasionally found in association with hemiplegia will be described later. *Forced movements* such as rolling round the longitudinal axis of the body sometimes result from lesions of the middle peduncle of the cerebellum.

Paralysis. Spasm and tremor are examples of excessive muscular action, while paralysis implies diminished muscular action. Paralysis of a muscle or a limb means that voluntary power to move it is lost or impaired; sometimes the term is restricted to pronounced defects

of movement, "paresis" being used to express minor degrees of weakness. Paralysis limited to one side of the body—the face, leg and arm being specially affected—is called *hemiplegia;* paralysis of one limb or one side of the face if dependent on cortical lesions is spoken of as *monoplegia.* When paralysis is limited to the lower limbs it is known as *paraplegia;* when it affects both sides of the body including all four limbs, the condition if dependent on brain disease, is called *diplegia, double or bilateral hemiplegia;* if the condition is due to spinal disease it is sometimes called *paraplegia cervicalis. Crossed or alternate hemiplegia* means paralysis of the limbs on one side, with paralysis of the facial, third or other cranial nerve on the opposite side of the body.

The degree of paralysis gives us information regarding the intensity of the lesion, while its distribution indicates to some extent the position of the lesion, which however cannot be accurately diagnosed until the nutrition and tone of the paralysed muscles have been determined.

When motor weakness is marked there can be no difficulty in detecting its presence, in defining its limits or in estimating its degree. It is quite otherwise however when paralysis is limited in area or is slight in degree. In such a case the observer requires to exercise the greatest care, and in order to avoid the risk of overlooking minor degrees of paralysis he ought to adopt as far as possible a definite plan of investigation. The general movements of the body should be observed before the finer or individual ones are systematically examined. Thus the ability of the patient to stand, walk, sit up or turn in bed, or to move his limbs, together with any disorders of these movements must be first investigated. Should any part of the body exhibit weakness, this ought to be next examined and subsequently the movements of other parts should receive careful attention. Each movement flexion, extension, abduction, adduction, and whenever possible each

muscle, should be separately tested and compared with those of the other side of the body.

In order to form a correct judgment on these matters it is necessary to have a thorough acquaintance with the action of the various muscles, and also with the best methods for putting each of them to the test. In the present work it must suffice briefly to allude to the examination of certain movements which most commonly need to be investigated.

Thus the strength of the hand muscles is ascertained by getting the patient to abduct and adduct the fingers, to touch the tip of his little finger with the end of his thumb, and to grasp one's hand or squeeze a dynamometer, an instrument by means of which a record may be kept of variations in the grasp at different times.

The strength of many muscular movements is best estimated by offering resistance to them. Thus the strength of the deltoid is ascertained by offering passive resistance to abduction of the arm; that of the pectoralis major by getting the patient to try and touch the opposite shoulder, while the observer grasps his wrist and tries to pull it away from the shoulder; that of the latissimus dorsi by offering resistance to what may be called the "coat-tail-pocket movement," *i.e.*, the patient puts his hand behind him as if to reach a pocket in a coat-tail, whilst the observer grasping the patient's wrist tries to drag the limb away from the spine. Similarly the power of flexion and extension at the wrist, elbow, ankle, knee and hip, of adducting and abducting the limbs, of the movements of the head and trunk as well as the special movements of the eyes, tongue, palate and larynx should be systematically investigated. (See section on cranial nerves.)

Attention may here be drawn to the importance of carefully testing the strength of the movements at the hip and the ankle in cases where there is reason to suspect the onset of disease in the central nervous system. Such suspicion is aroused by the common complaint of a tired feeling in walking or of weakne

in one or both legs in getting up stairs. On examination the gait may be quite normal and the patient when seated may perform all movements of the legs with apparent vigour. But if resistance is offered to each individual movement weakness is sometimes detected, and it may be limited to the hip or the ankle. Slight weakness in flexion at the hip is constantly overlooked, yet it sometimes occurs as an isolated symptom and its recognition may lead to a diagnosis of early myelitis or of disseminated sclerosis. Hence it seems desirable to give the following methods of examination:—

Strength of hip flexors. The patient, seated in a chair, is told (1) To raise the knee as high as possible, then the observer, placing his hand just above the knee, tries to push it down, the patient being told to resist the movement as much as possible (see fig. 31); (2) To try

Fig. 31.

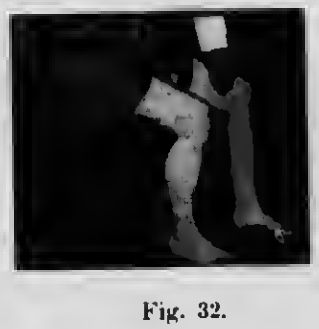


Fig. 32.

and raise his knee while the observer's hand presses down upon it (see fig. 32). These tests should be repeated two or three times, one limb being carefully compared with the other.

Strength of ankle dorsi-flexors. Here inspection is of the first importance. The patient, with the feet well advanced, and symmetrically placed, is told to raise the fore part of the foot as high as possible, whilst keeping the heel on the ground. In this way the slightest difference in the degree of dorsi-flexion of the ankles can be readily seen, as, for example, a slight dropping of

the outer part of one foot from weakness of the peronei —a very common defect. Should there be no obvious difference between the heights of the two feet, passive and active resistance by the hand on the dorsum of the feet should be carefully tried in order to detect any difference in strength between the dorsi-flexor muscles of the ankles (see fig. 33).

Fig. 33.

During the above examination the nutrition and tone of the paralysed muscles will not have escaped notice. Any marked change in the muscular tonus of a limb has a considerable influence on its movements: it may be detected by passive movements and by testing the knee-jerk and the other tendon phenomena. The limb to be examined should be moved at its various joints in all directions. Normally this can be done without difficulty, providing the patient offers no resistance. When however the tonicity of the muscles is increased the observer feels resistance which varies in degree from the slightest possible stiffness to extreme rigidity, which he may be unable to overcome. But when the tonicity is reduced the affected limb can be moved with abnormal ease and may be put into positions which are quite impossible in the case of a healthy person. This *hypotonus* as it is called is often a striking feature in locomotor ataxy.

The tendon reactions, of which the knee-jerk is the most important, vary in degree according to the condition of muscular tonus. For example a slight exaggeration of the knee-jerk is often a valuable indication of increased tonus in the quadriceps or vastus

internus, even when no stiffness of the leg can be detected by passive movements; such exaggeration would suffice—if associated with muscular weakness—to place the case in the group of spastic paralysis and to suggest a lesion of the upper motor neurons.

In the regional diagnosis of paralysis the tone and the state of nutrition of the muscles is of the greatest importance and should receive the closest attention, for they afford reliable indications as to the position of the lesion. Thus when the lesion is in any portion of the upper segment of the motor path the tonus of the paralysed muscle is increased, a marked increase being expressed by muscular spasm or rigidity of the affected limb, a slight increase merely by exaggeration of the tendon reflexes or by an alteration of the plantar reflex known as the extensor response, or Babinski's sign. On the other hand when a lesion affects the lower neurons the paralysed muscles are flaccid and tend to undergo a progressive atrophy, the degree of wasting varying with the intensity and duration of the lesion. The condition of paralysed muscles as regards nutrition and tone constitutes the best basis for a clinical classification of cases of paralysis, enabling us to separate most of them into the great groups of atrophic and spastic paralyses and thus to determine at once which neurons—the lower or the upper—are implicated.

Some apparent exceptions to this rule will be mentioned later (see p. 130).

In addition to the two simple groups of paralysis, a third group is constituted by cases in which an atrophic and a spastic paralysis are found combined in the same patient. This may be termed the group of *mixed paralyses*: for example, amyotrophic lateral sclerosis.

ELECTRICAL EXAMINATION.

For the electric testing of muscles and motor-nerves it is necessary to have a Faradic coil and a galvanic battery, the latter being provided with a current reverser

or commutator and a galvanometer graduated in milliampères.

The patient should be placed in a good light and the electrodes as well as the skin of the part under investigation should be thoroughly wetted with a solution of salt in hot water. One electrode of large size is to be placed on some indifferent spot, as the back of the neck or the sternum; while the other of smaller size is successively applied over the trunk of a nerve and over the individual muscles, especially at the "motor points" that is where the nerve branches enter the muscle.

It is best to begin with the Faradic current and then take the reaction with each pole of the galvanic battery. In either case the current should at first be feeble and then it should be gradually increased till a slight contraction of the muscle is obtained; the strength of the current, as indicated by the galvanometer, may be recorded.

Muscular contraction is caused by stimulating the nerve trunk or its nerve endings in the muscle, and with the galvanic current by stimulating the muscle-substance itself. Thus in certain stages of destruction of the lower neurons, contraction can only be produced by applying the galvanic current directly to muscular tissue—no reaction occurring on the applications of either current to the nerves. If a healthy muscle is stimulated by galvanism it contracts more readily on making or closing, than on breaking or opening the current, and a weaker current produces a closure contraction when the kathodal or negative pole is placed on the muscle than when the anodal or positive pole is applied to it. In normal conditions the order of contractions produced by a gradual increase in the strength of the current is as follows: KCC (kathodal closure contraction); ACC (anodal closure contraction); AOC (anodal opening contraction); KOC (kathodal opening contraction). As a rule the strength of current necessary to produce opening contractions is very painful and for practical purposes it is sufficient to observe the order of

closure contractions only; this in health being represented by the formula KCC>ACC.

ALTERED ELECTRICAL REACTIONS IN DISEASE. Changes may occur either as regards quantity or quality, the latter being of greater importance than the former.

Quantitative changes. Increased excitability of nerve and muscle is observed in many cases of tetany and occasionally in hysteria, hemiplegia and tabes. A diminution of electric excitability has been met with in arthritic muscular atrophy, in the myopathies and in the atrophy of disuse. In myasthenia gravis a temporary loss of excitability occurs after the application for a time of the Faradic current—this is called the myasthenic reaction.

Quantitative and qualitative changes. The reaction of degeneration or R.D.

If a muscle is cut off from its trophic centre in the medulla or cord, or if the centre itself is destroyed, the nerve degenerates and the degenerative atrophy spreads to the muscles supplied by the nerve. For one or two days the nerve excitability may be increased; it then begins to diminish equally to both currents, and by the end of the second week reactions to galvanism and Faradism are usually completely lost. The muscles behave in a similar way to the Faradic current, but to the galvanic current there is at first a diminished excitability and then during the second week a marked increase which continues during the third or even fourth week when it gradually diminishes. Instead however of the brisk contraction seen in a healthy muscle the movement is slow and prolonged, and if the current is continued the contraction is apt to persist and become tetanic. Further qualitative changes are noticed, ACC being equal to or greater than KCC, whilst KtOt' may be more readily obtained than AtOC.

In what is known as partial R.D. the nerve excitability to both currents is preserved and sometimes the muscles act normally to Faradism; but they show

decided changes both as regards quantity and quality to galvanism.

Many varieties of altered electrical reactions occur, and when the departure from normal is not great it may be difficult to say whether or not degenerative atrophy has commenced; its presence may be safely inferred if muscular contraction is distinctly more sluggish than in health.

The presence of R.D. is a sure indication of a lesion in some part of the lower motor neurons: thus it occurs in peripheral neuritis, in anterior poliomyelitis and in bulbar paralysis. It gives us however no direct information regarding the nature of the lesion, nor can any definite relation be traced between the variety of R.D. and the degree of paralysis.

PERVERTED MUSCULAR ACTION—DISORDERS OF EQUILIBRIUM.

For a voluntary movement to be successfully performed it is necessary that each muscle brought into play should contract in such a way and to such a degree that its strength is accurately proportioned to that of its fellows. An exact balance must be maintained between the contractions not only of the prime movers but also of their antagonists. If the adaptation is imperfect, in other words, if there are errors in the correlated contractions of the various muscles employed, the resulting movement is clumsy, jerky and disorderly. Such a condition is called *incoordination* or *ataxy*, and may occur without any detectable alteration in the strength of the individual muscles concerned. Nevertheless it is not always easy to assign definite limits to the use of the term ataxy, for an accurate adjustment of muscular action is necessarily impaired by weakness or spasm of any muscle required for a particular movement and there is sometimes a difficulty in deciding whether a defective movement is due to an alteration in the strength of certain muscles, or solely to a want of coordination in their respective actions. For example,

in some cases of "writer's cramp" when there is no obvious weakness or spasm of any of the muscles used in writing, it may be difficult to give a satisfactory explanation of the muscular irregularities displayed in the attempt to write.

Ataxy is most conspicuous in tabes dorsalis. In a well marked case ineffectual and disorderly movements of the legs are observed when the patient is walking, or when he tries to touch the knee of one limb with the toe of the other foot, or to describe an imaginary circle with the foot. The unsteadiness is increased by closure of the eyes; thus a tabetic patient standing with his feet close together, and his eyes shut will totter and sway and even sometimes fall to the ground. This is called Romberg's symptom and is most marked when sensation is lost in the soles of the feet, but it also occurs when the cutaneous sensibility is quite normal; moreover normal muscular power may be associated with extreme ataxy. In slight degrees of incoordination the effort to maintain equilibrium may be only evinced by the irregular tightening of the tendons on the dorsal aspect of the feet.

Reeling movements are observed in cerebellar disease, the patient swaying from side to side or from front to back; frequently the gait resembles that of a drunken man. The incoordination differs from that in tabes for it is not increased by closure of the eyes, and may not be detectable in the movements of the legs when the patient is lying down.

Vertigo is a disturbance of the sense of equilibrium; the patient feels as if going to stagger or that surrounding objects are oscillating or moving in a particular direction, when no such movement exists. In uncomplicated vertigo consciousness is not lost but it may be obscured, hence the terms giddiness and dizziness are sometimes applied to the condition. Severe vertigo is sometimes accompanied by actual reeling or falling to the ground. Although frequently met with in diseases of the brain, vertigo results from many other causes,

consequently its significance has to be determined mainly by a consideration of its associations. Thus it may result from disease of any part of the auditory apparatus; from errors of refraction or weakness of one of the ocular muscles, and it is common in many forms of dyspepsia. In these cases it is probable that the symptom depends on defective conduction of centripetal impressions to the cerebellum, especially of those from the semicircular canals. As regards intracranial lesions vertigo is especially related to disease of the cerebellum and its peduncles, but it is met with when other parts of the brain are affected.

SENSORY SYMPTOMS.

These may be either subjective or objective. Subjective sensations comprise the presence of pain or of dysaesthesia in the absence of an external stimulus.

Pain. The significance of pain varies greatly, partly according to its locality, but chiefly in consequence of the varying susceptibilities of the nervous system in different individuals. It is necessary to ascertain its exact position and distribution, its characters, its relation to time and movement, and above all to consider its associations. Occasionally violent pain leads reflexly to certain objective phenomena, as muscular twitchings, pallor or redness of the skin, or altered frequency of the pulse; even delirium may ensue during a severe attack of pain, and unconsciousness may follow it.

The situation of pain in relation to visceral disease is of much interest and importance. The sympathetic fibres supplying the viscera are derived from certain spinal segments and the pain of visceral disease is referred from these segments along sensory nerve fibres to definite areas of skin. It has indeed been demonstrated by Mackenzie and Head that certain definite and constant areas of cutaneous tenderness, each area having a maximal region in which there is pain, may be found in association with various visceral diseases.

Dysæsthesia or Parasthesia. These terms include

a number of abnormal subjective sensations, such as numbness and tingling, "pins and needles," crawling, pricking, smarting, or painful feelings of heat or cold. These sensations often constitute the initial symptoms of affections of the nerves, cord or brain and may exist for some time before objective changes in the cutaneous sensibility can be detected.

Various cephalic sensations may here be mentioned, such as a feeling of pressure on the head or of heat or fulness in the head. These symptoms, which may be associated with a little mental confusion, or with impaired memory, are common in cases of neurasthenia and will be further considered under that heading.

Objective Sensory Symptoms. These comprise (1) a diminution or loss of the normal sensibility of a part, *anæsthesia*. (2) An excess of the normal sensibility of a part *hyperæsthesia;* when diminution or excess of sensibility is limited to the application of painful stimuli the terms *analgesia* and *hyperalgesia* are sometimes employed. (3) Acceleration and retardation of sensory perception. (4) Perversion of the cutaneous sensibility—that is the production by an external stimulus of a feeling different to that experienced in health: the variation may apply to the character of the sensation, as when a prick is felt as a burning, or to its position as when a patient refers an impression on one side to a corresponding place on the opposite side of his body—*allocheiria;* or when he feels a single touch as two or three points, *polyæsthesia*. (5) Touch paralysis, or *astereognosis*, that is an inability to recognise the nature of objects by tactile impressions even when the cutaneous sensibility appears to be normal. (6) Modifications of the perception of position owing to changes in the joint sense and in the feeling of active muscular contraction.

In applying the tests necessary for the investigation of the various forms of cutaneous, muscular and arthritic sensibility, the following rules should be observed. The patient's eyes must be covered and he should be told to

say "yes" immediately he feels a touch or other stimulus and to indicate the part touched by placing his finger upon it. He should also state the nature of the stimulus whether a prick, a touch, a hot or a cold object: or he may be required to describe the position of a limb, when his perception of this sense is being investigated. In testing the cutaneous sensibility corresponding points on the two sides of the body must be compared, otherwise slight anaesthesia on one side might escape notice. Such examination is often attended with difficulty, for the observer has to rely on the intelligence, honesty and goodwill of the patient, and must be constantly on his guard against erroneous statements whether intentional or unintentional.

Tactile sensibility is best tested by lightly stroking the skin with a camel hair brush, or a tuft of cotton-wool; the finger should not be used owing to the difficulty in avoiding pressure, the sensation of which is not conveyed by the nerves to the skin but by those to the muscles. Sensation to pain should be tested by pricking the skin with a pin. Sensation to temperature is conveniently examined by applying to the part test-tubes containing water at different temperatures. Tactile discrimination is investigated by means of a pair of compasses with blunt points, the object being to determine the minimal distance at which the two points are recognised as two. This distance varies in health in different parts of the body, being for example 2·3 mm. at the tip of the fingers, and as much as 70 mm. on the arms and thighs.

The rapidity of sensory conduction is determined by noting the interval between a pin prick and the signal given by the patient directly he feels it. A long interval, even several seconds, is often observed in cases of locomotor ataxy.

The sensibility of muscles is tested by squeezing or by pressing on them. It may be increased as in peripheral neuritis or diminished as in tabes.

The sense of active muscular contraction may be

investigated by suspending a series of weights to the part to be examined. In health an addition of one-third to the weight originally applied is usually detected. On this sense along with the joint sense we depend for recognition of posture. The condition of these senses is determined by asking the patient, whose eyes are covered, to move a limb into a certain prescribed position to touch the tip of the nose with his forefinger, to describe an imaginary circle with his foot; or a limb grasped by the observer is moved into various positions the patient being asked to state the position after the limb has been brought to rest, or to imitate it with the limb of the opposite side.

The *vibration sense* is tested by placing a vibrating tuning fork upon the tibia or other accessible bony surface. In health a distinct thrill is felt, but in tabes and some other diseases, this osseous sensation may be lost.

Anæsthesia presents variations both in character and degree as well as in distribution. Thus it may be limited to touch, to pain, to temperature, or to the impulses produced by movement, or it may involve every possible external stimulus in equal or unequal degree. According to Head a qualitative difference is to be noticed between the anæsthesia due to peripheral and to central lesions. In the latter case, as when the sensory tracts in the spinal cord are damaged, we have to deal with diminished sensibility to touch, to pain, to all degrees of temperature, and to the perception of the position of a limb. But in peripheral lesions, as when a sensory nerve is diseased, we have to deal with epicritic and protopathic sensibility; we must not speak of loss of sensibility to temperature but of loss to certain degrees of temperature, for in protopathic anæsthesia loss of sensibility to the extremes of heat and cold is associated with loss of sensibility to painful stimuli, whereas in epicritic anæsthesia there is a want of perception of the finer grades of temperature in association with loss of sensibility to the lightest touch and of

the power to discern the doubleness of two points as tested with the compasses. During the recovery of a diseased nerve the protopathic sensibility is the first to return, hence at a certain stage in the process of repair this variety of sensibility may be found to be normal when the epicritic is completely lost. Sometimes an inaccurate localisation of cutaneous impressions is observed. This error, called *topoanæsthesia*, may occur in hysteria in the form of allocheiria. Another variety has been noticed by Horsley and Russell in cases of cortical disease: one error is in a proximal direction that is the patient refers the stimulus to a point higher up in the limb; another error is lateral or axial, that is the patient refers a touch say on a finger to the radial or ulnar side of it, in other words towards the preaxial or the postaxial side of the hand.

The distribution of anæsthesia is of great importance in diagnosis and should be carefully mapped out. When it affects the lateral half of the body including the limbs and half the face the condition is called *hemianæsthesia*, and the lesion is situated in the opposite side of the brain above the level of the pons, involving the cortex, the internal capsule or possibly the crus. Hemianæsthesia is a common symptom in hysteria, the sensory portion of the cortex being the seat of some change the nature of which is unknown. Sometimes the face is affected on one side and the limbs on the other side of the body; this is called *crossed hemianæsthesia* and is due to a unilateral lesion in the dorsal aspect of the pons.

When anæsthesia affects the legs and the lower part of the body it is sometimes called *paranæsthesia* and the level of the body to which the anæsthesia extends is a guide to the upper limit of the lesion.

In other cases impaired sensation is limited to the area of distribution of particular nerves or it is distributed in bands or patches corresponding to certain spinal roots. In peripheral and spinal diseases the boundary line of anæsthesia is often parallel to the long

axis of the limb, whereas in cerebral disease it tends to be at right angles to it. In contrast to this "segmental" variety is the anæsthesia of multiple neuritis, or that resulting from cortical disease, which fades in intensity as we pass from the hand towards the shoulder or from the foot towards the hip.

When anæsthesia is profound and its area abruptly limited, showing no correspondence to the anatomical distribution of cutaneous nerves or their roots its hysterical nature should be suspected.

Hyperæsthesia also varies much in degree and in distribution: it may be slight or excessive and may affect the skin, the organs of special sense, or the muscles and other deep structures. In slight cases pricking or pinching the skin or muscles causes more pain than in health, in severe cases slightly squeezing a muscle or merely stroking the skin is painful. Such intense hyperæsthesia is prominent in the multiple neuritis set up by arsenic or by alcohol. A zone of over-sensitive skin is commonly found at the upper level of spinal lesions, whilst bands of hyperæsthesia along the limbs indicate irritation of the posterior roots, as from caries or tumour of the spine. Hyperæsthetic spots not corresponding to the distribution of any particular nerves or roots occur in hysteria and sometimes pressure on such tender areas will excite an hysterical outbreak. A neuralgic area is often the seat of considerable tenderness, the hyperæsthesia being commonly most intense at certain spots, called "points douloureux," which correspond to the foramina of exit of branches of the affected nerve.

THE REFLEXES.

A reflex action is effected by means of an afferent sensory nerve, a cellular centre and an efferent motor nerve. Disorders of reflex action occur when any one of these parts is irritated or destroyed, or when the centre is cut off from the controlling influence of higher centres. In health, reflex movements are caused by stimu-

lation of the skin or of accessible mucous membranes; or by excitation of tendons, fasciæ or periosteum. The former are called superficial, the latter deep reflexes.

The superficial include the cranial and the cutaneous reflexes. The chief CRANIAL REFLEXES are: The contraction of the palate caused by irritation of the fauces; spasm of the facial muscles caused by irritation of the fifth nerve; sneezing and lachrymation caused by irritation of the nasal mucous membrane; coughing caused by irritation of the laryngeal mucous membrane; closing of the eyelids caused by irritation of the conjunctiva; contraction of the pupil to light, and its dilatation by irritation of the skin of the neck.

The CUTANEOUS REFLEXES consist of quick muscular contractions which are best excited by a gentle stimulation of the skin as by stroking it with a feather or the finger. The following are the most important, and may be usually obtained in healthy persons, especially in children.

The plantar reflex. This when present consists of movements of the toes and depends for its integrity on the reflex loops through the lower end of the cord at the level of the second sacral nerve. If stimulation of the sole is severe or if the reflex excitability is exaggerated, movements of the foot and leg may ensue, indicating extension of the wave of irritation to the upper end of the lumbar enlargement. Two varieties of this reflex are now distinguished, namely the flexor and the extensor responses. The former occurs in health, the latter in certain diseases.

The flexor response, in its simplest form consists of flexion of the four outer toes and sometimes of the great toe, preceded it may be by contraction of the tensor vaginæ femoris and other hip muscles. *The extensor response* or Babinski's phenomenon, which in health is only obtained in the infant, consists of extension of the great toe followed sometimes by extension of the other toes and eversion of the foot. Its presence signifies some interference with the pyramidal system. It is

best obtained by drawing the thumb nail or the end of a stethoscope slowly and firmly along the outer side of the sole.

The gluteal reflex consists of contraction of the gluteal muscles, obtained by stroking the skin over the buttock and depends upon the integrity of the loops through the fourth and fifth lumbar nerves. The *cremasteric reflex* by which the testicle is drawn up when the skin at the upper and inner part of the thigh is stimulated, demands the integrity of the first and second lumbar nerves.

The abdominal reflex consists of contraction of the abdominal muscles, and is produced by stroking the skin from the costal arch downwards. The umbilicus is drawn towards the stimulated side: this indicates integrity of the arc at the level of the ninth or tenth dorsal nerve. *The epigastric reflex* consists of a dimpling of the epigastrium on the side stimulated, and is induced by stroking the skin of the chest downwards from the nipple. Its presence requires the integrity of the reflex arcs from the fourth to the seventh dorsal nerves.

The bulbo-cavernous reflex is of value as an indication of the condition of the reflex arc about the level of the third sacral segment. It is obtained by placing a finger behind the patient's scrotum and pressing upwards, whilst the glans penis is pricked with a pin, when if the reflex arc is intact a brisk twitch will be felt in the bulbous part of the urethra.

In forming an opinion regarding the clinical value of the cutaneous reflexes the following points should be borne in mind: (1) That they are usually more marked in children than in adults, and in women than in men and that their strength varies much in different normal individuals; (2) that the cremasteric reflex is often absent in elderly men; (3) that repeated observations are often necessary before the presence or absence of the abdominal reflexes can be determined, which indeed may be unobtainable, even when there is no reason to suspect disease. The presence of a cutaneous reflex

indicates that the reflex arc upon which it depends is intact. Absence of a cutaneous reflex or its diminution, which is shown by slowness as well as by weakness of movement, points to a defect in some part of the reflex

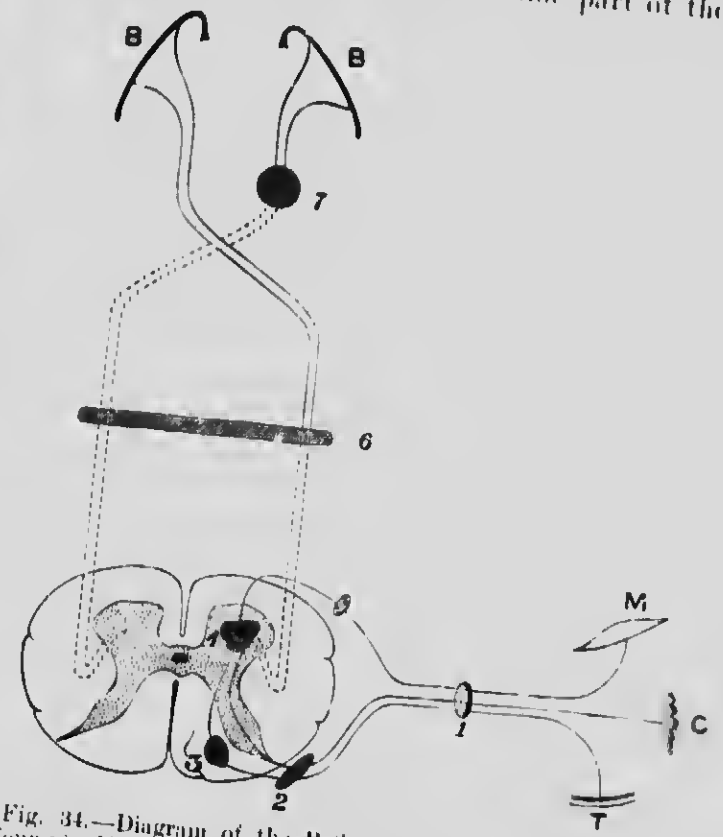


Fig. 34.—Diagram of the Reflex Functions of the Spinal Cord (Young.) M, muscle; C, skin; and T, tendon. B B, the cortices of the cerebral hemispheres, with their attached pyramidal tract fibres connecting the cortical giant cells with the ganglion cells of the anterior gray horns. 1, lesion of the peripheral nerve, causing atrophic paralysis, loss of sensation, and abolition of both kinds of reflexes; 2, lesion of the posterior root (pachymeningitis), causing loss of sensation and abolition of both kinds of reflexes, but no paralysis; 3, lesion of the posterior root-zone (tabes dorsalis), causing loss of the tendon reflex; 4, lesion of the anterior gray horns (poliomyelitis), causing atrophic paralysis and loss of both kinds of reflexes, but no sensory disorder; 5, lesion of the anterior root (pachymeningitis), causing the same symptoms as 4; 6, transverse lesion of the spinal cord, causing a spasmodic paralysis of the lower extremities, with excess of the tendon reflexes; 7, focal lesion of the cerebral hemisphere, causing hemiplegia of the opposite side, with excess of the tendon reflexes on the paralysed side, but with diminution of the cutaneous reflexes. The dotted lines indicate descending sclerosis of the pyramidal tracts.

are. Thus in disseminated sclerosis loss of the abdominal reflexes is an early and a valuable indication that the grey matter of the cord is involved, or that there is implication of the pyramidal fibres above the corresponding segmental levels.

Exaggeration of a cutaneous reflex indicates that some portion of the arc is irritated or that it is partially separated from the control of a higher centre. Thus the abdominal reflexes are increased when the dorsal roots are unduly stimulated as in cases of pachymeningitis and of early tabes, and commonly too when the cord above their level is diseased. But a complete lesion of the cord will cause abolition instead of exaggeration of the reflexes at a lower level.

The KNEE-JERK is the forward jerk of the foot and leg which in a healthy person is produced by smartly striking the ligamentum patellæ with the tips of the fingers, or with the edge of the ear-piece of a stethoscope or with a percussion-hammer. The jerk is caused by sudden contraction of the quadriceps or possibly the vastus internus and to obtain it, it is essential that these muscles should be made tense. This is usually accomplished by crossing the leg to be tested over the other or by getting the patient to sit on a chair or a table. In stout people who cannot easily cross one leg over the other the operator should support the limb to be examined by passing his hand beneath the thigh and grasping the opposite knee. Sometimes the knee-jerk cannot be obtained because the patient is unable properly to relax the flexors of the knee. This tension may often be overcome by getting the patient to hold up his head, and to interlock the bent fingers of each hand and to pull strongly at the time the ligamentum patellæ is being struck. It is also useful to push down the patella with one hand whilst the ligament is struck with the other. Reinforcement of a feeble knee-jerk is also obtained by getting the patient to grasp and squeeze the observer's biceps whilst the jerk is being elicited.

Ankle-Jerk and Ankle-Clonus. In health tapping the tendo Achilles, the knee and ankle being bent, may cause contraction of the calf muscles with sudden extension of the ankle. In tabes this jerk is sometimes lost before the knee-jerk. When the pyramidal tract is diseased the above procedure may produce to and fro movements at the ankle joint in other words clonus. But this is more readily excited by lightly grasping the fore part of the foot and suddenly pressing it upwards towards the tibia; there on relaxing the pressure, rapidly alternating flexion and extension at the ankle take place which continue so long as the tension of the tendo Achilles is maintained. The movements are uniform and occur from six to nine times a second. A spurious form of ankle clonus, consisting of a few imperfect movements, feebly sustained and irregular in rhythm, is sometimes obtained in cases of hysteria and neurasthenia. It is quite different to the clonus met with in spastic paralysis, and it is not associated with an extensor plantar reflex. Paroxysms of violent tremor in the so-called "*spinal epilepsy*," sometimes affect the legs in cases of spastic paralysis; such clonic spasm may be arrested by grasping the toes and bringing them suddenly and forcibly into plantar flexion.

Wrist and Elbow Jerks. In many persons a smart tap over the lower end of the radius produces flexion of the elbow. The movement is usually due to contraction of the supinator longus, but if the reflex is exaggerated the biceps is also brought into action. Similarly tapping over the triceps tendon or sometimes over the lower end of the ulna will cause extension at the elbow.

These reflexes have a like pathology to that of the knee-jerk; thus they are increased in cases of spastic paralysis, and diminished or abolished in cases of atrophic paralysis. Their intensity however varies in health more than the intensity of the knee-jerk, hence their diagnostic value is more difficult to estimate. A unilateral excess or diminution of the wrist and elbow jerk would however be a sign of great importance.

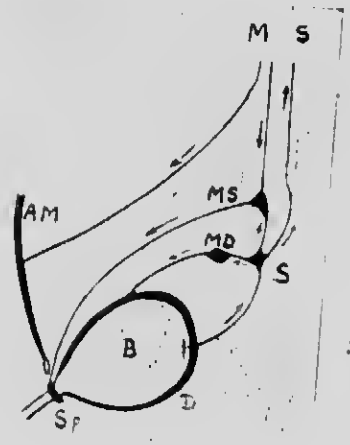
The Jaw Jerk, usually absent in health is elicited by percussing the chin whilst the patient's mouth is half opened or by tapping a spatula placed between the teeth. It is often easily obtained in amyotrophic lateral selerosis.

Kernig's Sign. In healthy persons if the thigh be flexed at a right angle with the trunk, the leg can be almost fully extended at the knee. But in cerebral meningitis it is impossible to extend the leg on the thigh owing to the marked contraction of the hamstring muscles.

The Organic Reflexes. The walls of the bladder and rectum contain muscular fibres to expel their contents, while at the orifice of each viscus there is a sphincter which being maintained in a state of tonic contraction prevents the continuous escape of their contents. It is stated that the lowest reflex centres for these viscera are situated in the hypogastric and hæmorrhoidal plexuses of the sympathetic. These centres are connected with the cord by means of the lowest spinal roots and are governed and brought under voluntary control by means of fibres passing between the cord and centres in the cerebral cortex.

When the lower end of the cord is damaged as by a lumbar myelitis the sphincters become permanently relaxed—the urine dribbles away as fast as it enters the bladder, and fæces escape as soon as they enter the rectum. When the cerebral influence is cut off as by a dorsal myelitis there is a tendency to *intermittent incontinence* of urine so that when the bladder is full the sphincter is reflexly relaxed and urine is involuntarily discharged. Similarly the fæces may be passed automatically or there may be constipation.

The loss of voluntary power leads to weakness of the detrusor vesicæ so that the bladder is never completely emptied and ultimately the detrusor becomes completely paralysed, the bladder is then over distended and urine dribbles away; this is *overflow incontinence*. Sometimes there is complete retention; this occurs



AM = Muscles of abdominal wall.
B = Bladder.
D = Muscular wall of bladder (detrusor).
Sp = Sphincter of bladder.
M = Motor impulse from brain to the abdominal muscles and the sphincter.
S = Sensory centre and sensory impulses to the brain.
MS = Motor centre for the sphincter at lower end of spinal cord.
MD = Motor centre for detrusor.
The centres MD and S are in the pelvic sympathetic ganglia according to Müller.

Fig. 35.—Centres for Bladder and Reflex Arcs. (Williamson.)

when the detrusor is paralysed but not the sphincter. Any retention of urine is a serious condition for residual urine is apt to decompose and set up cystitis which may lead to serious kidney disease.

The condition of the sphincter ani can be ascertained by introducing the finger into the rectum. When the anal centre or the lower end of the cord is destroyed the finger feels a complete and persistent relaxation of the sphincter. But if there is disease at a higher level cutting off voluntary control, the introduction of the finger leads to firm tonic contraction of the sphincter.

VASO-MOTOR AND TROPHIC CHANGES.

Many diseases of the nervous system are associated with obvious changes in the superficial tissues, and in the muscles, bones and joints. Of these the most frequent and striking is *atrophy of muscular tissue* which ensues in lesions of the lower neurons. The more acute and irritating the disease the more rapid is the degeneration of the nerve fibres and the greater the muscular atrophy. Moreover the rate and degree of the nutritive changes vary in different muscles and in different fibres of the same muscle. Many variations are met both as regards the intensity and the distribution of the muscular

atrophy, a description of the more important ones will be given later (see Section V). How are these changes brought about? A paralysed limb is of course unable to move, and when a limb is kept at rest its muscles waste,—witness the condition of the muscles of a fractured limb. But such wasting is slight in degree and general in distribution: whereas the atrophy due to damage to the lower neuron is severe and is limited to groups of muscles; moreover the R.D. is usually obtained on electrical examination. Hence it is impossible to avoid the conclusion that the atrophy depends on a loss of healthy nerve impulses to the affected part. The next question is, are there special nerve fibres which preside over muscular nutrition, in other words are there trophic nerves? These have not yet been discovered, and there appears to be no evidence in favour of their existence.

Changes in the skin and its appendages. Here also the changes vary in degree and in rapidity of development according to the intensity of the disease. Thus in acute cases the temperature of the affected part is raised, its vessels are dilated and there is a tendency to the formation of vesicles and bullæ; the slightest irritation or pressure may lead to vesication or even sloughing of the skin. These changes, indicating irritation of nerve tissue, occur in cases of acute disseminated myelitis and sometimes in cases of hæmorrhage into the cord or the brain.

Rapid sloughing of the skin called acute decubitus or bedsore is difficult to prevent by the most careful attention on the part of the nurse. Its common site is over the great trochanter and gluteal region in hemiplegia, and over the sacrum in cases of paraplegia. Occasionally the sloughing process extends down to the bone and if this occurs, when the bedsore is in the sacral region, an infective meningitis may be set up.

Herpes zoster, which is due to inflammation of the posterior root ganglia, illustrates the failure in nutrition which may ensue when the control of the nervous system

is removed. Similarly facial herpes is caused by changes in the Gasserian ganglion.

Slight degrees of nerve disturbance are represented by transient erythema or by urticaria, or by small white atrophic patches of skin in the course of affected nerves. More chronic changes result from slow degenerative changes in nerve tissue. The affected part tends to become cold, pale and livid, and in long standing cases of disease of the peripheral nerves or of the posterior roots the condition known as "*glossy skin*" is often present. It occurs also in old cases of hemiplegia. The skin of the hands, which are most commonly affected, becomes red, thin, smooth and shiny, while the finger tips get thin and pointed owing to wasting of the subcutaneous tissue.

Of other skin lesions which appear to be related to disturbed nerve function may be mentioned leucoderma, scleroderma and some purpuric eruptions. The hyperkeratosis of the palms and soles seen in arsenical neuritis, the perforating ulcer of tabes, the painless whitlows which develop in some varieties of syringomyelia, the destructive lesions of leprosy and the symmetrical gangrene of Raynaud's disease are also primarily due to imperfections in the nerve impulses going to and from the affected parts. The hair and nails may also suffer; thus the hair may fall off or lose its pigment, whitish hairs being sometimes found in the track of a neuralgia; whilst the nails may be striated, curved and brittle, and sometimes they are shed.

The bones and joints. Spontaneous fractures, owing to rarefaction of the bones, sometimes occur in tabes, syringomyelia and in general paralysis. Injuries of nerve trunks have been followed by swelling and thickening of the bones. The neuritis of leprosy, however, is often attended by atrophy of the bones of the limb and a similar atrophy is common in infantile paralysis. The condition of a limb in which a retrograde change has taken place in parts originally well developed must be distinguished from a limb in which

there has been *arrest of growth*. For example, a destructive lesion of the motor part of the cortex in infancy will hinder the growth of the limbs on the opposite side of the body, and hence at a later period of life, they will be shorter and thinner than their fellows, the bones as well as the soft tissues being more or less arrested in their development.

Painful swelling of joints sometimes follow lesions of the spinal cord or the peripheral nerves, and may occur in cases of hemiplegia. In tabes and syringomyelia the larger joints are liable to trophic changes, which generally develop and progress without pain or febrile reaction. There is a rapid effusion into the joint which is often considerably enlarged; the swelling may last for a long time and then subside. Sometimes the joint recovers completely, but as a rule the articular ends of the bones become atrophied and then, owing to relaxation of the ligaments and feebleness of the surrounding muscles, spontaneous luxations frequently occur. The atrophy is accompanied by new formation of bone at the ends of the bones and in the ligaments and other structures around the joints.

THE SPECIAL SENSES.

The sense of Smell is tested as follows:—The patient's eyes being shut substances such as camphor, assafœtida, or oil of cloves which affect the olfactary nerve alone should be held to each nostril in turn, the other nostril being closed with the finger; substances like ammonia or acetic acid which irritate branches of the fifth nerve within the nose should be avoided. Anosmia or loss of the sense of smell is most commonly due to nasal catarrh, polypi or other local disease. It occurs also in disease of the fifth nerve owing to trophic changes in the nasal mucous membrane, and also in cases of facial paralysis from loss of power in sniffing. All these conditions must be excluded before attributing anosmia to lesions of the olfactory nerve.

Taste. The tip, edges and back of the tongue as well

as the palate should be separately tested with sugar, quinine, common salt and dilute acetic acid, the patient whose tongue should be protruded being told to make a sign when he perceives any taste. Sweets and bitters are best appreciated at the back of the tongue, saline and other substances at its tip and edges. A weak galvanic current, which produces a sour or metallic taste is useful for mapping out an area of loss of taste. Diminution or loss of the sense of taste called ageusis may be produced by interruption of any part of its path (see fig. 16), and sometimes by a lesion involving the tip of the temporo-sphenoidal lobe.

Paragensis or perversion of taste, hypergeusis or increased sensation of taste and subjective taste sensations are sometimes present in cases of hysteria and of insanity. Occasionally they constitute the auræ of epilepsy.

Hearing. Disorders of hearing are much more frequently due to disease of the *sound conducting* apparatus, viz., the external auditory canal and the middle ear than to disease of the *sound perceiving* apparatus, the labyrinth, auditory nerve, and its cerebral path. A description of the methods of examining the ear will be found in special works; here it must suffice to point out the importance of distinguishing between deafness due to disease of the conducting apparatus and that due to disease of the perceiving apparatus. This is done by an investigation of the a nd bone conduction and of the ratio between the Normally a vibrating uning fork is heard longer when held close to the xternal meatus—air conduction—than when placed on he mastoid or other adjacent bone—bone conduction. When the sound conducting apparatus is damaged, air nduction is impaired but bone conduction remains rmal or becomes increased, whereas when the sound receiving apparatus is damaged both aerial and bone nduction are much less than normal, and as deafness reases conduction through the bone fails before that ough the air. Deafness from disease of the labyrinth

is identical with that from disease of the nerve, and to distinguish between them we have to rely on associated symptoms, such as paralysis of the facial nerve, which occurs in cases of deafness due to disease of the auditory nerve.

Sight is to be investigated in respect to acuity of vision, the colour sense and the extent of the visual field. Acuteness of vision is estimated by the patient's ability to recognise certain standard letters at a given distance. If vision is very defective the patient may be unable to see even the largest type; he may be able only to count the outspread fingers at a short distance, or possibly only to distinguish light from darkness. Colour perception is tested by getting the patient to match a skein of wool of a certain colour with all the skeins of a similar colour which are present in a mixed collection of every colour and shade. It is also useful to ask the patient to name certain colours shown to him but it must be remembered that a patient may not know the names of colours or he may give the correct name although the colour is not perceived. The extent of the field of vision can be roughly measured as follows: The patient should cover one eye and look steadily with the other at the observer's nose, who then holds up a finger in a plane with his face and at some distance from it. The observer now gradually brings his finger nearer to his nose, noting the distance at which the patient begins to see it. This should be done on both sides and from above and below. Any decided contraction of the field may thus be detected, as also may central scotoma for red and green, if small pieces of paper of these colours are used in place of the finger tip. In patients who are semi-conscious, deaf or unable to speak any decided defect in the field may be considered probable if the patient does not look in the direction of the finger when placed in various positions, or if he does not blink when menaced by a blow on one side or the other. In order to obtain an accurate chart of the visual field it is necessary to use the perimeter, an instrument in

which an arm shaped like a quadrant of a circle and graduated in degrees moves round a central pivot on which the patient's eye is fixed. A white or coloured object is then moved along the arm, which is placed at various angles, and the points at which the object ceases to be seen mark the limits of the field.

In a normal eye the field for the object is larger than that for colour; the fields for colour diminish in the following order: white, blue, yellow, red, green, violet. It is usually sufficient to test the visual field for red and green. The following are the defects to be observed:—

1. *Central Amblyopia*, or diminished activity of vision in the central part of the field accompanied with a central scotoma for red and green is generally caused by the abuse of tobacco.

2. *Concentric contraction of the field.* This is a converse condition to the preceding, the peripheral parts of the field being blind while the central portions are normally active. This occurs in atrophy of the optic nerve; it is also a common symptom in hysteria. Occasionally in cortical disease, probably from damage to the angular gyrus, there is dimness of sight affecting the peripheral parts of the field of the eye on the side opposite to that of the lesion.

3. *Hemianopsia* means loss of one half of the visual field not due to intraocular disease. The commonest variety is *homonymous hemianopsia*, the right or left halves of the fields of both eyes being lost. Loss of the left half-fields implies loss of function of the right halves of the retinæ and vice-versa.

As a rule the condition depends on a lesion either of some part of the visual path behind the chiasma or of the visual centre itself, namely the cuneus.

A second variety is *temporal hemianopsia* which is due to blindness of the nasal half of each retina, and is produced by a lesion affecting the middle of the chiasma, as from pressure, or from a localised meningitis.

A third variety is *nasal hemianopsia;* this is a rare condition and is due to injury to the uncrossed optic fibres by a lesion involving each side of the chiasma.

A fourth variety is *vertical hemianopsia*, in which there is blindness of either the upper or the lower halves of the fields.

4. *Quadrantic hemianopsia*, or blindness of corresponding sectors of the visual fields.

5. *Scotomata*, or localised areas of defective vision; a central scotoma means blindness of the macular region.

Disorders of vision are also produced by various changes in the eyeball, as opacity of the lens, or disease of the choroid, retina or optic nerve. Many of these conditions are revealed by means of the ophthalmoscope, an instrument of the greatest importance in the diagnosis of nervous disease. The chief conditions to look for are optic neuritis, optic atrophy, tubercle of the choroid, disseminated choroiditis, albuminuric retinitis, retinal hæmorrhage, and the signs of embolism of the central retinal artery.

SPEECH AND ARTICULATION.

The subject of aphasia is considered in Section XII. A few preliminary remarks, however, may here be made with the object of drawing the student's attention to some points of similarity between the nervous mechanism for language and that for a limb. In both cases this consists of an outgoing or motor, and an ingoing or sensory path, and in both cases the *outgoing path* is composed of upper and lower neurons. The upper neurons for speech are constituted by cortical cells situated in the third frontal convolution, and chiefly in that on the left side and by fibres which proceed from these cells to end in fine branches in the pons, medulla, and cord near the cells of the lower neurons. The latter cells form the nuclei of origin for

nerve fibres which run in the facial, vagus and spinal nerves to the muscles used in articulation, phonation, writing and various gestures. Now these muscles just as those of a limb may be affected by tremor, spasm, paralysis or inco-ordination, each of which will tend to cause some disorder of speech. Moreover the character of a paralytic disorder of speech will vary, as does that of a limb, according to the neurons, whether the upper or lower, which are damaged. Thus when the upper neurons of a limb are involved paralysis tends to affect movements rather than muscles, whereas when the lower neurons are involved paralysis tends to pick out individual muscles. Similarly as regards speech: lesions of the upper neurons are expressed by paralysis of the special movements by which articulation is accomplished; lesions of the lower neurons by paralysis of individual muscles used in articulation. In the former case, as in motor aphasia, if speech is not absolutely lost, any words uttered are correctly pronounced. In the latter case the constituent elements of words are imperfectly pronounced, or entirely escape utterance; such a condition is called dysarthria or anarthria.

The *ingoing apparatus for language* is constituted by the eye and ear and in the blind by the sense of touch, with their sensory paths and centres. This apparatus is made use of when one person tries to understand the language of another by listening to his utterances, by reading his writing or by watching his gestures. Deafmutism is an example of damage to one portion of the sensory path, namely the auditory; the patient cannot speak because he cannot hear; whilst sensory aphasia results from damage to the sensory centres for speech. In one variety called *word blindness* the patient although able to see objects and letters has lost the power of reading words. In another variety called *word deafness* the patient whilst able to hear sounds is unable to understand spoken language.

MENTAL SYMPTOMS AND DISORDERS OF CONSCIOUSNESS.

Mental symptoms mainly depend on disturbance of the cerebral cortex either by lesions directly involving it or by the indirect effect of lesions at a distance from it. They comprise exaltation and perversion of ideas, and failure of the mental powers. A stage of exaltation may be observed in chronic alcoholism and is frequently a marked feature in general paralysis of the insane. Perversion of mental processes is met with in various forms of insanity, in the delirium of fever and in that associated with certain forms of intracranial disease. The chief factors of delirium are illusions, hallucinations and delusions. An illusion is a false perception excited by an actual sensory impression. An hallucination is a false perception which arises without any sensory impression; objects are seen or voices heard in the absence of external realities. A delusion is a false belief or idea, a perversion of judgment.

Mental failure is indicated mainly by defect of memory, also by deficient power of attention and by defects in the moral sense.

Loss of consciousness varying in degree may come on suddenly or gradually. A partial loss in which a patient while apparently oblivious to his surroundings can be roused for a few seconds is called *stupor*. A complete abolition of consciousness is called *coma*: this is usually associated with stertorous breathing and a tendency to death by asphyxia. Coma may result from many causes, the chief of which are:—sudden lesions in any part of the brain especially hæmorrhage; acute or chronic diseases involving a considerable portion of the cerebral cortex, either directly as in meningo-encephalitis, or indirectly as a result of increase of intracranial pressure from tumours, hydrocephalus, or other compressing agency; an abnormal condition of the blood circulating through the brain as in diabetic coma, uræmia, or from the presence in excess of certain poisons as opium or alcohol.

Examination of the Cerebro-Spinal Fluid obtained by Lumbar Puncture.

The examination of a small quantity of cerebro-spinal fluid withdrawn from the sub-arachnoid space has proved of considerable value in the diagnosis of many diseases of the nervous system. The fluid is withdrawn by means of a lumbar puncture. This is effected by the insertion of a large hypodermic needle, about three inches in length, in the interval between the third and fourth or between the fourth and fifth lumbar vertebræ, that is well below the level of the spinal cord. The intervals named are respectively just above and just below a line joining the highest points of the iliac crests: such a line crosses the tip of the fourth lumbar spine.

The patient should lie upon his left side or sit in a chair, with the head and shoulders bent well forwards. After thoroughly sterilising the skin, the needle, previously boiled, should be inserted about half an inch to one side of the middle line and directed slightly upwards and inwards until the point is felt to be free: in children the puncture is made nearer to the middle line. Before making the puncture the skin may be frozen by means of a spray of ethyl chloride; in children a general anæsthetic is often advisable.

The fluid is allowed to escape drop by drop into a sterilised test tube until 3 or 4 c.cm. have been collected. The needle is then withdrawn and a collodion dressing applied to the wound. After the operation the patient should be kept in bed for twenty-four hours: this lessens the risk of the occurrence of any serious symptoms. In some cases the patient suffers from vertical headache, which is often of a peculiarly disagreeable type. Dangerous symptoms have sometimes followed the withdrawal of a large quantity of fluid, especially in cases of intracranial tumour in which, owing to increased pressure, the fluid escapes in a jet instead of drop by drop.

Normal cerebro-spinal fluid is clear like water. It has a specific gravity of 1·006 and a slightly alkaline reaction; it contains chlorides and traces of serum-globulin, cholin, albumose and a substance (probably dextrose) which reduces Fehling's solution. With the exception of a few endothelial cells and an occasional lymphocyte it contains no organised elements.

In disease the following changes are observed: The fluid is *bile-stained* in cases of severe jaundice. It is *blood-stained* in cases of intra-cranial hæmorrhage, and of injuries to the spinal cord; in these conditions the fluid is equally tinged with blood. But when blood has accidently entered the fluid as by puncture of an arachnoid vein, the first few drops are most tinged, and after the use of the centrifugal apparatus blood corpuscles are found at the bottom of the tube, whilst the supernatant fluid is clear. *Turbidity* of the fluid is observed in purulent and in cerebro-spinal meningitis, and to a less degree in tuberculous meningitis, although in this disease the fluid may be quite clear.

The nature of any cellular elements that may be present is determined by centrifugalising a little of the fluid and examining the deposit microscopically.

Lymphocytes, or mono-nucleated cells, are found in considerable numbers in general paralysis, in tabes, in cerebro-spinal syphilis, in the early stages of tuberculous meningitis, in post-basic meningitis and in the chronic stage of cerebro-spinal fever.

Polymorpho-nuclear cells are found in acute forms of meningitis, and in the acute stage of cerebro-spinal fever.

In rare instances tumour cells have been found in the fluid obtained from cases of malignant disease of the spinal cord or its membranes.

Micro-organisms may be found in the puncture fluid taken from cases of meningitis; their detection is of great diagnostic importance. The tubercle bacillus is present in tuberculous meningitis, the diplococcus intracellularis in cerebro-spinal fever, whilst staphylococci,

streptococci and pneumococci are found in other varieties of meningitis.

It will be noticed that whilst the tubercle bacillus and the syphilitic virus are associated with lymphocytosis, the infective organisms as the diplococcus intracellularis, and the pyogenetic organisms produce polymorphonuclear leucocytosis.

SECTION IV.

The Spastic Paralyses. Diseases of the Upper Motor Neurons.

In spastic paralysis weakness is associated with increased muscular tonus and hence the affected limbs present varying degrees of rigidity. The excess of tonus may be so great that the muscles feel as hard as boards even when the limb is at rest, and flexion and extension of its joints may be difficult or impossible; or the excess may be so slight that it is only shown by a very slight increase of resistance to passive movements or possibly merely by an exaggeration of the deep reflexes. Another peculiarity of spastic paralysis is that every part of the affected limb presents some degree of weakness and that although some muscles are weaker than others, it is rare for any of them to be entirely unaffected. In this respect it differs from atrophic paralysis in which usually there is a picking out of particular muscles or of groups of muscles.

A spastic paralysis indicates that the functions of some part of the upper motor neurons are impaired or abolished. The condition of the paralysed limb does not necessarily vary with the site of the lesion, *e.g.*, the spastic condition of the legs produced by a dorsal myelitis may be identical with that seen in children as a result of bilateral disease of the cortex. The position of the lesion is only partially determined by the distribution of the paralysis, and mainly by the presence or absence of other symptoms. According to distribution spastic paralysis may be hemiplegic, monoplegic, or paraplegic in type.

CHAPTER I.

HEMIPLEGIA AND ITS VARIETIES.

In the ordinary type of hemiplegia the face is paralysed on the same side as the limbs. For two or three weeks after the onset of the attack the paralysed limbs are flaccid, but sooner or later they become more or less rigid and the deep reflexes are exaggerated. A well marked case of old hemiplegia is one of the best examples of spastic paralysis for the student to begin with. He should observe the different degrees of paralysis presented by the affected muscles, and should especially consider the method by which an anatomical diagnosis is reached.

Steps of reasoning as to site of lesion. (1) The spastic paralysis is evidence that the lesion is in the upper segment of the motor path. (2) The association of the facial with the limb paralysis indicates that the lesion is in the brain part of the motor path. (3) Anatomical knowledge shows that the lesion must be on the side of the brain opposite to that of the paralysis and above a point at which the decussation of the fibres for the face, arm and leg occurs, that is above the middle of the pons. (4) The upper part of the pons is an unlikely spot because the fifth nerve is unaffected. (5) The crus is also excluded, for as a rule a lesion there would lead to paralysis of the eye muscles supplied by the third nerve. (6) We are now limited to a consideration of the internal capsule and the hemisphere above it. We have seen that the motor fibres for the face, arm and leg diverge as they approach the cortex, hence the nearer the lesion is to the cortex the more likely is it that the paralysis will be limited to one limb or to one side of the face. Anatomy then refers the lesion to the only remaining part of the motor path, namely the anterior two-thirds of the posterior limb of the internal capsule. There the motor fibres are close together, and there in

actual practice the lesion is most commonly found in the ordinary type of hemiplegia.

Muscles affected. The degree of paralysis varies in different places: some muscles seem to escape altogether, others are merely weakened, whilst a third group may be completely paralysed. The law underlying this variation is that the more unilateral or special a movement, the greater is its paralysis, while the more

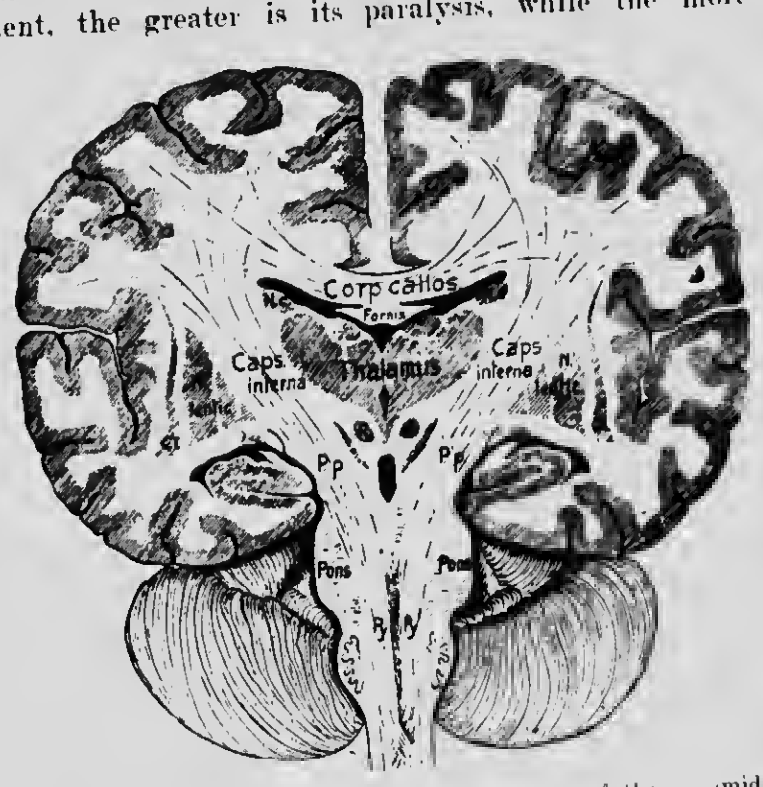


Fig. 36.—Section of brain to show the course of the pyramidal tracts through the internal capsule, the crus, pons and medulla: Cl., claustrum; Pp., crus; R., nucleus ruber. (Villiger.)

bilateral or automatic a movement the less is its paralysis. The explanation appears to be that bilaterally associated muscles can be stimulated by both cerebral hemispheres whereas special movements are represented only in the opposite hemisphere. Thus the arm, which is habitually used without its fellow suffers more than the leg and the special movements of the fingers and

thumb are more paralysed than the movements at the elbow and shoulder.

No weakness can be detected in the muscles of the eye or the larynx, and it is impossible to appreciate any difference in power between the two masseters. The tongue when protruded deviates towards the paralysed side owing to the action of the healthy genio-hyoglossus. With regard to the face its lower muscles are conspicuously paralysed, while the upper ones may entirely escape. As a rule, however, some weakness of the orbicularis palpebrarum can be recognised if, when the patient tries to keep his eyes tightly closed, the observer with his thumb forcibly raises the upper lid. The paralysis of the lower facial muscles is best seen when the patient tries to show his teeth: it may pass unnoticed when he smiles, for in all probability emotional movements are innervated from either hemisphere.

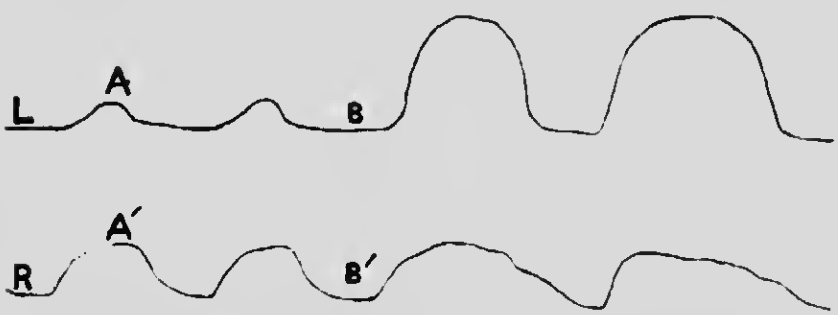


Fig. 37.—Tracings of respiratory movements in a case of right hemiplegia; L, left side; R, right side; A—B, and A′—B′, automatic respiration; the larger curves, forced respiration.

The thoracic movements during a deep or forced inspiration are usually diminished on the hemiplegic side, but in quiet automatic breathing it is curious to observe that (as first pointed out by Hughlings Jackson) the greater expansion may be on the paralysed side (see fig. 37). In estimating this difference it must be remembered that in health the right side moves a little more than the left side of the chest.

Posture of the limbs. In old standing cases of hemi-

plegia, the limbs, owing to the association of rigidity and contracture with the paralysis, assume certain fixed attitudes, which vary in different cases. The usual positions are as follows: In the upper limb there is adduction of the arm and pronation of the forearm with flexion of the elbow, wrist and fingers, especially of the distal phalanges. In the lower limb extension predominates; the thigh is adducted, the foot shows talipes equino-varus. Such 'late rigidity' lessens during sleep and may be partially overcome by placing the limb in warm water. Eventually it passes into structural rigidity which is necessarily persistent. These postures are well seen when the patient is standing, then too it is seen that the shoulder on the paralysed side is lowered and advanced forwards. In walking, the affected leg, which is almost fully extended, is swung outwards and forwards with a semi-circular sweep, the toes scraping along the floor; this movement is necessary for progression, owing to the rigidity of the limb and the paralysis of the dorsi-flexors of the ankle which prevents the lifting up of the anterior part of the foot.

Another peculiarity is observed in lateral walking: when the patient walks sideways the paralysed side is advanced in front of the other, for in this position the pelvis being more readily tilted towards the sound side the affected foot is more easily raised from the ground.

Other Symptoms. Sensory symptoms are absent or inconspicuous. Sometimes the patient suffers from aching pains or from numbness and tingling in the paralysed limbs, but definite anæsthesia is uncommon in the type under consideration: occasionally there is 'touch paralysis' but this is more common when the lesion is near the cortex.

The wrist, elbow and knee-jerks are exaggerated and frequently ankle-clonus can be obtained. In some cases the knee-jerk on the *healthy* side is unduly irritable, and this may be associated with obvious weakness of the leg, and even of the arm. The cremasteric and the superficial abdominal reflexes are enfeebled or lost on

the paralysed side, while the plantar reflex is of the extensor type.

As a rule the nutrition of the muscles does not suffer, but occasionally muscular atrophy occurs especially in the hand muscles, when it is often conspicuous in the abductor indicis. This wasting may be due to changes in the spinal motor cells owing to their not receiving proper impulses from the upper neurons, or to an associated peripheral neuritis.

Coldness, lividity and sometimes œdema affect the extremities of the paralysed limbs; even more severe trophic changes may occur, as blisters filled with dark serum, or sloughs over points of pressure, as over the trochanter, gluteal region, and the malleoli. Inflammation of the larger joints is occasionally met with.

In many cases the mental condition becomes gradually impaired; the memory is defective, the disposition is altered and there is an abnormal tendency to emotional disturbance, the patient laughing or more usually crying on the slightest provocation. This post-hemiplegic dementia is probably less the result of the focal lesion than of an associated and extensive degeneration of the cerebral vessels.

Nature of the Lesion. Omitting cases of infantile hemiplegia, which are considered later (see p. 99), the common lesions involving the motor fibres of the internal capsule are hæmorrhage and softening, the latter being a result of embolism or of arterial thrombosis. Occasionally the lesion is a tumour pressing on the capsule, but in such a case paralysis develops very slowly, one limb being affected some days or weeks before the other; and as a rule one or all of the classical symptoms of cerebral tumour are present, viz., headache, vomiting, and optic neuritis.

In a young person, embolism from heart disease and thrombosis from syphilitic endarteritis are the common causes of hemiplegia. An occasional cause of thrombosis is the blood-state associated with the post-puerperal condition. In persons over forty, hæmorrhage or

thrombosis from atheroma are usually found post mortem and frequently in association with chronic renal disease. In hæmorrhage the attack of hemiplegia comes on suddenly, in thrombosis it is often preceded by certain premonitory symptoms such as headache, vertigo, or transient palsy of face, speech or limb. The artery involved is the lenticulo-striate branch of the middle cerebral.

It must not be forgotten that the vascular lesions common in youth may also occur in elderly persons, and may thus complicate the pathological diagnosis. For example a man of fifty-five has hemiplegia apparently caused by thrombosis from atheroma, yet the post mortem shows that the softening which caused his hemiplegia was due to syphilitic endarteritis.

Fig. 38.—Showing the attitude of the arm in a case of athetosis, following a temporary attack of hemiplegia and hemianæsthesia.

Course. In some cases the hemiplegic weakness rapidly subsides, so that in a few weeks or even in a few days there may be little evidence of the previous paralysis, except perhaps as regards the finer movements of the hands. In other cases the paralysis is more or less permanent; it is usually impossible to foretell whether hemiplegia will be transient or lasting. The rule is that some movements gradually regain power whilst others remain permanently paralysed. The face and tongue recover before the leg, and the leg recovers

before the arm, and sometimes so completely that weakness of the dorsi-flexors of the ankle is the only detectable paralysis. In the arm the shoulder recovers before the elbow; movements at the elbow recover before those of the wrist and fingers, but rarely completely, whilst the latter are usually considerably and permanently paralysed. Occasionally irregular spasmodic movements develop on the affected side (see fig. 38). These are more common in cases of infantile hemiplegia and will be described under that heading.

Hemiplegia with Hemianæsthesia. In the ordinary type of hemiplegia, anæsthesia, if present at all, is slight or transient. When it is well-marked and permanent, and affects the face as well as the limbs on the paralysed side, the lesion involves the hinder end of the internal capsule which is supplied by the lentieulo-optic artery. Owing to the closer proximity of the leg than the arm fibres to the sensory tract, paralysis of the leg is more marked than that of the arm, but it is never so great as the paralysis of the arm in the ordinary type of hemiplegia. Hemianopsia is frequently present on the paralysed side, owing to involvement of the fibres of optic radiation: the functions of the other special senses may also be impaired on that side.

In *hysterical hemiplegia* crossed amblyopia may be present but hemianopsia does not occur. The condition is further distinguished by the following points:—The hemianæsthesia is often profound; the paralysis is rarely complete and is variable at different times, moreover the face and tongue are usually spared; the plantar reflex is absent or is of the flexor type.

Crossed Paralysis. In this form, paralysis of the limbs on one side is associated with paralysis of a cranial nerve on the opposite side of the body, and the lesion involves the origin of that nerve. Thus simultaneous paralysis of the right limbs and the left seventh or sixth nerve indicates a lesion in the lower part of the left side of the pons; when the face is affected, its upper part is more paralysed than in the ordinary type of

hemiplegia. Right hemiplegia with anæsthesia and weakness of the masseter on the left side of the face indicates a lesion in the middle of the left side of the pons; right hemiplegia with paralysis of the left third nerve a lesion of the left crus cerebri. These types of paralysis are more frequently caused by a neoplasm than by a vascular lesion; a common lesion involving the crus and the third nerve is a gummatous meningitis.

Treatment. The objects to be aimed at in the treatment of the limbs in hemiplegia are (1) to prevent deformities caused by the contraction of the stronger muscles, as well as stiffness of the joints; and (2) to restore as much power as possible to the paralysed limbs

The tendency to rigidity and the development of contractures and adhesions in the joints can be lessened by frequently changing the posture of the paralysed limbs; this is especially important during the first few weeks after the seizure. The arm must not be allowed to lie constantly flexed or the leg constantly extended, otherwise there is a danger of the limbs becoming fixed in these positions. Peripheral irritation, especially cold, also tends to increase the rigidity; it is therefore desirable to protect the limbs from such irritation by keeping them wrapped in cotton wool.

Massage, galvanism and passive movements are often of great service; they may be commenced two or three weeks after the attack and should be employed daily for a long period of time. The circulation of the limbs may be stimulated by sponging with warm salt water followed by vigorous rubbing.

As soon as any part recovers power, the patient should be encouraged to move it frequently and to perform daily such exercises as are practicable. In this way co-ordination may be improved and the range of movement gradually increased.

The treatment of the underlying conditions, embolism, thrombosis or hæmorrhage is considered later (see Section XV.). Should syphilis be suspected the administration of potassium iodide in large doses and mercurial inunction must be instituted as soon as possible.

CHAPTER II.

INFANTILE HEMIPLEGIA.

The common causes of hemiplegia in the adult, namely embolism, thrombosis and hæmorrhage are occasionally operative in the young child. Thus embolism as a result of endocarditis produced by scarlet fever, chorea, or acute rheumatism may produce hemiplegia even in young children; sometimes it occurs apart from endocarditis, the embolus being derived from a thrombus in a dilated left auricle. Thrombosis from syphilitic endarteritis, a common cause of hemiplegia in the adult, also occurs, though very rarely: the common type of cerebral syphilis in the child is a cortical meningo-encephalitis which leads to a spastic diplegia rather than a hemiplegia (see Section XXI.).

Hæmorrhage from a ruptured artery is practically unknown in infants, rarely it occurs in older children. But hæmorrhage from rupture of distended venules is not uncommon during the first two years of life: it takes place on the surface or less frequently in the substance of the brain. Such venous hæmorrhage may be caused by whooping cough, severe vomiting, or diarrhœa, but its commonest cause is a series of convulsions sometimes the result of an acute disease as measles, scarlet fever or pneumonia. Very often, however, the convulsions cannot be traced to any definite illness. But whatever be the cause of the venous hæmorrhage it may be followed, according to its position and extent, by a diplegia or a hemiplegia. Occasionally hemiplegia may be traced to a difficult or abnormal labour, especially when there has been injury to the head from the use of forceps. As already mentioned, the convulsions which so frequently usher in infantile hemiplegia may occur apart from any obvious illness, and although venous hæmorrhage following a series of fatal convulsions has been demonstrated post mortem, it is by no means clear that it is a common cause of

infantile hemiplegia. In fact the initial pathology of hemiplegia occurring during the first five years of life to which the name infantile hemiplegia is commonly applied, is still obscure.

Examinations of the brain made a long time after the onset of the disease have revealed a sclerotic atrophy or a porencephalus. The atrophy may affect a group of convolutions or even the whole of one hemisphere. The convolutions are atrophied and are firm and hard to the touch; there is an associated increase of the subarachnoid fluid and sometimes a compensatory dilation of the underlying ventricle. The condition of porencephalus, that is a cavity or cyst at the surface of the hemisphere, may be associated with atrophic sclerosis or it may occur independently. Sometimes there is evidence, in the presence of the products of degenerated blood pigment, that the cavity is the result of an old hæmorrhage; in other cases the porencephalus is probably a sign of defective development.

The nature of the initial morbid process which leads to the sclerotic atrophy probably varies in different cases. Doubtless in some cases it is a thrombosis of superficial cortical veins, of the superior longitudinal sinus or of several small arteries, whilst in other cases it may be hæmorrhage or embolism; but it is highly probable that in the majority of cases an encephalitis is the primary lesion, and that the acute inflammatory process is closely allied to if not identical with that of acute poliomyelitis. This view is supported by the clinical association of the two diseases, by their tendency to occur more frequently at the same season of the year, and by their occasional occurrence together in epidemic form.

Symptoms. Two modes of onset, which is always acute, have been observed. In most cases the child is seized with convulsions and loss of consciousness, which may last from a few hours to several days. The hemiplegia which is apparent when the child regains consciousness is usually complete, but it may be partial at first, and become more absolute after several attacks of

convulsions. In other cases convulsions do not occur, hemiplegia being ushered in by fever, vomiting and coma. Occasionally the onset is marked by strabismus and ocular palsies.

Should the patient survive the initial symptoms, the paralysis may gradually disappear; usually, as in adult hemiplegia, there is a partial recovery only, the leg improving more than the arm, and the shoulder more than the hand and fingers. But little weakness remains in the face; there may, however, be overaction and a tendency to grimaces. The contractures which ensue tend to deformities similar to those seen in the adult, with the exception that the fingers, instead of being flexed at all the joints, as in the common type of hemiplegia, are flexed at the metacarpo-phalangeal joints and extended at the phalangeal ones.

Fig. 39.—Showing athetotic spasm of the hand in a case of old infantile hemiplegia.

Another characteristic of infantile hemiplegia is the liability of the paralysed limbs to be the seat of perverse movements; they may be affected by tremor, by choreiform movements or more commonly by a slow mobile spasm which is called athetosis. These abnormal move-

ments are usually limited to the arm, the leg being rarely affected. As the patient gets older the bones of the paralysed limbs, and sometimes those of the paralysed side of the face, show an arrested development, so that the affected limbs are smaller in all their dimensions than their fellows. The face, too, becomes asymmetrical, being diminished in size on the paralysed side. The diminution in size is noticed as high as the eyebrow, but above the eyebrow the forehead is often less prominent and the parietal bone flatter on the opposite side—that is on the side of the lesion, the less prominent side corresponding to the atrophied hemisphere.

Recurring convulsions may date from the onset of the illness or may only come on some months or years later. At first they are usually limited to the paralysed side of the body, and are not always accompanied by loss of consciousness. After a time however they tend to become general, but even then they often begin on the paralysed side. Sometimes the patient does not suffer from convulsions but is subject to attacks of unconsciousness (petit mal). The intellectual faculties are usually impaired and there may be considerable dementia. The occurrence of epileptic seizures tends to increase any imbecility that may be present. True aphasia is rarely present, but if the child had begun to talk before the onset of the disease its speech usually becomes impaired and in some cases permanent speechlessness ensues. When the left cortex is seriously damaged various forms of aphasia may result and may be permanent. But in the child as a rule the function of speech is gradually taken up by the right side of the brain.

With regard to the reflexes, the superficial are generally diminished on the hemiplegic side, whilst the plantar is of the extensor type. The wrist and the knee-jerk are exaggerated and ankle clonus is often present; sometimes however owing to extreme rigidity of the limbs these signs are difficult to elicit.

Prognosis. It is rare for the patient to die during the initial illness. When this stage is passed the prognosis

is good both as regards duration of life and improvement of the paralysis. In some cases the hemiplegic limbs regain power rapidly, in others very slowly, but even when there is severe paralysis with marked rigidity a certain degree of improvement may be predicted if treatment is properly carried out. Recurring convulsions, athetosis and much mental impairment are unfavourable symptoms, especially as they hinder attempts at re-education of the movements of the paralysed limbs.

Treatment. During the initial illness the patient should be kept in bed, ice being applied to his head: the administration of a brisk purge is also desirable. As soon as the acute symptoms have subsided massage of the paralysed limbs may be commenced and should be systematically carried out for many months. When some power has returned, physical exercises suitable to the condition of the affected limbs should be prescribed and every effort made to direct volition so as to rectify imperfect movements. To this end exercises in front of a mirror are sometimes useful, for the child can then make an effort to correct any observed errors in position or in movement. Such re-education of the limbs is impossible when there is much mental deficiency, which however may often be improved by special education, patiently persevered in for a long time.

In spite of every care, contractures and deformities frequently develop and may require tenotomy and other surgical treatment.

CHAPTER III.

SPASTIC DIPLEGIA.

This implies a spastic paralysis of all four limbs and sometimes of the lower facial muscles, which is dependent on disease of the motor centres or the motor paths on both sides of the brain. It may be produced

by lesions of the pons or medulla which implicate the pyramidal tracts. In such cases involvement of some of the cranial nerves is the prominent feature, spasm and paralysis of the limbs being usually slight or moderate in degree, although the paralysis, especially of the legs, tends to increase. The condition also occurs as a result of pressure on the pyramidal tracts, as from a tumour of the cerebellum. Here also the spastic weakness is not conspicuous, and is overshadowed by incoordination of movement, headache and other signs of an intracranial tumour.

A double hemiplegia may also occur from a lesion of each internal capsule. Thus a man had right hemiplegia from thrombosis, the result of syphilitic endarteritis, and during partial recovery from the paralysis he was seized with left hemiplegia. But such cases are rare. The most typical forms of spastic diplegia occur in infancy in consequence of cortical disease which according to its extent and position gives rise to many variations in the distribution and in the relative proportion of spasm and paralysis, and also to variations in kind and degree of other symptoms—mental defects, perverse movements, convulsions—which are usually found in association with the spastic paralysis. The following account applies to the commonest variety of infantile cerebral diplegia or "birth palsy" as it is sometimes called.

Congenital Cerebral Diplegia.

The symptoms of this disease are usually observed a few days or weeks after birth, and rarely for the first time after the third year. It is noticed that the infant is backward either as regards its mental development or the movements of its limbs, which already may present an unusual stiffness. Frequently it is remembered that from birth the infant was never quite natural, thus indicating that the condition was congenital: some difficulty in sucking or in swallowing was present, or nystagmus, strabismus, or some perverse movement or

unnatural attitude was observed. Attempts at walking or talking are defective, indeed these acquirements may never be learnt. In a well marked case spastic weakness affects nearly all the muscles of the body, rigidity being usually more prominent than paralysis. The legs are more affected than the arms, and the arms than the

Fig. 40.—Photograph of a case of congenital cerebral diplegia, showing imbecility, and rigidity of the legs. (Lapage.)

face. The thighs are rotated in and adducted, the knees rigidly flexed and the heels drawn up by spasm of the calf muscles. The arms are rigidly adducted, and flexed, and the hands and fingers present various spasmodic deformities. The chin as a rule is pressed down upon the chest, and the head and trunk are bent forward, the spine being arched backwards. When the facial muscles are involved, the expression is often a fixed one; the palpebral fissures are wider than natural and the mouth tends to remain widely open. The above description applies to the most common attitude, which however varies in different cases; sometimes the head is retracted and the legs are rigidly extended at all their joints.

Perverse Movements are often present in cases of diplegia, especially when rigidity is slight. They may

take the form of tremor, sometimes similar to that seen in disseminated sclerosis, or of quick shock-like contractions—called "choreic," or of slow irregular muscular contractions—"athetosis." Overaction of the facial muscles, and irregular movements of the protruded tongue are also observed, while in severe cases all the muscles of the body may be implicated.

Fig. 41.—Boy with small conical head, internal squint, defective intelligence, and spastic limbs. Almost daily convulsions from age of six months till death at age of two years.

Epileptiform convulsions are frequently met with in the subjects of spastic diplegia. Sensory disturbance, apart from pain on passive movement of the stiff joints, is usually absent. The knee-jerks are exaggerated, and unless the limbs are too rigid, ankle clonus can often be elicited. The abdominal reflexes are sometimes absent, while the plantar reflex is frequently extensor in type.

The mental impairment varies greatly in degree; every transition is met with between slight backwardness and profound amentia. Speech may never be acquired, or the child may learn to talk at a much later period than usual, and in an imperfect fashion. A slurring and stammering articulation associated with facial grimaces is very common. If the disease comes on after the child has learnt to talk, speech is generally completely lost. Sometimes there is a difficulty in

swallowing: probably it depends on rigidity rather than on paralysis of the muscles concerned in the act.

Fig. 42.—Rigidity of the legs and talipes equinus, in congenital cerebral diplegia. (Ross.)

Convergent strabismus, nystagmus, and primary optic atrophy (from degeneration of the ganglion cells of the retina) are other symptoms that occur in a considerable number of cases. According to the preponderance of certain symptoms different *types* have been distinguished, viz.:—

(1) The generalised type, which corresponds to the above description. (2) The paraplegic type in which the spasticity is limited to the lower limbs, the arms being normal, or the hands showing occasional clonic spasms. (3) The perverse movement type—cases of bilateral athetosis and of so-called congenital chorea. (4) The mental type—cases known as congenital spastic idiocy in which only slight rigidity of the limbs is associated

with considerable or complete dementia. Between these types every variety of combination is met with.

Etiology and Pathology. In the large majority of cases the disease is congenital in origin; very rarely the disease appears to be acquired after the third year,

Fig. 43.—Photograph of boy showing spasm of face, fingers and arms; he could not speak nor completely close his mouth. The condition followed a febrile illness, of obscure nature, attended by unconsciousness.

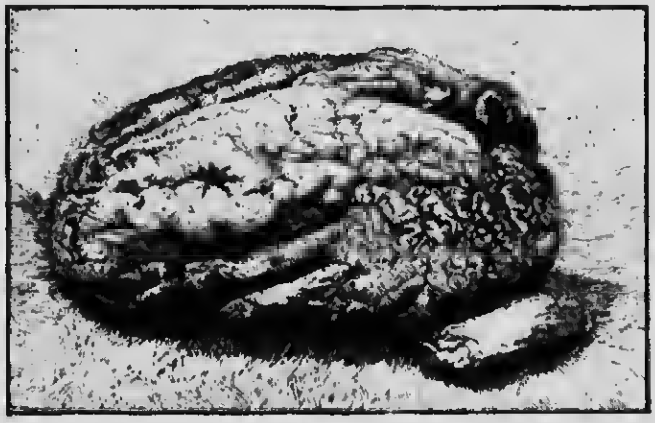


Fig. 44.—Brain from Case Fig. 41, showing a deep sulcus behind the frontal lobe, and imperfectly formed convolutions behind the sulcus. The cerebellum was uncovered as in the figure.

its symptoms coming on gradually or being ushered in by a febrile attack (see fig. 43) or by a series of convulsions. Both in the congenital and in the post-natal

cases similar changes have been found in the brain when examined some time after the onset: they consist of sclerosis and atrophy of the convolutions of both hemispheres, the surface of the brain having an appearance similar to that of a walnut kernel (see fig. 44). This sclerotic atrophy is usually striking in the Rolandic area, but its exact position and extent vary somewhat in different cases: sometimes the prefrontal lobes are markedly affected when amentia is the prominent symptom.

Cysts on the surface of the hemisphere are sometimes present and a distinct cavity may connect the arachnoid sac with the lateral ventricle: this is called *porencephalus*.

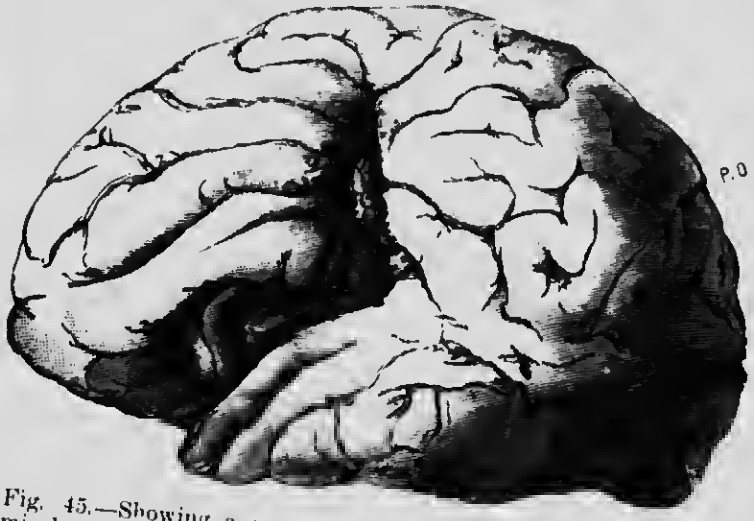


Fig. 45.—Showing a porencephalus cavity in the left cerebral hemisphere, taking the place of the central convolutions; there was a similar cavity in the right hemisphere; both cavities opened into the lateral ventricles. (Ross.)

With regard to the early lesions which lead to the above changes, there has been difference of opinion. Meningeal hæmorrhage occurring at birth, thrombosis or embolism in the cortex, and cerebral hæmorrhage leading to the formation of cysts have been described, but it seems probable that in most cases degeneration of the cortical cells is the primary lesion, and that this is

started by some toxic agent which is derived from the mother during intrauterine life. In favour of this view may be mentioned the frequency with which the mother has suffered from ill health during pregnancy; this may be the result of overwork and privation, of syphilis, or of some acute disease. When the symptoms do not appear until after the third year, they are sometimes preceded by an acute illness, but it is doubtful whether this is entirely responsible for the early cortical changes; it may have lit up a latent pathological condition which dated from birth.

Diagnosis. We have seen that the anatomical substratum of "birth palsy" is degeneration of the cortex, hence we might expect that other causes of such a condition would give rise to a like symptomatology. There is one group of cases especially which requires mention, namely cases of hereditary syphilis in which a meningo-encephalitis occurring in early life has produced sclerosis of the cortex with resulting degeneration of the pyramidal tracts. In both the syphilitic and the congenital diplegic cases, there is an association of spastic limbs, with mental deficiency and a liability to convulsive seizures. A diagnosis between the two groups is made by the presence or absence of the signs of late hereditary syphilis, namely notching of the upper central incisor teeth with narrowing of their cutting edges, symmetrical keratitis, deafness and scars at the angles of the mouth.

In some cases of "birth palsy" evidence of spasm in the arms is slight or is completely absent; then if convulsions do not occur and mental backwardness is not very obvious the spastic paraplegia may suggest disease of the cord. The only likely cord affection in young children is compression myelitis from caries of the vertebræ; as a rule this would be readily diagnosed by the undue prominence of one or more vertebral spines and by rigidity in movement.

Exceptional cases of "birth palsy" may present a resemblance to disseminated sclerosis; this affection

however is very rare in childhood and as a rule would be easily distinguished by the presence of nystagmus or of tremor.

The practical rule in all cases of spastic paraplegia occurring in children, other than those due to spinal caries, is to take the cerebral cortex into consideration, to direct attention to the mental condition, to any tendency to spasm in the arms, and to make enquiries as to the occurrence of convulsive attacks.

Prognosis. This depends on the severity of the symptoms, that is on the degree of the mental defect with that of the paralysis and spasm. In many cases some improvement may be expected both in the mental condition and in the ability to stand and walk.

Treatment. This must be carried out on similar lines to those laid down for infantile hemiplegia. Individual attention and special training are needed both for the development of the mind and for the correction of motor defects. The tendency to convulsive attacks may often be lessened by the administration of bromides or of borax: in the author's experience the latter drug appears to be more efficacious in the convulsions of "birth palsy" than in those of ordinary epilepsy.

CHAPTER IV.

MONOPLEGIA.

This term is usually applied to partial varieties of hemiplegia which depend on lesions situated in or near the cortical motor area, so that according to the seat of the lesion there may be a crural, a brachio-crural, a brachial, a brachio-facial, a facial or a labio-glossal monoplegia. The paralysis is usually partial in degree; the distal portions of the limbs are more affected than the proximal, and then often present some impairment of tactile sensibility or of the muscular sense. A distinguishing feature of monoplegia is its tendency to be associated with attacks of partial epilepsy. The clonic

spasms always begin locally; they may then spread to the whole of the affected side and ultimately, in some cases, to the opposite side. The site of the initial spasms together with the distribution of the paralysis, are indications as to the position of the lesion. Thus labio-glossal paralysis suggests disease of the lowest part of the ascending frontal convolution on both sides; brachial spasm and paralysis a lesion of the middle third of the ascending frontal on the opposite side, and so on (fig. 2). The common lesions leading to monoplegia are gummata, new growths and softening from arterial thrombosis.

The dependence of a monoplegia on cortical disease is usually indicated by its association with attacks of clonic spasms; when these are absent, as in the case of a slowly growing tumour, we have to rely on other symptoms such as the presence of optic neuritis. Sometimes the diagnosis from a peripheral neuritis has to be made; thus a patient may suffer from weakness and numbness of the distal portion of one arm. In such a case exaggeration of the wrist and elbow jerks, as compared with the other arm, would point to a cortical lesion, while their absence would be evidence in favour of neuritis.

Lastly, weakness of a limb with marked contractures may seem to originate in severe emotional disturbance; if so it is usually associated with manifestations of hysteria.

CHAPTER V.

SPASTIC PARAPLEGIA.

This implies that both legs are affected by paralysis and rigidity, the two elements varying much in degree in different cases. The condition indicates that the upper neurons for the legs are involved, but apart from a consideration of associated symptoms the exact position of the lesion cannot be determined. On

anatomical grounds it is likely that it is situated where the pyramidal tracts are near together, that is in the cord or possibly in the medulla or pons. The next most likely place would be the cortex, for there the centres for the legs are adjacent. These considerations are borne out in practice. We find that in the child the lesion may be either in the brain or in the cord; thus, as already mentioned, the spastic paralysis of a birth palsy may be limited to the legs. In the adult, however, when spastic paralysis affects the legs the lesion is nearly always in the cord. At both periods of life a slight spastic paraplegia may depend on involvement of the pyramidal tracts in the brain as by tumours of the medulla, pons or cerebellum. Spastic paraplegia occurs in a large number of spinal affections but it is most frequently found to depend on disseminated sclerosis or on a dorsal myelitis, either the result of compression, as from caries, or of independent origin. Very rarely it depends on degeneration limited to the pyramidal tracts, when the disease is called primary lateral sclerosis. In this affection spastic paraplegia is unattended by any other symptoms; for this reason it is useful to describe it first. On clinical grounds it seems desirable to consider disseminated sclerosis in the present section, but it is obviously inconvenient to separate dorsal myelitis from other varieties of myelitis. For analogous reasons other conditions in which a spastic paraplegia occurs are described in their appropriate sections.

Primary Lateral Sclerosis.—Primary Spastic Paraplegia.

By this is meant a degeneration of the pyramidal tracts in the spinal cord, occurring spontaneously and independently of myelitis or other local disease; it may be a result of nutritional changes in the cortical motor cells from which the pyramidal tracts arise and on which their nutrition depends. The occurrence of such a disease is still doubtful. Certainly the majority of

cases of uncomplicated spastic paraplegia in the adult afford indications sooner or later which prove that the disease is disseminated sclerosis; whilst the cases which occur in children are for the most part examples of cortical disease (see infantile diplegia).

Morbid anatomy has not thrown much light on the question. Still in spite of the absence of conclusive pathological data, it is nevertheless true that very exceptionally cases of uncomplicated spastic paraplegia

Fig. 46.—Showing lateral sclerosis of the spinal cord; A, A, A, degenerated pyramidal tracts. (Charcot.)

are met with which, in their development and subsequent course, as tested by observations extending over many years, do not justify any other diagnosis than that of a primary lateral sclerosis. Moreover, in cases of this kind, as in some observed by Erb, degenerative changes have been practically limited to the pyramidal tracts; the only other associated lesions being a very slight sclerosis of the direct cerebellar tracts and of Goll's columns.

The apparent exciting causes have been wet, cold, over-exertion and concussion of the spine. Syphilis may be an occasional cause for it sometimes leads to an analogous condition (see p. 117). The symptoms usually first appear between the ages of twenty-five and forty-five. Sometimes a neuropathic heredity can be traced: when this is the case it is possible that the pyramidal tracts, owing to congenital weakness, are particularly prone to degenerate when exposed to the action of toxins or other adverse influences.

Symptoms. The earliest symptom is weakness of the legs which may be equally affected, or the one leg may be more affected than the other. Hence the patient has

a difficulty in walking, and is easily tired; his legs feel weary and stiff, and his gait becomes dragging and difficult. On examination, weakness is usually first detected either in the flexors of the hip or in those of the ankle. The paralysis gradually spreads and increases in intensity, and is accompanied by muscular spasm, which sooner or later passes into permanent rigidity. The leg is rigidly extended; there is marked contraction of the adductors of the thighs and the feet are in the position of talipes equino-varus.

The knee jerk is much exaggerated and ankle clonus is readily elicited. The plantar reflex gives the extensor response; the abdominal reflexes are usually normal. The patient is often much troubled by cramps and twitching of the muscles, and tremor may be severe enough to shake the whole body. In rare instances the arms have also been affected, sometimes becoming rigidly extended and pronated.

Apart from pain in the back, sensory symptoms are absent throughout the whole course of the disease; the functions of the bladder and rectum are unaffected, and there are neither vaso-motor nor nutritive disturbances.

The course of the disease is very slow; sometimes it comes to a standstill and remains stationary for twenty years; occasionally distinct improvement is observed. As a rule however the patient gradually becomes quite helpless. Death generally results from some accidental cause, or from intercurrent disease.

Diagnosis. The diagnosis of primary lateral sclerosis can rarely be made with any certainty, and then only after observations extending over many years. As a rule the sclerosis is either the first stage of subsequent more extensive disease, or it is already associated with other lesions, the symptoms of which are not yet manifest. The probability of the case being one of disseminated sclerosis should always be prominently recognised –slight nystagmus or slight tremor are easily overlooked, so also are modifications of the superficial abdominal reflexes, which may be the first indications

that the functions of the grey matter are interfered with. The spinal column should also be carefully examined for signs of caries. Wasting of the small muscles of the hand would suggest that the case was one of amyotrophic lateral sclerosis. Attention should also be directed to the presence of slight sensory symptoms such as girdle pains or hyperæsthesia or areas of partial anæsthesia in the legs—signs that myelitis may be present.

Treatment. In the early stages moderate exercise is beneficial but over-exertion is harmful. When there is much spasm, rest in bed is desirable: the annoying spasmodic contractions of the muscles may be kept in check by hot baths, or by the application of hot bags to the spine. Antipyrin, or the bromides may be needed to give relief. Veronal, two grains once or twice daily, is also frequently useful. Above all, abundant sunlight and fresh air are needed to preserve the general health.

Familial Spastic Paralysis.

This is a rare condition which is characterised by the presence of spastic paralysis in several members of the same family and sometimes in different generations. The disease appears to be directly transmitted to children through either parent: it affects males and females equally, and its symptoms are usually prominent at an early age.

At first stiffness of the legs and clumsiness in walking are observed; at a later period there may be complete spastic paraplegia together with, in some cases, a spastic weakness of the arms. The face is not involved and in uncomplicated cases the mental condition is good: the cranial nerves are not affected. The deep reflexes are exaggerated and this exaggeration may precede the paralysis by several years. There is neither incoordination nor anæsthesia. It is stated that in some cases bulbar symptoms have occurred, and in others mental lethitude and optic atrophy. It is doubtful whether such cases should be included in the present group. By

post mortem examination alone, can a distinction be drawn between a large number of cases of family disease which present every transition between an uncomplicated spastic type and congenital diplegia on the one hand, and Friedreich's disease and "hereditary cerebellar ataxy" on the other.

It is believed that the disease depends upon some congenital weakness of the pyramidal cells of the cortex cerebri which leads to early degeneration of the pyramidal tracts, their distal portions being most affected. Degeneration of Goll's columns has also been found.

Erb's Syphilitic Spinal Paralysis.

This variety of spastic paralysis was first differentiated from primary lateral sclerosis in 1892 by Erb. It is a less common type of spastic paraplegia from syphilis than that which follows acute myelitis.

The disease is much commoner in males than in females and usually occurs between the ages of twenty and forty. The onset is very gradual and as a rule occurs within five years of infection.

The early symptoms are a tired feeling in walking, and weakness of the legs, together with signs of disturbance of the functions of the bladder. The latter in some cases are very prominent. Constipation and a dull aching pain in the back are often troublesome symptoms. The symptoms of the fully developed disease are: Spastic paresis of the legs, absolute paraplegia being rare; ankle clonus and exaggerated knee-jerks, together with Babinski's reflex; slight changes occur in the cutaneous sensibility, the temperature sense being often conspicuously affected. Kowalewsky has drawn attention to the strong convulsive contractions which occur when the legs are touched with warm or cold objects. Retention of urine is usually present; sometimes a spastic incontinence is observed. Cases of this disease often improve under treatment and complete recovery may take place; partial recovery however

is more frequent, some weakness of the bladder or the legs remaining.

But few post mortems have been made. Erb says that the lesion is an incomplete myelitis of the lower dorsal region with secondary degeneration downwards in the lateral, and upwards in the posterior columns. Endarteritis and degeneration of the arteries of the cord and meninges have also been described.

The presence of bladder and of sensory symptoms distinguish Erb's paralysis from primary and from amyotropic lateral sclerosis. In the latter affections muscular rigidity is also a more marked feature. Muscular atrophy, prominent in amyotropic lateral sclerosis, does not occur in Erb's paralysis.

CHAPTER VI.

DISSEMINATED SCLEROSIS.

This is a common disease at least in English practice. The characteristic symptoms are: spastic paraplegia, intention tremor in the arms, nystagmus and scanning speech. Optic nerve atrophy is often present.

Etiology. An indirect neuropathic inheritance may sometimes be traced: very rarely two or three members of one family have been affected. The disease is rare in children and in elderly people and occurs most frequently between the ages of twenty and thirty. Its exact causation is not known. Sometimes it has followed one of the infective fevers, especially enteric and influenza, also rheumatism and pneumonia. Depressing mental influences, over fatigue, blows and other injuries have been noted as the immediate antecedents to the early symptoms of the disease: so also have pregnancy and parturition. These and other etiological factors suggest that the disease is due to the action of some toxic agent on the nervous system, which is predisposed to be affected by it, in consequence either of

previous injury or disease, or of some congenital abnormality.

Symptoms. The manifestations of the beginning and the development of the disease vary widely in different cases. Sometimes the onset is quite sudden and may take the form of a hemiplegic attack, or an outbreak of hysteria. Sensory symptoms, as numbness of the extremities, or partial anæsthesia of one side of the face, may be first noticed. In one of my cases numbness and blueness of the fingers, suggesting Raynaud's disease, came on suddenly, lasted three months, then disappeared, and was followed by spastic weakness of the legs and other symptoms of disseminated sclerosis. In other cases visual defects or optic atrophy may precede other symptoms for many months or even a few years.

Probably in the majority of cases weakness and heaviness in the legs is the most striking, if not the earliest symptom noticed by the patient. In such a case on testing the strength of the legs slight weakness of the flexors of one hip, or of one ankle is often found; the knee-jerk is unduly irritable, while the plantar reflex may be either normal, absent, or may show the extensor response. Frequently such weakness, when met with in young women who appear emotional and self-centered, is put down to hysteria. This however should be excluded as a cause of paresis of a group of muscles, such as the flexors of the hip. The paresis may pass away either suddenly or gradually and then return and progress, or be replaced by weakness elsewhere. After a period which varies from weeks to months paralysis of the legs becomes conspicuous at least as regards flexion at hip, knee and ankle; it is associated with ankle clonus, exaggeration of the knee-jerk and Babinski's reflex. There may be no anæsthesia and no bladder disturbance.

The diagnosis of such uncomplicated spastic paraplegia is lateral sclerosis, and in the young adult this is frequently the first stage of disseminated sclerosis. In many such cases, a careful examination will reveal

slight lateral nystagmus. Slight tremor and a change in articulation may also be observed, or these symptoms develop later; but although tremor is common, typical intentional tremor and still more scanning speech may never appear and at any rate are less common than is stated in many text-books. Occasionally, however, tremor and nystagmus are prominent, while paralysis and spasticity of limb are less conspicuous. The course of the disease is very variable; occasionally its progress is arrested, and exceptionally complete recovery occurs. Frequently periods of arrest are followed by relapses until finally the patient becomes helpless, intelligence is clouded and the sphincters are paralysed. Then bulbar paralysis, or septic disease of the kidney, or pneumonia or other intercurrent malady quickly leads to a fatal termination.

The various symptoms that are met with in the fully developed disease may now be described.

Speech. In typical cases this is scanning and syllabic, that is the syllables of words are unduly separated and accentuated. The change is well brought out when the patient tries to pronounce polysyllabic words like perambulator, Manchester or Constantinople. Such a striking disturbance of articulation however is much less common than minor degrees of impediment. Sometimes the voice is husky and monotonous.

Ocular phenomena. Nystagmus affecting both eyes is a common symptom. Most frequently it is horizontal, sometimes it is also vertical and rotatory. In many cases it is represented by only a few jerks in extreme lateral tension of the eyeballs, and it may be present on one side only. In rare cases paralysis of the external rectus or of some of the other ocular muscles has been observed, but it is commoner to hear of a history of diplopia or squint than to be able to detect actual weakness of any ocular movement. The pupils are usually unaffected, sometimes they react imperfectly to accommodation. Disturbances of vision are common and at first are often transient; remissions and relapses occur

just as in the case of limb paralysis. Loss of vision frequently progressive, rarely proceeds to complete blindness. Pallor or decided atrophy of one or both optic discs occurs in 50 per cent. of cases of the fully developed disease: it is generally more pronounced on the temporal half of the disc. In some cases the atrophy accounts for the loss of vision: in other cases visual defects occur without ophthalmoscopic changes, when they may depend on a retro-bulbar neuritis.

The field of vision sometimes shows an irregular restriction, or there may be a central scotoma for white and colours, or for colours only. Central scotomata are common in the optic atrophy of disseminated sclerosis, but rare in that due to tabes (see fig. 150, page 542).

Motor disorders. In addition to the spastic paraplegia, which varies much both as regards the degree of paralysis and of rigidity, there may be weakness in the arm, but this rarely attains much intensity. Rarely there is atrophy of the small muscles of the hand or of other groups of muscles, owing to invasion of the spinal anterior horn by islets of sclerosis. Rarely paralysis may be detected in the tongue, the masseters, the palate or the vocal cords.

Tremor. When present, jerky tremors produced by voluntary movements are characteristic features of the disease. They may be absent from first to last. Frequently they are limited to the arms, while occasionally they affect every part of the body. In the latter case when a patient is lying in bed no tremor can be seen, but the head shakes when raised from the pillow, tremor of the trunk is observed when the patient sits up, and may affect the legs so severely that standing becomes impossible. But as a rule tremor is only conspicuously seen in the arms, and is best brought out when the patient takes a glass of water in his hand and endeavours to bring it to his mouth; the tremor, slight at first, increases in violence during the progress of the action and the water is often spilt before the glass reaches the mouth. Tremor may be conspicuously

shown, and often at an early period of the disease, in the hand-writing or in the movements of sewing.

As the disease advances and paralysis becomes more marked, the tremors tend to disappear.

Ataxy may be combined with the muscular weakness; in some cases the incoordination resembles that of tabes in others that of cerebellar disease. A spastic-ataxic gait is almost as common as a purely spastic-paretic one.

Reflex action. The wrist, elbow and knee jerks are usually increased and this exaggeration may be observed when the limb paralysis is slight. Ankle clonus is generally present. The superficial abdominal reflexes are diminished or absent; this defect is often an early symptom, and is one of the most characteristic signs of the disease. Of still greater importance, especially as regards the diagnosis of this serious disease from hysteria, is the presence of the extensor type of plantar reflex, which can often be elicited in the earliest stages of the disease; occasionally however at this period and at others the response is flexor in type.

Sensory disturbances. The cutaneous sensibility, normal at first, may become diminished at a later period. The loss is usually partial and not extensive, and is most marked in the hands and feet. Rarely hemianæsthesia occurs; this may be caused by an islet of sclerosis in the cerebral sensory path, or may depend on an associated hysteria. Subjective sensations of numbness and tingling in the extremities are common; they may also be experienced in the face and in other parts of the body. Sometimes headache and attacks of vertigo cause the patient much distress.

Sphincters. Slight loss of control over the sphincter of the bladder, causing a hesitation or a precipitancy in micturition, is not uncommon as an early symptom, but much interference in the functions of the bladder and rectum is usually only present at a late period of the disease. The sexual functions, too, remain normal for a long time; in some cases they become impaired or lost, in others they show over-excitability.

Psychical disorders. Mental irritability, emotional disturbance, undue complacency and other indications of slight mental changes are common, but actual unsoundness of mind is rare. In the final stage of the disease enfeeblement of the mind may be conspicuous and may pass into complete dementia.

Apoplectiform or epileptiform seizures are occasionally observed, they are analogous to those which occur in general paralysis of the insane.

Vaso-motor and trophic disturbances may be present, in the form of œdema of the feet, blisters on the skin, swelling of the joints, changes in the growth of hair or nails, or altered secretion of sweat. The condition known as erythromelalgia has been observed. Muscular atrophy, a rare phenomenon, is an indication that the lower neurons are implicated.

Morbid anatomy. Patches of sclerosis are found scattered through the brain and spinal cord in what appears to be a most random fashion. The peripheral nerves may also be involved especially the optic nerves, including the optic chiasma. In the central nervous system the islets of sclerosis are found chiefly in the white matter, but the grey matter does not escape and indeed that of the basal ganglia is a common site for the sclerotic process. The patches vary in size: they are irregular in shape, and greyish in colour, being usually a little darker and more translucent than the normal grey matter of the cerebral cortex. Recent patches are

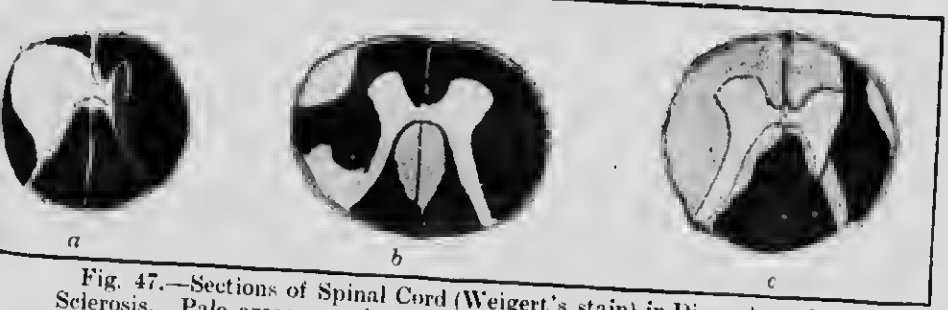


Fig. 47.—Sections of Spinal Cord (Weigert's stain) in Disseminated Sclerosis. Pale areas = patches of sclerosis; dark portions = normal white matter: (a) upper cervical region; (b) mid-cervical; (c) lowest cervical. Note absence of ascending degeneration. (Williamson.)

soft and gelatinous, old ones are firm in texture. In addition to their irregular distribution two other features are peculiar: (1) The patches have a sharp outline and there is an abrupt transition from diseased to healthy tissue. (2) It is rare to meet with ascending or descending degenerations as results of interruption of the long conducting tracts in the cord. By this last feature the disease is distinguished from disseminated myelitis, and from multiple syphilitic lesions, in which such degenerations are well marked.

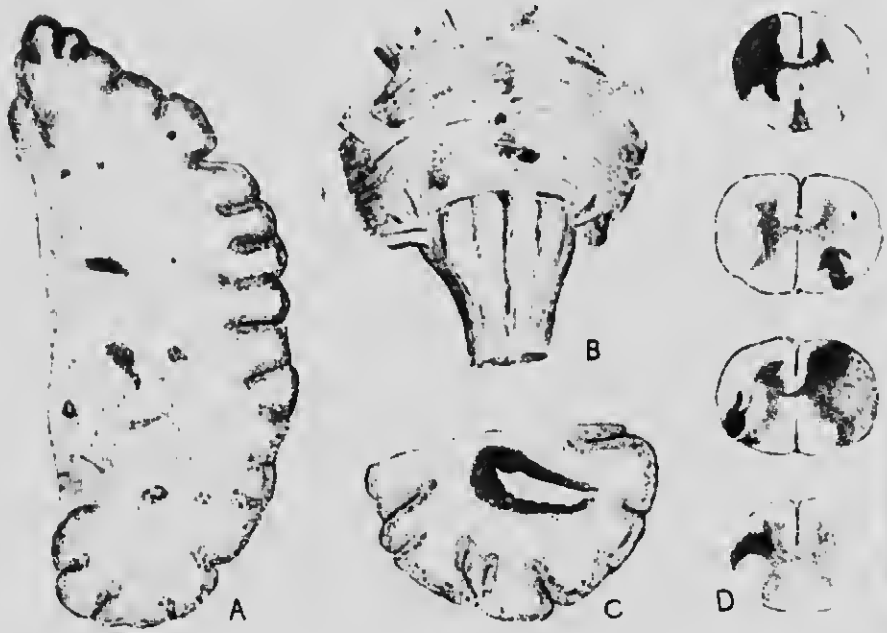


Fig. 47a.—Insular sclerosis. A, Centrum ovale of right hemisphere. B, Pons and medulla. C, Peri-ependymal sclerosis around the descending cornu of the lateral ventricle. (A, B, and C from an original case). D, Sections of the spinal cord to show the varying distribution of the sclerosis at different regions. (Gowers, after Leyden).

Under the microscope the sharply defined character of the diseased areas is striking—they are seen to be made up of proliferated neuroglia, and of wasted nerve fibres. The wasting is due to a narrowing or complete

disappearance of the myelin sheaths; the axis cylinders persist although ultimately they too may succumb.

The walls of the blood vessels may be either normal or thickened and show hyaline degeneration. In patches of recent origin the perivascular lymph sheaths are often widened and filled with round, granular fatty cells.

Pathology. It is still uncertain whether proliferation of the neuroglial tissue is the primary change, or whether it occurs as a secondary consequence of the wasting of the nerve elements, and even if we knew which change began first we could not speak with certainty as to its causation.

Some authorities believe that disease of the blood vessels is the initial lesion as marked vascular changes are sometimes present, and occasionally there is a certain correspondence between the extent of the area of sclerosis and the distribution of a diseased artery. But this is not common, and the vessels supplying the diseased area may be quite healthy. Another and more probable view is that the circulation of some toxic substance in the blood vessels and lymphatics irritates their endothelium and leads to the extravasation of toxic lymph into the surrounding nerve tissue; as a consequence the myelin sheaths of the nerve fibres degenerate.

If this toxic hypothesis is correct, it seems likely that the myelin degeneration will begin before the proliferation of the neuroglia. A congenital defect in the development of the myelin sheaths has been suggested as a predisposing cause.

Diagnosis. Typical cases are easy to recognise but aberrant forms of the disease often present difficulties. It is a good rule to think of disseminated sclerosis in all cases of spastic weakness of the legs, unattended by sensory symptoms and weakness of the bladder.

Hysteria. It is in the early stages of disseminated sclerosis that the diagnosis of hysteria is so frequently made. The error is avoided by repeated careful

examinations and by remembering that the beginnings of disease in the nervous system are frequently associated with, and overshadowed by hysterical manifestations. Inability to perform a complicated movement such as walking, apart from weakness of any particular group of muscles may be due to hysteria but weakness of the flexors of the hip or the ankle suggests a definite local lesion, and even if recovered from, it indicates the necessity of keeping the patient under observation, in order that sustained ankle clonus, Babinski's reflex, slight nystagmus, or volitional tremor, may be early detected. The presence of central scotomata or of optic atrophy would also exclude hysteria.

Ataxic paraplegia. In this disorder the condition of the limbs is similar to that observed in some cases of disseminated sclerosis, and for a time the diagnosis may be doubtful. Indeed it seems probable that a large number of cases of ataxic paraplegia ultimately prove to be varieties of disseminated sclerosis, or represent an early stage of the disease known as subacute combined degeneration of the spinal cord.

As to other maladies with which a temporary difficulty in diagnosis may be experienced especially in the early stages of the disease, it is sufficient to mention tabes, general paralysis, paralysis agitans, cerebral diplegia, and tumours of the cerebellum, the optic thalamus, crus and pons, and to refer the reader to their respective descriptions.

It is important to again draw attention to the fact that occasionally optic atrophy and visual failure are early symptoms of disseminated sclerosis, and for some time may be unattended by any other symptoms of the disease. A central scotoma and pallor of the temporal half of the optic disc developing in one eye, partially subsiding and subsequently developing in the other eye, are suggestive symptoms especially if the patient is young and has not suffered from syphilis, lead poisoning, or alcoholism. In such a case slight weakness of the

legs, Babinski's reflex and loss of the abdominal reflexes would strongly suggest the presence of the disease.

Prognosis. If the lesions in this disease are initiated by toxins either produced within the system or introduced from without it is reasonable to believe that their action might cease owing to their supply being either limited or cut off, or to their power being destroyed by the development of antitoxins.

That this does occur, if only in a partial degree, is indicated by marked improvement in the symptoms or by their complete disappearance. The rapid oscillations of many of the cardinal symptoms which may be so frequently observed in disseminated sclerosis are difficult to account for. At present no better explanation can be offered than that suggested by Dixon Mann who puts the matter as follows:—"Assuming that an autogenous toxin is the causal agent by which disseminated sclerosis is developed, it may be further assumed that in the early stages of the disease, the toxin is produced intermittently, and that it may derange the function of the implicated nerve structures before any recognisable anatomical changes are produced. Such disturbance of function would occur in waves corresponding to the intermittent evolution of the toxin, and would subside as each batch of toxin was exhausted."

Sometimes a complete and long remission of all the symptoms occurs, and very rarely the remission is permanent, complete recovery being established.

The natural history of the disease makes one wonder why recovery does not occur more frequently. Unfortunately, however, a downward progress, although often interrupted by periods of improvement, is the rule, and death occurs a few years after a definite diagnosis has been made. Occasionally life is prolonged for fifteen or twenty years; on the other hand the disease may run a rapid, uninterrupted course, death taking place within two years, occasionally even in a few months.

The duration of life is dependent not only on the occurrence and duration of the periods of remission.

but on the care bestowed on the patient in regard to nursing and general treatment. For life tends to be shortened by the occurrence of bedsores, or of severe bladder disturbance in which there is grave risk of cystitis and pyonephrosis. The appearance of bulbar symptoms is of the gravest significance.

Treatment. Until some antidote is discovered either empirically or through investigations as to the nature of the toxin, we can only do our best to retard the progress of the disease by placing the patient under the best hygienic conditions. To this end, residence in a warm climate, where much sun and good air are attainable, is desirable. Marked improvement often follows rest in bed for a few weeks, and in all cases it is desirable for the patient to remain in bed till noon daily. The general health should be maintained by nutritious food, cod liver oil and occasional tonics. Hydropathic treatment is sometimes of service, while the tendency to contractures and rigidity may be lessened by massage and passive movements. Arsenic, nitrate of silver, and quinine seem to have done good in certain cases, but as yet we do not know of any remedy that appears to influence the course of the disease. Many observers regard arsenic as one of the most useful drugs in the palliative treatment of disseminated sclerosis, and quite recently F. Buzzard, bearing in mind the resemblance between this disease and cerebro-spinal syphilis and the probability that it may be caused by some organism allied to the treponema of syphilis, has suggested a trial of Erhlich's "606" preparation.

In a few cases the author has thought that the administration of suprarenal extract has done good, and two patients testify strongly to its beneficial effects. In other cases the administration of calcium chloride has been followed by improvement. We have also reason to believe that exposure of the spine to the action of the Roentgen rays is worthy of trial.

SECTION V.

The Flaccid and Atrophic Paralyses.

In this variety of paralysis, muscular tonus is lowered; the affected limb feels looser and its range of passive movements is greater than normal. In some cases, as in Landry's paralysis, there may be no detectable wasting of muscular tissue, but in most cases of flaccid paralysis muscular atrophy sooner or later becomes a notable feature. But whether accompanied by muscular

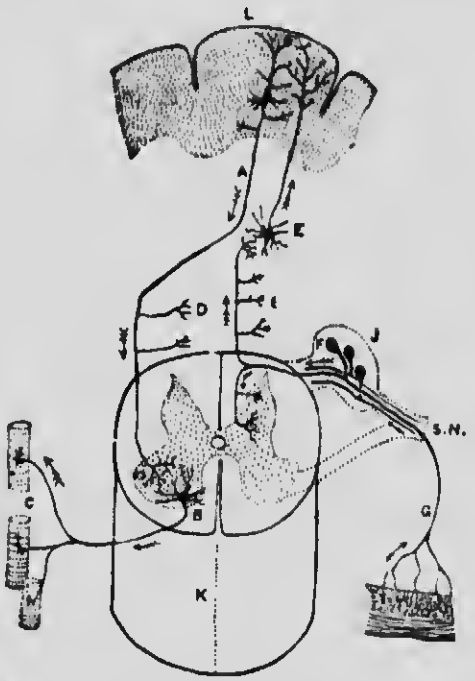


Fig. 48.—Diagram of the connection established by a ganglionic and a motor neuron. (Ramón y Cajal.) A, fibre coming down from a pyramidal cell in the motor area of the cerebral cortex; B, motor cell in grey matter of spinal cord; C, muscle-fibres; D, Collateral branch from the pyramidal fibre; E, cell in the medulla oblongata sending its axon upwards to the cerebral cortex; F, cells in the spinal ganglion; G, peripheral process of ganglionic cell ending in skin; I, collateral branches of central process of ganglionic cell; J, spinal ganglion; K, spinal cord; L, cerebral cortex; S.N., spinal nerve.

atrophy or not, a flaccid paralysis indicates an affection either of the lower motor neurons, or of the muscles themselves; in the former case the paralysis is sometimes termed degenerative spino-neural and bulbo-neural; in the latter primary myopathic. Two apparent exceptions to this rule may be mentioned:—1. Limbs just stricken with paralysis as in hemiplegia may be quite flaccid for a few days after the stroke, although the lesion involves the internal capsule, a part of the upper neurons. 2. A complete transverse lesion of the cord above the lumbar enlargement, abolishing the functions of the upper neurons for the legs, gives rise to a flaccid paraplegia, whereas an incomplete lesion, as in most cases of myelitis, is associated with a spastic paraplegia. But even in these cases the muscular flaccidity may really depend on temporary abolition of the functions of the lower neurons owing to their being completely cut off from all cerebral impulses.

The diagnosis of the position of the lesion—the part of the lower neuron affected—is based partly on the distribution of the paralysis but mainly on the presence or absence of other symptoms. The *character* of the atrophic paralysis offers no distinguishing features; it is essentially the same whether the cell, the fibre or its terminal branches are diseased. Thus atrophy of the small muscles of the hand may be due to disease of the anterior horns, the first dorsal root, or the ulnar nerve: and from an examination of the muscles only it would be impossible to say which of these portions of the lower segment was affected. Complete absence of sensory disturbance would be in favour of the anterior horn, while the presence of anæsthesia along the inner side of the arm or the hand would indicate a root or a nerve affection.

The distribution of paralysis, often helpful in regional diagnosis, cannot alone be relied on. For example an atrophic paralysis of the extensors of the wrists causing double wrist-drop, is characteristic of multiple neuritis but occasionally it is produced by anterior poliomyelitis.

Here again we have to fall back on a consideration of sensory and other symptoms.

The motor path for the muscles supplied by cranial nerves also consists of two parts constituted by upper and lower neurons. In some cases as when the lower neurons of the seventh or the fifth nerve are involved atrophy of the muscles supplied by them can be detected, but in other cases as when the lower neurons of the third, fourth and sixth nerves are diseased the atrophic condition of the eye muscles cannot be observed during life.

All forms of atrophic paralysis being due to disease of the lower neurons or the muscles it is necessary for the student to begin the problem of diagnosis by trying to determine in any particular case whether the muscular atrophy is dependent on an affection of nerve cells, of nerve fibres or of muscular tissue. In the accompanying table—under the headings of cells, roots, nerves and muscles—the chief causes of atrophic paralysis are mentioned. It is useful for reference, but it does not correspond in arrangement with the order in which the several varieties of atrophic paralysis will be described. For example, it is obviously convenient to consider the peripheral nerves together, and lesions of the roots under the diseases producing them.

ATROPHIC PARALYSIS THE CHIEF CONDITION; NO SENSORY DISTURBANCE.

Lesions of motor **cells** in :—

1. Spinal cord.
   Anterior poliomyelitis, acute and chronic.
   Progressive muscular atrophy.
   Amyotrophic lateral sclerosis.
2. Floor of aqueduct of Sylvius.
   Ophthalmoplegia.
3. Floor of fourth ventricle.
   Bulbar paralysis.

Lesions limited to anterior **roots**,– rare.

Lesions of motor **nerves**, cranial and spinal.

Lesions of **muscles**:—
Pseudo-hypertrophic paralysis.
Erb's juvenile form and other types of idiopathic muscular atrophy.

ATROPHIC PARALYSIS ASSOCIATED WITH SENSORY OR OTHER SYMPTOMS.

Lesions of motor **cells** and of **other elements** in the cord.
Various forms of myelitis.
Hæmorrhage.
Tumours.
Syringomyelia.
Lesions of **roots**:—
Meningitis, cerebral or spinal.
Tumours.
Caries of spine.
Lesions of **plexuses**.
Lesions of **mixed nerves**:—
Single nerves, cranial or spinal.
Multiple neuritis.

CHAPTER I.

LESIONS OF THE SPINAL NERVES.

The commonest cause of a lesion affecting a single nerve or a plexus of nerves is injury; the commonest cause of multiple neuritis is some poison in the blood. Occasionally, however, the effects of a toxin are limited to a single nerve, thus alcohol which often tends to a wide-spread neuritis may produce a mono-neuritis only. Similarly in regard to the poisons of gout, rheumatism, diabetes, enteric fever and other diseases.

In most cases of toxic neuritis the nerve fibres are primarily affected and present changes similar to those of Wallerian degeneration; this is the *parenchymatous* variety of neuritis.

In most cases of traumatic neuritis the nerve fibres are secondarily affected in consequence of an inflammatory overgrowth of the connective tissue which supports and surrounds them; this is the *interstitial* variety of neuritis. Occasionally interstitial neuritis appears to be set up by exposure to cold and wet, or by gout or rheumatism.

Local lesions of the peripheral nerves are most frequently caused by wounds, contusions, pressure, muscular strains, fractures or dislocations of bones, tumours or extension of inflammation (syphilitic or other) from the surrounding tissues and organs. Accidental injuries of nerves are most common about the wrist, the ulnar, median or radial nerve being completely or partially severed by a knife, broken piece of glass or other sharp object.

The **Symptoms** produced by a complete lesion of a mixed peripheral nerve may be motor, sensory, vasomotor and trophic. If, however, the lesion is incomplete, as for example that due to compression, the sensory fibres of the nerves suffer much less than the motor fibres and paralysis may be the only manifestation of the injury, although as a rule a careful examination will reveal slight changes in epicritic sensibility.

Interruption of impulses along motor nerve fibres is immediately followed by paralysis, partial or complete, according to the intensity of the lesion. The affected muscles are flaccid, they become atrophied and give the electrical reactions of degeneration. Loss of muscular tone is shown not only by flabbiness of the muscles and their lack of resistance to stretching, but by the loss of the tendon reflexes.

Proliferation of connective tissue accompanies atrophy of the muscular fibres, and if the latter do not soon regenerate they are gradually replaced by the new connective tissue which may lead to the development of contractures and deformities. Very rarely muscular spasms and twitchings occur as early symptoms: occasionally they may be due to irritation of motor fibres,

but usually they are reflex in origin being produced by excitation of sensory fibres.

Sensory symptoms vary much in character and intensity according to the nature and severity of the lesion; for example, when the nerve is being slowly compressed they are insignificant or absent, when it is inflamed they are often severe. In the latter case intense pain may radiate from the region of the affected nerve into other nerve territories. It is of a tearing, boring or burning character, is often continuous, though there may be remissions and paroxysmal exacerbations which are apt to be worse at night. Frequently there is hyperalgesia of the whole area of distribution of the nerve. The sufferings of the patient are increased by movement of the limb and by pressure over the affected nerve. Paræsthesia, in the form of numbness, tingling, or 'pins and needles,' is generally associated with some degree of anæsthesia; it occurs more frequently in cases of toxic degenerations than in traumatic lesions. Loss of sensation which is prominent in cases of acute neuritis is usually less extensive than the anatomical skin distribution of the nerve; this is due to the overlap which exists within adjacent nerve areas.

The relations between the areas of epicritic and protopathic loss vary according to the situation of the lesion whether in the nerve, the plexus, or the nerve roots.

In the first case the area of protopathic loss is less than that of epicritic loss, and between the margins of the two areas, painful stimuli cause an unusual amount of pain which is widely diffused. In the second case it is almost equal to it, whilst in the third case the area of loss of protopathic sensibility may be larger than that of the epicritic. If the cutaneous branches of a nerve are alone involved there is no loss of sensation to contact and deep pressure, and the sense of position and of movement of the part are retained.

In addition to muscular atrophy there may be trophic changes in the skin, nails, hairs, bones and joints.

These changes are more likely to be prominent when the nerve fibres are severely affected, and in areas of total analgesia. The skin becomes thin, smooth, glossy, inelastic and tightly stretched over the affected part. Blisters and bullæ may develop and lead to ulceration; whitlows and gangrene also occur. The nails are retarded in growth and become brittle, corrugated and furrowed. Occasionally extensive nerve lesions lead to fragility and atrophy of the bones and to effusions into the joints of the affected limb. Vaso-motor and secretory phenomena are manifested by changes in the colour, secretions and temperature of the paralysed part. During the first few days after the lesion there may be redness of the skin, hyperidrosis and a local rise of temperature; at a later period the skin may be pale or blue, and dry, and the local temperature subnormal.

Course and Prognosis. The progress of peripheral nerve paralysis depends upon the nature and severity of the lesion, that is upon the likelihood of regeneration of the damaged or divided fibres.

In slight cases of neuritis, such as those produced by contusion or pressure, complete recovery may take place within a few weeks. In severe cases, in which the muscles are atrophied and give the reaction of degeneration, many months may elapse before sensation and motor power are regained.

If the nerve is completely divided and the separated ends are subsequently sutured, restoration of sensation and motor power may gradually take place. After two or three months there is a gradual diminution in the area of total analgesia, and in about six months all forms of protopathic sensibility are regained. At this time epicritic anæsthesia is still complete and radiation of severe pain is produced by a pin-prick or other painful stimulus. The return of epicritic sensibility rarely begins earlier than six months and is seldom complete until a year or eighteen months after the physiological continuity of the nerve fibres has been established. It has been pointed out by Head and

Sherren that restoration of sensation does not take place apart from regeneration and union with the central nervous system; there is no gradual encroachment on the anæsthetic area by the surrounding healthy nerves.

The time required for the restoration of the motor functions varies with the distance of the lesion from the periphery of the limb. Sherren says that after division of a nerve at the wrist perfect power may be regained within a year, but if at the elbow or in the plexus, not for two years.

Diagnosis. Difficulties occasionally arise in the diagnosis of lesions of the peripheral nerves from those of the spinal cord or its nerve roots; from arthritic muscular atrophy, from hysterical conditions and from Volkmann's ischæmic contraction. Only the two latter affections will be here noticed.

Hysterical paralysis with contractures may follow any form of injury. As a rule paralysis and anæsthesia are both present but either may be found alone. The peculiarities of the anæsthesia are that all forms of sensibility, including the perception of pressure, are *equally* affected, and over the whole area of anæsthesia, the upper limit of which often surrounds the limb at the level of a joint. The weakened muscles are flaccid; their electrical reactions are normal. When contractures are present they involve the muscles on both sides of the limb, thus an attempt to further flex a rigidly flexed elbow is resisted by contraction of the triceps. In association with hysterical paralysis there may be contraction of the field of vision.

Volkmann's ischæmic contracture usually results from the injurious pressure of tightly-applied bandages or splints. The distal portion of the limb becomes painful and swollen, and after a time its muscles get firm and rigid, the result being a limitation of passive movement and some loss of voluntary power. There is no anæsthesia, and the electrical reactions of the muscles are normal unless the condition is complicated by injury to the nerves supplying the affected part.

Treatment. This will largely depend on the nature and degree of the lesion. When a nerve has been divided its two ends must be sutured together as soon as possible. In cases of toxic neuritis and indeed in all cases of nerve injury, whether operation has been deemed advisable or not, the regeneration of the nerve and the restoration of its functions are materially aided by the use of local warmth, massage and electricity. Massage of the paralysed muscles, systematically and carefully performed, is of the greatest importance. Electrical stimulation of the muscles is also useful, the galvanic current being the most effective when the muscles do not react to the faradic current.

Contractures and deformities may be prevented by passive movements of the paralysed part and by the application of splints, with the object of preventing the overaction of muscles which act in opposition to those which are paralysed; for example if the extensors of the wrist and fingers are paralysed, the joints should be kept extended by a splint placed along the forearm and hand.

It is also necessary to protect the part from accidental injuries or other sources of irritation which might lead to the development of trophic changes.

The Phrenic Nerve.

The phrenic nerve may be implicated in disease or fracture of the spine, in lesions—hæmorrhage, syphilitic meningitis, tumours—of the third and fourth cervical segments or of the third and fourth cervical roots. Rarely it is injured or compressed by wounds, aneurysms or tumours in the neck or thorax. Bilateral paralysis may occur in alcoholic, diphtheritic and other forms of multiple neuritis.

When both phrenics are affected the diaphragm may be completely paralysed. This is indicated by a hollowing of the epigastrium which is drawn in instead of being protruded during each inspiration, whilst during expiration it advances instead of receding as in normal breathing. Overaction of the lower intercostals is also

frequently observed, the margins of the thorax moving far more than in ordinary breathing. The impaired diaphragmatic movement can be demonstrated by an *x*-ray examination.

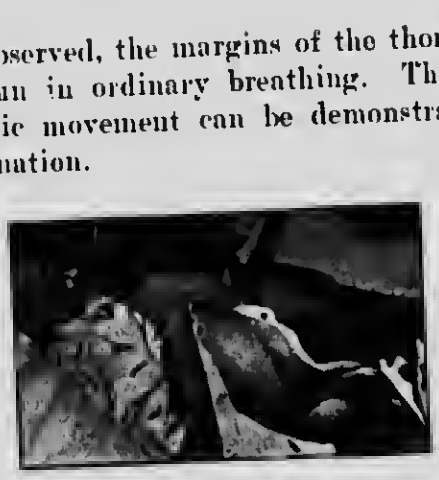


Fig. 49.—Early total paralysis of diaphragm. (Reynolds.)

In unilateral paralysis of the diaphragm these signs are only present on one side and are difficult to recognise; the condition causes little discomfort.

Respiration even in complete bilateral paralysis is not necessarily accelerated during rest, but dyspnœa begins with the slightest exertion and any concomitant affection of the lungs is rendered much more serious.

The Long Thoracic Nerve.

Paralysis of this nerve may be caused by wounds in the axilla or the supraclavicular fossa, by carrying heavy weights on the shoulder, or occasionally by excessive muscular exertion with the arm raised, probably owing to forcible contraction of the scalenus medius muscle through which the nerve passes. The condition occurs more frequently in men than in women, and on the right than on the left side. Sometimes the paralysis appears to be caused by a toxic neuritis as from diphtheria or influenza; sometimes no cause can be discovered.

The resulting paralysis of the serratus magnus, which may be preceded by pains in the neck and about the shoulder blade, is shown as follows:—When the arm is hanging by the side of the body the scapula is slightly raised and its lower angle is tilted towards the vertebral column. The patient has a difficulty in abducting the

arm above the horizontal level, and during the movement the scapula still more approaches the spine and also projects from the trunk. If he holds the arm horizontally forwards the "winging" of the bone becomes more marked and between it and the chest wall there is a deep fossa into which the observer's hand can be placed, so as to feel the inner surface of the scapula.

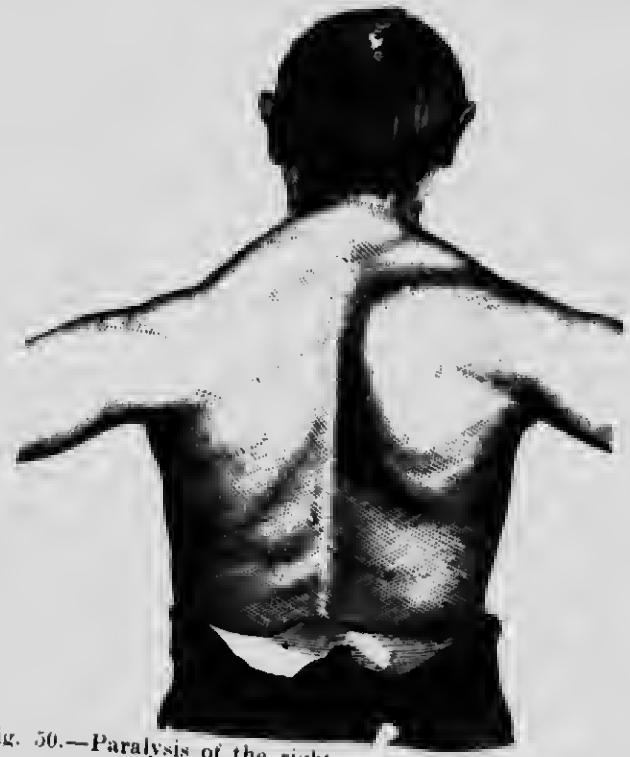


Fig. 50.—Paralysis of the right serratus magnus. (Ross.)

The arm can only be raised to a vertical position when the observer pushes the angle of the scapula outwards and fixes it in that position. If, with the arm thus raised, a deep inspiration is taken, the digitations of the serratus magnus are seen to be less marked on the paralysed than the healthy side.

Absolute rest to the arm should be ordered, the elbow being supported in a sling in order to raise the shoulder. The usual treatment by massage and galvanism must be carried out.

CHAPTER II.

PARALYSIS OF THE MUSCLES SUPPLIED BY THE BRACHIAL PLEXUS.

The brachial plexus is formed by the anterior divisions of the four lower cervical nerves, the first dorsal nerve and a part of the fourth cervical; it occupies the posterior triangle of the neck and the subclavicular fossa. Lesions may involve the whole of the plexus, but usually they affect only a portion of it. As a rule they are traumatic in origin; thus the plexus may be injured by stabs in the neck, by dislocation of the shoulder, by fracture of the clavicle or by the callus which forms after the fracture. Some of the fibres of the plexus may be torn by forcible dragging of the arm upwards and backwards as in clutching at some object when in the act of falling, or by a similar malposition of the arm during surgical operations upon the breast, or during the process of delivery "obstetrical paralysis."

Sometimes the plexus is compressed by tumours, a subclavian aneurysm, or a cervical rib. Occasionally its fibres are affected by a toxic or an infective neuritis.

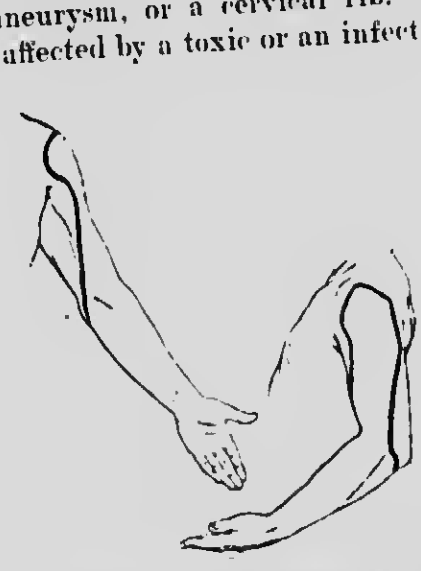


Fig. 51.—The distribution of anæsthesia to all forms of cutaneous stimuli after rupture of the cords of the brachial plexus. (Head.)

Symptoms. In a complete lesion of the whole plexus all the muscles of the arm are paralysed with the exception of the levator anguli scapulæ, the rhomboids and the serratus magnus, and in some cases the clavicular portion of the pectoralis major. The affected muscles are flaccid and undergo a progressive atrophy. There is anæsthesia of the limb with the exception of the tip of the shoulder, which is supplied by the cervical plexus, and the axillary aspect of the arm, which is supplied by the intercosto-humeral nerve. Vaso-motor and trophic changes may ensue, and sometimes arthritis of the elbow joint. As a rule the branch of the first dorsal nerve to the sympathetic is involved, when the pupil becomes small and the palpebral fissure narrowed. But paralysis more limited in distribution is much commoner, presenting many variations in different cases. Two types call for special description: they may occur as primary palsies, or as residual palsies of a total plexus paralysis.

The upper arm type or Erb-Duchenne paralysis.

In this variety, which is usually the result of a fall on the shoulder, there is paralysis of the deltoid, biceps, brachialis anticus and supinator longus, and sometimes of the supinator brevis. These muscles are supplied by the fifth cervical root, the fibres of which may be injured either before or after joining the sixth root to form the upper trunk of the plexus. When both these roots are implicated, paralysis of the clavicular portion of the pectoralis major and of the subscapularis may also result. The arm and forearm hang close to the side, with the forearm extended and pronated. The patient is unable to abduct the arm, to flex the elbow and to supinate the hand. When the subscapularis is affected, the power of inward rotation at the shoulder is impaired, and if atrophy of the muscle ensues, the movements of the scapula against the ribs may be accompanied by a grating noise.

As a rule there is little or no anæsthesia; occasionally epicritic sensibility is impaired on the outer aspect of the arm and forearm.

The lower arm type or Klumpke's paralysis.

This name is given to the atrophic paralysis which results from a lesion of the eighth cervical and first dorsal roots either before or after their union to form the lower trunk of the plexus. The muscles affected are the intrinsic muscles of the hand and the flexors of the wrist and fingers. The hand assumes a claw-like shape and its grasping power is diminished or lost; in the attempt to grasp, the hand is drawn back by the unopposed action of the extensors of the wrist. Numbness or distinct anæsthesia affects the inner side of the hand and forearm. In some cases the oculo-pupillary fibres are implicated, when myosis and narrowing of the palpebral fissure will be observed.

Fig. 52.—The area of anæsthesia following a lesion of the inner cord of the brachial plexus. (Head.)

The **prognosis** of injuries to the brachial plexus is worse than that of injuries to peripheral nerves, and it is worse in cases of supra-clavicular than in cases of infra-clavicular lesions. These differences are mainly due to differences in causation; laceration, a common plexus-lesion, being more serious than compression, a common cause of damage to a peripheral nerve.

The **treatment** of plexus-lesions is mainly surgical and operation should not be delayed after the complete reaction of degeneration has been detected in the

paralysed muscles. Both before and after operation, and also in cases of toxic neuritis, massage and electricity are serviceable in maintaining the nutrition of the muscles.

Cervical Ribs.

Supernumerary ribs are sometimes developed in connection with the seventh cervical vertebra and very rarely with the sixth. They may be rudimentary, consisting of a mere elongation of the transverse process of the vertebra; or more or less complete, fusing with the first dorsal rib, or articulating with the sternum. If the rib is short and runs directly outwards, a prominence may be felt in the neck, but pressure symptoms are rare. If it is long, and curves round the posterior triangle towards the sternum, symptoms may be caused by pressure on the brachial plexus and the subclavian artery, both of which structures pass over the rib.

The presence of a cervical rib is attended by symptoms, only in about five to ten per cent. of all cases. As a rule the symptoms are unilateral although the malformation is usually bilateral.

The symptoms occur more frequently in females than in males. Generally they appear in early adult life; in a few cases not until late in life. Their occurrence in youth is probably related to the completion of ossification, in old age to changes in the shape of the spine and thorax.

The nervous symptoms, which affect the right arm more commonly than the left, correspond to those produced by pressure upon the lower trunk of the plexus; namely pain and anæsthesia along the inner borders of the forearm and hand, together with paralysis and atrophy of the intrinsic muscles of the hand, and sometimes of the flexors of the fingers and wrist. Definite paralysis however is much less common than sensory disturbance, and in many cases the only symptom is pain along the ulnar borders of the forearm and hand, and to a less degree in the shoulders or neck.

The woll-nigh constant absence of symptoms pointing to a lesion of the sympathetic fibres, namely narrowing of the palpebral fissure and contraction of the pupil, indicates that the lower trunk of tho plexus, rather than its constituent roots, is involved.

Compression of the subclavian artery is shown by a weak radial pulse, the strength of which is increased by raising the arm, and so removing the sharp curve in the vessel where it crosses the rib.

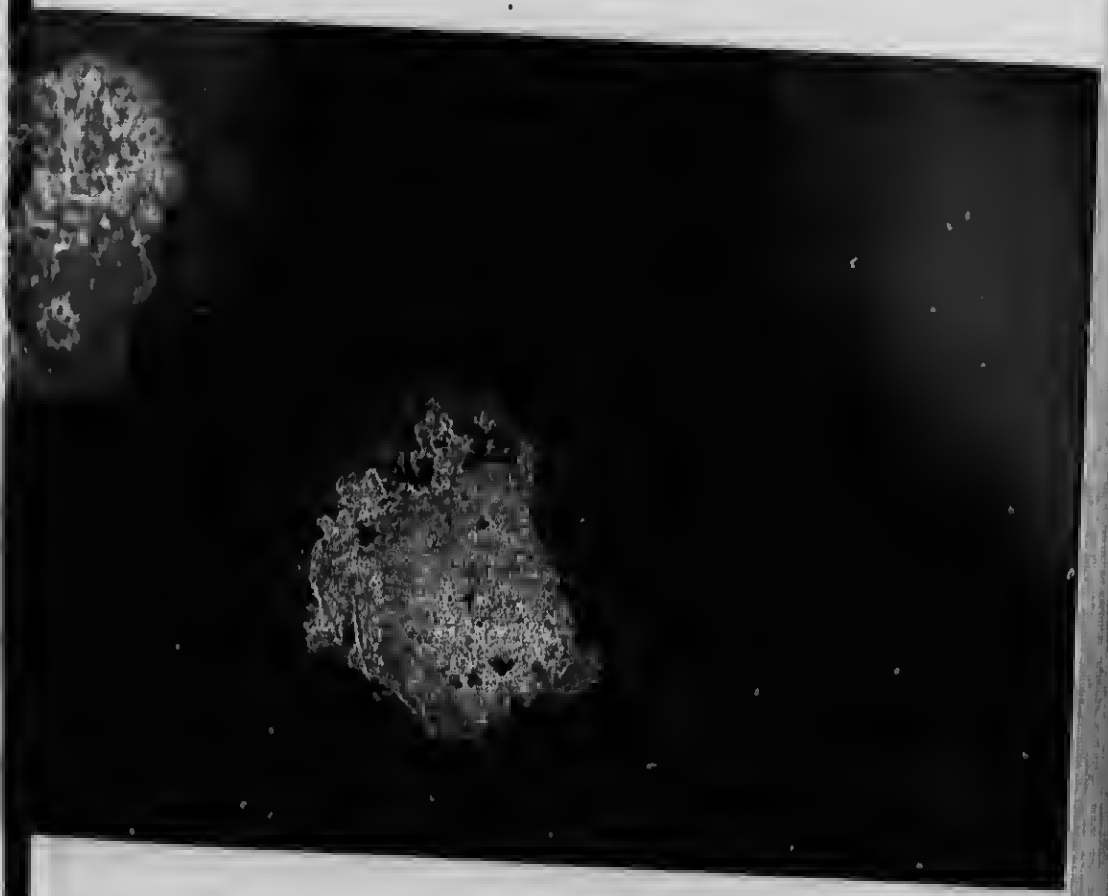
Fig. 53.—Photograph of hands in a case of cervical rib showing atrophy of small muscles of right hand.

Diagnosis. The presence of a cervical rib has been often overlooked; it is therefore of great importance to think of the possibility of this abnormality in every case of atrophic paralysis affecting the muscles supplied by the first dorsal root alone, or along with those supplied by the eighth cervical root, and especially if the paralysis is associated with inequality of the radial pulses.

It must be remembered that sometimes both hands are affected and that occasionally sensory symptoms are inconspicuous or even absent. In such cases, progressive muscular atrophy would naturally first suggest itself; but this diagnosis must not be made until the patient's neck has undergone a radiographic examination, by which the presence of abnormal ribs would be easily revealed.

The treatment consists in the removal of the super-

PLATE III.



Radiogram showing cervical rib on right side; the same case as fig. 53.

humerary rib, an operation which always relieves the pain and frequently leads to restoration of power in the wasted muscles.

The Circumflex Nerve.

This nerve supplies the deltoid and the teres minor muscles and sends branches to the shoulder joint. It may be paralysed by injuries to the shoulder especially when dislocation of the joint occurs. Occasionally an isolated neuritis of this nerve is met with in diabetes, lead poisoning, typhoid fever, after exposure to cold, or as a result of syphilis. The movements of elevation and outward rotation of the arm are paralysed; sensory disturbances may be represented by pain about the shoulder and anæsthesia over the insertion of the deltoid. When this muscle becomes atrophied the shoulder joint may become so loose that a deep groove can be felt between the head of the humerus and the glenoid cavity; sometimes however adhesions form in the joint.

Bilateral paralysis of the deltoid has been known to follow sleeping on the back, with the hands clasped behind the head.

The *Supra-Scapular Nerve*, which supplies the supra- and infra-spinatus muscles, is often involved along with the circumflex in injuries at the shoulder. Paralysis of the supra-spinatus impairs the power of raising the arm, paralysis of the infra-spinatus that of outward rotation of the humerus.

The Musculo-Spinal Nerve.

This nerve is most commonly injured as it winds round the humerus, sometimes in consequence of fracture of the bone, but more frequently as a result of compression between the bone and some hard substance, as when a drunken person passes the night in a chair with his arm hanging over the back of it.

In such a case there is paralysis of the extensors of the wrist, the long extensors of the thumb and fingers, and the supinators. Hence the wrist is dropped, the

hand is pronated, the fingers are flexed and the thumb is flexed and also adducted. The patient is unable to raise the hand or extend the fingers at the metacarpo-phalangeal joints, but he can straighten the fingers at the inter-phalangeal joints by means of the unaffected interossei. Supination of the forearm is impossible, unless the elbow is flexed when the movement can be performed by the biceps. Owing to the flexed position of the hand the grasp is weak but if the observer hyper-extends the wrist, the flexors are placed at a better advantage and grasping power is much improved.

When the nerve is compressed in the axilla as by fracture or dislocation of the humerus, or by the use of unsuitable crutches, the triceps becomes paralysed, when power to extend the elbow is lost.

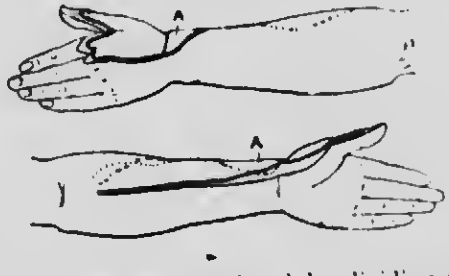


Fig. 54.—Loss of sensation produced by dividing the radial and external cutaneous nerves near the elbow. The thick line bounds the area insensitive to prick, the thin line that to cotton-wool. Both lines are dotted wherever the borders of the loss were not sharply defined. The triangle marked A was insensitive to prick, but sensitive to cotton-wool. (Head.)

The sensory symptoms vary with the position of the lesion. When this is in the upper arm above the origin of the external cutaneous branches, all forms of sensibility—epicritic, protopathic, and deep—are impaired or lost over the back and outer side of the thumb, and over the back of the hand opposite the first and second fingers. When the lesion involves the musculo-spinal nerve below the origin of the external cutaneous branches, anæsthesia is limited to the distribution of the radial branch, namely the outer side of the thumb, and to epicritic sensibility: frequently no sensory defect can be detected.

The Ulnar Nerve.

This nerve may be injured in any part of its course. At the elbow it is often damaged by fractures or other injuries; occasionally it is dislocated from its groove on the posterior surface of the internal condyle. At the wrist it is frequently divided by accidental wounds. A rheumatic, gouty, or syphilitic neuritis sometimes affects the nerve. It occasionally suffers in consequence of certain occupations, such as cigarette-making, and glass-working, which entail constant movements at the elbow.

When the lesion is situated at or above the elbow flexion of the wrist is feebly performed and is accompanied by deviation of the hand to the radial side. Owing to paralysis of the interossei and lumbricales there is inability to abduct and adduct the fingers, to flex their metacarpo-phalangeal joints or fully to extend their other joints. The latter defects are least marked in the index and middle fingers, because their lumbricales are supplied by the median nerve. The patient is also unable to flex the distal phalanges of the little and ring fingers; indeed all the movements of the little finger are lost, and so also is true adduction of the thumb.

The paralysed muscles gradually become atrophied; after a time the hypothenar eminence disappears, and deep furrows appear between the metacarpal bones, whilst little more than a fold of skin can be felt between the thumb and index finger.

When the lesion is situated near the wrist the fibres to the flexor digitorum profundus escape, and then owing to the unopposed action of this muscle and of the extensors of the fingers, the hand assumes a claw-like shape, the first phalanges being hyperextended and the last two flexed. This deformity, in severe and protracted cases, may be accompanied by backward dislocation of the first phalanges and forward dislocation of the other phalanges.

Cutaneous anæsthesia may be found over the palmar and dorsal aspects of the little and adjacent half of the ring fingers, and on the ulnar side of the hand: epicritic

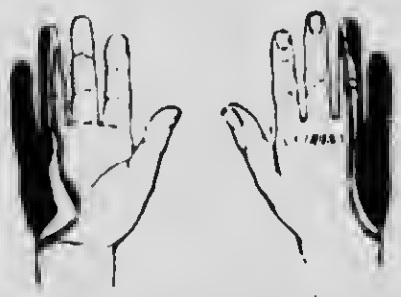


Fig. 55.—The area of anæsthesia following complete division of the ulnar nerve. The black is the area of protopathic and epicritic loss, the zone between the black area and the continuous line is the area of epicritic loss. (Head.)

is usually in excess of protopathic loss; deep sensibility is lost or retained according as the lesion is above or below the origin of the dorsal cutaneous branch.

The Median Nerve.

The median nerve may be involved in fractures of the bones of the arm or forearm, but more frequently it is injured by accidental wounds in the region of the wrist. Sometimes it is affected as a result of various occupations, as in carpenters, and professional golfers.

Fig. 56.—The area of sensory loss after complete division of the median nerve near the elbow. (Head.)

When the lesion is at, or above the elbow, the hand is slightly supinated and cannot be pronated; in trying to

compensate for this defect the patient rotates the shoulder inwards. Flexion of the wrist is feeble and incomplete and is accompanied by adduction of the hand owing to the unopposed action of the flexor carpi ulnaris. The patient is unable to abduct, oppose or perform any of the more delicate movements of the thumb, which is kept in an extended and adducted position. He is also unable to flex the proximal interphalangeal joints of the fingers and the distal joints of

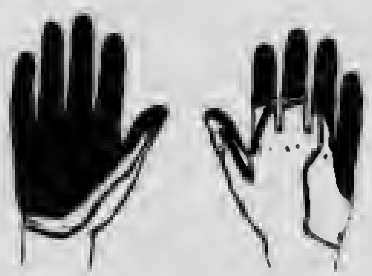


Fig. 57. Showing the loss of sensation produced by complete division of both median and ulnar nerves. The total area is contained within the continuous black line. (Head.)

the index and middle fingers. The interossei being unaffected, the metacarpo-phalangeal joints can be flexed whilst the unopposed action of these muscles on the distal joints tends to their hyperextension, and even to their subluxation.

When the lesion is at the wrist joint, paralysis affects only the muscles of the thumb and the two outer lumbrical muscles. Cutaneous anæsthesia, which is variable in character, intensity and distribution, may affect the palmar surfaces of the thumb, index, middle and adjacent half of the ring fingers, the radial portion of the palm, the dorsal surface of the two distal phalanges of the first two fingers and the radial side of the ring finger. The area of protopathic loss is usually less than that of epicritic; deep sensibility is retained.

CHAPTER III.

PARALYSIS OF THE MUSCLES SUPPLIED BY THE LUMBAR AND SACRAL PLEXUSES.

With the exception of the sciatic nerve, the nerves of the lower limb are less commonly injured and are less subject to disease than those of the upper limb.

The *lumbar plexus* supplies the cremaster, the flexors and adductors of the hip joint and the extensors of the knee, while its sensory branches receive impressions from the skin of the lowest part of the abdomen, the front and sides of the thigh and the inner side of the leg and foot.

Isolated paralysis of this plexus is very rare: it may be damaged by tumours of the retro-peritoneal lumbar glands, the kidney or the cæcum, or by a psoas abscess. Very rarely it has suffered from neuritis, which may have ascended from the sacral plexus along the lumbo-sacral cord.

The Anterior Crural Nerve.

This nerve has been injured by fractures of the pelvis or of the femur, by wounds in the groin or thigh, and by manipulations for reducing congenital dislocation of the hip; other causes are psoas abscess and tumours in the pelvis.

The most prominent symptom is loss of power to extend the knee, together with loss of the knee-jerk. The quadriceps becomes wasted; an atrophic paralysis also affects the sartorius and pectineus but does not give rise to any noticeable symptoms. If the lesion is high enough within the pelvis, the iliacus is sometimes involved; the psoas usually escapes, hence power to flex the hip is impaired but not abolished.

Sensory disorders extend over the lower two-thirds of the thigh, on its anterior and inner aspects, and over the inner side of the leg and foot.

The Obturator Nerve.

An isolated injury of this nerve rarely occurs; it may be damaged during a protracted labour, by pelvic growths or by an obturator hernia.

The power to adduct the thigh and to rotate the hip outwards is impaired; it is difficult or impossible for the patient to cross the affected leg over the other leg. There is no definite loss of cutaneous sensibility.

The *sacral plexus* innervates the extensors and rotators of the hip, the flexors of the knee and the foot muscles; its sensory fibres supply the skin of the gluteal region, the back of the thigh, the back and outer side of the leg and most of the foot.

Paralysis of the whole or part of the plexus may be caused by tumours in the pelvis or by inflammation, or by caries of the vertebræ.

As a rule the symptoms are those of a partial affection of the sciatic nerve; if the upper roots of the plexus are involved, the gluteal muscles and the outward rotators of the hip may also be paralysed, whilst if the lower roots are involved there may be anæsthesia in the distribution of the small sciatic nerve.

The Great Sciatic Nerve.

Apart from primary neuritis the sciatic nerve may be implicated in pelvic growths or it may be injured by wounds, dislocation of the hip, and fractures of the pelvis or the upper end of the femur. It suffers from gunshot injuries more frequently than any other nerve. Of its two main divisions, the external popliteal is more prone to suffer than the internal popliteal.

If the lesion of the great sciatic is near the notch, all the muscles of the leg and foot are paralysed, as well as the flexors of the knee. The patient is unable to offer any resistance to extension of the leg and finds that his power to walk is much impaired. There is anæsthesia on the outer side of the leg, and the greater part of the foot.

The External Popliteal Nerve.

This nerve may be injured as it passes round the fibula by fracture of the neck of this bone, by wounds, or by an unduly tight bandage.

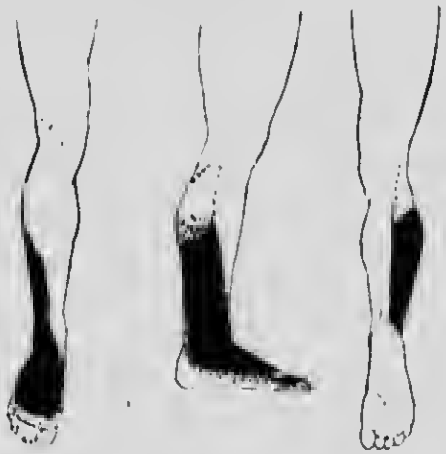


Fig. 58.—Areas of protopathic and epicritic loss after division of the external popliteal nerve. (Head.)

The symptoms produced by such an injury are:—Paralysis of the tibialis anticus, the extensors of the toes and the peronei muscles. Hence the foot is dropped and inverted and the toes are flexed. Degenerative atrophy of the affected muscles ensues and after a time, if the internal popliteal nerve is uninjured, contracture of the calf muscles leads to talipes equinus, and contracture of the interossei to flexion of the metatarso-phalangeal joints. Anæsthesia in the outer side of the leg and the dorsum of the foot is sometimes present.

The Internal Popliteal Nerve.

Owing to its deep position behind the knee this nerve is rarely injured. Should its functions be abolished, the popliteus, the calf muscles, the tibialis posticus, the flexors of the toes and the intrinsic muscles of the sole become paralysed.

The results are: That inward rotation of the flexed knee may be difficult, and that the patient is unable to stand on tip-toe, to extend or invert his ankle and to flex his toes. The paralysis of the interossei leads to a claw-like deformity of the foot, whilst contractures of

Fig. 59. Areas of protopathic and epicritic loss after division of the posterior tibial nerve. (Head.)

the muscles supplied by the unaffected external popliteal nerve produce talipes calcaneo-valgus. The sensory loss, which varies in different cases, may affect the sole and the outer side of the foot. Deep insensibility is not affected.

Lesions of the Cauda Equina and the Conus Terminalis.

The extreme lower end of the spinal cord, conical in shape, is called the conus medullaris or cones terminalis. The base of the conus has been artificially fixed at the level of the interval between the first and second lumbar vertebra and between the origin of the second and third sacral nerve. Surrounding the conus and arising from it are the nerve roots from the second lumbar segment downwards. Collectively these roots constitute the cauda equina which gradually becomes smaller in size as the successive roots part from it to penetrate the dura mater and emerge from their corresponding intervertebral foramina. The arrangement of these roots and their relation to the vertebræ are shown in fig. 60. It will be

seen that the upper lumbar roots are situated peripherally and run a short course within the canal; whilst the lower sacral, situated mesially, have a long course.

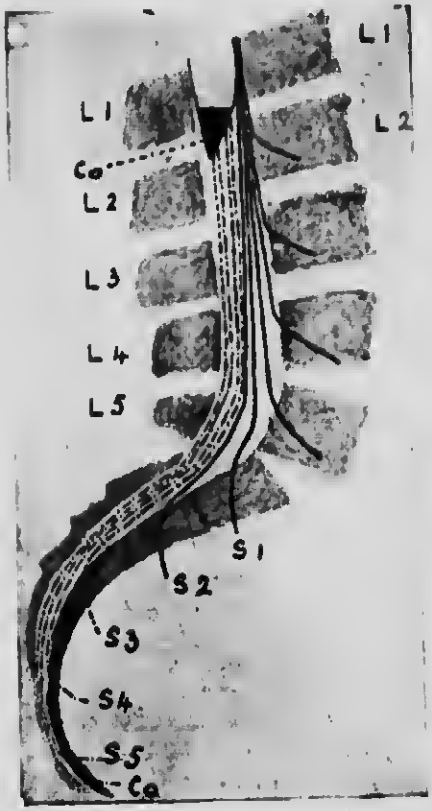


Fig. 60.—Vertical section of lumbar vertebræ and sacrum, showing termination of spinal cord, and nerves of cauda equina in vertebral canal. Deep black = termination of cord. CO = conus terminalis. L 1 to 5 = lumbar vertebræ. S 1 to 5 = sacral roots. Broken lines = nerves from conus, S 3 to 5, and coccygeal nerves = CO.

Etiology. Lesions of the conus or the cauda equina may be caused by: Fractures or dislocations of the vertebræ; hæmorrhage as a result of injury; penetrating wounds; meningitis, especially the gummatous variety; or tumours arising from the bones, membranes or nerve roots—neuroma, sarcoma, glioma, endothelioma, lymphangioma, hydatid cyst.

Symptoms. The clinical picture presented by disease of the *cauda equina* varies much in different cases, according to the nature and situation of the lesion. In cases of injury the onset is sudden or rapid, whilst in cases of tumour the development of symptoms is gradual.

When the lesion is situated at the level of the second lumbar vertebra and involves all the nerve roots of the cauda equina, the muscles of the lower limbs are paralysed and wasted and reveal changes in their electrical excitability; there is anæsthesia of the lower limbs below the groin, which affects also the buttocks, the perineum and the external genital organs; the plantar reflexes are lost and so also are the knee and the tendo-Achilles jerks; the bladder and rectum are paralysed and sexual power is lost.

Fig. 61.—Photograph of a case of tumour, involving the cauda equina, showing marked atrophy of the leg muscles.

Such complete and extensive paralysis is rare; as a rule the symptoms are related to the distribution of the sacral roots, or to these in conjunction with one or two of the lower lumbar roots. Thus it is common to find an atrophic paralysis of the muscles below the knee, wasted buttocks, and anæsthesia of the skin supplied by the first, second, and third sacral and fifth lumbar roots.

When the cauda equina is damaged below the exit of the second sacral nerves, muscular power in the leg muscles is retained, paralysis being limited to the bladder and rectum with loss of sexual power: there

may be a saddle-shaped area of anæsthesia in the gluteal region and anæsthesia of the perineum, scrotum and urethra; the knee-jerks and the plantar reflexes are preserved.

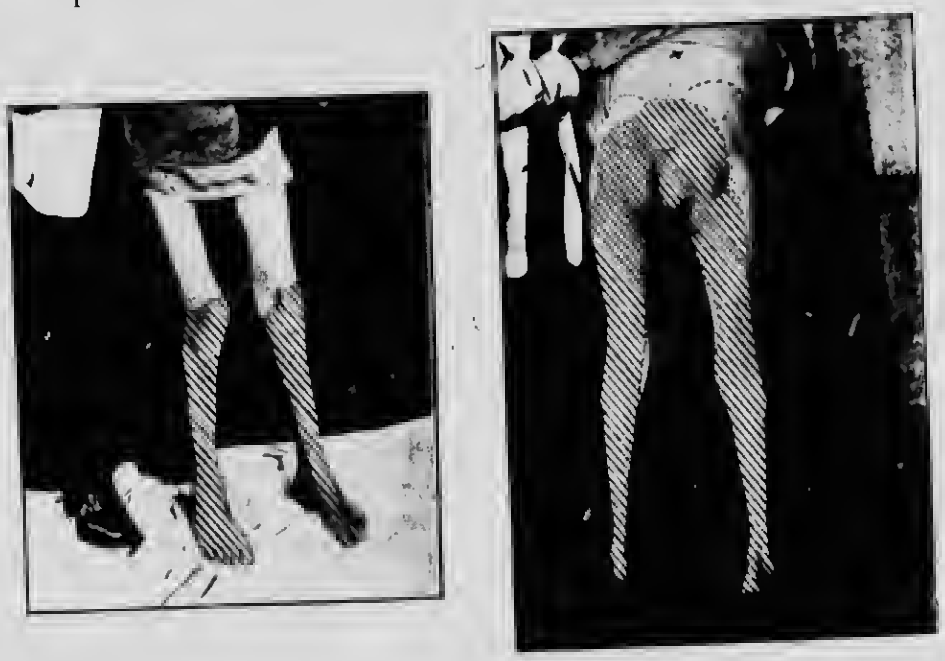


Fig. 62.—Photographs showing diminished sensation of the areas supplied by the 4th and 5th lumbar, and the 1st, 2nd and 3rd sacral roots, in a case of fracture of the second lumbar vertebra.

When the lesion is limited to a lower level, the functions of the genital organs, or those of the bladder and rectum may escape; and if the coccygeal nerves are alone implicated, the only symptoms may be paralysis of the levator ani, and anæsthesia about the anus and perineum.

Limitation of symptoms to parts innervated by the sacral roots cannot be always explained by the position of the lesion, for it is found that when there is only partial compression of the cauda equina, the most mesially placed roots, namely the sacral, are those which suffer the most. Thus in some cases of injury at the level of the second lumbar vertebra, signs of implications

of the third, fourth, and fifth lumbar roots may be absent or insignificant.

Furthermore a want of symmetry in distribution of the symptoms affecting the lower limbs is frequent. For example, in one of my cases, which came under observation fifteen months after a fracture of the third and fourth lumbar vertebræ, the left leg was nearly normal as regards both power and sensation, whilst the right leg gave evidence of damage to the fifth lumbar and first and second sacral roots, there being anæsthesia over the outer aspect of the leg and an atrophic paralysis of the glutei and the ankle and foot muscles, together with slight weakness of the hamstrings; the knee-jerk was present, the ankle-jerk and the plantar reflex were abolished. It is remarkable that in this case the bladder and the rectum were not affected.

It is usually stated that the *conus terminalis* contains centres for the sensory innervation of the bladder, urethra, penis, scrotum, anus and lower part of the rectum; for erection of the penis and ejaculation, and for discharge of the contents of the bladder and rectum. Müller however believes that the automatic centres for the bladder, rectum and genital organs are situated in the pelvic sympathetic ganglia. In support of this view Bálint and Benedict state that lesions of the conus cause loss of voluntary control over the bladder and rectum, but not paralytic incontinence.

The symptoms that have been observed in cases of disease of the conus are similar to those which occur in lesions of the cauda equina situated below the exit of the second sacral nerves; namely, paralysis of the bladder and rectum; loss of sexual power; anæsthesia of the urethra, bladder, lower part of the rectum, perineum, scrotum (testicular tenderness being preserved) and of a saddle-shaped area in the gluteal region. The legs are free from paralysis; the deep and superficial reflexes are normal.

If a lesion in the conus should extend upwards in the cord there may be paralysis of the glutei and the

muscles below the knee, with anæsthesia down the centre of the posterior aspects of the lower limbs, and over the soles of the feet, and with loss of the ankle-jerks and plantar reflexes.

Diagnosis. It is important to distinguish between affections of the lower end of the cord and the cauda equina because surgical intervention, while useless in the former class, may be beneficial in the latter.

In favour of a lesion of the lower end of the cord are: A sudden or rapid onset of symptoms; signs of injury to the last dorsal or upper two lumbar vertebræ, as revealed by inspection, palpation and the use of the Rœntgen rays; absence of much pain; symmetrical anæsthesia which may be of the dissociated variety; rapid development of muscular atrophy, and the early onset of bed sores.

In favour of a lesion of the cauda equina are: gradual onset of symptoms; signs of injury to the lower lumbar vertebræ or upper part of the sacrum; severe pain in the back, perineum and genitals, and radiating along the course of the sciatic nerves; and pain produced by flexing the hip with the leg extended; want of symmetry in the distribution of anæsthesia and paralysis; slow development of muscular atrophy and of other trophic disturbances; with a tendency to remission of the symptoms especially as regards the condition of the bladder and rectum.

Diseases of the sciatic or of other nerves to the legs are distinguished from disease of the cauda equina by the absence of bladder and rectal disturbance and by the unilateral distribution of symptoms in the lower limbs.

Treatment. Apart from the use of mercury and the iodides in syphilitic cases, the treatment of disease of the cauda equina is mainly surgical. Tumours have been removed with good results, and operations for fracture-dislocation of the spine, in many cases, have been also successful.

CHAPTER IV.

MULTIPLE NEURITIS.

The term multiple or peripheral neuritis is applied to a group of cases characterised by the presence of certain symptoms, which are distributed for the most part to the distal portions of the limbs, and which are known to depend on disease of the peripheral motor and sensory neurons. As a rule the symptoms are more or less symmetrically situated on the two sides of the body.

The possible causes of the disease are very numerous; it would seem as if any poison freely circulating in the blood or lymph might at times produce its chief effects on the peripheral nerves. Thus multiple neuritis may be caused by lead, arsenic, mercury, copper, phosphorus, or silver; by alcohol, ether, bisulphide of carbon, dinitro-benzol, aniline, carbon monoxide, sulphonal, and other drugs. It may also be caused by the micro-organisms which produce specific diseases or by their toxins, for example, those of diphtheria, influenza, enteric and other fevers; of pneumonia, erysipelas, gonorrhœa, syphilis; of the various forms of septicæmia (including puerperal infections), and malaria. Of beri-beri and leprosy it is an essential part. It occurs too in rheumatism, gout, and diabetes. It seems probable that cases of neuritis found in association with anæmia, pregnancy, cancerous and other forms of cachexia, and with gastro-intestinal disturbance, or after over-fatigue and exposure to cold and wet, also owe their origin to some toxic agent.

The action of the poison is not necessarily limited to the peripheral nerves; frequently it affects all parts of the nervous system, though in an unequal degree and distribution; partly because the nature of the poison varies, and partly because its selective action is modified by the varying susceptibilities of individual portions of the nervous system in different persons.

Slight forms of peripheral neuritis are common;

severe forms are less common and are probably more frequently brought about by prolonged over-indulgence in alcohol than by any other cause. In many cases there is evidence of a combination of factors, as of alcohol with lead, or with arsenic, or occasionally with pulmonary tuberculosis.

It is to be specially noted that cases of multiple neuritis are not uncommon, in which it is impossible to discover a cause and in which the previous health of the patient had presented no observable disturbance.

A well marked case of multiple neuritis is characterised by the presence of motor, sensory, and vaso-motor symptoms; frequently one or other of these groups predominates and sometimes may exist alone. Such variations are partly due to variations in the action of different poisons, although it must be clearly recognised that no particular set of symptoms is exclusively related to a particular poison.

For these reasons it seems desirable first to consider multiple neuritis in relation to its principal causes and subsequently to discuss the various clinical types which the disease may assume.

ALCOHOLIC NEURITIS.

The prolonged abuse of alcohol is probably the most frequent cause of peripheral neuritis. The effects it produces may not be exclusively due to the direct action of the poison on the nerve-structures; they may be also due to its general depressing influence which, by lowering the resistance of the nerve-tissues, exposes them to the attacks of micro-organisms and their toxins.

The disease is one of adult life occurring most frequently between thirty and forty years of age; the severer forms are common in women, but the milder forms are met with as frequently in men.

Symptoms. Very rarely the onset of paralysis is quite sudden; as a rule it sets in insidiously and is usually preceded by certain premonitory symptoms which have existed in varying degrees of intensity for many

weeks or months. These symptoms, which may persist throughout the disease, are disorders of tactile sensibility such as numbness and tingling, or "pins and needles" in the fingers and toes; vaso-motor disorders consisting of coldness and sometimes of hot, burning sensations in the extremities; muscular spasms in the form of tremors and twitchings of the limbs, together

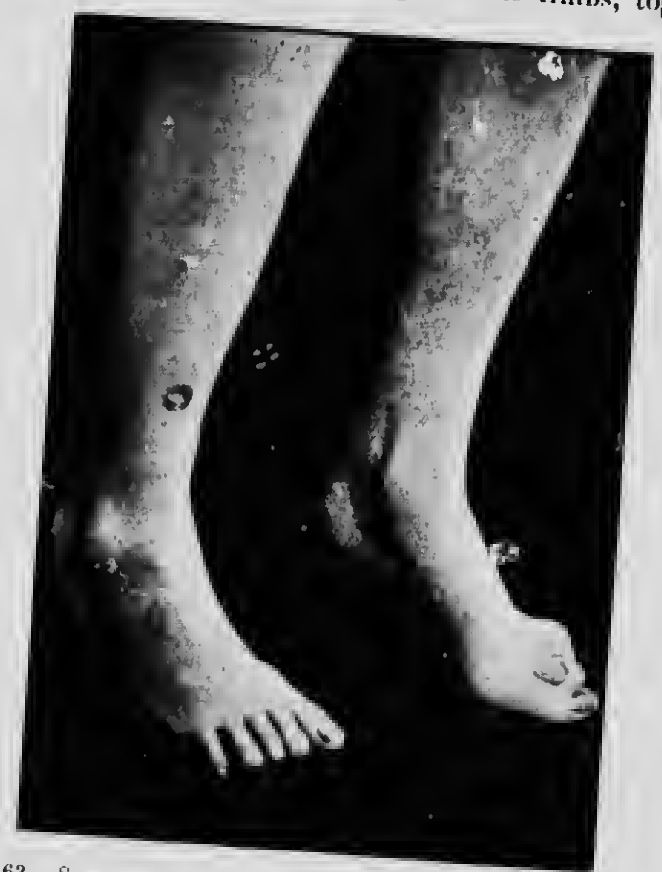


Fig. 63.—Spasm of the calf muscles in the early stage of alcoholic neuritis.

with cramps, which are most severe in the calves of the legs. These cramps are especially troublesome at night or in the early morning, when the patient is often obliged to get out of bed and press his toes on the floor and rub his calves before the spasm relaxes.

Sensory disorders. In addition to paræsthesiæ,

patients also suffer from pains of various kinds which occur most commonly and severely in the early stages of the disease. They may be aching, burning, or shooting in character. Sometimes paroxysms of darting pains occur in the limbs, or in connexion with the viscera; thus the patient may suffer from griping intestinal pains with lead colic, or from gastralgia which when associated with vomiting may resemble the gastric crises of tabes.

The plantar nerves are very liable to be affected, and then the soles of the feet are tender to pressure and cause distress in walking; superficial tenderness may be found also in the forearms and legs; and sometimes accessible nerve-trunks, such as the musculo-spiral, the ulnar or popliteal are unduly sensitive. But the most frequent and prominent sensory disorder is muscular hyperæsthesia. This is usually most marked in the calves, but it may also be present in other muscles: sometimes even moderate squeezing of the muscles is intolerable.

Cutaneous anæsthesia is usually present, varying much in intensity, character and extent. As a rule it is partial, but it may be complete: it is most marked over the distal portions of the limbs and gradually diminishes in intensity towards the knee and elbow. Hyperalgesia may be combined with diminished sensibility to touch.

Motor Disorders. In addition to symptoms of morbid irritation of muscular tissue, namely tremor, twitching and cramps, the further progress of the disease is exhibited by muscular weakness, which usually begins in the distal and gradually spreads to the proximal portions of the limbs, even in some cases to the muscles of the trunk. At first the patient notices that he is easily fatigued, or that he is losing his spring in walking: he may also find that he cannot execute certain special actions with the fingers, such as buttoning his clothes, as well as formerly. Subsequently the extensors of the toes, the dorsi-flexors of the ankles and the

extensors of the wrist and fingers become paralysed, giving rise to a double wrist- and double ankle-drop. The affected muscles are soft and flaccid and they undergo pro-

Fig. 64.—From a case of advanced alcoholic paralysis showing ankle-drop and pendulous condition of the anterior part of the foot, with flexion of the toes. (Ross.)

gressive atrophy. The thenar and hypothenar eminences become flattened and deep grooves appear on the back of the hand between the metacarpal bones. The back

Fig. 65.—Alcoholic paralysis showing double wrist-drop.

of the forearm is also flattened, and the triceps may be wasted; the deltoid, biceps and supinator longus are usually not much affected.

The calf muscles may be wasted as well as the muscles on the anterior aspect of the legs; in advanced cases all the muscles below the knees become greatly atrophied. The thigh and pelvic muscles may also suffer though usually in a less degree; when they are severely affected the patient becomes bedridden.

The peculiar gait of alcoholic paralysis, depends chiefly upon the flatness of the feet and the weakness of the dorsi-flexors of the ankles. To prevent the toes trailing along the ground the patient raises his feet unusually high by flexion of the knee and hip. The undue elevation of the knee at each step makes the gait resemble that of a high stepping horse, hence the name *steppage gait*. In severe cases the muscles of the back and abdomen are affected and occasionally those of the neck also.

The further advance of the disease is indicated by implication of the muscles of respiration. The diaphragm is first attacked; should the intercostals become feeble, expansion of the chest is impaired and life is seriously endangered. Slight feebleness of the facial muscles is not uncommon in alcoholic neuritis, but decided paralysis is rare. As a rule the muscles of the eyeball are spared; but in a few cases, nystagmus, ptosis, weakness of the external recti, and even the Argyll-Robertson phenomenon have been observed.

True *ataxia* is of rare occurrence in multiple neuritis. In the large majority of cases the disorders of gait and other motor defects are due to muscular weakness and not to inco-ordination. The latter, however, does occasionally occur, and has been observed when there was little or no paralysis.

Reflexes. Loss of the knee-jerk is one of the most valuable signs of multiple neuritis; but it must be remembered that the knee-jerk in the early stage, and in the milder varieties of the disease, is frequently exaggerated. The loss often persists for some time after the paralytic symptoms have disappeared. The cutaneous reflexes are generally enfeebled or abolished;

occasionally they persist and especially when hyperalgesia is prominent.

The *sphincters* as a rule are not affected: when mental disturbance is severe the patient may pass urine involuntarily or he may suffer from retention. Occasionally, however, there is evidence that the sphincters are affected. When this is the case the coexistence of spinal disease should be suspected, although it is possible that in some cases the nerves of the bladder may participate in the neuritic affection.

Trophic and vasomotor changes. The subjective feelings of heat or cold in the extremities are sometimes associated with perceptible changes; thus the fingers and toes may be white and cold: or red, dry and hot, or livid and moist. In rare cases the three stages characterising Raynaud's disease have been observed in succession. Profuse sweating is not uncommon; it may be general or local, as of the feet and hands.

Œdema is a noticeable feature in many severe cases of alcoholic paralysis. Generally it is dependent on cardiac dilatation from muscle failure; but it may occur apart from this, and when no feasible explanation other than disease of vasomotor fibres can be given. In the latter case the œdema is generally limited to the lower extremities and to the backs of the hands: but in cases of alcoholic heart-failure the dropsy is often widespread, and erratically distributed.

In chronic cases, the skin of the hands and fingers and sometimes of the soles of the feet becomes thin, smooth and shining, but true "glossy skin" which occurs after severe local nerve lesions is not common in multiple neuritis. The nails and hair tend to become dry and brittle; the former may be furrowed longitudinally and may grow irregularly. Bedsores and perforating ulcers are rare.

Pain in the joints together with redness and swelling sometimes occur during the course of neuritis. More chronic arthritic changes, especially in the joints of the

fingers and wrist may also occur and may end in restriction of movement, and even in permanent deformity.

Mental symptoms. Psychical disorders are so common in alcoholic neuritis that they may be regarded as part of the disease. Failure of memory for recent events is one of the earliest symptoms, and is usually associated with "disorientation," or loss of appreciation of time and locality. Thus a patient may be unable to tell the day of the week or to say whether he is in a hospital or at his own home. He cannot retain any fresh impressions in the memory; if an object is shown to him or a word repeated to him, he is unable a few minutes later to recall the word or to remember the object. Ultimately the memory becomes a complete blank; and a patient who has been lying helpless in bed for weeks may give a circumstantial account of a walk in the morning, of the public-houses he visited and of the boon companions he met by the way. This polyneuritis psychosis or Korsakow's psychosis, from the observer who first carefully described the condition, rarely occurs in neuritis produced by other poisons than alcohol.

Morbid Anatomy. In a well marked case of alcoholic paralysis pathological changes are frequently found in the brain and spinal cord, but most constantly in the peripheral nerves. They may involve both the connective tissue and the nerve fibres, but the latter always suffer most severely: the condition is mainly one of parenchymatous neuritis, interstitial neuritis being slight or absent. The changes are most intense in the

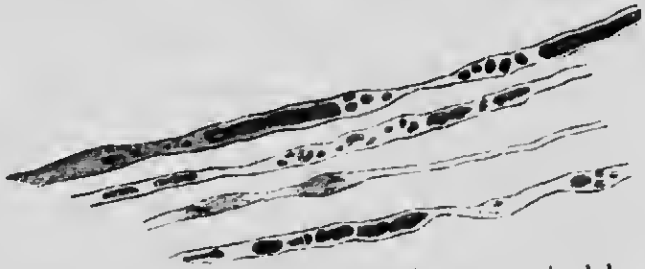


Fig. 66.—Fibre. of the anterior tibial nerve, stained by osmic acid. (Ross.)

terminal branches to the muscles and skin becoming progressively less marked towards the larger branches; the trunk and roots of the nerve are often quite healthy. The branches of the musculo-spiral and those of the anterior tibial nerves are usually most affected. Degenerated fibres have also been found in the phrenic and vagus nerves.

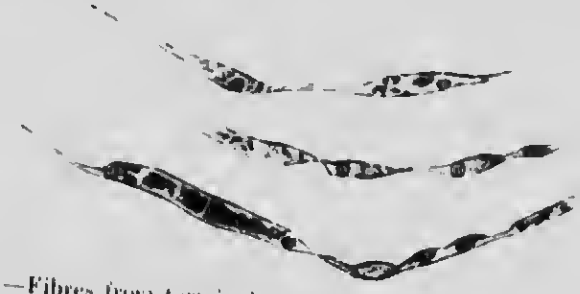


Fig. 67.—Fibres from terminal muscular branch of phrenic nerve, stained by Weigert's method. (*Ross.*)

The muscles in connexion with the diseased nerves are pale and wasted and their fibres are reduced in size. Sometimes there is overgrowth of the connective tissue of the muscles; this interstitial myositis is chiefly found in cases of chronic neuritis, but it may develop in acute cases. Probably in some instances it is due to the direct action of the poison on the muscular tissue, whilst in others it is secondary to the neuritis.

Some of the blood vessels in the nerves and muscles may show thickened walls with surrounding exudations: these changes may account for the observed irregularity in the distribution of changes in the nerves and muscles.

The spinal cord is often quite healthy. In some cases of alcoholic neuritis the cells of the anterior horns have been unduly pigmented or vacuolated; disappearance of Nissl's granules has been observed, together with a lateral displacement of the cell-nucleus. These changes may be secondary to those in the peripheral nerves, or both cells and nerve-fibres may be affected simultaneously. Degenerated fibres have been found in the posterior columns and occasionally in other parts.

Similar degenerative changes are also common in the large cortical cells of the brain, and to these cortical changes the mental disorder is doubtless closely related.

ARSENICAL NEURITIS.

Peripheral paralysis is an occasional result of poisoning by arsenic. It has followed the taking of a single large dose of arsenious acid, but as a rule it gradually appears along with other symptoms of arsenical poisoning after small or moderate doses of the drug have been taken for some time. Thus neuritis has occurred in patients with chorea who have taken daily doses of arsenic for several weeks. Such cases, however, are not common, and many patients suffering from anæmia, chorea or epilepsy, have taken ten to fifteen minims of liquor arsenicalis daily for a long period, without any signs of neuritis.

But although neuritis does not commonly follow the repeated taking of small doses of arsenic, the result, judging from the epidemic of multiple neuritis which occurred in 1900, appears to be different when arsenic is taken in combination with alcohol. In that epidemic the presence of arsenic in the beer was traced to the sulphuric acid used in the preparation of glucose and invert sugar, from which the beer was brewed. Now although many of the sufferers from neuritis had taken large quantities of beer, one or two gallons daily, others had taken not more than three or four pints of beer daily. In the former case the amount of arsenic taken would be considerable, equal to fifteen to thirty minims of liquor arsenicalis a day. In the latter case the daily dose in terms of liquor arsenicalis would not be larger than three to four minims; such a small amount of arsenic by itself would scarcely suffice to produce neuritis; it is more reasonable to assume that the disease was due to a combination of the two poisons, the toxic action of beer being increased by the presence of even a small quantity of arsenic.

Symptoms. In many cases symptoms of multiple

neuritis are preceded for a few days or weeks by other symptoms of arsenical poisoning, such as vomiting, diarrhœa, conjunctivitis, laryngeal and bronchial catarrh and various skin lesions. In other cases these symptoms have been slight or absent, and a sudden or a gradual development of neuritic symptoms has been the only apparent result of the poison. Occasionally neuritis has developed very rapidly after taking a single poisonous dose of arsenic. As a rule, however, the onset of paralysis is gradual and is preceded by subjective sensory symptoms which usually are prominent and persistent.

In most respects the symptoms of arsenical are similar to those of alcoholic neuritis. Thus the paralysis is ushered in and accompanied by marked sensory disturbances. The patient complains of severe darting, tearing and aching pains in the limbs, of paræsthesia in the fingers and toes and frequently of burning sensations in the palms and soles. Painful cramps, and cutaneous and muscular hyperæsthesia are other striking features. To these irritative sensory phenomena, anæsthesia and muscular weakness in the distal portions of the limbs are quickly added.

As a rule paralysis attacks the lower before the upper limbs. Just as in alcoholic cases, the extensor muscles of the hands and feet are principally implicated, and there may be complete hand- and foot-drop. Atrophy of the weak, flaccid muscles quickly ensues and after a time tends to be extreme. In severe cases the atrophic paralysis spreads from the distal to the proximal portions of the limbs, until finally all their muscles are more or less involved. The muscles of the trunk and in exceptional cases some of the muscles innervated by the cranial nerves become affected: paralysis of the palate, the oculo-motor muscles, those of the face, and of mastication have been recorded, but such affections are extremely rare. In severe and advanced cases the distribution of the paralysis is almost universal: it may involve the diaphragm and the intercostal muscles.

Tremors and spasmodic twitchings are sometimes present; athetoid movements of the fingers and hands have been observed. The tendon reactions are usually lost; the superficial reflexes are frequently present.

Every variety of anæsthesia may occur, loss of sensibility to touch, pain and temperature, either separately or combined. Frequently anæsthesia is associated with considerable hyperalgesia. The muscular sense is often affected; there may be a true ataxia. Occasionally ataxia is a dominant feature and the case may resemble one of tabes.

The sphincters are rarely affected; in a few cases incontinence of urine occurred when the mental condition was normal, probably as a result of concomitant disease of the spinal cord, but possibly owing to neuritis of the lower sacral nerves. The mental condition, when the influence of alcohol can be excluded, is usually normal.

The course and duration of arsenical neuritis are similar to those of the alcoholic variety. The disease is rarely fatal; in many cases recovery is complete, usually within a few months. Sometimes recovery is only partial owing to the development and persistence of contractures, and occasionally to permanent joint changes.

Morbid Anatomy. In the few cases of arsenical paralysis in which the nervous system has been microscopically examined a simple parenchymatous neuritis has been found, together with chromatolysis in the cells of the anterior horns, and in one case degeneration of the columns of Goll. Judging from investigations, both experimental and pathological, it seems probable that no part of the nervous system is exempt from the influence of arsenic.

Dixon Mann has suggested that the affinity of keratin tissue for arsenic may be the cause of the initial stage of neuritis, and may also explain the occasional presence of brain symptoms, for neuro-keratin exists in the grey

and white matter of the brain, and forms a sheath round the axis-cylinder and the white substance of Schwann.

Diagnosis from alcoholic neuritis. This is based less on any peculiarities in the neuritic symptoms than on the other effects of the two poisons, alcohol and arsenic. Of these the skin lesions produced by arsenic are of

Fig. 68.—Keratosis of soles of feet. (Reynolds.)

great importance. The palms and soles are often red and swollen and may be bathed in perspiration; sometimes the condition of the feet resembles that of erythromelalgia. Hyperkeratosis of these parts is a characteristic feature, the skin being thickened and

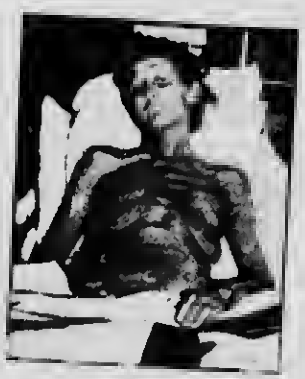


Fig. 69.—Pigmentation with branny desquamation. (Reynolds.)

covered with scales; this change is slow in development and is of long duration. Frequently pigmentation occurs, preceded or not by various erythematous eruptions. There is either a diffuse bronzed discolouration like that

of Addison's disease (the mucous membranes, however, being spared), or the pigmentation is punctiform or in circumscribed spots. In a few cases herpes zoster on the body or limbs has been found.

In cases of neuritis produced by arsenic alone it is rare to meet with signs of cardiac failure or of psychical disturbance. A decided mental change with delirium and defective memory is strongly in favour of poisoning by alcohol.

With regard to the neuritic symptoms, cutaneous and muscular hyperæsthesia are more constant and more severe in arsenical than in alcoholic cases. In arsenical cases also there is a greater tendency to wide distribution and rapid progress of paralysis, to rapid atrophy of the muscles with fibrillar contractions and to the occurrence of ataxia.

A doubt as to the arsenical nature of a case of neuritis may be solved by an examination of the urine, the epithelial scales and the hair for arsenic.

LEAD NEURITIS.

The paralysis that results from lead poisoning is usually bilateral and symmetrical; most commonly the extensor muscles of the wrist and fingers are attacked, but sometimes other groups of muscles become involved, while occasionally the paralysis is generalised and attacks nearly every muscle of the body.

Paralysis may be the first and only manifestation of saturnine poisoning, but generally it occurs in persons who have been subject to attacks of constipation, colic, headache or vomiting, and who present a peculiar form of anæmia, as well as a blackish-blue line at the junction of teeth and gums. Other effects of lead are arterio-sclerosis, nephritis and a granular degeneration of the red blood corpuscles.

Frequently, local sensory phenomena precede and usher in the paralysis, just as in other forms of toxic paralysis. Thus the patient may complain of numbness and tingling in the extremities, of cramp in the calves,

of dull aching pains in the joints, or of sharp pains along the course of the nerves. These sensory prodromata are usually slight in degree; they are often entirely absent, and certainly never present the constancy and intensity of those met with in connection with alcoholic paralysis. Sometimes paralysis comes on acutely, as during an attack of colic; at other times its development is very slow, increasing difficulties in the execution of certain movements being gradually noticed by the patient; but most commonly it develops in a subacute manner.

The paralysed muscles undergo rapid atrophy, and exhibit the reaction of degeneration. In some cases the area of paralysis is greater than that of altered electrical reactions; whilst in others the converse holds, that is muscles which are not manifestly weak give the partial reaction of degeneration.

Fibrillary contractions occasionally accompany the muscular atrophy. Tremor, however, is more common. It is fine in quality, and is increased by voluntary movement, and especially by fatigue; usually limited to the upper extremities, it may become general in aggravated cases.

Alterations in the cutaneous sensibility are usually absent in cases of lead paralysis; but when the muscles supplied by the external popliteal are paralysed, a band of anæsthesia may frequently be detected on the outer aspect of the leg; in the common wrist-drop type of paralysis loss of tactile sensibility is rare. Sometimes an ill-defined zone of partial anæsthesia may be discovered on the posterior aspect of the forearm, hand, or thumb, or, as I have observed, in the territory of the circumflex nerve when the deltoid is paralysed. A diminution of sensibility to the faradic current, however, is constantly present; much stronger currents can be borne over paralysed than over healthy parts, and the same difference holds with regard to the nerve trunks, a patient bearing the application of a strong current to the musculo-spiral better than to the median.

or ulnar nerves. The knee-jerks present no constant relation to the distribution of the paralysis; they are frequently normal, and may be either exaggerated or lost when paralysis is limited to the upper limbs.

The different localisations of lead paralysis may be described under the following headings:—

(1) *The Common or Wrist-drop Type.* This is by far the commonest variety of lead paralysis. The common extensor of the fingers is usually the first muscle to be affected. Its weakness is shown by inability to extend the first phalanges of the two middle fingers; when these are passively straightened the patient can readily extend the distal phalanges by means of the unaffected interossei and lumbricales.

Fig. 70.—Showing wrist-drop in a case of lead paralysis.

Then the special extensors of the index and little fingers, the extensor secundi and primi internodii pollicis are successively involved; soon after, the extensors of the wrist become weak, and ultimately the characteristic attitude of lead paralysis is assumed. The hand, semipronated, is dropped and forms a right angle with the forearm, the fingers are slightly flexed and the thumb is drawn inwards, towards the palm. The hand

is usually inclined towards the ulnar side, and power to extend it becomes completely lost. The flexors of the fingers are unaffected, yet their action is much interfered with by the weakness of the extensors; when, however, the hand is raised, the flexors can act with vigour.

The extensor ossis metacarpi or abductor pollicis is usually spared, or if affected it is not till some time after the other extensor muscles have been paralysed. The supinator longus also escapes, but with those two exceptions all the muscles innervated by the musculo-spiral nerve are attacked in this common variety of lead paralysis.

Fig. 71.—Showing atrophy of the muscles of the forearm and of the interossei, as well as paralysis of the extensors of the wrist, in consequence of lead poisoning.

(2) *The Upper Arm Type.* In this form there is paralysis of the deltoid, biceps, brachialis anticus and supinator longus; sometimes the supra- and infra-spinatus are also involved, while in rare cases paralysis of the pectoralis major is superadded. As a rule, paralysis of this group of muscles succeeds that of the extensor muscles of the forearm, but it may be primary, and then the deltoid is usually the first muscle to be attacked. In this type, changes in the electrical reac-

tions and atrophy of the muscles are much less marked than in the ordinary type of paralysis.

Sometimes paralysis and atrophy of the supinator longus—the other muscles of the upper arm remaining normal—accompany the common form of "wrist drop" paralysis.

Fig. 72.—Showing atrophy of the small muscles of the hand in lead poisoning.

(3) *The Aran-Duchenne Type.* This form is characterised by paralysis and atrophy of the small muscles of the hands, namely, the interosseous muscles and the thenar and hypothenar eminences. Atrophy is always well marked, and accompanies rather than succeeds loss of power. This type may be the primary and only

manifestation of lead poisoning, but more frequently it complicates the classical type of paralysis. It is named from the close resemblance the condition and attitude of the hands bear to the type of progressive muscular atrophy described by Aran and Duchenne, and which is known to depend on degeneration of the ganglion cells of the anterior horns.

In some cases paralysis and atrophy are partial in distribution, and affect the abductor pollicis and the first dorsal interosseous muscle more than the other muscles. The preponderant affection of these muscles has been attributed to their over-use in particular occupations. Thus it is frequent in file cutters, especially in those who with the left hand hold the chisel between the first phalanx of the thumb and the metacarpal bone of the index finger.

(4) *Peroneal Type.* Paralysis of the muscles of the lower limbs is not a common result of lead poisoning, and when present it either complicates paralysis of the arms, or forms part of a more generalised paralysis. The muscles chiefly affected are the long extensor of the toes and the peronei, while the tibialis anticus, although supplied by branches from the same nerve, viz., the external popliteal, usually escapes, just as the supinator longus does in the upper limb. Sensory phenomena often precede and accompany the paralysis. The patient may suffer from neuralgic pains in the knees and joints of the feet, or from numbness and tingling along the outer aspect of the legs. Sometimes there is hyperæsthesia either cutaneous or muscular, less commonly anæsthesia in the territory of the external popliteal nerve. This type of paralysis is more frequently met with in children than in adults.

Affections of the Cranial Nerves. Bilateral amblyopia may occur without ophthalmoscopic changes, and apart from nephritis; sometimes it is transient, sometimes permanent. Two kinds of neuro-retinitis are met with, an acute and a chronic form. The latter occurs in old

cases of lead poisoning in which the kidneys are affected, and is identical with albuminuric retinitis. The acute form is characterised by great swelling of the disc and by the presence of hæmorrhages. These changes may disappear under treatment, or may set up optic atrophy: the latter also occurs apart from preceding signs of neuritis.

Paralysis of the ocular or of some of the laryngeal muscles has been occasionally observed. Sometimes the facial nerve becames affected, and in one case of extensive lead paralysis under my care there was bilateral facial palsy.

Generalised Forms. In chronic cases of lead poisoning, when paralysis has existed for some time it occasionally happens that the paralysis slowly or quickly extends to all the muscles of the limbs, and sometimes to those of the trunk. In some of these cases the widespread atrophic paralysis is associated with exaggeration of the tendon reflexions and with Babinski's reflex: the morbid anatomy is probably that of amyotrophic lateral sclerosis. In other cases the deep reflexes are lost and recovery slowly takes place, when the presumption is that the general muscular atrophy was due to a motor neuritis (see p. 195).

Cerebral Disturbance. Hysterical manifestations may develop from lead poisoning, just as they do from the effect of syphilis or other toxic agents. The hemiplegic weakness and hemianæsthesia that sometimes occur are probably of this nature. But the more frequent cerebral phenomena are delirium, coma, and epileptiform convulsions; these symptoms may occur at an early period of lead intaxication; sometimes indeed they are its first indications. Optic neuritis often accompanies this acute cerebral disturbance: death may occur from the severity of the convulsions or the depth of the coma.

Chronic cerebral disturbance may succeed the acute form or may develop gradually. Sometimes a condition resembling general paralysis is met with, and it is

stated that this disease may be caused by lead poisoning; usually, however, the condition referred to is curable. Cerebral hæmorrhage and softening also occur, as results of arterio-sclerosis produced by lead.

Morbid Anatomy. Lead is remarkable in picking out motor nerves and principally those which supply extensor muscles. This peculiar selective influence is most strikingly shown in the common wrist-drop variety of paralysis, where the posterior interosseous branch of the musculo-spinal nerve is alone attacked. Degenerative changes are found, which are most intense in its intramuscular twigs and become slighter towards the proximal end of the nerve. Similar peripheral nerve changes are found in other varieties of lead paralysis.

In most cases of lead paralysis an examination of the cord has given negative results. In a few cases chromatolysis and slight atrophy of the ganglion cells in the anterior cornua have been found; occasionally more intense atrophy has taken place. Such degenerative changes may be specially related to atrophic forms of lead paralysis, as in the Aran-Duchenne type, but in one case of this kind examined by Madame Dejerine Klumpke the spinal cord was quite healthy, while extensive changes were found in the nerves of the brachial plexus and their terminal branches.

It is probable that some of the cerebral symptoms met with in cases of lead poisoning are due to the direct action of lead on the cortical cells. In a case of fatal lead encephalitis observed by Mott, minute hæmorrhages were found in the brain due to the breaking down of the cortical vessels. Goadby and Goodbody conclude from experiments on animals "that the essential and primary action of lead intoxication is the production of minute and microscopical hæmorrhages in various portions of the body, including the nervous system," and that lead paralysis is due to the presence of minute hæmorrhages in the peripheral nerves.

DIPHTHERITIC NEURITIS.

It is stated that some form of paralysis occurs in about 15 per cent. of all cases of diphtheria, but in all probability the percentage is higher, if account be taken of the slighter indications of paralysis, such as a nasal quality of voice, dimness of vision, diplopia, loss of the knee-jerk and slight weakness in the movements of the hands or feet. The early use of antitoxin is said to diminish the tendency to paralysis, thus Rolleston states that the frequency of post-diphtheritic paralysis varies from 4·9 per cent. when antitoxin is given on the first day, to 31·4 per cent., when its administration is delayed until the fifth day.

No definite relation can be traced between the severity of the throat affection and the degree or the extent of the paralysis; certainly many slight cases of faucial diphtheria are followed by well marked paralysis; sometimes indeed paralysis is the first evidence of diphtheria, any throat affection having escaped observation.

Paralysis may follow cutaneous diphtheria in the absence of any throat affection, and it is probable that in some cases of neuritis apparently due to a septic wound, the latter may have been infected by the poison of diphtheria.

Symptoms. A characteristic feature of diphtheritic paralysis, which usually appears about a fortnight after infection, is the early involvement of the soft palate. In slight cases this may be the only paralysis present; in other cases it is followed by paralysis of the ciliary muscle, whilst in a third series there is a subsequent affection of the pharynx, larynx, or limbs.

Paralysis of the soft palate is indicated by a nasal twang of the voice, by regurgitation of liquids through the nose during swallowing, and by an inability to suck, gargle or blow out the cheeks. On examination, the soft palate looks looser and sometimes lower than

normal, and it is not raised during phonation. Its reflex is diminished or lost; its surface may be anæsthetic.

Paralysis of the ciliary muscle causes loss of power to accommodate for near objects, so that the patient is unable to read small print. As a rule the pupils react both to light and accommodation. Sometimes the external rectus is paralysed on one or on both sides; occasionally other ocular muscles are affected.

The muscles of the pharynx may become paralysed when further difficulties in swallowing occur and food may enter the glottis. In some cases the recurrent laryngeal nerve is affected, giving rise to paralysis of one or both vocal cords; the voice is hoarse or lost; there is inability to cough and the mucous membrane is insensitive.

Paralytic symptoms in the limbs may develop about the same time as the palatal and ocular paralysis, but generally they develop at a later period. They comprise weakness and inco-ordination of movement, paræsthesia and anæsthesia. The muscular weakness affects the distal portions of the limbs and generally is slight in degree; occasionally it is pronounced, and there may be decided double wrist- and ankle-drop. The legs are affected before the arms, and the latter may escape altogether. Sometimes the paralysis spreads to the proximal muscles of the limbs and even to the muscles of the trunk and neck. The diaphragm may become paralysed, and should the intercostals be also implicated serious difficulties in breathing will occur, causing a danger of death from asphyxia. Another most serious complication is acute cardiac failure, the heart paralysis being probably due to a neuritis of the vagus.

The limbs are flaccid, and their paralysed muscles waste and show degenerative electrical changes.

It frequently happens that ataxia is a prominent symptom; it may precede or attend paralysis or may be present when there is no paralysis. The inco-ordination may affect the finer movements of the hands and

fingers, or be limited to the movements of the legs; sometimes the gait resembles that of tabes, sometimes it shows a combination of ataxia and paralysis.

The motor disorders are nearly always accompanied and usually preceded by some disturbance of sensation. As a rule this is slight in degree, consisting of numbness and tingling and partial anæsthesia in the distal portions of the limbs; exceptionally anæsthesia is severe and wide-spread. Shooting pains in the limbs are not common, and there is rarely any undue tenderness of the nerves or the muscles.

The knee-jerks are lost even in cases where no weakness of the legs can be detected; the loss may continue for many weeks after all other symptoms of diphtheria have disappeared. Exaggeration of the knee-jerk may precede its loss and rarely it may be present throughout the whole period of paralysis.

The bladder and rectum are rarely affected.

A serious complication is the occurrence of bulbar crises. During the second or third week of diphtheritic paralysis a patient who suffers from paresis of the palate, loss of power to accommodate and slight weakness of the legs may be suddenly seized with the following symptoms:—The voice becomes weak and hoarse and the cough loose, ineffectual and noiseless. The respiration, previously natural, may give warning of approaching danger; it is not necessarily rapid, but inspiration is sudden, deep and forcible and expiration short and weak; and mucus accumulates in the air passages. The crises which occur and which are frequently fatal, are marked by sudden and complete paralysis of deglutition, by complete aphonia, by alarming dyspnœa, and sometimes by repeated and uncontrollable vomiting. The pulse-rate rises to 140 or 150, and the temperature to 102° or 103°.

Another most serious complication is acute cardiac failure; the heart paralysis as well as the bulbar crises are probably due to a disorder of the vagi or of their centres.

Course. The duration of diphtheritic paralysis is very variable. When it is limited to the palate and the ciliary muscle recovery usually occurs in two or three weeks, but when the paralysis is widely distributed over the body some months may elapse before it entirely disappears. Recovery is generally complete, very rarely partial.

Death may result from either respiratory or laryngeal paralysis; from heart failure; from pneumonia, in consequence of the entry of small particles of food into the windpipe and, rarely, from the impaction of a morsel of food in the larynx.

Morbid Anatomy. The most constant change in diphtheritic paralysis is a parenchymatous neuritis of the nerves which supply the affected muscles; in some cases there is also interstitial neuritis which may be associated with nodular swellings due to inflammation and œdema of the connective tissue. These changes have been found in the nerves to the palate, the pharynx, the larynx, the limbs and the eye-muscles, and also in the cardiac plexus.

Diphtheritic bacilli have not been detected in the degenerated nerves, and it seems certain that the neuritis is set up by a toxin produced by the bacilli.

In some cases of diphtheritic palsy degenerative changes have been observed in the cell bodies of both motor and sensory neurons.

Influenza and Neuritis.

During an epidemic of influenza affections of the peripheral nerves are exceedingly common; isolated neuritis of almost any spinal or cranial nerve may be met with, whilst multiple neuritis, though less frequent, also occurs. In many cases the multiple neuritis is of the ordinary mixed type, in other cases ataxia rather than paralysis is the prominent feature. Occasionally neither ataxia nor sensory symptoms are present, and the condition is one of widely-spread uncomplicated flaccid paralysis. Such cases usually run a favourable

course, and ultimately muscular strength and nutrition are completely restored. It is therefore assumed that the paralysis is not of central origin, but is due to a multiple neuritis in which the motor fibres of the nerves are predominantly or exclusively selected by the toxin of influenza.

Tuberculosis and Neuritis

There is abundant evidence that the peripheral nerves taken from tuberculous subjects, frequently exhibit changes typical of parenchymatous neuritis. Such neuritis may be latent, or may give rise to symptoms which vary in different cases. In some cases muscular atrophy is prominent; in others sensory phenomena—hyperæsthesia, anæsthesia and neuralgia—are present: very rarely the character and distribution of the motor and sensory symptoms are those of a typical multiple neuritis. The neuritis may depend on lowered vitality, rather than on the direct action of tuberculous toxins; moreover the influence of alcohol, which is sometimes taken in considerable quantities by phthisical patients, must not be overlooked.

Puerperal Neuritis.

Cases of neuritis occasionally occur which appear to depend on poisons the result of disturbed metabolism during pregnancy, or of infective or other changes in the blood occurring after labour, whether premature or at the full time. It is often impossible to determine the nature of the poison, or to exclude the influence of anæmia or of the cachexia, which is so frequently associated with the puerperium. The possibility of alcohol must also be considered.

During pregnancy neuritis of every degree of intensity may occur; it is especially apt to be severe in cases of obstinate vomiting. In its mildest form it may be represented only by pains, paræsthesia and hyperæsthesia in the limbs; in severe forms by paralysis and

anæsthesia in the distal portions of the limbs—that is a multiple neuritis of typical character. These symptoms may pass away before labour, or more commonly they continue and become marked for a time after labour.

Neuritis beginning after the child is born may be localised to a single nerve or may affect many nerves. Some of the cranial nerves occasionally suffer, and in a few cases there has been a close resemblance to post-diphtheritic paralysis.

As a rule the prognosis is favourable, and complete recovery takes place; occasionally there has been a fatal issue. In a case reported by Korsakow and Serbski, parenchymatous neuritis was found in the limb nerves, in the lumbar and sacral plexuses and in some of the cranial nerves; there was also an increase of neuroglia in Goll's columns and in the lateral columns of the spinal cord.

Gonorrhœal Neuritis.

Symptoms of a local or of a multiple neuritis have occasionally developed during the course of a gonorrhœa. Neuritis of a single nerve may depend on an invasion of gonococci into its sheath, but it is probable that a multiple neuritis is caused by their toxins rather than by the organisms themselves.

In a case reported by Raymond the paralysis, in the rapidity of its development and its distribution, resembled Landry's disease; the muscles of both sides of the face were paralysed as well as those of the limbs and trunk.

Septicæmia.

Symptoms indicative of multiple neuritis occasionally follow the blood-poisoned state which occurs as a result of some injury or local inflammation. Very rarely the disease has followed suppuration in the lungs, pleura or bladder.

The possibility of a wound being invaded by the organisms of diphtheria must not be overlooked.

Erysipelas.

Multiple neuritis is a rare sequela of erysipelas. It has followed erysipelas associated with wounds, or of the face in the absence of visible abrasion. In a severe case under my care, death occurred from respiratory paralysis.

Syphilis.

Syphilitic neuritis mainly affects the cranial nerves, and is then of the interstitial variety. Sometimes one of the spinal nerves is affected, as the circumflex or the sciatic. A multiple parenchymatous neuritis is excessively rare. A few cases during the secondary period

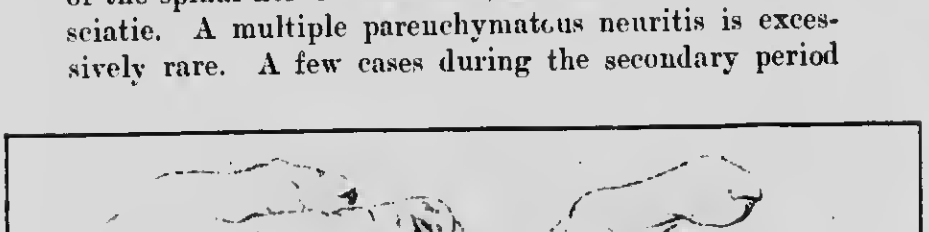


Fig. 73.—Trophic changes in a case of multiple neuritis. (R. W. Taylor.)

of syphilis have been recorded, and in some of them it is probable that the neuritis was caused by the circulation of a toxin derived from the specific organism. In other cases it was difficult to exclude the influence of alcohol or of mercury. T. Buzzard has recorded two cases of almost universal paralysis as instances of peripheral neuritis caused by syphilis. A case reported by R. W. Taylor is remarkable for the extensive derangement of the cutaneous sensibility and for the mutilations of the fingers and toes (see fig. 73). In this case it is possible that syphilis was complicated by leprosy.

Senile Neuritis.

Symptoms of slight neuritis are sometimes present in old persons, and degeneration of the peripheral nerves has been found post mortem. This degeneration may be explained partly by the malnutrition of old age, and partly by a deficient supply of blood to the nerves owing to atheroma of the arteries.

Cancer.

Very rarely symptoms of peripheral neuritis have been observed in the course of carcinoma, when all other known causes could be excluded, the assumption being that the etiological factor was a toxic agent derived from the altered metabolism associated with the cancerous cachexia. Characteristic symptoms have occurred in cases of cancer of the stomach, and in one case, degenerative changes were found in the nerves.

Rheumatism and Neuritis.

The muscular atrophy and paresis which may follow any lesion of a joint are constantly observed in cases both of acute articular rheumatism and of rheumatoid arthritis.

Localised neuritis also occurs. The ulnar nerve is especially prone to suffer; this is shown by the atrophic paralysis of the muscles supplied by the nerve, by impaired cutaneous sensibility in its territory, and sometimes by thickening and tenderness over the trunk of the nerve. There is also evidence that other nerves of the brachial plexus, and that branches of the lumbar and sacral plexuses are occasionally attacked.

It is of significance to note that these peripheral nerve symptoms may occur in a limb *quite free from joint irritation*, as during early convalescence from rheumatic fever. Such an occurrence strongly suggests that the neuritis is directly due to the rheumatic poison and does not depend on an extension of inflammation from the joint to adjacent nerves.

Very rarely the distribution and character of the symptoms correspond to those of a typical multiple neuritis.

Gout and Neuritis.

Gouty subjects often suffer from numbness and tingling in the finger tips, and these symptoms appear to be easily excited by small quantities of alcohol in persons who inherit a strong tendency to gout. This susceptibility makes it difficult to decide how far symptoms of peripheral neuritis in a gouty subject are to be attributed to the influence of alcohol or to that of gout. Nevertheless there is satisfactory evidence that a localised neuritis, and even rarely a general peripheral neuritis may be set up by some poison in the blood produced by gout.

Diabetic Neuritis.

It is common to meet with neuralgia in diabetes, especially in the territory of the sciatic nerve. Doubtless in many cases neuralgia depends on a chronic form of neuritis, and may be associated with trophic changes in the skin and muscles. Sometimes paralysis occurs, the thigh muscles being most frequently involved. Cases of ulnar and brachial neuritis are also met with. Facial paralysis which, in contrast to other forms of diabetic paralysis, is apt to develop quickly, has been observed. Retrobulbar neuritis and paralysis of some of the ocular muscles are other rare complications. Symptoms of multiple neuritis, similar to those due to alcohol, are exceptionally rare. The legs are more affected than the arms, and the territory supplied by the external popliteal nerve is most frequently involved. Sensory symptoms, especially severe pains, occur; ataxia is sometimes a noticeable feature. Perforating ulcers in the feet have been observed; frequently they are accompanied by severe pain, in this respect differing from tabetic ulcers.

Slighter cases of neuritis are less rare; they are represented by pains, cramps and tenderness of the calf

muscles, numbness and tingling in the legs and slight weakness. Frequently the only objective sensory symptom is loss of sensation to the vibrating tuning-fork (Williamson). Loss of the knee-jerk is another notable manifestation, occurring in about 30 per cent. of cases of diabetes. The sign may exist alone or be associated with neuritic symptoms. The tendo-Achilles reflex is also usually lost and the loss may, as in tabes, precede that of the knee-jerk. It is probable that the loss of these reflexes is due to disease of the peripheral nerves, but Williamson has found the nerves normal when the knee-jerks were absent. It is to be noted that the knee-jerk may return and vary in intensity from time to time, the changes having no relation to variations in the severity of diabetic symptoms. Nor can any relation be traced between the frequency of neuritis and the amount of sugar present in the urine. In many cases the percentage of sugar has been small, even as low as one per cent. Moreover, when owing to a strict diet, sugar has entirely disappeared from the urine, the neuritic symptoms have persisted. The morbid product leading to the neuritis is unknown; it is not derived from the decomposition of sugar, and is probably the result of the perverted metabolism and auto-intoxication which goes on in a diabetic subject.

Malaria and Neuritis.

A large number of cases of paralysis occurring in connection with malarial fever are on record, and in many of them the evidence is in favour of a neuritic origin. Sometimes the neuritis is limited to a single nerve either cranial or spinal, sometimes to the motor nerves supplying the muscles on the front of the legs, or, as in a case described by Brandt, to the brachial plexus.

In other cases symptoms of a typical multiple neuritis affecting all four limbs have been observed; occasionally the paralysis has been almost universal in distribution. The paralytic symptoms may develop quickly

during a fit of ague or soon after it. The attack may be the only one, or the paralysis may occur intermittently or periodically. Occasionally it has been associated with impairment of speech, of the special senses or of the mental functions.

Symptoms of multiple neuritis have also developed in persons who have been exposed to the influence of malaria but have not suffered from an attack of fever.

The diagnosis of malarial neuritis is based on the etiology, and on the malarial cachexia, together with the presence of enlargement of the liver and spleen, and on the detection of the plasmodia in the blood.

The prognosis is usually favourable; one fatal case is recorded by Luzatto, who in addition to a parenchymatous neuritis found chromatolytic changes in the cells of the anterior horns.

Leprous Neuritis.

The neuritis of leprosy is peculiar, it being typically adventitial. The morbid process probably begins in the sheath of the nerve and in the sheaths of the fasciculi. These parts become thickened, and ultimately the development and growth of connective tissue in the interior of the fasciculi, and even between the nerve fibres, lead to slow wasting of the nerve elements. This hyperplasia of connective tissue is caused by the direct irritation of the bacilli, which are found in groups in the interstitial tissue. As this grows and shrinks, the bacilli gradually become destroyed. In the early stages of the disease small bulbous enlargements may be found on the nerves, owing to local thickenings of the connective tissue.

Symptoms. The symptoms consist of muscular wasting and anæsthesia in association with patches of pigmentation and pallor of the skin, and mutilations of the fingers and toes. Distinct enlargements may frequently be felt along the course of various nerves. The paralytic phenomena generally first appear in the distal portions of the extremities; in the upper limb

the territory of the ulnar nerve is usually first and most severely affected: in the lower limb it is the outer side of the leg and foot. One of the peculiar features of this disease is the irregularity in the distribution of the symptoms, in consequence of the unequal way in which the fibres of the peripheral nerves are diseased. Thus anæsthesia may not correspond in area to the distribution of any particular nerve, or the knee-jerk may be obtained when the extensors of the knee are partially paralysed. This irregularity in the distribution of the symptoms sometimes makes a case of anæsthetic leprosy present a close resemblance to one of syringomyelia, especially as in some cases the anæsthesia consists in a loss of sensation to pain and temperature whilst sensation to touch is preserved.

In syringomyelia, however, the anæsthesia is more widely distributed, and there are no enlargements of the nerves. In leprosy there is no spastic paraplegia and the bladder and rectum are not affected. The diagnosis is made certain by the detection of the leprosy bacillus in the subcutaneous tissues.

Beriberi or Kak-ké.

The chief symptoms of this disease depend on a multiple peripheral neuritis, which usually runs a chronic course, although, like other forms of peripheral neuritis, it may occasionally develop with great rapidity and severity.

The earliest symptoms, according to Pekelharing and Winkler, consist in a slight degree of the reaction of degeneration in the peroneal nerves and the dorsi-flexors of the ankles. About the same time or a little later certain subjective sensations are experienced, such as heaviness and numbness in the legs, tingling, creeping, or burning sensations, together with palpitation and undue excitability of the heart. Then a progressive paralysis and atrophy set in: the dorsi-flexors of the ankle are first attacked, and soon give the complete reaction of degeneration: subsequently the calf muscles,

the extensors of the knee, and other muscles of the lower limbs become affected. The trunk muscles may be involved, and in severe cases the muscles of the upper extremities; the extensors of the wrist being usually the first to be affected.

The face often suffers and sometimes the muscles of the eyeball and larynx. The atrophic paralysis is associated with marked sensory disturbance. Tactile sensibility and the muscular sense are (according to Scheube) prominently affected; sensibility to pain is usually preserved, but the temperature sense and the cutaneous sensitiveness to faradism are often diminished.

Œdema is one of the characteristic features of the disease. In some cases it is very slight and limited to the legs and ankles, but in other cases it spreads widely, until at length it becomes general anasarca, and involves also the serous cavities, especially the pericardium. Patients suffering from beriberi are anæmic, and complain of palpitation and dyspnœa, symptoms largely due to cardiac weakness and dilatation of the right side of the heart.

The severity of the disease varies in different epidemics, the mortality varying from two to sixty per cent. Death commonly results from cardiac failure, or from paralysis of the diaphragm and intercostal muscles.

The peculiar features of beriberi are the association of dropsy and signs of cardiac failure with symptoms of a multiple neuritis. Dropsy and dilatation of the heart are not uncommon in alcoholic paralysis, but they are of less constant occurrence than in beriberi. The former condition is also distinguished by the presence of severe pains and muscular hyperæsthesia, and by the fact that the arms are often paralysed as soon as the legs.

The most constant *morbid changes* in beriberi are:—Degenerative neuritis affecting the nerves of the extremities and also in some cases those of the trunk, heart and larynx; together with an excess of fluid in the cellular tissue, and in the pericardium and other serous cavities.

Bacilli similar to those of splenic fever have been found in the blood, heart and other tissues. They are believed to enter the body through the alimentary canal, and are probably derived in some way from the consumption of rice. It is probable that a toxic substance, produced during the growth of the organisms, is the cause of the neuritis; it appears to have a special affinity for the cardiac branches of the vagus.

Progressive Hypertrophic Interstitial Neuritis.

This is a rare disease which generally occurs in children, and shows a tendency to affect more than one member of a family. The disease runs a slow and progressive course, and is characterised by the following symptoms: Ataxia of the four limbs with muscular atrophy; lightning pains; marked disturbances of sensation with delay in its transmission; hypertrophy and hardness of all the nerve trunks of the limbs that are accessible to palpation; nystagmus; myosis and the Argyll-Robertson phenomenon; kypho-scoliosis and talipes varus. The tendon reflexes are lost; the sphincters are not affected.

Necropsies, made in a few cases, have revealed pronounced hypertrophic sclerosis of the limb nerves, and of the anterior and posterior roots of the spinal cord; together with sclerosis of the posterior columns presenting the same distribution as in tabes. The overgrowth of the fibrous tissue in the nerve bundles and nerve sheaths leads to atrophy and disappearance of the nerve fibres. The lesions are usually more marked in the nerves and roots of the lower limbs than in those of the upper limbs and the trunk.

CLINICAL TYPES.

In the preceding account of multiple neuritis it will have been noticed that the symptoms vary to some extent with the nature of the poison producing them. It is now desirable to consider the subject from a

clinical standpoint, briefly reviewing the chief types of the disease that are met with, irrespective of its causes. These are: (1) The common or mixed type, in which motor, sensory, and vasomotor phenomenon are present in various combinations. This has been adequately described under the headings of alcoholic and arsenical neuritis. (2) The motor type. (3) The sensory type. (4) The ataxic type. (5) The vasomotor type.

The motor type. Of this there are two possible varieties, namely, the spasmodic and the paralytic. The existence of the *spasmodic* variety is open to question. The occasional presence of exaggerated knee-jerks (showing increased muscular tonus) in the early stages of alcoholic and other forms of peripheral neuritis, and the presence of tremors and active spasms, sometimes throughout the whole course of the disease suggest the question: Are there cases of multiple neuritis which are clinically expressed by a spasmodic condition of the extremities and by no other prominent symptoms? In other words, may the whole malady be represented by morbid irritation of motor nerve fibres? It is impossible to give a positive answer to these questions. The author has observed such cases, in which it seemed that the disease was arrested before destruction of nerve fibres, giving rise to paralytic phenomena, had taken place. Moreover it cannot be denied that the distribution of the spasms in tetany together with their occasional association with other symptoms of neuritis, strongly suggests the possibility that this disorder may be a variety of an irritative form of peripheral neuritis (see p. 434).

The *paralytic* variety in which paralysis is the dominant feature, sensory and other symptoms being absent or inconspicuous. An atrophic paralysis characterises the common variety of lead palsy, and localised forms of motor neuritis occur as a result of other poisons, as alcohol, those of influenza, diphtheria, and of enteric fever.

With regard to cases of widespread motor paralysis

which have been observed in connexion with lead poisoning, diphtheria, influenza, and some other toxic agencies, the diagnosis is often very difficult, and especially so where the course is acute and rapid. The distal portions of the limbs are mainly involved; the affected muscles are flaccid, the knee-jerks are lost, and sensory symptoms are slight or absent. Some of these cases end fatally owing to invasion of the respiratory muscles; others survive, and the affected muscles undergo a progressive atrophy. There is pathological evidence that the morbid changes may be limited to the intramuscular branches of the peripheral nerves, or to the anterior horns of the spinal cord, or may involve the whole of the lower motor neuron. It may therefore be accepted as a fact that occasionally an uncomplicated generalised motor neuritis does occur.

The Sensory Type. The slighter degrees of multiple neuritis are represented sometimes by motor defects, such as spasm or weakness of some of the special movements of the fingers and thumb, but far more commonly by various sensory disturbances, such as numbness and tingling in the fingers and toes, and slight anæsthesia; frequently a careful examination will reveal impaired sensation over the tips of the fingers or the sides of the hands. In many cases, especially those of alcoholic or arsenical origin, there is considerable muscular hyperæsthesia. The knee jerks are increased, diminished or lost. These may be the only symptoms present; or they may be accompanied by slight weakness of the movements of the digits, or possibly of those of the hands and feet. Even when alcohol has been taken in large quantities for a long time, the manifestations of neuritis may be very slight. They occur also in gout and in diabetes; in these diseases decided paralysis and anæsthesia are rare. The neuritis of leprosy is largely sensory. Frequently no cause can be discovered, when the symptoms although not increasing in severity tend to persist, or they improve for a time and then relapse.

The ataxic type: neuro-tabes peripherica. As already stated ataxia is not a common symptom in multiple neuritis, even when cutaneous and muscular sensibility are profoundly affected; and further, marked degeneration of terminal nerve fibres to both skin and muscle has been found in cases which did not present any signs of incoordination during life.

Occasionally ataxia is a prominent symptom and is attended by only slight signs of weakness or of impaired sensation. In some cases the development of ataxia is rapid. When it is associated with loss of the knee-jerk and with sensory disturbances the condition may strikingly resemble that of tabes. One distinction is the condition of muscular sensibility, which unaffected or diminished in tabes is often greatly exaggerated in multiple neuritis.

The vasomotor type. A gradual transition may be traced between the vasomotor phenomena present in many typical cases of multiple neuritis and those which characterise Raynaud's disease. In one case recorded by the author, and proved microscopically to be a genuine instance of multiple neuritis, gangrene of the feet developed during the last two months of life. Moreover degenerative neuritis has been histologically demonstrated in cases presenting typical symptoms of Raynaud's, either when the ordinary symptoms of peripheral neuritis were present, or when they were absent. On the other hand histological examination of the peripheral nerves in many cases of Raynaud's disease has failed to reveal any abnormal changes.

On the whole it seems probable that an unusual prominence of vasomotor symptoms in multiple neuritis is due to the fact that vasomotor fibres have been especially selected by the poison which has set up the disease; and that there are certain varieties of Raynaud's disease in which the symptoms are mainly the result of a degenerative neuritis.

DIAGNOSIS.

There can be little difficulty in diagnosing a well marked case of multiple neuritis. The symmetrical distribution of the flaccid paralysis and the sensory disorders to the distal portions of the limbs, the tenderness of the muscles and nerves, and the freedom of the bladder and rectum from disturbance of their functions are characteristic features.

The chief difficulties in diagnosis are met with (1) in the early stages of the affection, (2) in acute cases of rapidly generalised paralysis, (3) when ataxia is a prominent symptom, and (4) when sensory symptoms are slight or absent.

1. The pains of the initial stage may be mistaken for those of rheumatism or neuralgia. Rheumatic pains are related to the joints rather than to the muscles or the nerves, and are not usually associated with numbness and tingling in the hands or feet, as are the pains of neuritis. The bilateral symmetry of the pains and numbness is a distinction from neuralgia, which is a unilateral disorder.

Voluntary power in the limbs may be impaired by *acute myositis*; but in this disease the muscles are rigid rather than flaccid, the limbs being often swollen from inflammatory œdema of the muscles and subcutaneous tissue; apart from pain there is no sensory disturbance, and there are no signs of real paralysis.

In the early stage of multiple neuritis the knee-jerk may be abnormally brisk, but the exaggeration soon gives place to diminution and loss, nor is it ever associated with ankle clonus or an extensor plantar reflex. Hence any suspicion of a commencing spastic paralysis is quickly dispelled. In neuritis an exaggerated knee-jerk is usually associated with sensory symptoms and sometimes with loss of the tendo-Achilles jerk, for this reflex may disappear before the knee-jerk.

A commencing *myelitis* of the lumbar enlargement will cause weakness of the legs and loss of the knee-jerk and thus simulate a peripheral neuritis, from which however it may be distinguished by loss of control over the sphincters and by the distribution of any anæsthesia present.

2. The diagnosis of acute rapidly progressive cases of paralysis, which is often very difficult, is considered in Chapter VI.

3. Ataxia is occasionally a prominent symptom in multiple neuritis, and if associated with sensory disturbance and loss of the knee-jerk, but with little or no muscular weakness, it may be difficult to exclude tabes.

Neuritis is however distinguished by the absence of changes in the pupillary reactions, of optic atrophy and of disordered micturition. In favour of neuritis also would be great tenderness of the nerves and muscles; in favour of tabes a history of syphilitic infection, and the presence of an excess of lymphocytes in the cerebro-spinal fluid. The superficial abdominal reflexes are frequently much exaggerated in early tabes: they are normal or diminished in multiple neuritis. Thoracic bands of anæsthesia are found in tabes, but not in neuritis.

4. Cases of multiple neuritis are sometimes met with in which sensory symptoms are slight or completely absent, the disease being represented only by an atrophic paralysis affecting mainly the peripheral portions of the extremities. Such paralysis might be due to an anterior poliomyelitis of the cervical and lumbar enlargements: as a rule however the paralysis is more irregularly distributed than in neuritis and may be more marked in the proximal than in the distal segments of the limbs. In favour of neuritis also would be the presence of ataxia however slight. Frequently too in cases of apparently uncomplicated motor neuritis the tuning fork may reveal loss of the vibrating sensation, an indication that some sensory fibres are involved.

PROGNOSIS.

When a patient suffering from multiple neuritis is placed under favourable condition, recovery—partial or complete—may be expected; and even when paralysis is widely distributed, and affects the diaphragm a hopeful prognosis is frequently justified. The duration of such severe cases is often considerable and some of the muscles, as those of the hand, and the peronei, may remain permanently weak and wasted. Contractures of the limbs and articular changes are also sometimes persistent. When psychical disturbance is severe and long-lasting, permanent mental weakness is to be feared.

Fig. 74.—Photograph showing permanent contractures of the flexors of the hip and knee resulting from alcoholic paralysis.

In estimating the danger to life we have to consider the general condition of the patient, the rapidity with which paralysis has spread, and the nature and severity of any complication that may be present. In alcoholic cases the vitality of the patient is often much lowered,

and his tissues may offer a feeble resistance to the attacks of tubercle bacilli, pneumococci or other toxic agents. The subjects of alcoholic neuritis are peculiarly liable to pulmonary tuberculosis, and any weakness of the respiratory muscles would constitute a predisposing factor.

Impaired action of the heart is another serious complication in alcoholic cases; cardiac muscle failure is a common cause of death. The liability of the vagus and phrenic nerves to be implicated in post-diphtheritic paralysis must also be remembered.

The prognosis is unfavourable when the development and extension of the paralysis are rapid. In these acute cases, for which often no adequate cause can be discovered, the mortality is high owing to the frequency with which the respiratory muscles are attacked, and the consequent tendency to pneumonia.

TREATMENT.

The first essential in every case of multiple neuritis, is to discover the cause and to remove it or stop its action as soon as possible. In cases due to alcohol the patient should be deprived of alcoholic drink in every form; the deprivation is rarely attended with danger, if suitable nourishment is administered and careful attention is given to the digestive organs. To ensure the complete withdrawal of alcohol it is usually desirable to put the patient in a home or hospital under the care of trained and trustworthy nurses.

In all except the mildest cases of neuritis, rest in bed is advisable. The patient is thus most easily protected from exposure to cold, the pernicious effects of movement are reduced to a minimum, and local treatment can be carried out most satisfactorily. The severest cases require a water-bed; this not only relieves pain better than an ordinary bed but by giving more uniform support to the body, lessens the risk arising from a weak heart or from paralysed respiratory muscles. For the

relief of tender nerves and muscles there is nothing better than the intermittent application of warm fomentations, the limb being subsequently wrapped up in a thick layer of hot cotton-wool. Occasional vapour baths, or warm packs, are also beneficial, but they should be used with caution when the action of the heart is much impaired.

As regards drugs, salicylate of soda and iodide of potassium appear to be of service. Neuralgic pains are often relieved by the administration of antipyrin, phenacetin, aspirin, and exalgin; when the suffering is intense a hypodermic injection of morphia may be necessary. In malarial neuritis quinine should be tried, and in anæmic or septicæmic cases large doses of perchloride of iron. When syphilis is suspected, mercury and iodide of potassium should be administered. The weak, dilated heart of alcoholism requires digitalis and strychnine: these drugs are also of service in severe cases of post-diphtheritic paralysis.

Great care must be exercised to prevent the development of contractures, especially those which depend on shortening of the calf muscles and the hamstrings: the limbs must be supported and maintained in a correct position by sand-bags and other means.

When the acute symptoms of the disease have subsided, recourse may be had to massage and electricity, remedies which are of great service in helping to restore the degenerated nerves and muscles. At first massage should be as gentle as possible, but after a time it ought to be regularly and vigorously applied. The patient should be encouraged to move the affected limbs against resistance, and to perform various forms of exercises.

From first to last, abundant fresh air and sunlight, and the careful administration of nourishment are of the greatest importance.

CHAPTER V.

ACUTE POLIOMYELITIS.

Infantile Paralysis and Acute Atrophic Paralysis of the Adult.

Poliomyelitis is an acute disease, which occurs in children. At first the paralysis may be wide-spread: subsequently it tends to disappear, except in one limb, some of the muscles of which undergo rapid wasting in consequence of destruction of the cells in the anterior horns of the spinal cord with which they are related.

Etiology. The disease occurs most frequently during the first three years of life, and especially between the ages of six and eighteen months. It is also met with in older children and sometimes in young adults; it rarely develops after the age of forty. The possibility of an intra-uterine attack is strongly supported by the history of a case recorded by F. Batten. Whilst the onset may occur at any time of the year, it is most common during the hot summer months.

In many cases the attack comes on suddenly without any warning. In other cases it appears to have been induced by injury; over-exertion; exposure to cold; or some infectious disease, notably scarlet fever, measles and whooping cough. That anterior poliomyelitis is itself an infective disease is shown by the fact that occasionally it attacks more than one member of the same family, and by the occasional occurrence of epidemics in which many cases of this disease have occurred in one locality within a short space of time. There is suggestive evidence that the disease is contagious, and that the incubation period is less than six days.

Symptoms. The clinical history of this affection may be divided into four periods, namely (1) the period of invasion, (2) the stationary period, (3) the period of regression and (4) the period of atrophy with deformi-

ties. It must, however, be understood that these periods are not abruptly separated from one another.

The period of invasion. Sometimes the child is put to bed apparently quite well, and in the morning one of its legs is found to be paralysed. Sometimes the development of paralysis is more gradual; at any rate the mother is unable to state when the limb first showed signs of weakness. Such a history is common, but it does not follow that initial febrile symptoms were absent; probably in many cases they were overlooked.

In a large number of cases the disease is ushered in by constitutional disturbance. The child is hot, fretful and heavy for sleep; the temperature is raised to 100 or 101°, sometimes to 104°. There are pains in the body and limbs; the latter are tender to pressure. The child is content to lie in bed, not crying to be taken on its mother's lap as young children usually do when suffering from ordinary ailments. Initial stupor may deepen into coma, or there may be delirium and sometimes convulsions.

The severity of the attack bears no relation to the degree or the extent of the paralysis; often this is not noticed until the acute symptoms have subsided. Incontinence of urine, however, may be present, and the careful observer may have been struck from the first with the limpness of an arm or a leg.

As a rule during this stage, which lasts from a few hours to a few days, paralysis develops with rapidity. Its maximum distribution varies greatly in different cases. Most commonly it is limited to one limb, the leg being affected oftener than the arm. Sometimes both legs and one arm are involved, while in severe cases paralysis attacks not only the four limbs, but also the muscles of the trunk and neck and even, though very rarely, those of the face or the eye.

The stationary period. For a time the initial palsy, having reached its maximum, remains stationary both as regards degree and extent. The affected parts are limp, the muscles being flaccid; sometimes hypotonus

is so marked that the affected limbs can be placed in almost any position. Many of the weak muscles react sluggishly to the faradic current; some of them may not respond at all. The electrical reactions indeed show every variation from a simple diminution of the normal response, to the complete reaction of degeneration. The faradic irritability of both nerves and muscles sinks quickly in those that are severely affected, and is usually abolished within a fortnight in muscles that are permanently paralysed.

The pain and hyperæsthesia often present in the initial stage may persist; more commonly they subside. The cutaneous sensibility is normal; rarely a diminution to pain and temperature is observed, an indication that the disease has extended beyond the anterior horn.

The sphincters of the bladder and rectum are not affected, except occasionally during the first few days.

The skin reflexes are usually lost at the level of the lesion, whilst the condition of the tendon jerks varies with the distribution of the paralysis. Thus the wrist and elbow-jerks are lost when the muscles of the arm are involved; the knee-jerk is lost when the extensors of the knee are paralysed, and the tendo-Achilles reflex when the calf muscles are paralysed. But even when paralysis is limited to the anterior tibial muscles, the knee-jerk is often absent at least for a time. When one leg only is paralysed, the knee-jerk of the other leg is present, and it may be exaggerated owing to extension of the disease to the pyramidal fibres. For the same reason an extensor plantar reflex is sometimes obtained.

The period of regression. After a stationary period which varies in duration from a few days to a few weeks, some of the paralysed muscles begin to improve and the improvement may progress for several months; in a few cases complete recovery takes place; but as a rule only some of the muscles are completely restored, the rest becoming more and more flabby and wasted.

The resulting permanent atrophic paralysis is usually situated in some part of the lower extremity, the leg

being more commonly affected than the thigh. The peronei, the extensors of the toes, and the tibialis anticus are most often involved; occasionally the calf muscles alone are attacked. Of the thigh muscles the psoas and iliacus, the glutei, and the quadriceps are most commonly affected; the flexors of the knee usually

Fig. 75.—Photograph of a case of acute anterior poliomyelitis in the adult.

escape. In the upper limb the shoulder muscles most frequently suffer: the deltoid alone, or in association with the spinati, the biceps and the supinator longus. Much less frequently the muscles below the elbow are involved, the extensors or the flexors of the wrist and fingers, and exceptionally, the small muscles of the hand (see fig. 76). In some cases the muscles of the back are also involved, as the erector spinæ, and the lower part of the trapezius. Very rarely the disease is limited to the dorsal portion of the spinal cord; thus in a case

recorded by Lapage the most prominent symptoms were paralysis of the abdominal muscles and atrophy of the lower part of the erector spinæ; the lower limbs were quite normal.

Fig. 76.—Infantile paralysis affecting in varying degree most of the muscles of both arms and both legs. The right arm and the left leg were most severely affected, the extensors of the wrist and fingers and the dorsi-flexors of the ankle being completely paralysed.

The period of atrophy and deformities. As already mentioned the definitely paralysed muscles undergo progressive atrophy; they may become so attenuated that the bones seem to underlie the skin. Sometimes the apparent bulk of a muscle is maintained by an accumulation of the subcutaneous fat.

The bones often participate in the trophic disturbance, and become smaller in every dimension. This reduction in size does not necessarily vary in relation to the degree of muscular atrophy; for example, the leg may be much shortened, when only one or two of its muscles are atrophied. The long bones become not only short and thin, but porous and friable and hence fractures may readily occur; the paralysed foot or hand is smaller than the sound one, and even the pelvis may

be arrested in its development. A radiogram of the affected bones shows that they have a smooth surface, having lost many of their processes and ridges.

Fig. 77.—From a case of old infantile paralysis. (Larmuth.)

The ligaments also waste and become relaxed; as a consequence the joints are loose and prone to dislocation. The skin of the affected limb is thin, smooth and inelastic, often mottled and bluish and distinctly cold to the touch. It is liable to be marked by slight injuries, and is often the seat of chilblains or of indolent ulcers.

The permanent deformities, so characteristic of this disease, are produced chiefly by the unopposed action and gradual shortening of the healthy antagonists of the paralysed muscles, and partly by the laxity of the articular ligaments, and the action of gravity.

Talipes equinus and equino-varus are the commonest deformities, while talipes vagus and calcaneus occasionally occur. Sometimes the knee becomes permanently flexed, owing to weakness of the anterior muscles of the thigh. When all the muscles of both legs are paralysed the patient may move about by crawling on his knees, dragging his small thin legs after him.

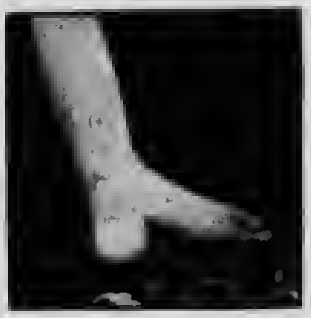


Fig. 78.—Talipes calcaneus from atrophy of the calf muscles in a case of infantile paralysis.

Lateral curvature of the spine may result from the attitude induced by a shortened leg or from unilateral weakness of the back muscles; lordosis may be caused by partial paralysis of the sacro-spinal muscles.

Fig. 79.—Showing dropped shoulder from atrophy of supporting muscles, in a case of acute poliomyelitis.

The most common deformity in the upper limb is due to the dropping of the humerus from the glenoid cavity,

owing to atrophy of the deltoid and other shoulder muscles; the arm hangs powerless by the side and dislocation at the shoulder is readily produced.

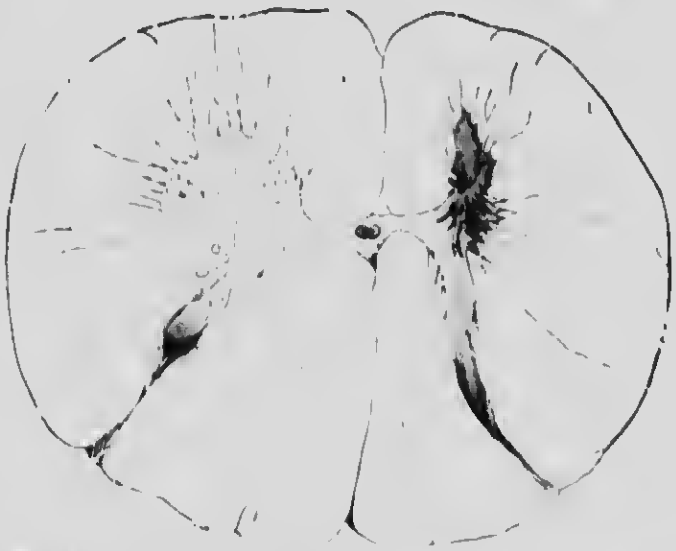


Fig. 80.—Section of cervical cord from a case of infantile paralysis of long standing, showing atrophy of the anterior cornu, and the white columns on the right side. (Charcot.)

Pathology. In old-standing cases of acute poliomyelitis the cord in certain localities is small on the affected side; this reduction in size is mainly due to shrinking of the anterior horn, the cells of which are atrophied or entirely destroyed, their place being taken by dense connective tissue. These sclerotic changes are vascular in origin and are not the result, as was formerly supposed, of a primary degeneration of the ganglion cells. If cases be examined at an early stage of the disease the branches of the anterior spinal artery are seen to be distended; some contain thrombi, while the surrounding grey matter and often too the neuroglia are infiltrated with blood cells and lymphocytes. The lymph spaces about the vessels and the nerve cells are filled with serum. The nerve cells themselves show degenerative changes which vary in

degree according to the duration and intensity of the disease. Degenerative changes may also be traced into the anterior roots, and into the muscular branches of the peripheral nerves, and sometimes into the white matter adjacent to the anterior horn, and into the grey matter posterior to it.

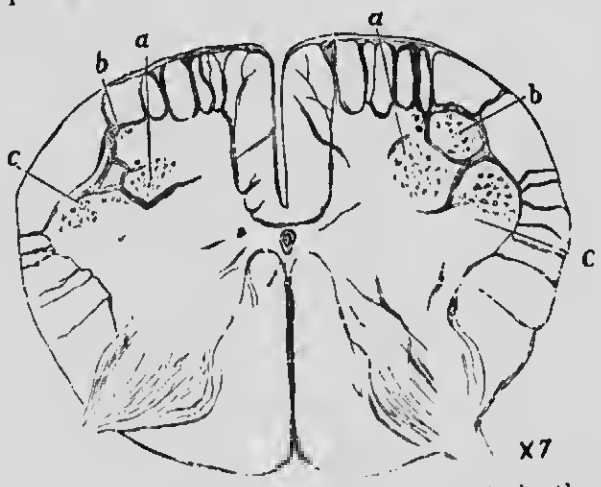


Fig. 81.—The letters a, b, c indicate respectively the central, antero-lateral, and postero-lateral groups of ganglion cells. On the left side the group b has almost entirely disappeared, causing a marked falling in of the circumference of the grey matter. The groups a and c are fairly well represented on the left side, but the cells composing them are not so numerous as on the right. The internal group has disappeared from both sides. (After Humphreys.)

In many cases the perivascular changes, as well as the intense cell-infiltration of the grey matter, are not limited to any particular portion of the cord but are found throughout its whole length, and sometimes in well-vascularized regions of the brain. The soft meninges, especially those of the lower part of the cord, may also show considerable cellular infiltration: this condition explains the lymphocytosis which is sometimes found on examination of the cerebro-spinal fluid obtained by lumbar puncture.

Although the causative agent of acute poliomyelitis is still unknown, there is suggestive evidence—afforded by a study of recent epidemics of the disease, as well as by the results of experiments—in favour of its being

a bacterial toxin. Thus it has been clearly proved that the disease can be transmitted from man to monkey by the injection of an emulsion of the diseased part of the spinal cord; and that it can be transmitted in the same way from one monkey to another.

F. Buzzard believes that the essential lesion of acute poliomyelitis is an inflammation of the interstitial tissue of the central nervous system, the effects of which are most pronounced in the grey matter of the spinal cord owing to its great vascularity and low resisting power.

A close relation between the disease and certain forms of acute encephalitis has been frequently noticed. The morbid anatomy of the two diseases is strikingly similar and there is abundant evidence that encephalitis may be excited by the causal agent of poliomyelitis. Thus there are recorded instances of the two diseases occurring in the same individual or in two or more members of the same family at the same time. In a family observed by W. Pasteur, seven children were attacked by an acute febrile disorder; two developed infantile paralysis, another had hemiplegia as a result of encephalitis, whilst two of the others suffered from nervous symptoms of a transient character.

Wickman has drawn attention to the frequency with which the disease affects other parts of the nervous system besides the spinal anterior horns. During epidemics of acute poliomyelitis he observed the following types in addition to the ordinary type of infantile paralysis: (1) a type resembling Landry's disease, (2) a bulbar or pontine type, (3) an encephalitic type, (4) a polyneuritic type characterised by much pain and local tenderness, (5) an ataxic type, (6) a meningitic type and (7) an abortive type, in which constitutional symptoms, identical with those of other cases in which paralysis developed, alone occur.

The occurrence of so many different forms of the disease shows that the microbe or its toxin, although it has a predilection for the anterior cornua, may attack

almost any part of the nervous system. It may enter the body through the gastro-intestinal tract, or possibly by the nasal mucous membrane or even through the middle ear.

Diagnosis. In the absence of obvious paralysis it is tempting to attribute the early febrile stage to gastro-intestinal disturbance, to rheumatism (when there is much pain), or to dentition, even when there is no direct evidence of these conditions. Such errors in diagnosis, as well as delay in detecting the first signs of paralysis, are avoided by remembering the possibility that a pyrexial attack may depend on anterior poliomyelitis, and that prostration does not produce actual immobility of a limb. When paralysis is detected the loss of Faradic irritability in the muscles indicates the nature of the case.

An atrophic paralysis of both legs might be due to lumbar myelitis, but then the paralysis would be associated with anæsthesia and usually with weakness of the bladder sphincter; a difficulty could only arise in the adult, for lumbar myelitis apart from caries is practically unknown in the young child.

Peripheral neuritis is rare in children; in the adult it would be distinguished from acute poliomyelitis by the presence of anæsthesia or other sensory disturbance and by the distribution of the paralysis.

The palsies of cerebral origin are spastic in nature: the tendon reactions are exaggerated, not diminished or lost.

Hip joint disease, infantile scurvy and other conditions in which pain interferes with movement are distinguished from infantile paralysis by the preservation of the knee-jerk and the absence of any real paralysis.

Our knowledge relating to spinal localisation together with that derived from the investigations of A. Bruce in regard to the localisation of motor functions to certain groups of nerve cells, enables us to forecast the position of the lesion in acute poliomyelitis with con-

siderable accuracy. For example, in the common type of the disease, where the muscles on the front of the leg are paralysed, we find that the postero-lateral group of nerve cells in the anterior horn at the level of the first and second sacral segments of the cord are mainly involved.

Prognosis. The disease is rarely fatal, and even in the stage of severe constitutional disturbance when cerebral symptoms are prominent or when there is paralysis of some of the respiratory muscles, the danger to life is usually more apparent than real. The paralysis having reached its maximum the question arises how much of it will be lasting. The answer is that both complete recovery and very slight paralysis of limited area do occur but only very rarely, the rule being that many paralysed muscles recover while others remain distinctly affected. Thus if at the outset both legs are paralysed, one leg may completely recover while in the other leg the peronei alone may be permanently paralysed; or permanent atrophy of the shoulder muscles of one arm may be preceded by complete paralysis of both arms, with weakness of some of the neck muscles. A consideration of the complete return of motor power in parts that were profoundly paralysed would lead one to infer that in some cases the wave of recovery would spread to every part that was initially affected. In one remarkable case under the author's care, complete paralysis of the limbs and many of the trunk muscles, which developed rapidly after a feverish attack, passed away within a fortnight, and at the end of three weeks the child was quite well; such a result is very rare.

In forming an opinion with regard to the restoration of power two indications are helpful, namely, the rate of improvement and the faradic irritability of the muscles. The earlier, improvement in power begins and the quicker its progress the better the prognosis. If at the end of three weeks muscles respond to the faradic current they will eventually recover, but if no

response is then obtained, some loss of power will be permanent.

The prognosis is bad if, in spite of careful treatment, the amount of paralysis has remained stationary for several months, and if the affected muscles show no response to any form of electrical stimulation.

Is the atrophied corpus, which is present in an old case of infantile paralysis, likely to become the starting point for fresh disease? The answer is that very rarely this may occur, for cases have been observed in which progressive muscular atrophy and amyotrophic lateral sclerosis appeared to be related to an attack of acute poliomyelitis occurring in infancy.

Treatment. A child suffering from a febrile attack from any cause should be put to bed and, if the attack is associated with paralysis, rest in bed should be continued for at least a month in order that inflammatory changes in the nervous system may have time to subside. The side or the prone position is better than the continued recumbent posture. The bowels should be freely opened by calomel, castor oil or liquorice powder. During the pyrexial stage the patient should keep to liquid food, and small doses of aconite or salicin may be given every four hours. When severe pain is present relief is afforded by placing the child on a water-bed and wrapping its limbs in cotton-wool; sometimes phenacetin or antipyrin is required and rarely an opiate may be necessary.

Mild counter-irritation to the spine by the application of a poultice made with one part of mustard and three parts of linseed meal will ease pain in the back and tend to lessen any rigidity of its muscles. If the patient suffers from severe headache which is not relieved by ordinary remedies, the withdrawal of a small quantity of cerebro-spinal fluid by lumbar puncture may give relief. For much restlessness and for convulsions, which occur in some cases, sodium bromide should be given. The condition of the bladder

requires attention, for sometimes there is retention of urine when catheterisation is necessary.

When the muscles of the thorax are involved, care must be taken to avoid the risk of bronchitis or pneumonia. Embarrassed respiration may be relieved by inhalations of oxygen; if life is threatened artificial respiration should be performed in the hope that the wave of respiratory paralysis may subside.

After the acute stage has subsided there is often much prostration, when great care is needed to ensure complete rest to the patient and to protect him from every form of disturbance.

Gentle massage of the paralysed parts may be commenced at an early period, but it is advisable to postpone electrical treatment for four or five weeks in order that complete rest may not be interfered with. As soon as the patient's general condition is satisfactory the flaccid wasted muscles should be systematically rubbed and kneaded twice daily, and with sufficient force to stimulate the circulation in the hope that an increased flow of blood and lymph will promote the nutrition of the affected part. The local circulation is also improved by daily sponging in warm salt water, followed by brisk rubbing and afterwards keeping the limb warm by cotton-wool or extra flannel clothing.

Passive movements are also of great benefit, and the patient should be encouraged to put forth as much voluntary power as possible. If unable to move the weak limb he should be told to move the healthy limb against resistance, for by so doing, movements are sometimes excited in the paralysed limb. As soon as any voluntary power is regained it is desirable to institute a source of muscular exercises: a well-selected series of movements should be prescribed and much attention given to their proper performance.

Electricity, although less valuable than massage, is an important agent in the treatment of infantile paralysis. It is highly improbable that its application to the spine can have any effect on the cord lesion, but

there is satisfactory evidence that its application to paralysed muscles will help to promote their nutrition. On examination it will be found that a certain number of muscles respond to faradism; these muscles will ultimately recover, and their recovery is hastened by the application of either the faradic or the galvanic current. Many of the muscles, however, do not react to faradism, and then galvanism is alone of service.

In applying galvanism it is convenient to apply one large flat electrode, well soaked in warm salt water, to the chest or back, whilst the other electrode, a small one, is stroked over the affected muscles. The stroking or treatment-electrode should be the pole—negative or positive—which causes the most active contraction. The weakest current that will cause a contraction should be used, and in order to avoid frightening the child it is desirable at first to make several applications of the electrodes while no current is passing. In this way the child, becoming accustomed to the apparatus, gains confidence.

A convenient method of applying galvanism to the lower limbs is to place each foot in a separate bath of warm water with the positive electrode in one bath and the negative in the other.

Too much stress cannot be laid on the importance of persevering with massage, electricity and active and passive movements of the affected parts for at least a year. It is astonishing how much restoration of power may sometimes be effected in a limb which at first seemed hopelessly paralysed.

During the treatment, attention must be paid to the position of the limb in order to check, as far as possible, the development of deformities. Thus if the lower limb is paralysed, the patient must not be allowed to lie with the knee and hip flexed. A dropped foot should be protected by a cradle, from the weight of the bed clothes, and it may be desirable to support the foot at a right angle to the leg by means of an "artificial muscle." The counteraction of other deviations may

usually be accomplished by the exercise of a little practical ingenuity.

It is doubtful whether any medicine has an influence on the disease; in some cases strychnine has seemed to be beneficial. General tonics, as iron, quinine and arsenic, and cod liver oil are also useful. Recently the taking of urotropin (hexamethylentetramin) has been advocated. In experimental poliomyelitis in monkeys, Flexner and Clark have demonstrated the presence of urotropin in the cerebro-spinal fluid soon after the administration of a large dose by the mouth. They have shown that when the virus of poliomyelitis is injected intracerebrally in monkeys, urotropin being already present in the fluid, and the drug is subsequently administered by the mouth, that in some of the animals so treated the incubation period is prolonged and the onset of paralysis is prevented.

When after prolonged treatment there appears to be no hope of further improvement, the skill of the orthopædic surgeon may be required to correct deformities, to support loose joints, and in other ways to minimise the effects of paralysis. Various mechanical appliances, tenotomy, resection of joints, tendon transplanting, nerve grafting and even amputation are measures which have to be considered in different cases.

CHAPTER VI.

ACUTE ASCENDING PARALYSIS—LANDRY'S PARALYSIS.

In the year 1859 Landry described a case of acute generalised flaccid paralysis in which the paralysis began in the legs and involved in rapid succession the arms, the trunk and some of the cranial nerves. Sensory disorders were not prominent, but tactile sensibility was diminished in the distal segments of the limbs and was completely lost over the tips of the fingers. The reflex

reactions were lost. The bladder and rectum were not affected. The patient died quite suddenly about a fortnight after the onset of the paralysis. A careful microscopical examination failed to detect any morbid changes in the spinal cord; the peripheral nerves were not examined.

Since that date many similar cases have been recorded and there has been much difference of opinion in regard to their pathology. Lesions have been found in some cases in the spinal cord, in others in the peripheral nerves or in both nerves and cord, while in a fourth group of cases no evidence of a morbid process could be detected in either situation.

The term Landry's paralysis has served as a convenient name for a number of cases of rapid generalised paralysis in which the pathological diagnosis during life was uncertain. Many authorities would exclude cases in which sensation is affected, and restrict the term to cases of uncomplicated motor paralysis presenting in other respects a clinical history similar to that described by the discoverer.

It is clear, from the presence of a flaccid paralysis and the absence of the reflexes that some portion of the lower motor neuron is the part mainly implicated. The question to be settled is, which portion is primarily affected, is it the cell body of the neuron or its peripheral axon, in other words, is Landry's paralysis an affection of the anterior horns or of the peripheral nerves.

Recent writers, for example F. Buzzard, appear to regard the disease as an acute toxic poliomyelitis, which is to be definitely separated from acute toxic polyneuritis. Now although it is impossible to determine the pathological nature of the case described by Landry it is clear that its symptoms correspond more closely to those of a multiple neuritis than to those of lesions limited to the anterior horns. Thus the cutaneous sensibility of the extremities was distinctly impaired, and Landry in his description of the disease

based on an analysis of ten cases, says that sometimes sensibility and motility may be *equally* affected.

In relation to this question the order in which the muscles are affected is of importance. Landry expressly states that the distal parts of the limbs are affected before the proximal parts, the arms before the trunk, and the diaphragm before the intercostals. Such an order is a characteristic feature of multiple neuritis, a feature which curiously enough F. Buzzard says does not occur in the disease which he describes under the heading of Landry's paralysis. In this disease he says that the order in which the muscles are attacked corresponds to the order in which they are innervated by the spinal cord. But surely if the term Landry's paralysis is to be retained it must be applied to cases presenting the clinical manifestations observed by the discoverer.

In our present state of knowledge it is often impossible in acute cases of ascending paralysis to distinguish between those due to acute changes in the peripheral nerves and those due to acute changes in the spinal cells. A description of the chief differences between the two conditions will be found in the section on diagnosis; the indications there given may help in making this distinction, which should always be attempted. In the clinical account of Landry's disease, the symptoms met with in a case thus classified should be minutely recorded irrespective of their assumed central or peripheral origin.

Etiology. The disease chiefly occurs between twenty and forty years of age and it affects men more often than women. In many cases it develops apparently in the midst of good health; in other cases it has followed exposure to extremes of heat or cold. Sometimes the probable cause has been one of the poisons known to produce peripheral neuritis; thus the disease has followed excesses in alcohol, or has occurred in connexion with some infective disorder.

Symptoms.—*The period of invasion.* In some cases the onset of paralysis is quite abrupt; in others it is

ushered in by certain premonitory symptoms, lasting a few hours, days or even weeks. The most frequent premonitory or initial symptoms are sensory in character, and consist of numbness, tingling, formications, and other paræsthesiæ which mainly affect the fingers and toes and the peripheral parts of the limbs. Pains of various kinds may also be present: thus there may be diffuse aching of a limb, or of the back; or acute pains shooting along the course of some of the chief nerves, or localised to one nerve, as the sciatic. Attacks of gastralgia or diarrhœa have also been noticed. The muscular masses of the limbs, especially the calves, may be painful and tender; and pain may be readily produced by any active or passive movement of the body. In other cases feelings associated with muscular weakness have been present; such as langnor, heaviness of the limbs, and fatigue after slight exertion. Occasionally the premonitory symptoms have been motor in character; weakness of the legs being present for a few weeks before the rapid advance of ascending paralysis. During the initial stage vaso-motor and secretory disorders have also been observed, such as dead, cold, or livid extremities, or clammy sweating of the palms and soles. It is probable that the premonitory stage tends to be longer when the disease is set up by the action of a definite poison like alcohol, or like the toxines which result from diphtheria or septicæmia, than when it occurs in persons who have not been exposed to the action of any recognisable toxic agent.

The period of paralysis. The sensory symptoms of the premonitory stage may persist; as a rule they are not conspicuous features. Sometimes the cutaneous sensibility is normal, sometimes it is distinctly blunted; occasionally marked anæsthesia has been observed. Cutaneous and muscular hyperæsthesia and tenderness on pressing over the nerve trunks have also been observed.

Disorders of sensation, however, although usually present in some degree, are completely overshadowed

in intensity by the motor paralysis which dominates the disease. Generally indeed, the first striking symptom is weakness of the lower limbs, one limb being often affected before the other. The weakness increases, and the power of walking or standing without support is quickly lost. When lying down the patient may still be able to move the toes or feebly to flex the ankle and knee; but in a few days, or even a few hours, every trace of motor power in the lower extremities may be abolished; they then lie flaccid and powerless, and the feet and toes assume the dropped position imposed upon them by the action of gravity and by the pressure of the bed-clothes.

Soon after the onset of weakness in the legs, or even simultaneously with this, the arms become attacked. At first the finer movements of the fingers and thumb are enfeebled; then the grosser movements of the wrist, elbow, and shoulder, until in a short time the upper limbs may become as completely paralysed as the lower. The muscles of the pelvis, loins, and abdomen are now invaded. The power of sitting up is soon lost; while, owing to the weakness of the abdominal muscles, the acts of coughing, sneezing, and defæcation become weak and ineffective. In many cases, too, the muscles which move the head are attacked, so that the power of rotating the head or of raising it from the pillow may be lost.

Further progress of the disease is usually indicated by implication of the muscles of respiration. As a rule the diaphragm is attacked before the intercostal muscles: when the latter are paralysed breathing is carried on by the accessory muscles of respiration. Soon these also fail and the patient after a short struggle dies from asphyxia.

Should the patient live for some time after embarrassment of respiration has set in, some of the muscles supplied by the cranial nerves may become paralysed. Those which preside over the movements of swallowing and speech are most commonly affected. Disorders of articulation depend in different cases on paresis of the

tongue, lips and soft palate. Sometimes phonation is affected; a feeble voice may depend on weakness of the muscles of respiration, but qualitative changes in phonation indicate paralysis of some of the laryngeal muscles. Sometimes facial paralysis, unilateral or bilateral occurs; more rarely paralysis of the masseters or of some of the eye muscles.

Psychical disorders are rarely present, and the patient who cannot move his body or limbs and can scarcely express himself by speech, may give evidence that his mental functions are unimpaired.

The paralysis is of the flaccid type but atrophy does not occur in acute cases which end fatally in a short time. In cases, however, which end in recovery, or in which life is prolonged for a few weeks, some of the muscles become wasted and may show altered electrical reactions.

In the paralysed parts all the deep and superficial reflexes are quickly lost, and in the fatal cases are lost at death. When recovery takes place the reflexes may slowly return, but not as a rule until all traces of paralysis have disappeared. Retention of urine and obstinate constipation occasionally occur, and are probably due mainly to weakness of expulsive efforts, owing to paralysis of the abdominal muscles and the diaphragm. Overflow of urine may occur as the result of a distended bladder.

It is exceptional for the disease to be ushered in, or accompanied by fever; towards the end of life the temperature may be raised and the pulse quickened. Profuse sweating, albuminuria and enlarged spleen have been observed in some cases.

Course and Prognosis. The usual course followed by Landry's disease cannot be better summarised than in the words of the discoverer. He says the first phenomena always manifest themselves at the extremities of the limbs, and most frequently at the extremities of the lower limbs. The paralysis pursues an ascending course, and attacks the muscles in an almost constant

order: namely, in the first place, the muscles which move the toes and feet, then the posterior muscles of the thigh and pelvis, and, lastly, the anterior and internal muscles of the thigh; in the second place, the muscles moving the fingers, the hand, the arm upon the scapula, and, lastly, the forearm upon the arm; thirdly, the muscles of the trunk; fourthly, the muscles of respiration, and finally, those of the tongue, pharynx, and œsophagus.

Sometimes the arms are attacked before the legs; sometimes both are attacked simultaneously; a few cases have been recorded in which the muscles supplied by the bulbar nerves were paralysed as soon as those of the upper extremities, or even before them—the muscles of the lower limbs being the last to be implicated.

The usual order in which the muscles become paralysed corresponds to that observed in multiple neuritis. But there are other cases in which the paralysis spreads from one part to another according to the order in which the muscles are innervated by the spinal cord; the trunk being affected before the arms, and the lower intercostals before the diaphragm. These cases probably depend on toxic changes in the ganglion cells of the spinal cord.

The rate of progress and the duration of the disease vary greatly in different cases. In fatal cases death generally occurs before the tenth day; it may take place in forty-eight hours, or not for two or three weeks. The mode of death is nearly always by asphyxia, in consequence of respiratory paralysis.

There can be no doubt that cases presenting symptoms identical with those of the fatal cases do occasionally end in recovery; a patient may lie completely powerless, with marked respiratory paralysis, and yet make a complete recovery. Paralysis may cease to advance at any stage of its progress; the muscles last attacked are the first to recover. The most favourable cases are those in which there is no paralysis of the diaphragm or of the

intercostals, and in which the loss of power, although widely distributed, is not complete.

Pathology. As already pointed out the condition of the nervous system in cases reported under the heading of acute ascending or Landry's paralysis is very inconstant. In one set of cases no changes were found, even when the cord and nerves had been carefully examined. In another series changes were found in the spinal cord; sometimes they were widespread, and similar to those met with in acute myelitis; sometimes they were limited to the anterior horns and similar to those found in the early stage of acute anterior poliomyelitis. F. Buzzard has observed the following: Early pericentral chromatolysis of the cells of the anterior horns and of Clarke's column, together with a loss of chromatin granules, and eccentration of the nucleus; the most marked cellular changes being usually found in the lumbar region. In some cases he observed a diffuse fatty change in the fibres of the spinal cord and to a less extent in the peripheral nerves; some of these fibres showed true Wallerian degeneration; neuroglial proliferation and vascular changes were not found.

In a third series of cases changes were found in the peripheral nerves only. Finally in a fourth series morbid changes were observed both in the central and the peripheral parts of the nervous system.

Kruger believes Landry's disease to be the terminal phase of a chronic multiple neuritis, which has spread by direct continuity to the cord, and thence to the bulbar nuclei. Recently Schweiger in a typical case ending fatally on the seventh day of the disease, found an intense interstitial neuritis in many of the nerves, especially the vagi, also proliferative changes in the connective tissue, and degenerative changes in the cells of the spinal ganglia. He believes that the toxin affects first the peripheral nerves and ascends through them to the cord.

With regard to the bacteriology of Landry's paralysis nothing definite has been made out. In many cases no

organisms have been found; in the blood of one case Roger and Josué found a diplococcus resembling that of pneumonia. From the blood of another case F. Buzzard obtained a micrococcus which when cultivated and injected into a rabbit produced a rapidly spreading paralysis. Buzzard believes "that if the disease is due to some bacterium, the latter does not infect the cord itself, but exerts its influence either through the blood or the lymphatic system."

Diagnosis. If, as I believe, the cases of paralysis described by Landry are composed of two groups, the one group depending on acute toxic polyneuritis, the other on acute toxic poliomyelitis, it is desirable and important especially as regards prognosis to enumerate the chief points of distinction between them, and secondly the features which characterise the usual variety of acute poliomyelitis.

In *acute polyneuritis* sensory symptoms, namely, paræsthesia, anæsthesia, tenderness of muscles and nerves, are usually present. Paralysis is more prominent in the limbs than in the trunk, and in their peripheral than in their proximal portions. Of the trunk muscles the diaphragm is especially liable to be paralysed. The facial is more frequently affected than any other cranial nerve.

In *acute toxic poliomyelitis* sensory symptoms are absent. The muscles of the trunk and the limbs tend to be equally and symmetrically affected by paralysis; the intercostals are often involved before the diaphragm. When the cranial nerves are involved, the muscles of articulation, deglutition, and phonation suffer first. Atrophy and electrical changes do not occur, whereas both may occur in the neuritic group.

In *acute anterior poliomyelitis* constitutional symptoms and pyrexia are often prominent early features. The patient may suffer from severe pains in the back and limbs; anæsthesia does not occur. There is frequently a want of symmetry in the paralysis on the two sides

of the body, or a muscle or a group of muscles in a paralysed region may escape.

Treatment. The patient should be kept at complete rest upon a water-bed and well protected from exposure to cold. A purge may be necessary. If any fever is present aspirin, salicylate of sodium, or a diaphoretic mixture is desirable. When breathing is embarrassed, owing to paralysis of the respiratory muscles, the administration of atropine and strychnine may afford relief by diminishing the bronchial secretion. Sometimes life is prolonged by artificial respiration and the inhalation of oxygen.

When the dangerous stage has passed, atrophy of the flaccid muscles may be prevented or reduced, and muscular power restored by the help of massage and electricity and the administration of strychnine and other tonics.

CHAPTER VII.

CHRONIC ATROPHIC PARALYSIS OF SPINAL AND BULBAR ORIGIN.

Under this heading it is desirable to group together the conditions known as progressive muscular atrophy, amyotrophic lateral sclerosis, progressive bulbar paralysis, and progressive ophthalmoplegia; both clinically and pathologically they may be regarded as different forms of the same disease. The essential lesion is a primary degeneration of motor neurons. In some cases degeneration is limited to the lower neurons, but in the large majority it involves the upper neurons, as well as the lower. The clinical features vary according to the group of lower neurons primarily affected, and according to the degree in which the upper neurons are also involved.

In progressive muscular atrophy some of the muscles of the limbs become weak and wasted; signs of spasticity are slight or absent. There is atrophy of certain groups

of cells in the spinal anterior horns, as a rule this is associated with a slight degeneration of the pyramidal fibres.

In amyotrophic lateral sclerosis, progressive wasting of muscular tissue is combined with decided spasticity of the limbs. These symptoms depend on degenerative changes affecting both the anterior horns and the pyramidal tracts.

In bulbar paralysis, atrophy begins in the muscles supplied by some of the lower cranial nerves; the nuclei of these nerves show a degenerative atrophy; frequently too there is sclerosis of the pyramidal fibres.

Similarly in progressive ophthalmoplegia, paralysis of the ocular muscles results from degeneration of the oculo-motor nuclei.

PROGRESSIVE MUSCULAR ATROPHY: CHRONIC ANTERIOR POLIOMYELITIS.

The progressive atrophic paralysis which characterises this disease is dependent on a gradual wasting of the ganglionic cells in the spinal anterior horns: this wasting is a result sometimes of chronic inflammatory, sometimes of simply degenerative changes. On this basis some authors attempt to separate cases of "subacute" and "chronic poliomyelitis" from cases of "progressive muscular atrophy," in which degenerative changes are supposed to occur apart from inflammation. Their reasons are that the former conditions are distinguished from the latter condition by the following clinical features: Loss of power precedes muscular atrophy; the muscles of the upper arm or those of the leg are first affected; the course of the disease is more rapid than in progressive muscular atrophy. But clinically every gradation may be traced between the two groups of cases, and on the whole it seems desirable (1) to consider their symptoms together, and (2) to enumerate the chief clinical types that may be distinguished.

As already mentioned the morbid process may be limited to the lower motor neurons, but more commonly it implicates the upper neurons as well; both systems of neurons are markedly affected in amyotrophic lateral sclerosis. But here again every transition may occur between a typical case of uncomplicated anterior poliomyelitis and one of amyotrophic lateral sclerosis. It is convenient to describe them separately, although it must be admitted that the separation is somewhat artificial.

One other point requires mention: The term "progressive muscular atrophy" is a clinical one, and strictly speaking ought not to be restricted to the present disorder, for a progressive atrophic paralysis occurs in other conditions, for example, in multiple neuritis and in the muscular dystrophies. The name however is of old-standing, and it would therefore be difficult to alter a term so generally accepted.

Etiology. The disease is one of adult life and usually begins between the ages of twenty-five and forty. Occasionally it occurs in childhood, when hereditary tendencies can be more frequently traced than when it occurs in the adult (see p. 236).

It is commoner in males than in females, and especially in those who do manual work. Sometimes the limb first attacked has received a severe strain or other injury; this would suggest a reflex origin as in the case of articular muscular atrophy, or that the injury to the limb has caused minute hæmorrhages or other vascular changes in the spinal cord: the disease may develop some time after the injury. Similarly the lesion left by acute poliomyelitis in infancy may be the starting point of degenerative changes occurring at a later period of life. Rarely the disease has followed measles, or some other infectious malady, also syphilis, acute rheumatism and lead poisoning.

Symptoms. The first thing usually noticed by a patient is weakness or wasting of some of the small muscles of the hand. Abduction and apposition of the

thumb are often the earliest movements to be affected: subsequently the abductor indicis becomes weak. Paralysis of these muscles is indicated by disability to perform delicate and complicated movements, such as buttoning the clothes or picking up a pin. The disease now spreads to the other interossei, to the lumbricales, and to the muscles of the little finger, when abduction and adduction of the fingers become difficult or impossible. Atrophy of these muscles, which may precede loss of power, is shown by the deep furrows between the metacarpal bones and by flattening of the thenar and hypothenar eminences. As a rule, one hand, generally the right, is invaded before the other, which in a few months, however, is also affected.

A characteristic deformity is soon produced by the weakness of the interossei, which permits the long flexor muscle to bend the phalangeal, whilst the long extensor hyper-extends the metacarpo-phalangeal joints; hence the hand has the appearance of the talons of a bird, and is called the *main en griffe*, or claw-hand. The thumb becomes rotated out, so that its palmar surface lies in a plane parallel to that of the palm; this is described as the monkey-hand. Weakness and wasting now spread up the arms, one limb being affected in advance of the other. Atrophy of the forearm muscles, the flexors being the first affected, leads to disappearance of the claw-shaped appearance of the hand; ultimately the hand may resemble that of a skeleton. At a later period the deltoid, the upper part of the pectoralis major, the biceps, the brachialis anticus, the spinati, the teres, and the subscapularis become atrophied; then the arms, thin and weak, hang like flails by the sides of the body.

When the serratus magnus is much affected and the arm is held out horizontally, the posterior border of the scapula projects like a wing. The rhomboids and the lower portion of the trapezius may be also involved, when the movements of the scapula will be still further impaired.

Figs. 82 and 83.—Photographs of the hand in a case of progressive muscular atrophy, unassociated with symptoms of lateral sclerosis, showing the characteristic claw-like deformity and the wasting of the thenar and hypothenar muscles.

Even when the disease is advanced and wide-spread the following muscles usually escape, namely the upper part of the trapezius, the latissimus dorsi, the triceps and the lower half of the pectoralis. As the disease advances the muscles of the neck, especially the extensors, tend to become involved: in extreme cases the head falls forwards, allowing the chin to rest on the sternum. Finally the muscles of respiration are attacked; and when, as is generally the case, the intercostals are mainly involved, respiration becomes purely diaphragmatic.

In very advanced cases, owing to wasting of both the muscular and the adipose tissues, the body becomes greatly emaciated, and the bones seem to be covered only by skin and fascia.

During the course of the disease fibrillary tremors may be observed in many of the affected muscles. These tremors are often seen in muscles which are not obviously atrophied: they then indicate that atrophy is about to occur. On striking the muscle, a slow and prolonged contraction occurs and fibrillary tremor is produced or, if previously present, is increased. In the affected muscles progressive electrical changes can be demonstrated presenting every variation from the partial to the complete reaction of degeneration.

The superficial reflexes are diminished or lost when the muscles upon which they depend are degenerated, or when the upper neurons are involved: in the latter case the plantar reflex may be extensor in type.

The knee-jerk may be normal, lost or increased. Its diminution or loss is concurrent with atrophy of the extensors of the knee. Its exaggeration, which is common, is a sign that the pyramidal tracts are involved.

Apart from occasional aching pains in the limbs sensation is not affected in any way. The visceral functions are rarely disturbed.

Clinical Types. (1) The *common type*, as above described, in which wasting begins in the small muscles

of the hands. In some cases the atrophy remains more or less limited to these muscles, but as a rule it spreads up the arms.

(2) *The shoulder type* in which the deltoid and other muscles about the shoulder as well as those of the upper arm become atrophied, while the muscles of the hand and forearm with the exception of the supinator longus (which is wasted) are spared. Many cases of this type owe their origin to injury and depend on vascular changes in the anterior horns—anterior poliomyelitis rather than progressive muscular atrophy: others to some chronic intoxication, as that of lead.

(3) The *forearm type*. In one variety the extensor, and in another the flexor muscles of the wrist and fingers are first affected.

(4) The *leg type*. Here the atrophic paralysis begins in the peronei and spreads to the anterior tibial group and often to the thigh and glutei muscles. In one case under the author's care, both lower limbs were completely paralysed, and according to the history weakness preceded the wasting, suggesting that the initial lesion was inflammatory in nature. The knee-jerk and the plantar reflex were lost. There were no other symptoms besides the atrophic paralysis.

Pathology. A microscopical examination of the spinal cord shows that the cells which preside over the nutrition of the affected muscles are atrophied and shrunk, some having entirely disappeared. The anterior root fibres are degenerated and so also are many of the fibres in the peripheral nerves which supply the atrophied muscles. It is remarkable that in some cases the degeneration is limited to the intra-medullary portions of the motor roots, the extra-medullary portions and the corresponding motor nerves being normal, or only slightly changed.

The morbid process may begin in the cells at the level of the first dorsal roots, as in the common variety: or at the level of the fifth cervical root as in the shoulder

type of the disease; or rarely in the lumbar enlargement, when the muscles of the legs are affected.

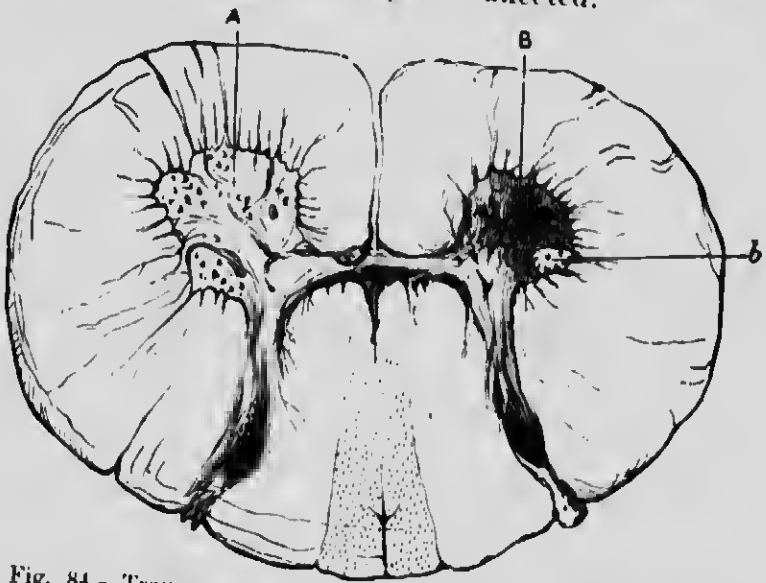


Fig. 84.—Transverse section of the cervical region of the spinal cord, from a case of progressive muscular atrophy. (Charcot.) A. Left anterior grey horn; the ganglion cells have persisted, but are much altered in appearance. B. Right anterior grey horn, almost complete atrophy of the cells, one group only (b) having persisted.

The anterior horns may be alone diseased; more frequently the pyramidal tracts are also degenerated, sometimes even in cases which present no spastic symptoms during life. Other portions of the cord do not always escape; frequently there is degeneration of the fibres of the antero-lateral column, especially of the commissural fibres which are adjacent to the anterior horns. Degenerative changes have also been observed in the spino-cerebellar tracts, in Clarke's column, and occasionally in Goll's column.

As to the cause of the changes in the motor cells, nothing is definitely known. Sometimes there is evidence, in the presence of dilated and thickened vessels, that the process is inflammatory; but frequently such evidence is wanting; the process then seems to be a simple degeneration of the affected neurons.

In a few cases—as the author can testify—the changes

appear to be started by irritation of sensory fibres as from injury to a limb. In relation to this origin the occasional development of progressive muscular atrophy in persons who suffered from infantile paralysis in childhood must be borne in mind, for such cases suggest that a local lesion may have some influence in starting the progressive atrophy.

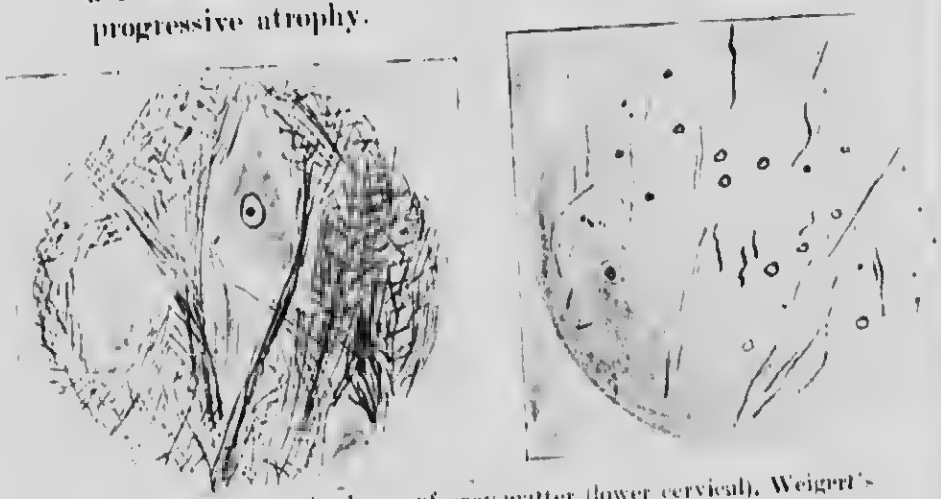


Fig. 85.—Anterior horns of grey matter (lower cervical), Weigert's stain. I. Normal, showing numerous fine nerve fibres and two nerve cells. II. Anterior horn in progressive muscular atrophy (same method of staining), nerve fibres very scanty, nerve cells completely degenerated. (Williamson.)

Diagnosis. Atrophy of the distal or of other muscles of the arms occurs in syringomyelia, in pachymeningitis and in tumours and caries in the cervical region of the spinal cord. But in these affections sensory symptoms are prominent, whereas they are absent in anterior poliomyelitis. In one form of lead paralysis the small muscles of the hands are picked out and there is no sensory disturbance; frequently too some of the arm muscles are also wasted, but as a rule the extensors rather than the flexors of the wrist are involved. In such cases the diagnosis may be difficult or impossible; indeed the lesion may be in the anterior horns, the blue line on the gums and other signs of lead poisoning being indications that lead is the cause of the poliomyelitis.

When progressive muscular atrophy begins in the

shoulder muscles the condition may closely resemble that of one of the myopathies. In the latter, fibrillary contraction of muscular fibres is absent and there is no reaction of degeneration. The distribution too of the atrophy presents differences; in poliomyelitis the latissimus dorsi, the lower half of the pectoralis major and the triceps often entirely escape, whereas they may be markedly atrophied in the myopathies, and so also frequently are the platysma, the levator anguli scapulae and the upper part of the trapezius, these muscles being spared in poliomyelitis. Enlargement and hardening of some portion of a muscle as the lower part of the deltoid, while its upper part is wasted is also often present in a myopathy. Exaggeration of the knee-jerk, not uncommon in poliomyelitis, does not occur in the myopathies.

The muscular atrophy occasioned by the presence of a cervical rib is unilateral in distribution, and is associated with sensory changes. A correct diagnosis is readily made by an *x*-ray examination.

Course and Prognosis. The disease is slowly progressive, and although periods of arrest may occur, life is not generally prolonged for more than from five to fifteen years. More rapid cases occur which end fatally in a year or two; in these cases fibrillary tremors in the muscles are usually marked and wide-spread. The variable duration of the malady depends to some extent on the involvement of the respiratory muscles and on the accompanying risks from bronchitis or pneumonia. A more hopeful prognosis may be given in cases of traumatic origin and in some of them, as the author has seen, complete recovery may ensue.

Treatment. Attention to the general health is of the first importance, for there can be no doubt that good feeding, fresh air and sunshine, together with tonics and cod liver oil have a decided influence in arresting the progress of the disease.

The hypodermic injection of nitrate of strychnine seems to have done good in several cases; at first one

hundredth of a grain should be administered daily, and this may be rapidly increased to one fortieth of a grain, or even more. During this treatment the patient must be carefully watched, and it is well to intermit the drug from time to time.

Massage and galvano-faradism, regularly employed, tend to arrest the muscular atrophy: systematic exercises are also useful.

The Hereditary Form of Spinal Muscular Atrophy in Children.

This is a rare type of progressive muscular atrophy which begins in the first year of life, and often attacks more than one member of the family.

The first symptom is symmetrical weakness of the flexors of the hip and of the quadriceps femoris: subsequently atrophy is seen which is accompanied by weakness. The atrophic paralysis then extends to the muscles of the trunk and of the shoulder-girdle; subsequently it may invade the distal portions of the limbs. Later, bulbar symptoms may appear. The knee-jerks are lost, and the wasted muscles give the reaction of degeneration. Fibrillary tremors are sometimes present: contractures may develop after a time. The diseased muscles never show enlargement.

The course of the malady is rapid and death occurs within a year or two, frequently as a result of paralysis of the respiratory muscles.

In only a few cases has a pathological examination been made. The following changes were found:—Atrophy of the anterior horn cells of the spinal cord, of the anterior roots and to a less degree of the motor nerves and their intramuscular branches; and slight sclerosis of the pyramidal tracts.

The disease is distinguished from acute poliomyelitis by its more gradual onset, by the absence of fever and convulsions and by its fatal termination. It differs from the peroneal form of muscular atrophy in the distribution of the atrophic weakness and in the absence of sensory disturbance.

AMYOTROPHIC LATERAL SCLEROSIS.

A slight sclerosis of the pyramidal tracts is not uncommon in progressive muscular atrophy; this gives rise to increased muscular tonus which may be expressed clinically merely by exaggeration of the knee-jerk. In the amyotrophic disease such sclerosis is more marked and more extensive, the result being that tonic muscular

Fig. 86.—Photograph showing atrophy of the interossei, as well as the result of the patient's greatest effort to extend his fingers.

spasm is as prominent a feature as muscular atrophy. The whole of the upper motor neuron may be involved as well as the lower neuron. Other reasons for separating this disease from progressive muscular atrophy are

its more rapid course, and the early appearance of bulbar symptoms.

Etiology. With one exception, namely, that it occurs more frequently in females than in males, the etiology of the disease, so far as it is known, is similar to that of chronic anterior poliomyelitis. Occasionally the disease affects more than one member of the same family; this would suggest a congenital weakness of the motor elements.

According to Collins the malady is most frequently met with amongst the working classes; some of his cases developed shortly after parturition.

Fig. 87.—Photograph showing the attitude of the foot: the same case as Fig. 86.

Symptoms. In its common form, the onset of the disease is gradual. The first noticeable symptom is weakness of the arms; in a few cases this is preceded by pain, or by numbness and tingling. As a rule the small muscles of the hand are first affected with weakness and wasting; then the muscles of the forearm and

arm are successively invaded, the order of their invasion corresponding closely to that of progressive muscular atrophy. The wasting is accompanied by fibrillary twitchings, and in course of time sometimes at an early period -by muscular tension and contractures, so that the limbs begin to assume deformed positions which may be permanent. The arms are adducted to the sides; the elbows and wrists are semi-flexed; the hands are pronated and the fingers strongly flexed into the palms. In some cases muscular resistance is readily evoked by attempts at passive movements, but it tends to diminish as the atrophy increases.

Fig. 88.—The hand in a case of advanced amyotrophic lateral sclerosis.

The deformities of the hand are similar to those of progressive muscular atrophy.

The atrophic paralysis may spread to the muscles of the trunk and neck; the head may fall forwards, allowing the chin to rest on the sternum. The electrical irritability of the wasted muscles is diminished to both

currents and sometimes a partial reaction of degeneration is obtained; at an advanced period all electrical contractility may be lost.

After a few months the lower limbs show signs of weakness, but their muscles instead of wasting become rigid. The knee-jerk is much exaggerated, and ankle clonus and Babinski's plantar reflex can be elicited. The gait is spastic in character; at a later period the muscles become so rigid that walking is impossible and the patient becomes helpless and bedridden. Still later the rigidity may be replaced by flaccidity and the muscles of the legs may undergo a diffused atrophy, but never to such a degree as in the arm-muscles.

The last stage of the disease is characterised by the presence of bulbar paralysis, some symptoms of which may indeed have made their appearance at an earlier period. Speech is usually first affected; then the tongue begins to waste and to show fibrillary tremors; the muscles of the lips are atrophied; the palate and lower facial muscles become paralysed, and also occasionally the muscles of mastication. Ultimately the patient succumbs from inability to swallow, from respiratory paralysis, from cardiac disturbance, or from pneumonia, sometimes the result of inhalation of particles of food.

One striking feature of the disease is the great increase in muscular irritability, as shown by the readily evoked muscular contractions produced by tapping. All the deep reflexes too show marked exaggeration, the jaw, wrist, elbow and knee-jerk. The cutaneous sensibility remains normal. The sphincters are unaffected, except possibly towards the end of the disease.

In addition to the above form of amyotrophic lateral sclerosis, other types are met with. Sometimes bulbar symptoms open the scene and are followed by an atrophic paralysis of the arms and a spastic paralysis of the legs. More rarely a spastic paraplegia is first developed. Every variation too in the relative propor-

tion of muscular rigidity and atrophy may be observed: some cases closely resemble progressive muscular atrophy, whilst in others the chief features are those of lateral sclerosis.

Pathology. The anatomical substratum of the disease is degeneration of the whole motor tract, the upper as well as the lower segment being affected. The motor nuclei in the medulla and pons undergo atrophy, just as do those in the ventral horns of the spinal cord. There is also a primary degeneration of the upper neurons; this probably begins in the most distal portions of the pyramidal tracts and then extends upwards through the medulla, pons, crus and internal capsule to the cortex, where changes have been found in the large pyramidal cells (Betz-cells) of the precentral convolution. The latter changes, however, are not necessarily secondary to degeneration of the pyramidal fibres, and they may be equally intense.

The wasted nerve elements are slowly replaced by connective tissue. The sclerosis is not confined to the pyramidal tracts; it involves other portions of the antero-lateral columns and sometimes the columns of Goll.

As suggested by the varying sequence of the symptoms, degeneration may begin in either segment of the motor path. In most cases it begins in the grey matter of the cervical cord; in other cases it begins in the bulbar nuclei, while in a third series of cases the lateral columns of the cord are first attacked. No doubt it frequently happens that degeneration begins simultaneously in the upper and the lower neurons, but advances more quickly in the one set than in the other. It is improbable that the degeneration spreads from the upper to the lower neurons or *vice versa*.

Diagnosis. Although no sharp boundary line can be drawn between progressive muscular atrophy and amyotrophic lateral sclerosis, typical cases of the latter are distinguished by the spastic condition of the legs, the increase of the deep reflexes both in the arms and the

legs, the tendency to bulbar paralysis and by the shorter duration of the disease. Most cases terminate fatally in from one to four years, whilst cases of progressive muscular atrophy may survive for ten, fifteen or even twenty years.

The differential diagnosis from other diseases is based on the lines already mentioned under progressive muscular atrophy (see p. 234).

Treatment. In addition to the treatment recommended for progressive muscular atrophy, special attention must be given to allaying the spasticity and to the prevention of contractures and deformities. These objects are best attained by the use of hot water, or hot-air baths, followed by massage and passive movements.

PROGRESSIVE BULBAR PARALYSIS: GLOSSO-LABIO-LARYNGEAL PARALYSIS.

Etiology. As a rule this disease, which is rare, occurs during the second half of life and especially after the age of fifty: occasionally it comes on at an earlier period and may even develop during childhood. In the latter case a family disposition is sometimes to be traced, but usually inheritance is limited to an association with other neuroses. In a large proportion of cases, no adequate cause can be discovered: in a few cases the disease appears to have been set up by emotional strain, exposure to cold or other debilitating influence, by lead poisoning, diphtheria, blows on the back of the neck or over-use of the muscles subsequently implicated.

Symptoms. The symptoms begin insidiously and creep on gradually. A slight indistinctness of speech is usually the first symptom to attract attention: it may only occur when the patient is fatigued. At first this is found to depend on a difficulty in pronouncing the consonants l, r, n, t, and s, and a little later in pronouncing o, u, p, b, and m. The former disability indicates a defect in the movements of the tongue, the latter in the movements of the lips. At an early stage

of the disease these defects are limited to the finer movements of the tongue and lips which are necessary for perfect articulation, but gradually the coarser movements of these parts become affected. The patient becomes unable to raise the tip of the tongue towards the hard palate or, after its protrusion, towards the nose; and gradually the power of protrusion, as well as the side to side, and up and down movements become more and more impaired. Increasing weakness of the orbicularis oris leads to inability to blow, whistle, or to compress the lips. Wasting is associated with the paralysis and accompanies it step by step. The tongue loses its plumpness; when grasped between the finger and thumb the thinness of its intrinsic muscles is at once recognised. Its progressive atrophy is also indicated by the fibrillary contractions of its fibres, the contractions occurring in a longitudinal direction: these together with the small size of the tongue, and the wrinkling of its mucous membrane give the appearance of a bag partly filled by worms. Wasting of the lips is not usually so conspicuous: in many cases they gradually get thinner and furrowed and appear to be lengthened: sometimes they are the seat of fibrillary contractions.

The muscles of the palate, and those concerned in swallowing and in phonation are also liable to be implicated. Paralysis of the palate gives a nasal resonance to the voice; when the paralysis is well-marked the palate is not raised during phonation.

Weakness of the tongue alone will cause difficulties in chewing and swallowing, as it hinders the rolling about of food in the mouth and its passage backwards to the pharynx; in order that food may be caught by the reflex movements of the pharyngeal muscles the patient after taking a drink has to throw his head backwards. If the soft palate is paralysed liquids regurgitate through the nose, while weakness of the epiglottic and pharyngeal muscles may permit food to pass into the glottis instead of into the œsophagus. Should this occur death from choking is threatened, for the power

of expulsion by coughing is usually very feeble. Paralysis of the adductors of the glottis is responsible for the ineffectual coughing, as well as for the imperfect phonation.

In some cases the masseters, the pterygoids and the temporal muscles are involved; then the difficulty in masticating food, due to paralysis of the tongue, is increased, for the teeth cannot be brought together nor can the mouth be perfectly closed.

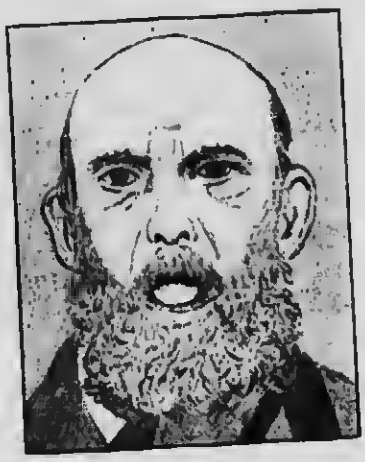


Fig. 89.—Bulbar paralysis of sudden onset, maximum protrusion of the tongue. (Gowers.)

Sooner or later the patient presents a characteristic appearance. The lower lip is loose and pendulous: the mouth is half open, and swallowing being difficult or impossible, the saliva accumulates and is constantly dripping from the angles of the mouth. The tongue is shrivelled and motionless, and speech is lost, or is represented by the slow monotonous mumbling of a few vowel sounds. The contrast between the lower and the upper part of the face is striking, for, except in rare cases, the muscles above the orbicularis oris are not involved: hence the naso-labial folds are well-marked and the orbicularis palpebrarum and the frontalis act in a normal manner.

For a time, the electrical reactions of the affected

muscles are normal; but when atrophy of the tongue is considerable, a partial or a complete reaction of degeneration is often obtained.

Sensibility remains unaffected throughout the whole course of the disease. The reflex irritability of the soft palate, of the pharynx and even of the larynx, is diminished or is abolished, and as the sensation of these parts is preserved, the loss of reflex action must be due to disease of the motor portion of the reflex arc. The tendon reflexes of the limbs are usually exaggerated; they may be normal. Sometimes a pronounced jaw-jerk is obtainable.

The intelligence remains quite clear to the last. The temper may be irritable, and patients often manifest a tendency to laugh or to cry on the slightest provocation; during an emotional attack, the immobility of the lower part of the face contrasts strongly with the activity of the eyes and the lively movements of the upper part of the face.

Course. The downward progress of the disease is sometimes interrupted by periods in which the symptoms seem to be in abeyance, or in which there is even improvement, but a fatal issue is almost invariable, and usually takes place within a couple of years. The patient gradually gets weaker and thinner, the emaciation being largely due to the insufficient quantity of food. Inanition alone may cause death; more commonly it is a contributory factor only, life being ended by respiratory paralysis, by cardiac failure, by suffocation owing to impaction of food in the glottis, or in consequence of a septic broncho-pneumonia set up by the inhalation of particles of food into the air-passages.

Pathology. The morbid changes are similar to those observed in progressive muscular atrophy, but the cells of the bulbar motor nuclei are affected, instead of those of the spinal nuclei. The degeneration affects the hypoglossal and spinal accessory nuclei the most severely; the nucleus ambignus (the motor nucleus for the combined vago-glossopharyngeal nerve), the facial, and the

motor nucleus of the trigeminus are also degenerated though usually in a less degree. In these nuclei the cells are wasted or completely destroyed: the neuroglia is proliferated, the proliferation being secondary to the cell-degeneration: the walls of the vessels are sometimes thickened. The nerve fibres proceeding from the affected centres are also wasted.

As a rule degeneration of the pyramidal tracts is found: occasionally it is absent, the lower neurons alone being involved.

Diagnosis. Paralytic symptoms, closely resembling those of chronic bulbar paralysis, may be caused by vascular lesions in the medulla implicating the nuclei of the cranial nerves. They are distinguished by their sudden onset, by their less symmetrical arrangement, and by their more variable course.

In the course of multiple neuritis, especially when caused by diphtheria, symptoms resembling those of bulbar paralysis may occur. But the history of the case, its rapid progress, the implication of nerves other than the cranial, and the quick recovery which generally takes place will serve as distinguishing features. Very rarely permanent paralysis of bulbar type has followed an attack of diphtheria; there is however no tendency, as in chronic bulbar palsy, for the lesion to progress.

Some cases of bulbar paralysis present a striking resemblance to myasthenia gravis. They are distinguished by: the absence of a tendency to remission and to relapse of symptoms: by the prominence of atrophy of the affected muscles, whereas in myasthenia there is little or no atrophy, although the loss of power may be great: by the presence of the reaction of degeneration, and by the absence of the myasthenic reaction (see p. 462).

Perhaps the greatest difficulty in diagnosis occurs in cases of "pseudo-bulbar paralysis." This condition may result from bilateral lesions involving either the lowest portions of the precentral convolutions, or the pyramidal fibres connecting those centres with the bulbar nuclei. The general history of such cases is

that the patient had an attack of hemiplegia from which he made a partial recovery: this is followed in a few weeks or months by hemiplegia on the other side, which results in paralysis of the lips and tongue; the deglutition being to some extent impeded.

Such a history of the development of bulbar symptoms, together with their character, usually prevents any mistakes in diagnosis. Moreover the upper neurons being affected, the paralysed muscles maintain their nutrition and tone and show no electrical changes: and reflex action is unimpaired. On the other hand, in the true bulbar cases, the paralysed muscles undergo a progressive atrophy; they show fibrillary tremors and give the reaction of degeneration.

Treatment. Little or nothing can be done to arrest the downward progress of this disease. Special attention must be given to the feeding of the patient: jellies, blanc-manges and other varieties of semi-solid food are usually swallowed more easily than either liquid or solid food. In the later stages of the disease and in all severe cases, much care is required to prevent food from getting into the larynx: frequently nasal feeding becomes necessary.

PROGRESSIVE OPHTHALMOPLEGIA: CHRONIC NUCLEAR PARALYSIS.

This variety of ophthalmoplegia is characterised by a slow and progressive development of paralysis of the eye muscles. The paralysis is the result of degenerative changes in the oculo-motor nuclei, changes precisely similar to those that occur in the nuclei of the lower cranial nerves in bulbar paralysis, and in those of the spinal nerves in progressive muscular atrophy. A difference however is to be noted as regards the relation of syphilis to the degeneration in the three types of paralysis. The virus of syphilis is a common cause of progressive ophthalmoplegia: it is only an occasional cause of chronic bulbar paralysis, while it has no etiological relation to progressive muscular atrophy.

The symptoms of ophthalmoplegia may exist alone; more commonly they are associated with manifestations of disease in other parts of the nervous system. Thus the condition often forms a part of tabes or of general paralysis of the insane; it is found in association with optic nerve atrophy, with bulbar paralysis, and in rare instances with progressive muscular atrophy.

Fig. 90.—Photograph of a case of total ophthalmoplegia, with double ptosis.

The first muscles to suffer are usually those that are associated in action, for example the elevators of the lids and the superior recti, or the external rectus on one side with the internal rectus on the other. In some cases there is a gradual extension to all the external muscles of both eyes, and sometimes also to the internal muscles—total ophthalmoplegia. In other cases paralysis remains limited to a particular movement of the eyes, or to a particular muscle; thus there may be isolated palsy of the ciliary muscles, or isolated loss of

the reflex action of the iris. The variety of paralysis, and its combinations are very numerous. Sometimes there is an associated weakness of the orbicularis palpebrarum; this indicates an extensive nuclear degeneration.

The disease is almost invariably progressive, and is ultimately fatal.

CHAPTER VIII.

PERONEAL MUSCULAR ATROPHY.

This is a slowly progressive form of muscular atrophy which presents a clinical affinity with the distal type of myopathy, and pathological affinities with both multiple neuritis and anterior poliomyelitis

As a rule the disease begins in childhood, and only rarely after the age of twenty-five. One or more members of a family may suffer from it; sometimes the malady may be traced through four or five generations. In many cases there has been a previous history of some acute specific infection, especially measles.

Symptoms. The first symptom to attract attention is weakness with wasting of the peroneal muscles, which allows the feet to drop, and to become inverted, so that the child walks on the outer side of his feet. About the same time, or perhaps earlier, the small muscles of the feet begin to atrophy; subsequently the wasting spreads to the muscles on the front of the leg and later to those of the calf. The next muscles to be affected are usually those of the lower thigh, especially the vastus internus, giving what is known as the bottle-shape (the neck of the bottle being downwards) to the thighs. The patient is able to walk at any rate for some time, his gait having the high stepping character.

After a period, varying usually from one to four years, the small muscles of the hand – the interossei and the thenar and hypothenar eminences- gradually become wasted; sometimes the muscles of the forearm are

also implicated; those of the shoulder, the neck and the back are generally spared. The fingers and toes assume a claw-like shape, the terminal phalanges being flexed and the proximal hyperextended; deformities of the feet also develop, talipes equinus and talipes equinovarus being the usual ones.

Fibrillary twitchings may be seen in the muscles; sometimes they are very marked. Electrical changes in the muscles are observed at an early stage of the disease, the most frequent change being a greatly diminished irritability, or a complete loss of response, to the faradic current; it is stated that the full reaction of degeneration may be present at an early period in peroneal muscular atrophy, whereas it is usually a late feature in progressive muscular atrophy.

The ankle-jerk is lost, frequently in the initial stage of the disease; the knee-jerk is preserved unless the extensor muscle of the thigh becomes involved, when it may be diminished or lost; rarely it is exaggerated. The superficial reflexes remain normal, though the plantar reflex is often difficult to obtain.

Pains and cramps in the legs are often complained of. In some cases the cutaneous sensibility remains normal, but as a rule patches of anæsthesia are present on the outer aspects of the legs, and over the soles of the feet. There is no undue tenderness on pressing the nerve trunks or on squeezing the muscles. The bladder and rectum are not affected.

In some cases of peroneal muscular atrophy, tabetic symptoms have been observed, namely, absent knee-jerks, slight ataxia, the Argyll-Robertson pupil and even perforating ulcers on the soles of the feet.

The disease runs a very chronic course; sometimes its progress is arrested. Life is not shortened, except by the advent of intercurrent disease.

Pathology. The following lesions have been found: A degenerative atrophy of the cells in the ventral horns of the spinal cord; degeneration of the intra-muscular fibres of the peripheral nerves, especially of those to the

peroneal muscles; changes in the cells of Clarke's column, and slight sclerosis of the posterior columns and the pyramidal tracts. Changes in the sensory peripheral nerves have also been seen.

Some observers regard the disease as a variety of chronic multiple neuritis, others as a form of chronic poliomyelitis.

Diagnosis. The characteristic feature of this disease is, the gradual onset and progressive development of muscular atrophy, which is symmetrically distributed to the distal portions of the limbs, and is usually associated with slight sensory symptoms.

In multiple neuritis anæsthesia is generally more marked and muscular hyperæsthesia is frequently present. Its course is more rapid than that of peroneal muscular atrophy, and after a variable time the weakness of the muscles ceases to increase and then begins to disappear; whereas in the peroneal affection the atrophy steadily progresses and is permanent.

In progressive muscular atrophy sensory symptoms are absent, and there are differences between the two diseases as regards age, mode of onset, and subsequent course.

Sensory symptoms do not occur in the myopathies; in many cases however the differential diagnosis of peroneal atrophy and the distal type of dystrophy is very difficult.

Treatment. As in other chronic forms of muscular atrophy the treatment mainly consists in the maintenance of the general health, and of the nutrition of the affected muscles by means of massage and electricity. In the early stages of the disease the administration of thyroid gland has been recommended. In the later stages, tenotomies and various orthopædic methods may be of great service.

CHAPTER IX.

THE MUSCULAR DYSTROPHIES: THE MYOPATHIES.

The muscular atrophies hitherto described depend on lesions of some part of the lower motor neuron. But in the present group, degeneration of muscular tissue comes on independently of any changes in the nervous system, and apparently in consequence of a congenital defect in the vital endurance of the muscle-elements. The manifestations of this defect may be present at birth, or may be delayed until puberty or even later.

The characteristic changes which have been observed in myopathic muscles are:—Atrophied and hypertrophied fibres, showing various regressive changes; proliferation of their sarcoplastic nuclei; a deposit of fat and an increase of connective tissue; and lastly a thickening of the walls of the vessels. Slight changes have also been found in the nervous system, namely, a reduction in the size and the number of the anterior cornual cells, together with atrophic changes in some of the fibres of the anterior roots and the motor nerves.

In all probability these neural changes are neither primary, nor coincident with the muscular disease, but are secondary to it, in consequence of injury to the terminal branches of the axis cylinders, as by pressure from proliferating connective tissue.

In some cases of myopathy congenital smallness or degenerative wasting of muscular fibres is alone present. in others the atrophy is associated with an interstitial overgrowth of fat and connective tissue. Corresponding to the predominance of one or other of these changes. cases may be separated into two classes. In the one class muscular atrophy is the distinctive feature; in the other, enlargement of muscles. Between the two classes, however, no trenchant distinction can be drawn. for there are many connecting links, and the relative proportion in which the pseudo-hypertrophied and the atrophied muscles are combined in different cases pre-

sents every variety. But whether enlarged, small, or natural in size, the affected muscles are weak.

The chief clinical types (following Batten) under which the symptoms of the myopathies will be described are:—

The pseudo-hypertrophic type.
The juvenile type (Erb)
The facio-scapulo-humeral type (Landouzy and Déjerine).
The distal type.
The simple atrophic type myatonia congenita or amyotonia congenita.
Myotonia atrophica.

PSEUDO-HYPERTROPHIC PARALYSIS.

Etiology. This is a disease of early childhood: frequently its first symptoms are observed when the child attempts to walk. Sometimes the onset is delayed until the fourth or the fifth year and rarely till after puberty, but in the latter case previous enlargement of muscles may have escaped notice. Boys are attacked much more frequently than girls, the proportion being about five to one. When present in females the disease often develops later, is less severe and less rapidly progressive.

Sometimes several members of a family are affected, and although the females usually escape they may transmit the disease to their sons.

Symptoms. As a rule the first symptom to attract attention is insecurity in standing or in walking: the child easily falls, gets up clumsily, and in going up stairs has to support himself by the banisters. These imperfections depend on muscular weakness, which sooner or later is associated with an alteration in the size of certain muscles.

Enlargement is commonly most conspicuous in the calves, which often feel firmer than natural. It may also be seen in the vastus externus, in the rectus femoris and in the glutei. The muscles on the front of the leg

and the flexors of the knee are not often enlarged; they may be weak and wasted. The flexors of the hip also are usually weak.

Next to the gastrocnemius and the soleus the infraspinatus most frequently shows enlargement: it is often firm as well as bulky and prominent. The deltoid, the supraspinatus and sometimes the triceps may be hypertrophied; the biceps is commonly wasted. The muscles of the forearm and hand are rarely involved. Occasionally a slight enlargement of the extensors of the wrist and fingers, or of the supinator longus has been observed, or the last-named muscle is wasted.

Fig. 91.—Photograph showing enlargement of the masseters in a case of pseudo-hypertrophic paralysis.

One of the most constant features of the disease is a bilateral atrophy or even an entire absence of the latissimus dorsi and of the lower part of the pectoralis major. In consequence of this the axillary folds are thin or practically absent, and it is almost impossible with the hands in the arm-pits to lift the patient up.

for his shoulders are unable to offer any downward resistance.

The erectores spinæ are often weak and sometimes enlarged. The muscles of the face and neck with the occasional exception of the clavicular portion of the sterno-mastoid remain unaffected. Occasionally the tongue and the masseters are thicker than normal; prominence of the latter is sometimes a noticeable feature (see fig. 91).

The relative degree and the distribution of muscular weakness give rise to characteristic defects in the attitudes and movements of the body. In *standing* the child preserves its balance by keeping the feet wide apart and by throwing back the shoulders and upper part of the body, so that a plumb-line dropped from the most prominent vertebral spines will fall behind the sacrum. This attitude is to compensate for the dropping forward of the pelvis owing to weakness of the glutei and erectores spinæ—marked lordosis being the result. In early stages of the disease, the lordosis disappears when the patient is in the sitting posture; in advanced stages it persists.

In *walking* the feet, widely separated, are lifted off the ground with some difficulty and the body is inclined first to one side and then to the other. This characteristic waddling gait is mainly effected by alternate contractions of the gluteus medius: these are necessary to enable the advancing foot to clear the ground. In some cases, when the flexors of the hip are not too weak, there is a "high steppage" gait, the foot being lifted by exaggerated flexion at the knee and hip.

In *rising* from the supine position on the floor, the weakness of the extensors of the knees and hips gives rise to a characteristic series of movements. The patient first rolls over in order to get on to his hands and knees; he then partly extends his knees and supports himself on his fingers and toes. He next moves his hands backwards until he is able to lift first one hand, then the other to his knees, when by gradually working his hands

Fig. 92.—Photographs showing the mode of rising from the ground in pseudo-hypertrophic paralysis as well as the lordosis, and the talipes equinus: the same case as Fig. 91.

up his thighs and pressing on the ground with the soles of his feet he is able to push himself up. A final jerk is generally necessary to extend the hips and throw back the shoulders in order to gain the erect posture. A close observation of this performance shows that a screw-like movement at the hips is associated with the simple extensor movements at the knees and pelvis.

As time goes on certain distortions may develop. The elbows and knees may become fixed in a flexed position. The spine may show a lateral curvature, or the back instead of being hollow becomes convex. Contraction and shortening of the gastrocnemius prevent the patient bringing the heel to the ground and gradually lead to talipes equinus and sometimes to subluxation of the ankle joint.

With an increase in the severity of these deformities together with the spread of muscular weakness the patient gradually becomes bed-ridden, and is helpless except perhaps as regards the movements of his hands. His limbs are very thin and muscles even such as those of the calf, which formerly were enlarged, are now small and wasted.

The electrical irritability of the muscles normal at first, shows a quantitative decrease to both currents when the disease is developed, the decrease corresponding to the degree of muscular change. The knee-jerk which may be normal at first gradually disappears as the weakness of the extensors of the knee increases.

Sensation is unaffected; weakness of the sphincters is very rare. The general health is not necessarily interfered with until the final period of the disease. The mental condition is generally normal, though occasionally there is a certain amount of incapacity.

As a rule the disease progresses very slowly and insidiously, but it is rare for the patient to reach adult life. Death often occurs about puberty in consequence either of exhaustion, or of some intercurrent malady as one of the acute specific fevers, or a lung affection, the weakened respiratory muscles being contributory fac-

tors. In a few cases the disease becomes arrested, especially when it begins late in life.

ERB'S JUVENILE FORM OF MUSCULAR ATROPHY.

In both this and the next variety of myopathy males and females are affected more equally than in pseudo-hypertrophic paralysis.

The juvenile form usually begins in the second decade of life. The muscles of the upper arm and thigh, together with certain muscles of the shoulder and pelvic girdle, undergo progressive atrophy; those of the forearm and leg are generally spared. Hence in a typical case there is a striking contrast between the size of the

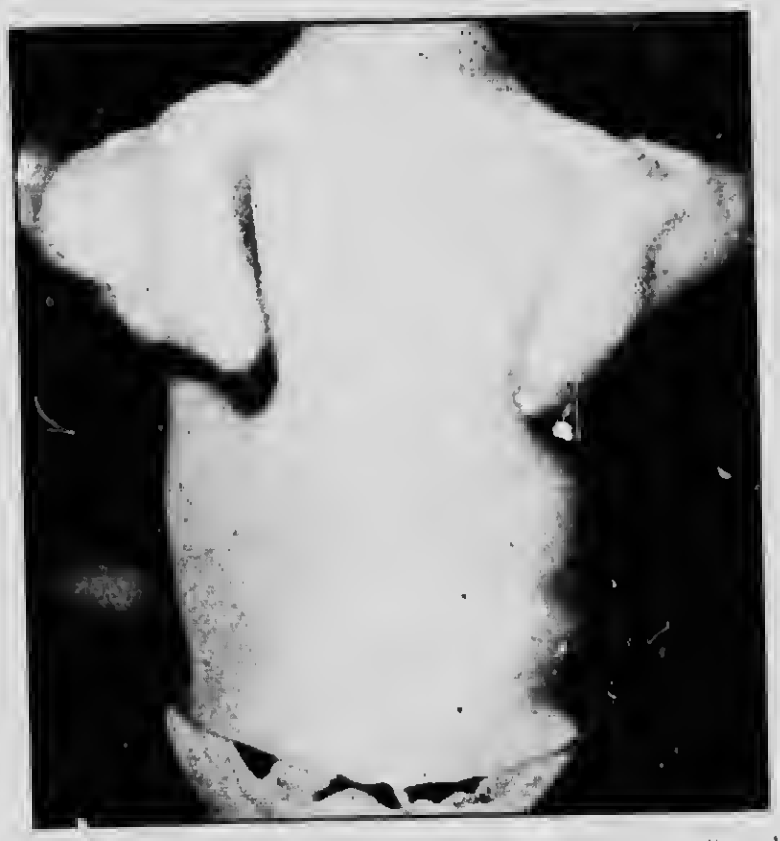


Fig. 93.—Photograph showing marked atrophy of trapezii, and projection of scapulæ owing to atrophy of serrati.

arm and the forearm, and between that of the thigh and the leg.

The biceps, triceps and supinator longus are the first to be affected, one arm being attacked in advance of the other, so that the amount of atrophy is not symmetrical. Subsequently wasting invades the latissimus dorsi, the lower part of the pectoralis major, the trapezius, the serratus magnus and the rhomboids. The whole of the trapezius including the highest clavicular fibres, may become atrophied; this is a distinction from the myelopathies in which the upper portion of the muscle escapes. The platysma is frequently atrophied; occasionally the sterno-mastoids are wasted. The deltoid and the pinati usually escape; sometimes they show enlargement. It is not uncommon to find wasting of the upper, and hypertrophy of the lower part of the deltoid.

When the patient raises his arms in front of him by means of the anterior fibres of the deltoids, the scapulæ, owing to atrophy of the serrati, project like wings, leaving a deep hollow on each side between the posterior border of the scapula and the spinal column.

At a subsequent period of the disease the glutei, the flexors of the hip and the muscles of the thigh especially the quadriceps extensor, become wasted and weak. Occasionally the anterior tibial muscles suffer, when dorsi-flexion of the ankle may be impaired or lost. Weakness of the erectores spinæ also is common and in conjunction with that of the glutei leads to spinal lordosis, which disappears when the patient sits down unless the recti abdominis are also involved.

The gait and the manner of getting up from the ground may be quite similar to those observed in pseudo-hypertrophic paralysis.

THE FACIO-SCAPULO-HUMERAL TYPE OF LANDOUZY AND DEJERINE.

This is a type in which weakness of the facial muscles begins to develop in infancy and may be present at birth. In other respects the distribution of the atrophy

is identical with that of the juvenile form, which in all probability is the same disease. In relation to this it is interesting to note that sometimes the face is involved after the limbs in the juvenile form: this I have observed in several cases.

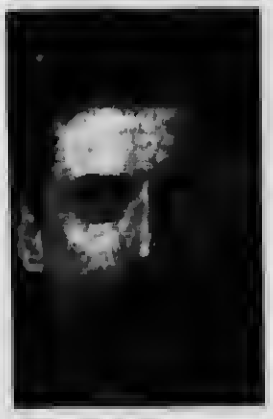


Fig. 91.—Photograph showing slight weakness of the muscles on the right side of the face in a case of the facio-scapulo-humeral type: the same case as Fig. 93.

The hereditary nature of the facio-scapulo-humeral type is sometimes shown by its presence in successive generations; in many cases, however, no other members of the family have been affected.

Fig. 95.—Showing extreme atrophy of biceps, triceps and upper part of deltoid.

The characteristic feature of this type is the early and marked involvement of the facial muscles. The

orbicularis oris and orbicularis palpebrarum are prominently affected, the patient being unable to close the eyes completely and to whistle or blow. The lips are everted, the lower lip projects and there is a certain similarity to the mouth of a tapir. The smile is often peculiar owing to weakness of the zygomatici: the mouth forms a straight line, instead of its angles being drawn upwards and outwards.

After the face the atrophy attacks the muscles about the shoulders and arms; subsequently the wasting may spread to the muscles of the back, hip and thigh. The distribution of the atrophic weakness in the trunk and limbs is the same as that in the juvenile type.

THE DISTAL TYPE.

The characteristic feature of this type is an atrophic weakness of the distal muscles of the limbs, especially the extensors of the wrists and fingers, the extensors of the toes and the dorsi-flexors of the ankles; the proximal muscles are not affected. Sometimes the face is implicated.

In some cases the condition is noticed in infancy, in others not until adult life.

Clinically the disease closely resembles the peroneal type of muscular atrophy; in the latter, however, sensory changes in the legs are often present and the facial muscles are never affected.

AMYOTONIA CONGENITA.

This is a rare infantile malady characterised by smallness and extreme flaccidity of the voluntary muscles, by loss of the deep reflexes and by a tendency to gradual improvement.

The condition is usually congenital, and is noticed at birth or shortly afterwards; in a few cases it has developed in a previously healthy infant, apparently as a result of diarrhœa or of acute bronchitis. Its cause is unknown; there is no evidence of either an hereditary or a familial tendency; the health of the parents has

Fig. 96.

Fig. 97.

Fig. 98.

Fig. 99.

Figs. 96 to 100.—Showing the attitudes in a case of amyotonia congenita, recorded by A. Moussous; attempts to regain the sitting posture from the position seen in Fig. 96 were impossible without help.

been good, including that of the mother during the gestation of the afflicted child.

The limbs, especially the lower ones, are the most severely affected; the trunk is often involved, the face but rarely.

The affected muscles are completely toneless but no localized atrophy nor hypertrophy can be detected. There is general weakness of the limb but no actual paralysis, voluntary power over individual muscles, although feeble, being retained.

In consequence of the great limpness of the muscles together with the relaxation of the ligaments, the limbs are flail-like at all their joints, and may assume the most curious positions. Thus the wrist and the ankle may be so over-extended that the metacarpus touches the forearm and the dorsum of the foot the front of the tibia. The distribution of the muscular flaccidity is symmetrical. The hands and feet are often unusually long.

"As a rule these children never learn to walk, but adopt some strange method of getting about; the child will roll round and round on the long axis of the body in order to get from one part of the room to the other, or will assume a squatting attitude, and it is to this peculiar attitude that the name 'frog-child' was originally applied by Dr. Head to one of my cases." (Batten.)

Contractures are apt to occur in the course of time, usually slight flexor ones at the knee and hip.

The muscles, while reacting normally to the galvanic current, show a lowered excitability to faradic stimulation which, when the amyotonia is severe, needs to be very powerful to obtain any response. It is remarkable that this is borne without complaint, although no loss of sensibility to any other form of stimulation can be detected.

The superficial reflexes are normal; the deep are lost, but return in cases in which considerable improvement takes place in the condition of the muscles. The

sphincters are never affected. The mental condition is normal and the child learns to talk at the ordinary age. The growth of the bones and the general bodily development are not interfered with.

The course of the disease is one of slow and progressive improvement which may be hastened by the persevering use of massage and passive movements. It is impossible to foretell the amount of improvement that may take place short of complete recovery, of which there is no recorded instance.

MYOTONIA ATROPHICA.

This is a very rare condition which, according to Batten, should be regarded as one of the myopathies; clinically it occupies an intermediate position between a typical myopathy and Thomsen's disease, for it is characterised by the association of muscular atrophy with a slow relaxation of the muscles of the extremities after voluntary contraction. This myotonic condition is most conspicuous in the flexor muscles of the hands, so that the patient finds it impossible to relax his grasp suddenly.

The atrophic weakness affects the facial muscles, the sterno-mastoids, the vasti of the thighs, and the dorsi-flexors of the feet, and occasionally the forearm muscles and the masseters and temporals.

The disease is a familial one and brothers are attacked more frequently than sisters. Its manifestations usually develop between the ages of twenty and thirty. The course of the disease is slow and progressive. In many cases muscular wasting is present for several years before the myotonic state appears; the latter may be very limited in distribution.

In one case a pathological examination was made by Steinert, who found degeneration of the posterior columns in the lumbar region; the muscles showed the changes found in other myopathic cases.

Diagnosis. Well marked types of myopathy are easy to recognise, but there are many aberrant forms in which it may be difficult to decide whether the muscular

atrophy depends on an abnormal condition of muscular tissue or on disease of the spinal anterior horns. In favour of a myopathy would be:—(1) The onset of the atrophy at an early age. (2) Its occurrence in more than one member of the family. (3) Its distribution: the progressive wasting of the muscles does not correspond as does that of a spinal amyotrophy to definite groups of nuclei in the spinal cord. (4) The absence of fibrillary twitchings and of a definite reaction of degeneration. (5) The condition of the tendon reflexes, which are never exaggerated and disappear in correspondence with the muscular wasting.

Prognosis. All forms of myopathy with the exception of amyotonia congenita present a slowly progressive downward course. Occasionally this is arrested and the disease remains stationary for many years. In a few exceptional cases partial or complete recovery has occurred. The prognosis is the most unfavourable in the pseudo-hypertrophic type, death usually occurring from some inter-current affection of the respiratory system before the age of twenty.

Treatment. No drug is known to have a specific action upon the affected muscles, or in any way to influence the course of the disease. Cod liver oil and various tonics should be given from time to time in order to maintain the health and strength of the patient.

The muscular condition may be improved and the progress of the atrophy delayed by massage, electricity and movements of the limbs both active and passive. Mild gymnastic exercises are useful, but any strain or fatigue must be carefully avoided. Walking should be encouraged as long as it is possible; a patient suffering from pseudo-hypertrophic paralysis tends to get rapidly worse when he has lost the power to walk. If inability to walk depends on contracture of the calf muscles tenotomy may be required.

When there is considerable atrophy of the muscles of the shoulder girdle, the power of movement in the arms is said to be improved by fixation of the scapulæ, their posterior borders being joined together.

CHAPTER X.

FACIAL HEMIATROPHY.

Facial hemiatrophy is a rare condition characterised by wasting of the skin, subcutaneous tissues and bones and to a less degree of the muscles of one side of the face.

Etiology. The atrophic changes usually begin before puberty, and only rarely after twenty years of age.

Fig. 101.—Photograph of a case of facial hemiatrophy.

They are more commonly found in females than in males. In the majority of cases no obvious cause can be discovered; in others the disease has followed an injury to the face, neuritis of the fifth cranial nerve, or some acute infectious disease. In a few cases a direct heredity has been traced.

Symptoms. As a rule the earliest changes are observed in the skin, a part of which becomes thin and

white or whitish-yellow. The affected patch increases in size and sometimes becomes fused with other similarly affected patches, until ultimately the whole of the skin on one side of the face has a thin and parchment-like appearance. The subcutaneous fat and connective tissue also waste. Should the change involve the hairy parts of the face, the hair may become thin and altered in colour, and may fall out. The growth of the bones is retarded, so that in old-standing cases the frontal, nasal and maxillary bones are much smaller than their fellows on the unaffected side. The nasal cartilages share in the atrophic process, and sometimes the cartilage of the ear also.

Fig. 102.

Fig. 103.

Fig. 102.—Photograph of a case of bilateral facial atrophy. Fig 103 is a photograph of the boy taken about two years before the onset of the disease was noticed.

In contrast to the atrophy of other tissues, the facial muscles usually remain unaltered, maintaining their normal bulk, strength and electrical excitability. In old-standing cases, however, the muscles generally become thinned, partly from disease and partly from absorption of the interstitial fat. The tongue may also show atrophic changes. The cutaneous sensibility and the sense of taste are not affected.

According to the distribution of the atrophic changes the following varieties may be distinguished: (1) Complete unilateral facial atrophy. (2) Incomplete unila-

teral atrophy. (3) Bilateral atrophy; and (4) cases with similar changes in other parts of the body.

The malady as a rule steadily progresses for some years, and then becomes stationary; it has no tendency to shorten life.

Its **pathology** is obscure; in one case Mendel found a proliferating neuritis of the trigeminal nerve. It is doubtful how far the cutaneous and osseous changes can be attributed to such a lesion. When we bear in mind that the atrophy is rarely if ever associated with sensory disturbances, it seems unlikely that it can be solely produced by lesions of a nerve, which is so largely composed of sensory fibres. There is a greater probability that the condition is due to an arrest of development during the period of growth; the cause of such arrest is unknown, by J. Hutchinson it is attributed to a morphœa of the trigeminal nerve.

The **treatment** consists in the application of massage and electricity to the facial muscles, and in the administration of general and nervine tonics, such as quinine, strychnine and arsenic.

SECTION VI.

Intrinsic Diseases of the Spinal Cord.

A Spastic or an Atrophic Paralysis is found, variously combined with other Symptoms. The Character and the Distribution of the Symptoms indicate a Focal Lesion in the Spinal Cord.

CHAPTER I.

MYELITIS.

It has been customary to regard the term 'myelitis' as synonymous with inflammation of the spinal cord, and to describe under the heading 'myelitis,' various groups of cases the symptoms of which are supposed to depend on variations in the acuteness and the distribution of the inflammatory process. There is, however, abundant pathological evidence to support the view that softening of a portion of the cord, which is the common lesion found in myelitis, is produced more frequently by other causes than by inflammation. Thus in 'compression myelitis' caused by spinal caries the degenerative changes in the cord are largely the result of evascularisation and ischæmia, due to slow vascular occlusion. Again in 'syphilitic and tuberculous myelitis' the destruction of the nerve elements is the result partly of gummatous or of tuberculous infiltration and partly of vascular changes. Lastly in 'primary myelitis' the diffuse softening or the degeneration, that is found, depends more commonly on the action of some toxic substance, or on blocking of the vessels by thrombosis, than on inflammation.

For these reasons some writers would limit the term myelitis to cases of true inflammation and apply the term *myelomalacia* to cases produced by the other causes.

mentioned. At present, however, it is convenient to retain the term myelitis for both classes, nor is there any etymological reason to the contrary. Moreover it is not easy, nor always possible to determine after death whether the lesion is due to inflammation, to arterial thrombosis or to a toxic degeneration. Even during life the manifestations of these different conditions may be similar, or identical.

In nearly all cases of primary myelitis the process is acute or subacute; chronic anterior poliomyelitis occurs and chronic transverse myelitis, but, excluding syphilitic cases, it is exceedingly rare. According to the distribution of the lesions, which corresponds to definite clinical types of the disease, and leaving out of consideration acute poliomyelitis, two chief varieties of myelitis may be distinguished, namely the transverse and the disseminated form. The latter, which is rare, is probably a true inflammation of the cord; the former is common and in a large number of cases is due to softening from vascular obstruction.

Etiology. Myelitis may occur at any age, but is most frequently met with between the ages of fifteen and forty. Anterior poliomyelitis occurs chiefly in young children.

Syphilis is one of the commonest causes of myelitis but inasmuch as the myelitis is usually associated with other changes, for example, gummatous infiltration, it seems desirable to consider syphilitic myelitis along with other forms of syphilitic affections of the spinal cord (see Section XXI.).

It is frequently impossible to discover any adequate cause of myelitis in cases which are not due to syphilis. Sometimes exposure to wet or to cold; alcoholism; overexertion; a strain to the back; or simple concussion of the spine has preceded the attack; it may be that one or other of these antecedents, by lowering the resistance of the tissues, has favoured the entry of micro-organisms into the system. A sudden suppression of the menses following exposure to cold may tend to a morbid

state of the blood, which may induce a myelitis, especially in the lumbar region.

Myelitis may be set up by adjacent inflammation such as meningitis; or by compression of the cord, as from caries or cancer. Myelitis, especially the disseminated variety, has been observed to develop during the course of, or after one of the infectious diseases, as small-pox, measles, typhoid, influenza, erysipelas, dysentery, malaria, gonorrhœa and the puerperal state.

Morbid Anatomy. In acute inflammatory conditions the morbid process is usually of considerable extent *diffuse myelitis*—or it may occur in scattered patches throughout a large portion of the cord—*disseminated myelitis*. At an early period the affected parts show engorged vessels, small hæmorrhages and extensive infiltration by leucocytes. Sometimes no thrombosis can be detected; frequently cell-infiltration is a more marked feature than changes in the nerve elements. When the initial lesion is vascular occlusion the process, though often extending across the cord, is limited to a few segments. This is *transverse myelitis*. In this variety some of the vessels have thickened walls and some are blocked with thrombi, but there is little if any emigration of leucocytes to be seen.

Both processes lead to softening and disintegration of the cord, hence at a late period it is impossible to determine whether the softening is a result of inflammation or of vascular obstruction. On section the cord bulges or is quite diffluent, and the distinction between white and grey matter is lost. If microscopical sections can be made the nerve cells are seen to be swollen, granular and fatty; the myelin sheaths of the nerve fibres are broken up and some of their disintegrated particles run together to form fatty-looking masses, while the axis-cylinders become enlarged and fusiform, and ultimately destroyed. Finally both cells and fibres disappear, the connective tissue increases in quantity, but in some cases this also may undergo destruction. Evidence of a varying degree of meningitis is also present; the

meningeal vessels are congested and the pia-arachnoid is infiltrated with cells; there may be a serous or even a purulent exudation.

When the process is less severe and of long standing, a dense proliferation of the neuroglial tissue occurs, which may lead to a definite sclerosis. The destruction of nerve fibres in the diseased segments cuts them off from their nutrient cells, and is therefore followed by degeneration of the distal portions of the several neurons, so that below the lesion a descending degeneration can be traced in the pyramidal tracts, and above the lesion an ascending degeneration in the posterior median and direct cerebellar tracts. Associated with these degenerations the neuroglia becomes thickened and a dense sclerosis gradually takes the place of the degenerated fibres.

Symptoms of Transverse Myelitis. The onset is usually rapid, and it may be quite sudden. The first symptoms may be sensory or motor, or they may relate to some disturbance of micturition. Early sensory symptoms are feelings of numbness and tingling, of fatigue and heaviness in the limbs; and sometimes of aching pains in the back or legs. Early motor symptoms are weakness and stiffness of one or both legs; the paralysis rapidly increases and becomes considerable in a few hours or days. In some cases the first symptom is retention or incontinence of urine. Shivering, pyrexia and other constitutional symptoms are usually absent or are but little marked.

Dorsal Myelitis. In this, the commonest variety of myelitis, there is paralysis of the legs together with the lower part of the trunk, the latter being often indicated by the upward movement of the umbilicus when the patient tries to rise from the recumbent posture. At first the limbs are flaccid and the knee-jerks show no decided change. But sooner or later the limbs become spastic, and the knee-jerks exaggerated; ankle-clonus can be elicited and the plantar reflex is of the extensor type.

The paralysis may be complete or partial. In the former case the patient is confined to bed and is unable to move any part of the lower limbs, or by a strenuous effort can only just cause a feeble movement of the toes or a flickering contraction of some of the thigh muscles. When the paralysis is partial the patient may be able to walk, either with or without assistance according to the severity of the lesion, but in either case some dragging of the feet is noticeable. If when seated in a chair the strength of the individual movements is tested it will be found that the flexor movements are distinctly weak. In slight degrees of myelitis, weakness may be limited to flexion at the hip or ankle; whilst in severer forms flexion of the knee is also involved and the extensor movements of the legs may become affected. The muscles preserve their normal bulk, occasionally they waste to a slight degree probably because they are not used. The electrical reactions are usually normal.

Anæsthesia is almost invariably present but its degree and distribution vary much in different cases. It may be complete, involving every form of sensation, affecting both of the legs and extending as high as midway between the umbilicus and the ensiform cartilage. When incomplete the legs are usually more affected than the thighs and differences in degree may be observed on the two sides, as well as over the lower part of the abdomen. Sensation to touch may be lost, when to pain it may be preserved. Sometimes the impairment of sensation is very slight and can be detected only on the outer aspect of the leg or over the lower part of the abdomen: its limits are then often difficult to define, and they may vary with each examination. A band of hyperæsthetic skin just above the level of the anæsthesia is often present, and sometimes there is slight tenderness over the corresponding spine which may be brought out by passing a hot sponge down the vertebral column. Frequently too at this level the patient has a sense of constriction round the body; this is known as the "girdle sensation."

The height of the lesion is also indicated by the superficial reflexes; thus in its common situation at about the eighth segment the epigastric reflex is present whilst the abdominal is absent. The functions of the bladder and rectum are frequently disturbed and owing to implication of their sensory tracts the patient may be unaware when urine and fæces are being passed. Sometimes there is retention of urine with overflow incontinence; sometimes there is frequent micturition with incomplete emptying of the bladder, and some dribbling of urine. As regards the bowel, constipation is the rule; this is either persistent or associated with occasional involuntary evacuations. Sexual power is frequently lost and priapism may be a troublesome symptom.

The urine is often alkaline; its alkalinity is increased in cases of retention, owing to decomposition in the bladder. Cystitis is liable to develop, when the urine will contain pus together with much mucus. Bed sores may form on parts subjected to pressure, especially over the sacrum and sometimes on the heels or in the region of bony prominences. Vaso-motor disturbances, including œdema of the limbs occur, and occasionally there is effusion into the knee joints.

The *progress* of such a case is variable; sooner or later a stationary stage is reached which lasts for months or years, but even during this stage slight variations in the symptoms occur from time to time: one day paralysis of a part is complete, the next day it is incomplete, the patient being able to move the toes or the foot. Anæsthesia is still more variable and may pass away altogether. Periods of improvement are common and sometimes the improvement is permanent: indeed may go on to complete recovery. More frequently relapses occur till ultimately the paralysis is complete and persistent. Muscular rigidity tends to increase and often becomes extreme, the legs being either rigidly extended, or firmly flexed owing to contraction and shortening of the flexor muscles. The

patient is often much troubled by twitching of the muscles and sudden spasmodic contractions of the legs. Occasionally "spinal epilepsy" is readily produced by slight stimulation, as by striking the patellar tendon.

Lumbo-Sacral Myelitis is much less common than the dorsal variety. There is weakness of the legs with modifications of their cutaneous sensibility; the dis-

Fig. 104.—Extreme permanent contracture in a case of myelitis.

tribution and degree of the paralysis and the anæsthesia vary with the position and extent of the lesion, which may be limited to the lumbar enlargement or may involve both lumbar and sacral segments. But in any case the abdominal muscles escape and the anæsthesia does not reach higher than the groin or a short distance above it.

Most of the paralysed muscles undergo a rapid

atrophy which is steadily progressive; some of them however, namely those which derive their nerve supply from spinal segments below the level of the diseased focus, may be in a spastic condition. For a similar reason the knee-jerk may be lost while ankle-clonus and the extensor type of plantar reflex may be easily obtained.

But if the lesion involves only the lowest segments of the cord the knee-jerks are present; the paralysis is altogether of the atrophic variety; the sphincters are relaxed, and there is loss of sensibility and of reflex action within the areas supplied by the sacral segments. Bed-sores are more liable to occur than in dorsal myelitis; they often develop at an early period of the disease and tend to become extensive and deep.

Cervical Myelitis. In this variety, which is rare, the condition of the lower limbs is identical with that observed in dorsal myelitis; but in addition to the spastic paraplegia there is an atrophic paralysis of some of the arm muscles, whilst impaired action of the lower intercostal muscles can usually be detected. A variable amount of anæsthesia is found over the legs and over the trunk, sometimes nearly as high as the level of the lesion. Its distribution in the arms as well as that of the muscular atrophy gives a clue to the spinal segments which are implicated. Thus a lesion in the lower part of the cervical enlargement would lead to a band of anæsthesia down the inner side of the limb, and to an atrophic paralysis of the hand and wrist muscles. In such a case the sympathetic fibres to the eye, which run in the first dorsal roots, are frequently implicated giving rise to contraction of the pupil and to a narrowing of the palpebral fissure. Sometimes the pulse is rapid; occasionally hyperpyrexia occurs. In rare cases optic neuritis has been observed. Myelitis at a level higher than that from which the nerves to the arms are derived is extremely rare. In such a case the neck muscles might be atrophied and the arms as well as the legs be affected with a spastic paralysis. Unless the lesion is

partial the diaphragm becomes paralysed and death from asphyxia quickly ensues.

Acute Disseminated or Acute Ascending Myelitis. In this variety paralysis and anæsthesia are more extensively distributed than in the transverse varieties; sometimes they develop simultaneously in the limbs and trunk; sometimes they pursue an ascending course the legs being first affected, then the abdomen and chest, and finally the arms and the diaphragm. In other cases the

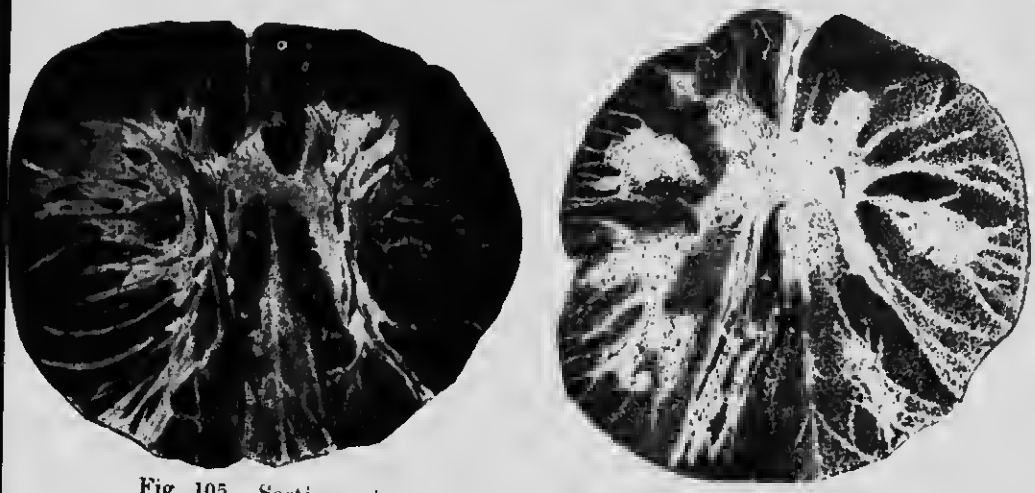


Fig. 105.—Sections of spinal cord in a case of acute disseminated myelitis (Dreschfeld.)

development is more irregular, thus paralysis of one leg may be followed by paralysis of one arm and later by paralysis of the other limbs. In a case recently under the author's care the development of the symptoms indicated the presence, at first, of foci in the lumbar and cervical enlargements, and later, of disease spreading upwards from these parts. Thus paralysis of the legs came on at the same time as paralysis of the extensors of the wrist and fingers; the paralysis was of the flaccid type and in the legs was associated with loss of the knee-jerks; there was also retention of urine with intermittent overflow. After a time the paralysis and anæsthesia gradually spread upwards to the trunk,

and from the hands to the shoulders till respiratory paralysis caused a fatal termination. This occurred about four months after the first symptoms, namely numbness and tingling in the hands and feet.

Frequently in these acute spreading forms the temperature is raised, and there is an early tendency to bed-sores and to cystitis which is often severe in type. Optic neuritis sometimes occurs and may precede the spinal symptoms; it probably depends on the toxæmia which has also caused the myelitis. In some cases the optic nerves show patches of inflammation.

According to F. Buzzard this form of myelitis is produced by a bacterial infection of the lymphatic system of the spinal cord and therefore may be regarded as a spinal lymphangitis.

In another variety called *disseminated encephalomyelitis* or by Leyden *acute ataxia*, areas of acute inflammation are found in the cerebral peduncles, and in the pons and medulla as well as in the cord. The symptoms are: Defects in articulation similar to those of disseminated sclerosis, tremor and ataxia in the limbs especially in the arms, and sometimes tremor of the head. Much paralysis is rare; sensory disturbance is slight or absent; the deep reflexes are usually increased and the gait is spastic. As a rule the mental condition is not impaired; the sphincters often escape. This disease occurs more frequently in children than in adults; in some cases it has commenced during the course of an infectious fever, such as measles, smallpox or typhoid.

Acute diffuse central myelitis, a rare condition, is characterised by a rapid loss of power and sensation in the limbs; the arms or the legs may be first attacked, or both may be affected simultaneously. The reflexes are lost, trophic changes develop rapidly, the temperature is usually high, and death occurs within a few days.

The sudden onset of severe symptoms during the early stage of any variety of myelitis would indicate a sudden

extravasation of blood into the cord. This condition is sometimes called *hæmorrhagic* or *apoplectic myelitis.*

Diagnosis. Transverse varieties of myelitis are usually readily distinguished by the presence of paralysis and anæsthesia below a definite level, together with disturbance in the functions of the bladder and rectum. Myelitis being present it is of great importance to determine whether it is primary or is secondary to vertebral caries, meningeal tumour or other lesion lying outside the cord. The compression myelitis produced by these causes is usually preceded for some weeks by root symptoms, whilst an examination of the back, which should be repeatedly made in all doubtful cases, will often reveal some deformity varying from a distinct angular curvature to a slight projection of one spine due to caries. In the absence of deformity local pain and tenderness on tapping over one or two vertebræ, together with rigidity of the back during various movements, are suggestive signs of caries. Examination with the *x*-rays, especially in cervical caries, is frequently of service. In compression myelitis, due to a tumour, root-symptoms are severe and signs of a primary growth may be discovered in some other part of the body.

If extra-spinal lesions can be excluded it is only occasionally that the diagnosis of a primary myelitis presents any difficulty. Hæmorrhage is characterised by the sudden onset of symptoms, complet aralysis of the legs or of all four limbs developing in a few minutes; by the absence of pyrexia and by the presence of severe pain in the back. It is only in hæmorrhagic myelitis, which probably depends on thrombosis of small vessels, that the onset would be equally sudden; but in this condition, premonitory symptoms often precede the paralysis and the temperature is raised.

Peripheral neuritis limited to the legs might be mistaken for a lumbar myelitis, but it does not lead to bed-sores nor to bladder symptoms; the anæsthesia has less defined limits than in myelitis, it is rarely so

marked and is usually symmetrically distributed to the peripheral portions of the limbs; in myelitis the sensory loss is bounded by lines which correspond to the limits of segmental areas. As a rule pains are more prominent in cases of neuritis, and the muscles may be tender to pressure.

Sometimes hysterical paraplegia presents a resemblance to dorsal myelitis; in both cases paralysis and anæsthesia may be complete or incomplete. A difference is to be noted as regards muscular rigidity and its variability at different times; also in hysteria the lifting of one leg from the bed does not raise the other with it, whereas this occurs in true spastic paraplegia. The presence of marked ankle clonus and of an extensor plantar reflex would exclude hysteria, but occasionally in myelitis these signs cannot be properly elicited. Moreover the two maladies may be associated: paraplegia due to hysteria may prevent the recognition of weakness of certain groups of muscles caused by myelitis. In such cases a single examination may not remove uncertainty, and many observations and a full consideration of every aspect of the case may be necessary before a certain diagnosis can be made.

Two other maladies sometimes create difficulties, namely, disseminated sclerosis and intramedullary tumour. A spastic paraplegia characterises the earlier stages of disseminated sclerosis, but as a rule it is uncomplicated, and its development is a gradual one. In exceptional cases, however, it is associated with slight anæsthesia and difficulty in micturition, when a dorsal myelitis may be closely simulated. The latter would be excluded by the presence of nystagmus or of intentional tremor in the hand. An intramedullary tumour may run its course without pain or any definite root symptoms; then its presence would be suggested by a slow and often unilateral development of paralysis.

It is always difficult to distinguish between acute inflammation and acute softening of the cord produced by thrombosis. Langdon believes that the following

points are in favour of softening: the absence of a history of preceding injury or of acute illness; the more sudden onset of paralysis and the absence of premonitory symptoms, of fever and often of bed-sores.

Prognosis. Broadly speaking, the prospect is better if the myelitis has followed influenza or some other infectious disease, especially syphilis, than if no definite cause can be traced. It is better, too, in the transverse than in the disseminated form. Of the transverse varieties the risk to life is greatest in cervical myelitis owing to the danger of respiratory paralysis; it is greater in lumbar than in dorsal myelitis because bed-sores and cystitis are more apt to occur and to be severe.

In exceptional cases, complete recovery occurs even when considerable paralysis has lasted for many months; in many cases some power is regained, whilst in others the patient remains bed-ridden without any prospect of recovery. In all forms of myelitis, the more absolute the paralysis and the anæsthesia, and the longer they have lasted, the less the chance of any recovery.

Bed-sores owing to the risk of septic infection, and cystitis owing to the risks of pyelitis and pyelonephritis, are always to be viewed with grave anxiety.

Treatment. From the very outset absolute rest in bed is essential, and it is better for the patient to lie on one side or in the prone position than on the back. Frequent changes in posture are desirable in order to prevent undue pressure on bony prominences.

If the myelitis can be traced to exposure to cold or to some infectious disorder, diaphoretic measures are advisable; profuse sweating may be induced by the conduction of steam under the bed clothes. The patient should be wrapped up in warm blankets; hot milk and barley water should be frequently administered. The bowels must be freely moved by the administration of five grains of calomel, followed by a saline aperient.

In every case, whether syphilis is suspected or not, it is advisable to give mercury either by the mouth or by inunction, for there is much evidence in favour of its

efficacy. Iodide of potassium, except in syphilitic cases, appears to be of little use. Alcohol must be prohibited; the diet should be light, nutritious and non-stimulating, so that butcher's meat is better avoided, at any rate for a time.

In order to prevent as far as possible the occurrence of bed-sores and cystitis—the two most dangerous complications of myelitis—great care and cleanliness are necessary. A water-bed is always advisable; the skin should be frequently sponged, and the bed sheets changed if at all soiled by the evacuations. If at any spot the skin is unduly reddened it must be washed with spirit lotion or with a solution of alum, then carefully dried and dusted over with oxide of zinc and starch powder. When a bed-sore has formed it should be dressed like any other ulcer, and kept as aseptic as possible. Carbolized vaseline and iodoform ointment are useful applications, or iodide of starch paste, if the sore is large and indolent.

When there is retention, with or without dribbling of urine, catheterisation is necessary. It is well to use a soft rubber catheter, the greatest care being taken to keep it thoroughly aseptic. In males, when there is incontinence of urine, a porcelain urinal, well padded in order to avoid undue pressure, must be kept constantly in position. in both sexes non-irritating antiseptic wool should be placed under the patient and around the genital organs in order to absorb any evacuations from the bladder or rectum. Should cystitis develop, the bladder is to be washed out twice a day with some mild antiseptic, such as a warm solution of boracic acid—fifteen grains to the oz. It is also desirable for the patient to take seven to ten grains of urotropin or of salol thrice daily.

Apart from syphilitic cases no drugs, with the possible exception of mercury, appear to have any influence in improving or arresting the morbid changes in the cord. It is possible that the exposure of the spine to the Roentgen rays may prove of service; the author has

employed this method in several cases, and in some apparently with benefit.

The general health should be maintained by the administration of quinine, strychnine and other tonics, by good nourishment and by cod-liver oil. Strychnine is of most value in atrophic paralysis, and should be avoided in spastic cases. Rigidity of the limbs is often relieved by sponging with warm water and by the use of passive movements, whilst a tendency to the development of contractures may be lessened by careful attention to the position of the limbs in bed. The severity of the reflex spasms which occur in the chronic stages is sometimes lessened by the administration of the bromides, or still better of hyoscine. The use of electricity should be restricted to the cervical and lumbar varieties of myelitis; in such cases galvanism together with massage of the atrophied muscles tends to arrest the wasting, and even to reduce it.

Acute Suppurative Myelitis: Abscess of the Spinal Cord.

This is a rare affection. Most frequently it occurs as a secondary complication to a distant septic process, such as bronchiectasis, gonorrhœa or infective endocarditis. It has been found in association with suppurative meningitis, and with tuberculous and carcinomatous disease of the vertebræ, probably as a result of the invasion of the damaged spinal segments by bacilli derived from a purulent cystitis or other source of infection. Suppurative myelitis may also occur as a primary condition.

The purulent collection is found in the central region of the cord, chiefly occupying a position in the anterior parts of the posterior horns, and the posterior columns; its longitudinal extent varies, sometimes it may be considerable, suppuration spreading far above and below the area of original disease.

The symptoms in primary cases are similar to those of acute transverse myelitis; in secondary cases to those of

an intramedullary tumour. Frequently the signs of abscess formation are preceded or obscured by the symptoms of meningitis. Some cases run an afebrile course, others are characterised by rigors and pyrexia.

The prognosis is very bad and death is not often long delayed.

CHAPTER II.

INTRAMEDULLARY TUMOURS.

The chief forms of tumour which begin within the spinal cord or which grow into it from the pia-mater are glioma, sarcoma, tubercle and gumma. Myxoma, myxo-sarcoma, glio-sarcoma and other varieties of new growth are also met with, though very rarely.

A glioma originates within the grey matter, often in the tissue around the central canal, and grows outwards into the white substance. The central canal is rarely invaded, as it is in syringomyelia. Sarcomata may be primary, or secondary to growths in the brain, spinal roots or meninges. Degenerative changes are apt to occur in both gliomatous and sarcomatous growths, leading to the formation of cavities in their central parts. Tuberculous tumours are found in association with meningeal tubercle; they also occur as solitary growths. Gummata are always present in connexion with the spinal blood-vessels, or with the pia-arachnoid membrane.

Symptoms. In many cases the clinical picture of an intra-medullary tumour is that of syringomyelia, in other cases it is that of myelitis. Occasionally, at any rate for a time, there is a close resemblance to anterior poliomyelitis.

Pain is rarely so prominent as in extra-medullary tumours; there may be aching in the back, but spinal tenderness and radiating pains are usually trifling or absent. Root symptoms, however, may be present and even severe when the growth invades the posterior horns.

Local anæsthesia of parts innervated by the affected segments of the cord, is an early symptom which may be followed by an atrophic paralysis, also of segmental distribution. Muscular atrophy is generally in excess of muscular weakness: it may be difficult to detect when the dorsal region is implicated, but it is conspicuous in the arms or in the legs when the tumour occupies one of its common situations, namely, the cervical or the lumbar enlargement. Thus a growth beginning in the grey matter at the level of the fifth cervical segment will produce a band of anæsthesia along the outer side of the arm, and wasting of the deltoid, biceps and

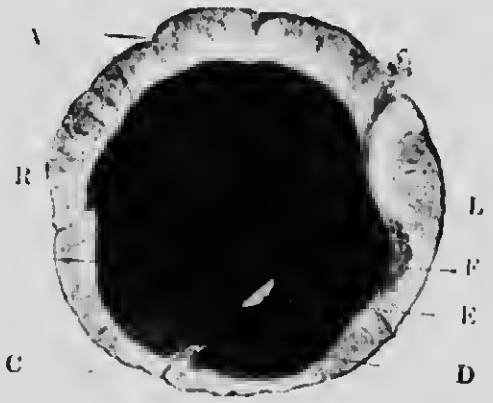


Fig. 106.—Gliomatous growth occupying a large portion of the spinal cord in the cervical region. A, posterior fissure; C, anterior fissure, D, new growth; E, central canal; F, anterior cornu; R, right, L, left side of cord.

supinator longus muscles. Such symptoms if unilateral in distribution at first, soon become bilateral.

Sooner or later other parts of the cord become affected resulting in paralysis and anæsthesia below the level of the lesion. Loss of sensation to pain and temperature may be noted, before tactile sensation become affected: this dissociated anæsthesia may be present only on one side, whilst motor paralysis affects the opposite side. (Brown-Séquard's symptom-complex.)

In very slowly growing tumours the nerve elements

may be thrust aside without being materially damaged, and a growth occupying the greater portion of a segment of the cord may be found post mortem when only slight and indefinite symptoms were present during life; or a particular variety of symptoms may be alone marked. Thus ataxia and loss of the knee-jerks may represent a growth involving the posterior columns; or severe atrophy of one arm, a growth in the cervical grey matter. In a case, under the author's care, of glioma in the lower three-fourths of the cervical cord, the only prominent symptom during the first four months was an atrophic paralysis of nearly all the muscles of one arm; sensory symptoms were absent and for a time the diagnosis of anterior poliomyelitis seemed to be the most probable one (see fig. 106).

Generally speaking, signs of an extensive unilateral lesion, together with the early occurrence of disturbance of micturition and the presence of Babinski's reflex, are suggestive signs of an intra-medullary new growth.

Atrophic paralysis of the muscles supplied by the sacral plexus occurs in tumours of the cauda equina; it is often preceded and accompanied by severe pain in the region of the sacrum, bladder and rectum and along the course of both sciatic nerves. Frequently also there is anæsthesia in the whole or a part of the area innervated by the sacral plexus.

Course and Prognosis. A gradual onset and a slow development and progress of the symptoms are characteristic features of a spinal tumour. The chronic downward tendency may be interrupted by periods, either of amelioration or of aggravation of symptoms; the sudden occurrence of the latter is often due to the development of myelitis. From this cause the lesion may extend completely across the cord, giving rise to absolute paralysis and anæsthesia and loss of all the reflexes in parts below the diseased area.

The final stages of a cord-tumour are similar to those of a transverse myelitis, death occurring as a result of asphyxia from respiratory paralysis, of septic infection

from a bed-sore, or of secondary kidney disease from cystitis. In cases of tubercle, death may be the result of pulmonary or of meningeal tuberculosis. The disease varies in duration, but rarely lasts longer than two or three years.

Diagnosis. Important features of an intra-medullary tumour are the slow and steady development of paralysis and of other symptoms indicating a focal lesion of the cord, without signs of an upward extension of the lesion and without signs of decided root implication. Dissociated anæsthesia and considerable muscular atrophy are other points in favour of a growth.

Some cases present a close resemblance to myelitis, but in this disease the onset of paralysis is usually much more rapid than in spinal tumour. Syringomyelia also may be closely imitated, but as a rule its course is much more prolonged: moreover, its presence is frequently characterised by the occurrence of painless whitlows, arthropathies and kypho-scoliosis.

A tumour of the cauda equina may be mistaken for sciatica; in the former the symptoms are bilateral, in the latter unilateral in distribution.

It is often very difficult to diagnose between an extra- and an intra-medullary tumour. In the former, pain and other root-symptoms are usually prominent, preceding the symptoms due to compression of the cord. In the latter case root symptoms are slight or absent, except when the growth is situated in the cauda equina.

Treatment. The treatment consists in the alleviation of pain, and in the prevention of sepsis, cystitis, pneumonia and other complications. When syphilis is suspected, antisyphilitic remedies should be given a trial. In some cases it is difficult to say whether the symptoms indicate a tumour beginning outside or inside the spinal cord; it is then justifiable to make an exploratory laminectomy.

CHAPTER III.

SYRINGOMYELIA.

Syringomyelia is a disease of the spinal cord, characterised anatomically by the presence of one or more cavities, which in the majority of cases are surrounded by a layer of embryonic neuroglial tissue; and clinically by dissociated anæsthesia, muscular atrophy and various trophic lesions.

Pathology. On post mortem examination the following appearances may be observed. The spinal meninges are usually normal, but sometimes the dura mater in the cervical region is thickened. The cord is often irregular in contour: it may be flattened from before

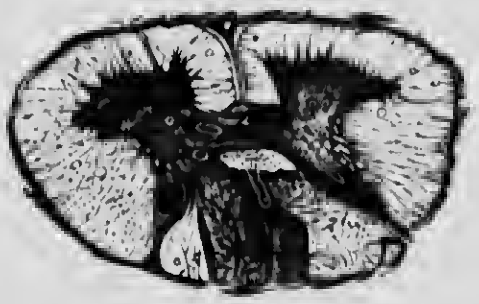


Fig. 107.—Transverse section of the spinal cord from the middle of the cervical enlargement, from a case of syringomyelia, showing a cavity behind the posterior commissure, and destruction of a large portion of the ganglion cells of the anterior grey horns. (from Leydon.)

backwards, or narrow in some places and bulging in others, the bulging portion being soft and fluctuating. The medulla also is sometimes flattened or reduced in size. The aqueduct of Sylvius may be widened and occasionally the ventricles are enlarged, with thinning of the surrounding tissue.

The cord during removal may rupture, and a clear or turbid serum may escape. On transverse section one or more cavities are exposed. As a rule there is only one cavity, which is situated in the lower cervical and upper dorsal region. In length and volume it presents many

variations; in some cases it is but short, in others it is very long and may even extend from the filum terminale to the medulla oblongata. Sometimes it occupies the greater part of a transverse section, sometimes it is so small as to be barely visible to the naked eye. It may occupy the place of the central canal, or, as more frequently happens, it is behind the canal lying in the posterior central grey matter, in the posterior horns or in the posterior columns. Frequently it invades the anterior horns, and exceptionally the whole of the cord appears to be destroyed, the cavity being surrounded by a thin layer of neuroglial tissue. The cavity may or may not communicate with the central canal: an existing or a past communication is indicated by the presence of cylindrical epithelium lining a portion of its wall. Sometimes a section of the cord shows two cavities, but one of them is usually a diverticulum of the other as may be seen by an examination of the sections.

The thickness of the neuroglial tissue which surrounds the cavity varies in different cases; its density is greatest nearest the cavity. Sometimes the cord appears to be infiltrated with gliomatous elements and the infiltration may extend above and below the limits of the cavity. In some cases proliferation of the embryonal glial tissue is marked and leads to the formation of distinct tumours, in which owing to breaking down of their substance, cavities are produced. The proliferated tissue is often rich in blood-vessels, and capillary hæmorrhages are not uncommon.

Secondary ascending and descending degeneration in the columns of the cord may occur. Degenerative changes are also found in the motor nerves with consequent atrophy of the muscles supplied by them.

The most probable explanation of the condition is that it depends on a congenital anomaly. This view is supported by the persistence of the embryonal tissue and the position of the cavity, in relation to a consideration of the facts of normal development. At a certain period of fœtal life a fissure-like cavity extends from

the centre of the cord backwards between the two rudimentary posterior columns. This becomes gradually narrowed by the growth of the posterior columns and as they first unite anteriorly to form the posterior commissure the cavity is divided into two parts. The anterior portion becomes the true central canal, whilst

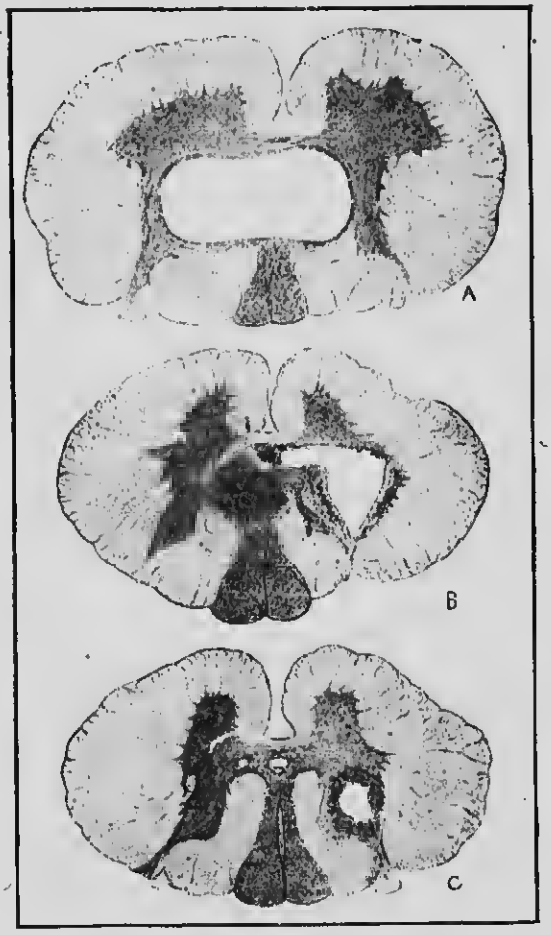


Fig. 108.—Syringomyelia, from a case in which there were tumours in the dorsal region of the cord, the cauda equina, and the pons varolii. A, mid-cervical region, showing a large oval cavity behind the grey commissure. B, first dorsal, showing a growth behind the posterior commissure, an irregular cavity occupying the posterior half of the grey matter on the right side, and portions of the growth outside the cavity. In C, this cavity is smaller and the growth outside it more abundant. (Gowers, after T. Harris.)

the posterior portion is gradually closed by the coalescence of the posterior columns, which occurs from behind forwards. If from any cause development is arrested before these processes are complete a cavity is left between the posterior columns which, if the posterior commissure is not formed will open into the central

canal. The embryonic cells around the canal may also persist and their subsequent proliferation lead to obvious tumour formation.

Some authorities believe that such proliferation is the essential lesion, and that it may be started by an irritant poison in the fluid of the central canal, or in the lymph channels of the posterior roots; and that a cavity is formed by the breaking down of the new-formed gliomatous tissue. It seems probable as Gowers suggests that there are two classes of cases: One class in which there is a congenital anomaly, and another in which there is central new growth which may be associated with new growths in other parts of the body. In the former class the clinical course is slow and prolonged, in the latter it is apt to be rapid and fatal.

It must be remembered that cavities in the cord are sometimes the result of haemorrhage, or of the breaking down of tissue the nutrition of which has been impaired owing to disease of its vessels. But as a rule such cavities do not resemble those of syringomyelia. Nevertheless it is possible that occasionally a central hæmatomyelia may be the starting point of gliomatous cavitation.

The term *hydromyelia* should be limited to cases of simple dilatation of the central canal which are unattended by changes in the surrounding tissue and by any clinical manifestations.

Etiology. Although usually depending on a congenital defect, the first symptoms of the disease rarely appear before the tenth year and most commonly between twenty and thirty. They may be excited by local changes in the lesion as a result of injury, or other agency. The disease appears to be nearly twice as common in males as in females. Signs of certain congenital anomalies are sometimes found in the subjects of syringomyelia, for example, the cicatrix of a spina bifida occulta, enlargement or other deformity of the hands and feet, or a hydrocephalic head.

Symptoms. A congenital cavity in the cord does not

necessarily give rise to symptoms, but if from any cause it becomes distended, or the surrounding tissue proliferates, the functions of the nerve elements may then be impaired, and give rise to symptoms which will vary in distribution according to the position and extent of the disease.

The two most characteristic manifestations are a peculiar variety of anæsthesia, and muscular atrophy, which usually affect the arms and upper half of the trunk. In many cases other symptoms are present, namely trophic lesions of the skin, bones and joints, and occasionally affections of some of the cranial nerves.

Sensory Symptoms. As a rule the patient's attention is first directed to his condition by finding that injuries such as cuts and burns are unattended by pain. In some cases this anæsthesia is preceded by spontaneous pains, or by feelings of heat or cold in the spine and limbs, and there may be hyperæsthesia to thermal and painful stimuli.

On examination of a typical case it is found that the cutaneous sensibility to pain and temperature is lost, whilst that to touch is preserved. This dissociated anæsthesia is accounted for by the more central position of the fibres for painful and thermal sensations as compared with those for tactile impressions. It is often irregularly distributed over the hands and arms, and is rarely symmetrical on the two sides. Its boundaries are generally sharply limited. In some cases sensation to heat is preserved when that to cold is lost, or conversely. In other cases, especially in an advanced stage, tactile sensibility is impaired or lost, and sometimes over areas which show no loss of sensation to pain or to temperature. This suggests an extension of the lesion to the posterior columns, when also the sense of position may be affected.

Interference with the conducting sensory tracts may produce loss of sensation below the level of the lesion: all forms of sensation may be impaired, but more commonly, one side of the cord being more involved

than the other, tactile sensibility is impaired on one side of the body and painful and thermal sensibilities on the other side.

Very rarely dissociated anaesthesia is limited to the lower limbs: this indicates a lesion in the lower portion

Fig. 109.—Showing spinal curvature in a case of syringomyelia.

of the cord. More frequently portions of the head and face are anaesthetic, the central portion of the face, including the eyes, nose and mouth being often spared.

Motor Symptoms. Muscular atrophy occurs in more

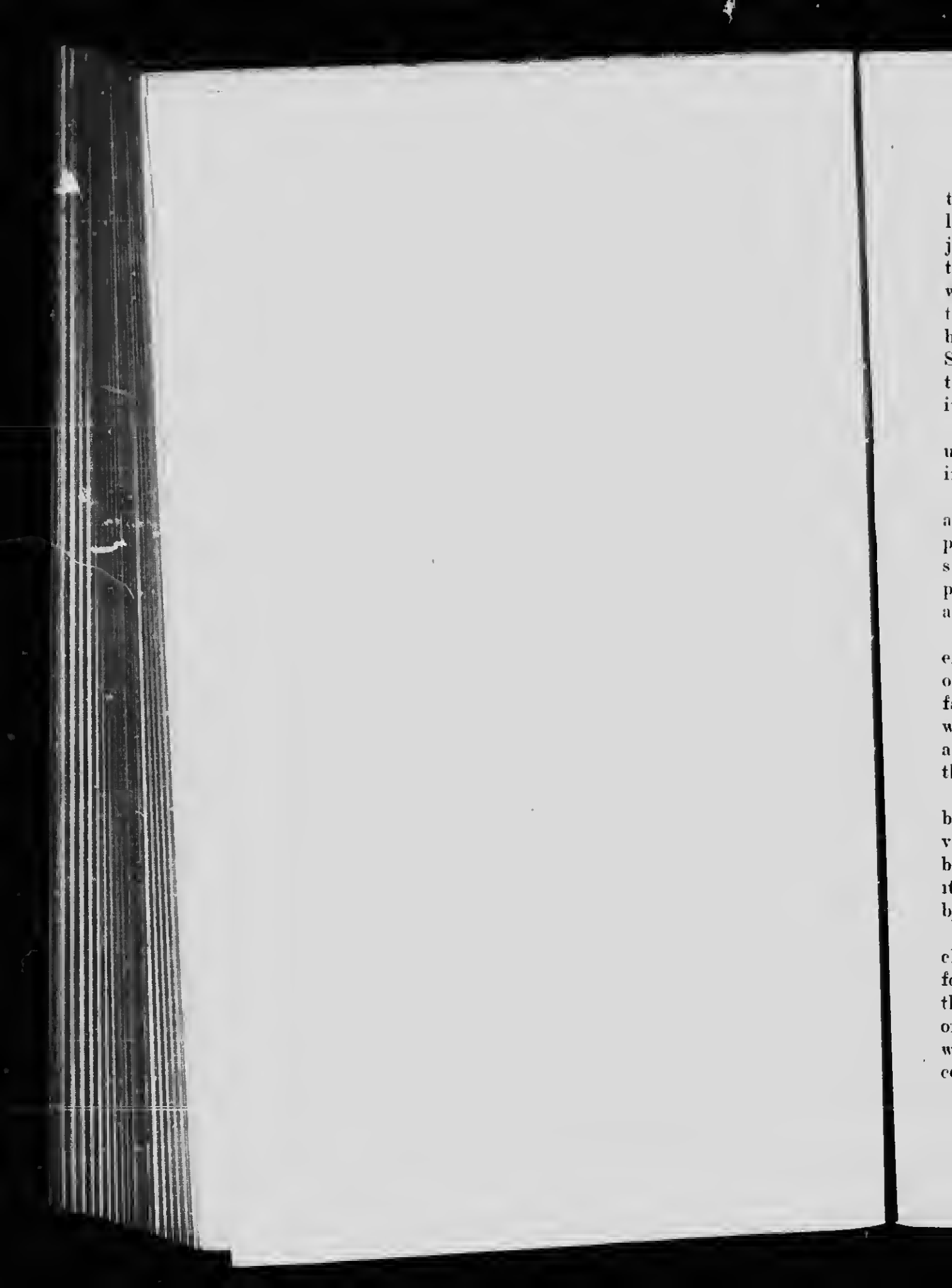
than one half of the cases of syringomyelia. Its distribution is very similar to that of progressive muscular atrophy, for the wasting usually begins in the small muscles of the hand and ultimately leads to the position known as the *main en griffe*. It then gradually spreads up the arm to the shoulder. Sometimes the shoulder muscles are the first to be affected. As the disease progresses the muscles of the trunk and neck may be invaded and occasionally the muscles of the lower limbs. Curvature of the spine, which is often present, is due to weakness of the back muscles, and in some cases to changes in the bones. Atrophy of the leg muscles is usually associated with loss of the knee-jerk, the lesion being in the lumbar enlargement. But in the ordinary type of syringomyelia there is spastic paraplegia, slight or moderate in degree, and the knee-jerk is exaggerated: these symptoms are due to compression of the pyramidal tracts in the upper portion of the cord. The atrophied muscles are necessarily weak, but the relation of paralysis to wasting varies in different cases. Fibrillary tremors are sometimes present: the mechanical and electrical excitability of the affected muscles is gradually diminished.

Vaso-motor and trophic changes. The hands and feet tend to get blue and cold; sometimes they are swollen and red. The skin may be thick and horny, or thin and glossy; it may be abnormally dry or covered with sweat. Acute inflammation of the skin of the affected part is easily set up by a slight injury, or it occurs independently of this: deep ulceration or even gangrene may ensue. Eczema, herpes and bullous eruptions also occur. The nails are often affected becoming furrowed, thickened and brittle. A striking disturbance is the formation of painless whitlows upon the fingers. A finger or thumb becomes swollen and œdematous: necrosis ensues and leads to loss of substance and sometimes to the separation of a terminal phalanx.

Changes in the *joints* are not uncommon; the shoulder, elbow, and wrist are most frequently involved, a con-

PLATE IV.

Skiagram of a case of syringomyelia, showing absorption of upper part of head of humerus, with eburnation of the bone: the shadowy part beneath the acromion indicates new formation of bone in the capsule.



trast to tabes, in which disease the joints of the lower limbs are principally attacked. The character of the joint lesion and its clinical course are similar in the two diseases. There is swelling, often considerable, which develops quickly without any pain. All the tissues are thickened, and ultimately the cartilages become eroded and the ends of the bones atrophied. Spontaneous fractures sometimes involve the bones of the arms: the process of repair is usually very slow and imperfect.

The *bladder and rectal functions* are rarely disturbed unless it be in the last stage of the disease, or in cases in which the sacral region of the cord is involved.

As the first dorsal segment of the cord is frequently affected it is not uncommon to find symptoms indicating paralysis of the *cervical sympathetic* on one or both sides, namely, a small sluggish pupil, narrowing of the palpebral fissure and defective secretion of sweat on the affected side.

In a few cases there is evidence that the disease has extended to the medulla and pons. Thus nystagmus, ocular palsies, dissociated anæsthesia of the head and face, paralysis of one side of the tongue and palate, with paralysis of one vocal cord, difficulty in swallowing and disorders of respiration, and irregular action of the heart have been observed in different cases.

Occasionally impairment of taste, smell or vision has been present. A peripheral contraction of the field of vision may depend on an associated hysterical condition, but sometimes it occurs apart from this, when possibly it may be explained by pressure on the cerebral cortex by a distended ventricle.

As a rule the **course** of syringomyelia is a very chronic one: its progress is slow and may be arrested for long periods, even twenty years. Death is usually the result of some complication, as cystitis or bed-sores, or an intercurrent affection as phthisis. Sometimes where there is much overgrowth of gliomatous tissue, combined with destructive processes, the symptoms

develop with rapidity and death occurs in a few weeks or months. Rarely it occurs suddenly owing to rupture of the cavity in the cord.

Varieties. Of variations from the ordinary type may be mentioned: 1. Cases in which muscular atrophy is the only conspicuous feature.

2. Cases presenting dissociated anæsthesia, but no muscular atrophy.

3. Cases in which the typical symptoms are present but are limited to one side of the body.

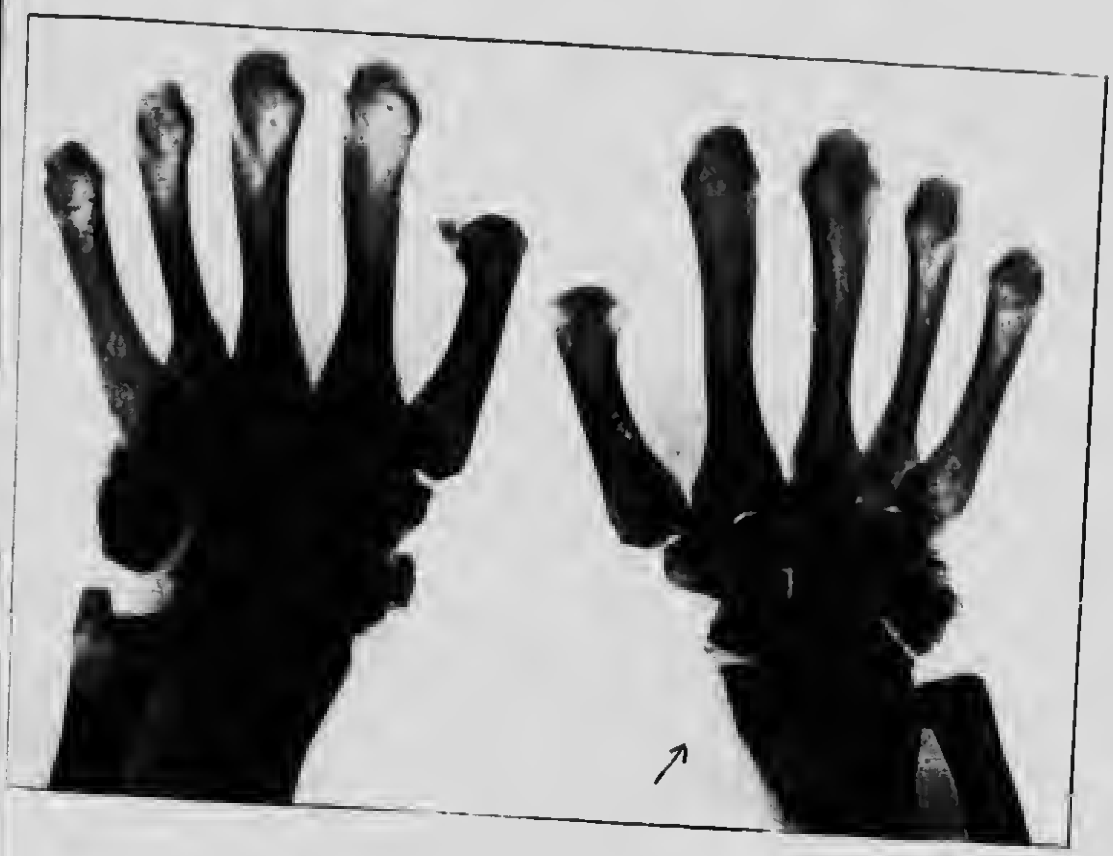
Fig. 110.—Photograph showing the position of the shoulders in a case of spasmodic syringomyelia.

4. The spastic type, characterised by intense and wide-spread rigidity together with exaggeration of the tendon reactions and an extensor plantar reflex. The rigid attitude of the body resembles that of paralysis agitans. The arms are firmly applied to the sides of the chest: the hands are hyper-extended and the three inner fingers are firmly flexed into the palm.

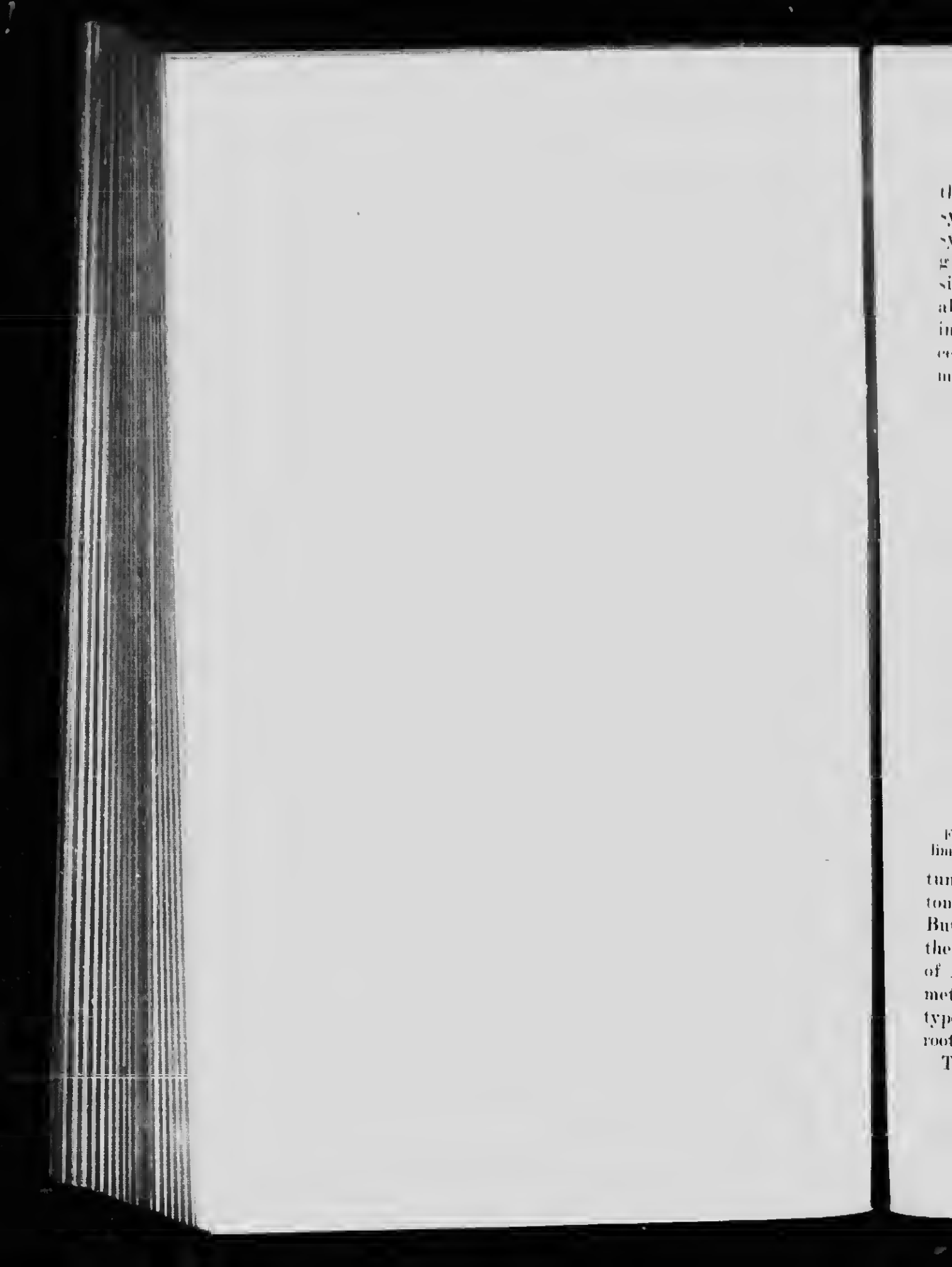
5. Morvan's type. In this variety the trophic changes in the extremities are unusually prominent. Very conspicuous often is a slow necrotic dactylitis which may destroy most of the small bones of the hands or feet, and thus lead to considerable deformity.

Diagnosis. The association in any case of dissociated anæsthesia, muscular atrophy and trophic changes in

PLATE V.



Skiagram of a case of syringomyelia, showing obliteration of the outlines of the right carpal bones by absorption, and by the formation of new bone; the arrow points to the normal bones of the left wrist; the same case as Plate IV.



the skin, bones or joints justifies the diagnosis of syringomyelia. But there are few diseases in which the symptoms present so many variations both in kind and grouping, and occasionally a certain diagnosis is impossible. Any one of the cardinal symptoms may be absent, at least for a time, and the diagnosis may involve a consideration of progressive muscular atrophy, cervical rib, disseminated myelitis, cervical pachymeningitis, spinal hæmorrhage and intramedullary

Fig. 111.—Photograph showing the rigid attitude of the body and limbs; the same case as fig. 110.

tumour. The two last named diseases may cause symptoms almost identical with those of syringomyelia. But in spinal hæmorrhage the onset is sudden, and there is a tendency to gradual improvement. In cases of intramedullary tumour the sensory loss is less symmetrical than in syringomyelia, and is of the peripheral type, whilst the local palsies correspond to certain nerve roots.

Tabes is occasionally simulated, especially when

syringomyelia affects the lower part of the cord; for in both cases there may be burning pains, dissociated anæsthesia, changes in the joints and loss of the knee-jerk. The presence of the Argyll-Robertson pupil,

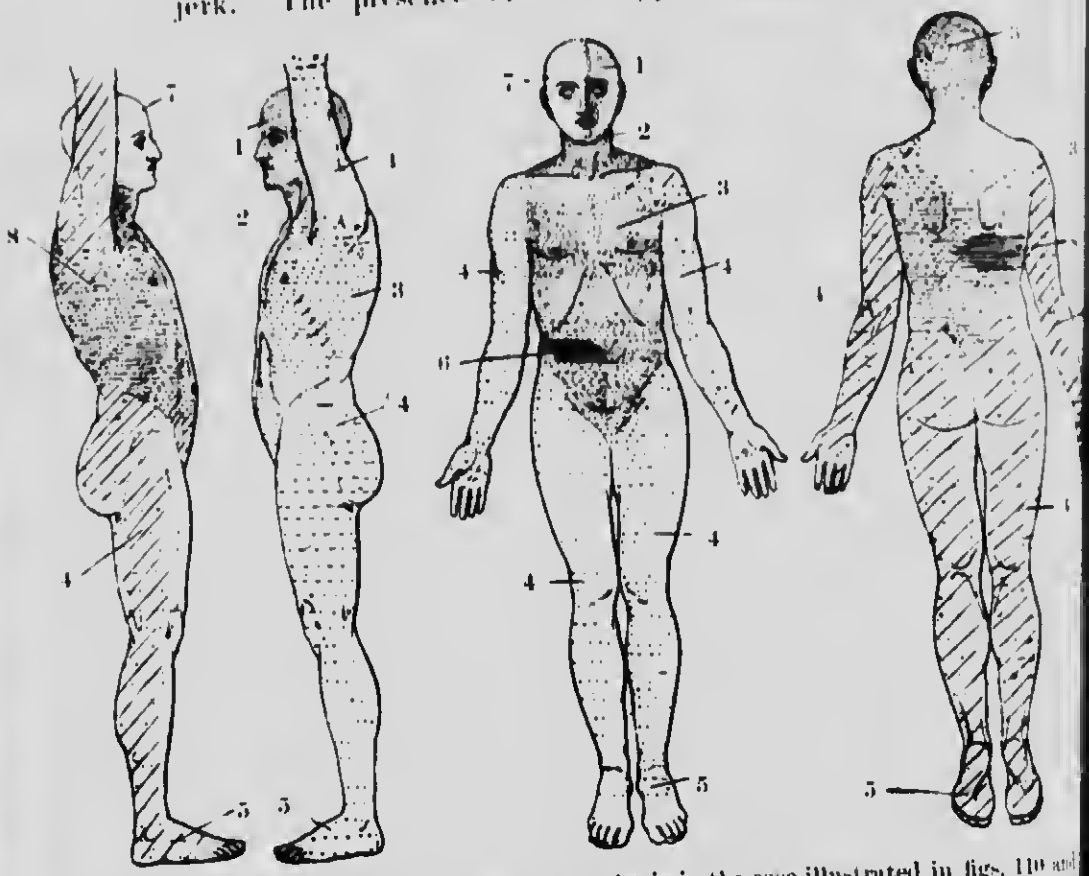


Fig. 112.—Showing the distribution of anæsthesia in the case illustrated in figs. 110 and 111. (1) Impaired sensation to pain; temperature sense normal. (2) Impaired sensation to temperature only. (3) Impaired sensation to both pain and temperature. (4) Slightly impaired sensation to both pain and temperature. (5) Very slightly impaired sensation to both pain and temperature. (6) Impaired tactile sensation. (7) Sensation normal. (8) Slightly impaired tactile sensation.

and of optic atrophy would be in favour of tabes; whilst the development of muscular atrophy in such a case would point to syringomyelia.

Some cases of the spastic type resemble primary lateral sclerosis, but in the latter disease sensory symptoms are absent.

The neuritis of leprosy leads to muscular atrophy and

to irregular areas of anæsthesia, but there are no signs that the functions of the cord are interfered with; frequently too a thickening of the nerves may be felt.

Prognosis. The disease is steadily progressive though periods of arrest sometimes occur. It involves no special danger to life, and many patients are able to follow their occupation for several years. No instance of recovery has been recorded.

Treatment. It is important to warn the patient of the risk of injury to the anæsthetic parts. He should also avoid overstrain of the arms.

No drug appears to have exerted much influence over the course of the disease; in some cases improvement has followed the use of the iodides. Belladonna and ergot have been recommended for painful spasms of the limbs and for weakness of the bladder.

Cases of syringomyelia have been reported in which the application of the x-rays to the spine has arrested and partially cured the disease. The treatment should be in the hands of an expert who will take care that the currents are not too strong, nor the sittings too frequent or too prolonged.

CHAPTER IV.

SPINAL HÆMORRHAGE: HÆMATOMYELIA.

Primary hæmorrhage into the spinal cord is a rare disease. Its rarity as compared with the frequency of cerebral hæmorrhage is explained as follows:—The spinal arteries are less subject than the cerebral to severe disease, partly because they are better supported by connective tissue, and partly because they are long and tortuous; they are therefore less affected by a high blood-pressure, which is such a potent factor in the production of degeneration and rupture of the cerebral arteries.

Secondary hæmorrhage may take place in areas of the cord already diseased in consequence of myelitis, tumours, or syringomyelia. Minute extravasations are

sometimes found after death in cases of severe convulsions, of tetanus and of diseases attended by much venous congestion.

Etiology. Primary hæmorrhage may occur at any age, but it is most common between the ages of twenty and forty. Men are more subject to the disease than women, owing to their more frequent exposure to injury and to muscular strain. Injury is the most frequent exciting cause, especially as a consequence of falls from a height, which cause concussion of the spine: there may, or may not be, an actual lesion of the vertebral column or of the meninges, as for example a partial dislocation with recoil (Thorburn). Injury during labour has caused hæmorrhage into the cord of the newborn child. Violent muscular exertion involving strain, or a sudden forcible bending forwards of the head has occasionally been an immediate antecedent: thus spinal hæmorrhage has suddenly followed a dive into a swimming bath. Very rarely it has occurred in consequence of severe vomiting during pregnancy. Prolonged exposure to cold, suppression of menses, venereal excess, and morbid blood states are other assigned causes.

Pathology. Primary hæmorrhage begins in the grey matter, and often is limited to it: this localisation is due to the greater vascularity and looser texture of the grey, as compared with the white substance. The whole or only one half of the grey matter may be invaded: the posterior or the anterior horn may be involved: sometimes the hæmorrhage surrounds the central canal. There is a much greater tendency for the extravasation to spread longitudinally than laterally, and this is especially the case as regards the posterior horns. The grey matter of the cervical and lumbar enlargements is more frequently affected than that of the dorsal region. Myelitis is sometimes set up in the vicinity of a hæmorrhage. It is stated that the blood becomes absorbed in about six weeks. At a later period the seat of the hæmorrhage is represented by a pigmented scar.

or a cyst-like cavity: possibly, too, the altered tissue may initiate the development of a syringomyelia or a central gliomatosis. On the other hand, a pre-existing cavity may be the underlying cause of a hæmorrhage, which may be set up by an apparently inadequate cause, such as strenuous exertion.

Symptoms. Except in cases of secondary myelitic hæmorrhage, it is rare to meet with numbness and tingling in the limbs or other premonitory symptoms. As a rule the onset of paralysis is quite sudden, its complete development occupying not more than a few minutes. Occasionally the maximum intensity of the symptoms is not reached for an hour or two; this would suggest a gradual escape of blood, a slight leakage being followed by more extensive extravasation.

Frequently the onset is marked by transient pain at the seat of the lesion; usually the pain is not severe, but sometimes it is intense and radiates along the areas innervated by the affected nerve roots. In one of my cases severe pain between the shoulders was followed in less than a quarter of an hour by complete paralysis of the hands and the lower limbs.

Consciousness is usually retained: occasionally it is lost for a brief period owing to shock from the abruptness of the onset or from the severity of the pain.

In many cases the fully-developed symptoms are those of a complete transverse lesion of the cord, namely, absolute loss of power and sensation below the level of the lesion, with paralysis of the bladder and rectum and loss of all the reflexes. At the level of the lesion there may be a band of hyperæsthesia around the body. The temperature, normal at first, may be slightly raised in the course of a few days, owing to the development of myelitis.

When the hæmorrhage is in the cervical enlargement, there is, in addition to a flaccid paralysis of the four limbs, loss of power in the abdominal and thoracic muscles, so that, if the fourth segment is spared respiration is carried on by means of the diaphragm.

It may be still further embarrassed by acute distension of the abdomen. When the lowest portion of the cervical region is involved oculo-pupillary changes occur, and the temperature of one side of the face may be higher than that of the other side.

After a week or two a change in the symptoms is observed. Some of the muscles of the still flaccid arms begin to atrophy and to show changes in their electrical reactions. The legs, however, become spastic, the tendon reactions are exaggerated and the extensor plantar reflex is obtained. Constipation and retention of urine, which characterise the early stage, may give

Fig. 113.—Distribution of anæsthesia in a case of hæmorrhage into the lower cervical segments of the spinal cord. (Ross.)

place to incontinence of both urine and fæces at a later period.

As regards sensation, the initial complete anæsthesia may be replaced by the dissociated variety in which tactile impressions are perceived, whilst sensations to pain, heat and cold are lost either on both sides, or on the side opposite to the paralysed limb, giving rise to Brown-Séquard's symptom-complex. In some cases the dissociated form of anæsthesia is present from the ontset.

When the lesion is in the dorsal region, the arms are not affected, but otherwise the symptoms are similar. When it is in the lumbar region the flaccidity persists, and certain muscles of the legs, according to the position

of the lesion, undergo progressive atrophy, and give the reaction of degeneration. The knee-jerks are lost; the sphincters in many cases are paralysed. In spinal hæmorrhage bed-sores are apt to form at an early period and to become severe. Cystitis is another serious complication.

Diagnosis. It is important to distinguish between a hæmorrhage into the spinal cord and one into the spinal membranes, for in the latter condition laminectomy may relieve the pressure, whereas in the former condition the operation can be of no benefit. Meningeal hæmorrhage is characterised by the violence of the pains in the back and limbs and by jerking of the muscles; spinal hæmorrhage by a rapid development of paralysis which is usually complete, and by dissociation of sensation.

Some cases of acute myelitis closely resemble hæmatomyelia; as a rule the onset of myelitis is less sudden, and it is preceded by premonitory symptoms and by slight pyrexia.

Dissociated anæsthesia is a striking feature of syringomyelia, but the onset of the disease is gradual, and its symptoms slowly develop as time goes on; whereas partial recovery takes place in many cases of hæmatomyelia.

Prognosis. The prognosis is favourable both as regards the duration of life and the improvement in the symptoms. Complete recovery is quite exceptional; the rule is that while some portions of the limbs regain sensation and power, other portions remain permanently anæsthetic and powerless. Death may occur at an early period as a result of paralysis of the respiratory muscles, or at a later period in consequence of a purulent cystitis, or of infection through a bed-sore.

Treatment. Absolute rest in bed for a few weeks is of the greatest importance; it is better for the patient to lie in the prone position than on his back. In a strong subject it is desirable to withdraw blood from the

back by cupping or by the application of leeches. Injections of ergotin have also been recommended.

Attention should be given to the condition of the bowels, and to the prevention of straining, as in coughing. The subsequent treatment is similar to that for myelitis. Every care must be taken to prevent cystitis and the formation of bed-sores. The restoration of function in atrophied muscles may be hastened by the employment of massage and electricity.

CHAPTER V.

COMPRESSION-AIR SICKNESS: CAISSON DISEASE: DIVER'S PARALYSIS.

This disease occurs in men who work under water in a highly-compressed atmosphere, either in diving dress or in metal cylinders called caissons. The men do not suffer during their exposure to the compressed air, but after their return to a normal atmosphere, that is, the illness comes on only after decompression; the essential factor is a sudden change from a high to a low atmospheric pressure. The higher the pressure, the longer the exposure, and the more rapid the return to a normal air-pressure, the greater is the risk.

Symptoms. For a time after leaving the water the man may feel quite well, but after a few minutes or longer he is suddenly seized with pains in the limbs or body which are quickly followed by paralysis. In mild cases the patient complains merely of slight pain, of tingling and numbness in the limbs and of heaviness and weakness in the legs. In severe cases intense neuralgic pains are referred to the legs, knees and hips, and sometimes to the arms. Pain in the back or in the epigastrium also occurs: when it occurs in the epigastrium it is often accompanied by nausea and vomiting. The legs become quickly paralysed, and there may be complete paraplegia with anæsthesia and loss of the

reflexes. Frequently the pains persist and may occur in parts which are completely analgesic.

When the paraplegia is incomplete the knee-jerks are present, and are sometimes exaggerated; the case then resembles one of dorsal myelitis.

Symptoms of cerebral disturbance are present in some cases; as a rule they are slight and transient. They comprise headache, vertigo, slight mental disturbance, deafness, temporary coma and a transient monoplegia or hemiplegia.

In most cases recovery takes place; sometimes in a few days, sometimes not for several months. In severe cases paralysis may be permanent. Occasionally death occurs shortly after the attack.

Pathology. The following changes have been found in the spinal cord, more especially in the posterior and lateral columns of the dorsal region:- Small fissures, surrounded by zones of parenchymatous myelitis, minute hæmorrhages, small areas of softening, and dilatation of the blood-vessels and the perivascular lymph sheaths. The spinal membranes and the nerve roots are not affected; the grey matter is usually normal, or may present slight changes, as vacuolation of its cells, and enlargement of the pericellular spaces.

The explanation of these changes is that during the period spent by the worker in the compressed air, the blood becomes surcharged with gases, and on a sudden return to the normal atmospheric pressure bubbles of gas escape from the small blood-vessels and rupture the nerve-tissues, or form emboli in the capillaries which lead to minute softenings.

Another possible cause of some of the symptoms of compressed-air sickness is the breathing of oxygen at a high tension. Heavy work is more easily performed in an atmosphere containing an increased percentage of oxygen, but beyond a certain limit, an increase of oxygen pressure is poisonous: this, as suggested by Lorrain-Smith, may be a factor in the causation of the disease.

Treatment. In order to avoid the risk of this disease the transition from a high to a low atmospheric pressure must be gradual; the period of decompression being regulated according to the duration and degree of compression.

The treatment of the symptoms is immediate re-compression; that is, the patient should again be put under increased pressure which is then gradually reduced. By this means relief is afforded to the pains, and the other symptoms may also be arrested or cured. When pain is severe morphia may be necessary. The treatment of the developed disease is that of an acute myelitis and its complications.

SECTION VII.

Spinal Disorders Characterised by the Presence of Pain.

CHAPTER I.

DISEASES OF THE SPINAL MEMBRANES.

The chief morbid processes that affect the spinal membranes are hæmorrhage, inflammation and tumours.

Hæmorrhage: Hæmatorrachis.

Meningeal hæmorrhage is very rare; the blood is extravasated either outside or inside the dura-mater, in the latter case it may be either between the dura and arachnoid, or beneath the arachnoid. Sometimes the extravasation is a simple extension from intra-cranial hæmorrhage. In other cases it is the result of injury to the spine, of severe convulsions, or even of excessive muscular exertion. Occasionally it occurs in connection with the hæmorrhagic diathesis, or in the hæmorrhagic forms of small-pox, yellow fever or other acute disease. Rarely it is due to the rupture of an aneurysm of the aorta or of one of the vertebral arteries into the spinal canal. Hæmorrhage takes place more commonly outside than inside the dura mater, and in the cervical region than in other parts; in some cases the cord is compressed.

Symptoms. These may be slight or indefinite, but usually they are marked and point to severe irritation of t' membranes and the nerve roots. Characteristic symptoms are: A sudden violent pain in the back, shooting pains along the course of the nerves, hyperæsthesia and various subjective sensations. Rigidity of the back and reflex muscular spasm usually accompany the pain. Weakness of the limbs and a varying

degree of anæsthesia may ensue—the arms being most affected when the cervical region is the seat of hæmorrhage.

There is no pyrexia; the absence of fever and the sudden onset of the symptoms distinguish meningeal hæmorrhage from meningitis. The greater frequency of severe and widespread pains distinguishes it from hæmorrhage into the cord, in which also paralysis is present from the onset.

A fatal issue is common and death occurs either quickly, or after a few days as a result of secondary meningitis. The most serious cases are those of cervical hæmorrhage owing to the risk of respiratory paralysis. Should the patient survive a week or ten days the prognosis becomes less grave, unless some complication has developed.

Treatment. The patient should lie on his face or side and should be protected in every possible way from disturbance of mind or body. Local venesection by leeches or cupping followed by an ice bag to the spine is advisable. Afterwards an aperient should be given: morphia may be required to relieve the pain. The question of laminectomy for removal of the blood should be considered, especially when life appears to be threatened.

Spinal Meningitis.

It is noteworthy that while spinal meningitis is frequently found at post mortem examinations it does not often give rise to distinctive manifestations during life. This is due partly to the fact that in the chronic forms of meningitis, which are common, the symptoms are generally slight or absent, and partly to the rarity of acute meningitis.

The symptoms of meningitis are mainly due to involvement of the spinal roots and, in one of the most frequent varieties of meningitis, namely, that which is associated with caries, they are rarely very conspicuous. Another fairly common variety is the

meningo-myelitis due to syphilis; in this variety root-symptoms are often more evident. (See Section XXI.).

It is true that symptoms indicating an affection of the spinal roots, especially of those in the cervical region, are not uncommon. Thus the physician is consulted for radiating pains down the arms which, from their distribution, obviously depend on irritation of the roots rather than of the peripheral nerves: on examination a tender spine may be detected as well as bands of slight anæsthesia or hyperæsthesia down the outer or inner aspect of the arms. In many of these cases the evidence is against meningitis and is in favour of an affection often rheumatic or gouty—limited to the roots: secondary involvement of the cord does not occur, and consequently complete recovery is the rule.

Pathologically we find that the spinal membranes, even the arachnoid, may be affected separately: but there is a tendency, especially when the inflammation is acute, for the process to spread from one membrane to the other. Leptomeningitis is a term applied to inflammation of the pia-mater or the pia-arachnoid: it may run an acute or a chronic course. Pachymeningitis or inflammation of the dura-mater occurs in two forms, namely, external when the inflammation affects the outer, and internal when it affects the inner aspect of the dura-mater. Sometimes meningitis is spoken of as external or internal: by the former is meant meningitis beginning outside the dura-mater; by the latter meningitis beginning within its sheath.

Acute Leptomeningitis.

Etiology. One cause of this rare disease is the tubercle bacillus; another cause is the diplococcus intracellularis of epidemic cerebro-spinal meningitis. In both cases as a rule the membranes of the brain are involved as well as those of the cord. Other rare sources of infection are: Pneumonia, septicæmia, gonorrhœa, ulcerative endocarditis and pelvic suppuration; the acute infections diseases as erysipelas, typhoid, scarlet

fever and influenza. Some cases of acute meningitis have immediately followed injury or exposure to cold: it may be assumed that a lowering of tissue resistance has permitted the invasion of micro-organisms.

Morbid Anatomy. Congested at first, the pia-arachnoid soon becomes opaque and thickened, and is covered with inflammatory exudation which may be semi-purulent. The inner aspect of the dura-mater is often involved and in places is adherent to the pia-mater. The spinal fluid is turbid and purulent and is increased in quantity. The nerve-roots may be surrounded with exudation, which sometimes invades the cord and sets up a marginal myelitis.

In tuberculous cases, grey granulations are found scattered in the exudation which is usually gelatinous and small in quantity: as a rule the condition is associated with tuberculosis of the cerebral membranes. In severe septic cases the pus around the cord is continuous with that at the base of the brain. Diplococci, pyococci and other micro-organisms have been found in the exudate.

Symptoms. The onset is acute and is characterised by rigors, pyrexia and pain in the back. The pain becomes severe and is intensified by movement and by pressure over the spinous processes.

Irritation of the posterior roots is expressed by lancinating pains which occur in paroxysms of great severity and radiate across the body or down the limbs according to the seat of the lesion. Irritation of the anterior roots gives rise to muscular spasms which involve the neck, trunk or limbs, so that there may be retraction of the head, opisthotonus, or rigidity of the limbs varying in degree at different times and in different cases. Cutaneous and muscular hyperæsthesia also are often conspicuous features. Implication of the vaso-motor nerves is shown by an erythema readily produced by stroking the skin; even large wheals may thus occur. Sometimes a crop of herpes develops.

The superficial reflexes and the tendon reactions are

generally exaggerated, and Kernig's sign can usually be elicited. The bowels are constipated and there is retention of urine.

After a few days or longer the symptoms of irritation may subside and in some cases are replaced by symptoms indicating destruction of the nerve roots or the cord. The former are more resistant to disease than the latter, hence paralysis and anæsthesia are generally the result of damage to the cord. The distribution and variety of the paralysis will vary with the position of the disease, whether in the cervical, dorsal or lumbar region (see myelitis). When the cervical portion of the cord is affected there may be considerable dyspnœa, deranged action of the heart and unequal pupils. When the membranes of the brain are also diseased, vomiting, headache, signs of implication of the cranial nerves, delirium or coma may be present.

The symptoms of meningitis vary according to its nature, being often slight in purulent and in tuberculous cases; in the latter they are often masked by the associated brain disease.

Course and Prognosis. Acute purulent meningitis is nearly always rapidly fatal. Mild cases of meningitis may recover completely; in other cases recovery is gradual and only partial, a variable amount of paralysis and anæsthesia remaining; the patient is often permanently crippled either by spastic weakness, or by atrophy of some groups of muscles. Some patients die from respiratory paralysis, others from the secondary consequences of bed-sores or of cystitis.

The earlier paralysis sets in and the severer the general symptoms, the worse is the prognosis. The chances are better in cases that follow slight injuries or exposure to cold than in those which complicate infective fevers, or depend on severe injury to the spine.

Diagnosis. This chiefly depends on the association of fever with symptoms of irritation of the spinal roots. In uncomplicated myelitis pain in the back is absent or inconspicuous.

The muscular spasms and rigidity of tetanus are similar to those of meningitis, but in tetanus, trismus is an early and a marked symptom; the muscular spasms are easily excited by external stimuli, and pyrexia is absent, at any rate at the onset; the skin is not hyperæsthetic.

The fluid withdrawn by lumbar puncture is of service in differentiating meningitis from other diseases, and in deciding as to its nature. The number of cells is often greatly increased; polynuclear leucocytes are found in non-tuberculous meningitis, mono-nuclear lymphocytes and sometimes tubercle bacilli in tuberculous meningitis. Streptococci would be in favour of purulent meningitis, whilst the diplococcus intracellularis would point to the cerebro-spinal variety, which in this country is more commonly met with in the sporadic than in the epidemic form.

Treatment. Perfect rest and quiet, careful feeding and nursing are of the first importance. A water-bed is usually advisable. Dry cupping to the spine should be employed at the onset; afterwards it is desirable to rub mercurial ointment into the back. For the relief of pain it is often necessary to administer morphia; in the severest cases inhalation of chloroform may be required. Should the acute symptoms subside, iodide of potassium is sometimes useful and tonics such as iron, quinine and strychnine are often beneficial. Counter irritation to the spine, hot douches and warm baths should now be recommended, whilst massage and electricity may do much to develop the muscles and lessen the contractures.

Chronic Lepto-meningitis.

Clinically it is scarcely possible to distinguish this variety from pachymeningitis, with which indeed it is frequently combined.

In a localized form it is found in vertebral caries, in traumatic affections of the spine and in myelitis which involves the periphery of the cord; the commonest form of meningo-myelitis is that due to syphilis (see Section XXI.).

Chronic meningitis is also found in tabes, in chronic alcoholism and in old age. Sometimes it follows the acute variety; in other cases it begins insidiously. The patient complains of pain and stiffness in the back, of paraesthesia and of a feeling of weakness and heaviness in the limbs. He may suffer also from radiating pains in the area of distribution of nerves the roots of which are implicated in the lesion. Cutaneous hyperaesthesia in the legs occurs: anaesthesia when present is usually slight, and is represented by a blunting of the sensibility of the feet and legs. Complete paraplegia is rare, but weakness of muscles followed by wasting is common and may be associated with contractures.

Localized Serous Spinal Meningitis.

Recently attention has been called, especially by Horsley, to a variety of chronic spinal meningitis which produces symptoms closely resembling those of a meningeal tumour. A circumscribed collection of cerebro-spinal fluid is found pressing on the cord, usually in the lower dorsal region. The condition generally occurs in adults; there is some evidence that it may be caused by syphilis, gonorrhœa, or influenza. Horsley states that the symptoms differ from those of a spinal tumour in the following respects: The pain is more diffuse in distribution: it first affects the whole of one limb, then spreads to the other limb, and finally goes up the back. The paralysis is not limited to a group of muscles, but it affects the whole limb uniformly, and is of the spastic variety.

The prognosis is favourable if operation is not too long delayed. Horsley recommends a simple laminectomy, followed by opening of the theca and washing out with a mercurial lotion.

Internal Pachymeningitis.

In leptomeningitis the inflammation often spreads to the inner surface of the dura mater. The latter is also involved in two other affections, namely, internal

hæmorrhagic pachymeningitis, and internal hypertrophic meningitis.

Internal Hæmorrhagic Meningitis, or Hæmatoma of the Dura Mater. This is a rare disease; usually it is found in association with a similar condition in the dura mater of the brain. A reddish-brown exudation covers the inner surface of the dura; it is composed of fibrin, connective-tissue and extravasated blood which may be enclosed in cyst-like cavities.

The symptoms are those of a slight meningitis, but they are often masked by those due to the accompanying cerebral condition. In many of the cases the patients

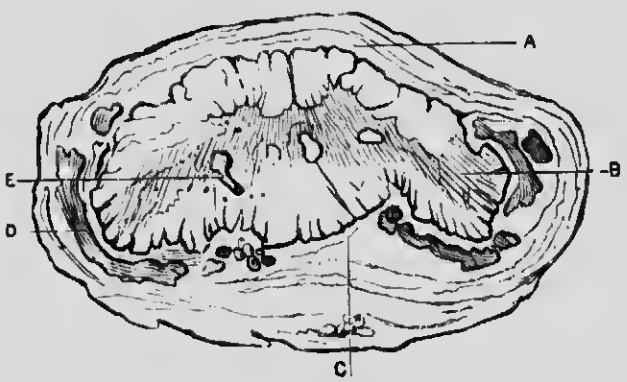


Fig. 114.—Transverse section of the middle of the cervical enlargement, from a case of hypertrophic cervical pachymeningitis. A, Hypertrophied dura mater; B, roots of the nerves traversing the thickened membranes; C, pia mater adherent to the dura mater; D, lesion of chronic myelitis; E, cystic formation in the grey substance. (Charcot and Joffroy.)

are addicted to chronic alcoholism, or suffer from general paralysis of the insane.

Internal Hypertrophic Meningitis. This especially affects the cervical region; hence it is often called hypertrophic cervical meningitis. Its causes are uncertain: exposure to cold, injury and syphilis have been mentioned.

The dura mater becomes greatly thickened and adherent to the neural arches, and to the pia-arachnoid which is also thickened. The new tissue compresses first the nerve roots and later the spinal cord: a surface

myelitis develops, and ultimately the whole transverse area of the cord becomes sclerosed at the level of the lesion. Some of the changes in the spinal cord may be primary; in syphilitic cases inflammation of the interstitial tissue of the cord may be concurrent with that of the membranes.

At first the patient suffers from severe pain in the neck and between the shoulders: the pain radiates to the head and down the arms. Muscular twitches occur, and there is usually much stiffness of the neck and arms. After a period of two or three months paralysis and anæsthesia develop. The muscles supplied by the affected roots undergo progressive atrophy, and as the lower portion of the cervical region is usually mainly involved the muscles supplied by the median and ulnar nerves are correspondingly paralysed; hence the wrist and the metacarpo-phalangeal joints are hyperextended whilst the terminal phalanges are flexed. Oculo-pupillary phenomena owing to involvement of the sympathetic fibres which run in the first dorsal roots are sometimes present. When the lesion is at a higher level the muscles supplied by the fifth root are often chiefly affected. At a later stage symptoms of compression of the cord are observed, namely, spastic paraplegia, diminished sensation in the legs and trunk, and disturbance of micturition.

Very rarely the lumbar region is the seat of this disease; then there are shooting pains down the legs, and at a later period some of their muscles become paralysed and atrophied. The sphincters are often involved at an early period.

The presence of severe pains in the neck, shoulders and arms distinguishes this affection from amyotrophic lateral sclerosis and from cervical myelitis. In vertebral caries the pains are rarely so severe, and there are usually signs of bone disease.

When syphilis is suspected, mercury and iodide of potassium should be administered; radiant heat or sulphur baths should also be tried. In all cases counter

irritation to the spine is beneficial; blisters or painting with iodine may do good, but the actual cautery is the most efficacious. When the pain is severe, phenacetin or morphia may be required; division of the nerve roots has also been recommended. In severe and protracted cases it may be necessary to consider the question of freeing the thickened membranes by a surgical operation.

External Pachymeningitis.

In this variety of meningitis the inflammation begins on the outer side of the dura mater. As a rule it is secondary to disease of adjacent structures, most frequently to tuberculous disease of the vertebræ. In such cases the process is chronic, and is of limited extent: the dura mater is thickened and its outer surface is covered by caseous or purulent material. Sometimes an acute and extensive pachymeningitis is set up by a bed-sore which has perforated the sacrum, or by an abscess in the neighbourhood of the spine.

The symptoms are similar to those of other forms of meningitis. In the acute variety the prominent symptoms are those of root-irritation combined with rigors and pyrexia. In the common chronic, localized variety, root-symptoms are usually slight; the chief manifestations are caused by a gradual compression of the spinal cord.

The treatment is mainly that of the primary disease.

CHAPTER II.

DISEASES GIVING RISE TO COMPRESSION OF THE SPINAL CORD: COMPRESSION MYELITIS.

Compression myelitis is a term applied to changes in the spinal cord that are the direct or the indirect result of encroachment on the vertebral canal; actual compression of the cord does not always occur.

The two most common causes of this condition are tuberculous caries and fracture-dislocation of the vertebral column; the latter belongs to surgery rather

than to medicine and will not be dealt with in this book. Other causes are:—Tumours in the spine or in the membranes, including hydatid cysts; aneurysm of the aorta; and very rarely caries due to syphilis; arthritis deformans and exostoses.

TUBERCULOUS CARIES.

Tuberculous disease of the spine develops most frequently in childhood, sometimes in middle adult life, and occasionally at a later period. In many cases a tuberculous inheritance can be traced, and very often the patient presents signs of tubercle in the lungs, the

Fig. 115.—Caries of spine (diagrammatic). On the left is a longitudinal section of the spinal cord and dura mater. To the right is a transverse section of the vertebral column, showing section of spinal cord and dura mater in vertebral canal. The carious part of the vertebra is darker in colour. Note the thickening of the dura mater—external tubercular pachymeningitis, which is causing compression myelitis. (Williamson.)

glands or in other parts of the body. Sometimes the disease appears to have been excited by injury to the back.

Pathology. At first tubercles develop in the bodies of one or two vertebræ and less commonly in the vertebral joints or the inter-vertebral cartilage. Caseous purulent material is formed which leads to breaking down and collapse of the bones, a frequent result being undue prominence of one or more spinous processes.

In most cases the tuberculous disease spreads to the

connective and adipose tissue outside the dura mater, the resulting formation being the usual cause of compression of the cord. In other cases the cord is directly compressed by displacement of the bones, or by an abscess which has formed at the seat of the lesion. The cord is not always narrowed; the degenerative changes in its nerve elements are probably mainly due, not to direct compression, but to œdema from obstruction of the venous and lymphatic circulation, and partly also to true inflammation.

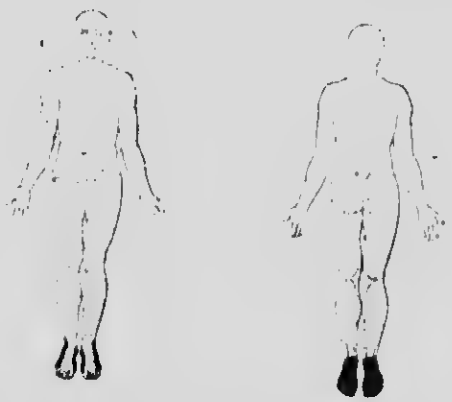
At first the cord is softer than normal; in advanced cases it is firmer than normal, owing to selerosis of the interstitial tissue, when the usual ascending and descending degenerations can be traced from the diseased area. It is probable that these later changes are uncommon. At any rate it is remarkable how frequently a long-standing paraplegia from caries will pass away either partially or completely. Such a result is quite in accordance with the usual pathological findings, namely, œdema and narrowing of the cord without actual destruction of its nerve elements. When the bone changes become arrested and the œdema subsides, the functions of the nerve cells and fibres may be gradually restored. The nerve roots are often irritated and compressed as they pass across the vertebral canal or through the inter-vertebral foramina.

Symptoms. Some of the symptoms produced by spinal caries are due to bone disease, some to implication of nerve roots, whilst others depend on changes in the spinal cord.

Bone symptoms. These which may be alone present, consist of pain and tenderness over the carious bones, rigidity of the back, and deformity of the spine; sometimes an abscess is present. The pain is increased by movement, by deep pressure on the spine and often by attempts to move it laterally. Pain is also caused by pressure on the head or the shoulders.

In cervical caries the head is often inclined to one side, and is held stiffly in order to avoid the pain

produced by movement. When spinal tenderness is not very obvious it may be evoked by passing a hot sponge or the kathode of a galvanic battery down the spine. Pain in the spine and rigidity of the back or neck precede, often by several months, any deformity. Sometimes the deformity never appears: frequently it is absent in cervical caries, which, however, may be associated with thickening of the surrounding tissues. An abscess may form near the disease and point backwards; sometimes pus finds its way into the mediastinum or descends in the sheath of the psoas muscle to the



Figs. 116 and 117.—Charts showing the impairment to touch, pain and temperature in a case of tuberculous caries of the eighth and ninth dorsal vertebræ. The darkly shaded parts of the feet represent total loss of sensation to cotton wool.

groin. A retro-pharyngeal abscess is sometimes started by cervical caries.

Root symptoms. These are rarely severe, and may be absent. In dorsal caries they are represented by a girdle pain and by a zone of cutaneous hyperæsthesia extending from the affected spot a variable distance round the trunk. In cervical and lumbar caries pains are felt down the arms or the legs and bands of hyperæsthesia may be traced along the limbs varying in distribution according to the roots affected. In some cases hyperæsthesia is succeeded by anæsthesia, or anæsthesia may co-exist with pain—anæsthesia dolorosa.

Herpes zoster is occasionally present, and this eruption, whether on the limbs or the trunk, should always suggest the possibility of spinal disease. When the lower cervical vertebræ are diseased the sympathetic fibres may be implicated, giving rise to contraction of the pupil and to sweating on one side of the face.

When the upper cervical vertebræ are diseased the phrenic nerve is liable to be affected; the spinal accessory and even the hypoglossal and the fibres from the vagus to the palate are involved in rare cases.

Spinal cord symptoms usually develop very gradually; sometimes they come on rapidly and occasionally quite suddenly.

When caries affects some of the dorsal vertebræ, there is a spastic paraplegia with a varying amount of disturbance of sensation and of the functions of the bladder and the rectum. In caries of the lower cervical vertebræ some muscles of the arms may be weak and wasted, whilst those of the legs may become weak and spastic; the intercostal muscles are also liable to be affected. In caries of the upper cervical vertebræ all the limbs may show spastic paralysis. In lumbar and sacral caries certain muscles of the legs, varying with the situation of the bone-disease, become paralysed and undergo a degenerative atrophy.

For further details relating to the symptomatology the reader is referred to page 272; the cord symptoms produced by caries being in every way similar to those produced by an uncomplicated transverse myelitis; it is to be noted, however, that much sensory disturbance, and especially complete anæsthesia up to the level of the lesion, is less common in compression than in ordinary myelitis; in compression also, there is less tendency to the formation of bed-sores and to paralysis of the bladder.

Course. In many cases the signs of vertebral caries and the symptoms of cord implication develop about the same time, but in other cases there is no correspondence between the development and course of the

bone and cord disease. A curvature or a prominent vertebral spine may exist for many years before paralytic symptoms appear: or the symptoms of compression myelitis may develop before the signs of bone mischief are obvious. In some cases paralysis passes away during the time that the spinal deformity is becoming more and more marked; on the other hand paralysis may persist after the carious process is healed.

Complete and permanent restoration of power is not uncommon: occasionally relapses occur. Death is generally the result of some complication—infection from a bed-sore; cystitis, pyelonephritis; or general tuberculosis.

Diagnosis. Compression myelitis from spinal caries is frequently curable, hence the early recognition of bone mischief is of great importance. In all cases of spinal cord disease the back should be carefully and repeatedly examined, in order that signs of caries or of other lesion outside the cord may not escape notice. Even apart from a definite vertebral prominence, localised pain and deep tenderness, together with rigidity of the back during movement are suggestive signs that the paralytic symptoms depend on disease of the bones. In many cases, and especially in cervical caries, examination with the Roentgen rays is frequently of service.

The motor symptoms of compression myelitis in the cervical region, namely, muscular atrophy in the arms and spastic paralysis in the legs are those of amyotrophic lateral sclerosis; but in the latter disease sensory symptoms do not occur.

Tenderness of the spine is met with in hysteria and in neurasthenia, but in these conditions it is not localised, nor is it limited to deep pressure as in caries. Sometimes the symptoms of hysteria co-exist with those due to a compression myelitis: then much judgment may be required to distinguish between them. In caries, root pains are occasionally felt only on one side, when they may be thought to indicate a simple

neuralgia; here again a careful examination of the spine may remove any difficulty.

Prognosis. It is always difficult to foretell the result in a case of paralysis from spinal caries. As a rule the outlook is more favourable in children than in adults and tends to become graver as life advances. Damage to the cervical or to the lumbar enlargement of the cord is more serious than when the dorsal segments are affected; in cervical caries there is the danger of respiratory paralysis, and in lumbar caries the possibility of trophic changes.

The power of recovery shown in young persons is sometimes astonishing: cases have been observed in which an almost complete paralysis of the limbs and of many of the respiratory muscles has entirely passed away, the patients making a complete recovery. As Gowers truly says there is no disease of the cord in which symptoms of equal gravity so often pass away.

In all cases of this disease the prognosis is largely dependent on the exact cause of the compression symptoms; frequently this can be determined by the use of the *x*-rays, when the probability of its successful removal by operation may be estimated with some degree of certainty. Thus the detection of an abscess situated near the cord suggests that its opening would be followed by recovery; whereas if the skiagram shows great compression by bone the prognosis will be graver, for the probability of success following an operation will be much less.

Treatment. A child suffering from spinal caries in combination with symptoms pointing to the involvement of the spinal cord or its roots, should be kept on its back in bed for at least a year, or until such symptoms have subsided. Whenever possible it is desirable to wheel the bed into the open air, and to carry out in every other way the general treatment for tuberculosis: this includes the administration of tonics, cream, cod-liver oil and other fatty substances.

In many cases this treatment, combined with methods

for extending and supporting the spine, is attended by excellent results; the tuberculous lesion becomes healed, the cord is relieved from pressure and the patient gradually regains the use of his limbs. During this course of prolonged rest, the condition of the bladder should receive constant attention and every precaution be taken against the formation of bed-sores.

In adults the results of rest and extension are rarely so favourable, and if no improvement is observed at the end of three months the question of laminectomy ought to be considered. In persons of advanced age spinal tuberculosis is an intractable disease and treatment is of little avail; frequently the disease progresses with rapidity and leads to a myelitic softening of the cord. In some cases an early laminectomy may afford relief.

For the best methods of dealing with abscesses found in association with spinal caries; for a description of the jackets and other kinds of support that are needed in different cases of this disease, and for the indications which render the operation of laminectomy advisable, the reader is referred to surgical works. It must suffice to say that laminectomy should be performed (1) when there is reason to believe that the compression symptoms are caused by a deep-seated abscess; the opening of such an abscess and the removal of necrosed bone often relieve the symptoms and promote a rapid and complete recovery; (2) when no improvement in the condition of the paralysed limbs has occurred after prolonged rest in bed; and (3) when life is threatened owing to the symptoms produced by pressure upon the upper cervical spinal segments.

The operation is contra-indicated when the pressure symptoms suggest that there is more than one focus of disease, and also in cases where there are signs of general tuberculosis or of a local tuberculosis affecting the kidneys or other organs. The operation is not devoid of risk; it may be followed by a renewed activity

of the spinal disease, or by the development of tuberculous meningitis.

In rare cases compression myelitis has resulted from aneurysm, exostoses and various affections inplienting the vertebral articulations.

An *aneurysm of the aorta* may erode the dorsal or the lumbar vertebræ and lead to compression of the cord or its nerve roots. If it ruptures into the vertebral canal complete paraplegia develops suddenly and death quickly ensues from ascending paralysis.

Exostoses growing from the bodies of the vertebræ may slowly compress the cord, but a diagnosis can hardly be made unless similar exostoses are found elsewhere.

The most common joint affection is *arthritis deformans*, which may lead to complete ankylosis of the vertebral articulations; in such a case there would be great rigidity of the neck or back. Sometimes the foramina are narrowed causing injury to the nerve roots, followed by radiating pains in the limbs or round the trunk; even slight muscular atrophy may ensue.

VERTEBRAL TUMOURS.

The bones of the spine may be the seat either of benign or of malignant growths; the latter occur more frequently than the former. Carcinoma of the vertebra is always secondary to a primary growth elsewhere, as in the breast, uterus, stomach or œsophagus. Sarcoma may be primary or secondary, single or multiple.

The malignant growth erodes and softens the vertebræ, and thus leads to collapse of their bodies, to spinal curvature and a general shortening of the vertebral column.

It may invade the muscles and other tissues outside the bone and form a swelling by the side of the spine: frequently too it narrows the inter-vertebral foramina, surrounds the dura mater, and compresses the nerve-roots and the spinal cord. Direct compression of the

cord is less common than in spinal caries; the paralytic symptoms are generally the result either of oedema of the cord, or of acute myelitis. It is rare for the growth to penetrate the dura mater and still rarer for it to invade the cord.

Symptoms. Symptoms may be caused by the bone disease, or by implication of the nerve roots or the spinal cord.

Vertebral symptoms. As a rule, but not invariably, pain and tenderness on pressure over the diseased bones are conspicuous features. The pain, often severe and boring in character, is increased by movement and may disappear when the patient is at rest in the recumbent posture; in many cases cessation of the pain during rest is a striking feature.

Curvature of the spine at the seat of the growth is sometimes present; it is usually more rounded than in caries. In some cases, especially at a late period of the disease, a swelling may be detected near the spine; this is especially noticeable when the cervical vertebræ are implicated.

Root symptoms. Pressure on, or invasion of, the posterior roots gives rise to severe pain; this is an early and a constant symptom. At first the root-pains may be slight and intermittent, but they gradually increase in intensity, and sometimes paroxysms of excruciating pain are produced by the slightest movement of the body. The pains radiate down the arms or the legs, or around the trunk according to the position of the lesion. Associated with them there may be areas of hyperæsthesia, whilst when the nerve-roots are destroyed corresponding zones of anæsthesia may be detected. Occasionally herpes zoster develops owing to disease of the posterior root ganglia.

Irritation of the anterior roots may be expressed by muscular spasms and twitchings, but as a rule these are slight or even absent. Destruction of the roots leads to an atrophic paralysis of the muscles supplied by them.

In many cases of malignant disease of the vertebræ, bone and root symptoms are alone present, and death occurs before any compression of the cord has taken place. In other cases *spinal cord symptoms* appear early and are steadily progressive. They are similar to those met with in spinal caries (see p. 320). Generally, they develop very gradually, but sometimes with great rapidity, in consequence either of displacement of bone or of an acute myelitis set up at the level of the growth; all power in the legs may be lost within twenty-four hours.

Fig. 118.—Chart showing the distribution of complete anæsthesia in a case of sarcomatous tumour involving the first two dorsal vertebræ; the dotted parts represent hyperæsthesia. In this case there was complete flaccid paraplegia.

Course. The course of the disease is steadily progressive, although the degree of pain and paralysis may lessen from time to time. In slowly growing tumours life may be prolonged for several years, but in cancer death usually occurs in from nine to eighteen months.

Diagnosis. The presence of a vertebral tumour may be diagnosed with certainty if paraplegia is found in association (1) with a painful deformity of the spine, the pain being increased by pressure and by movement; (2) with severe root pains; (3) with evidence of a primary growth in some other part of the body, and (4) with increasing weakness and emaciation. In some cases the diagnosis has been confirmed by the use of

the *x*-rays. It is in cases in which signs of compression of the cord do not appear even after pains in the spine and along the roots have existed for many months that the diagnosis is difficult. Repeated examinations of the neck and of the spinal column, together with an enquiry as to the pre-existence of a growth in the breast or other part, will do much to throw light on the matter.

In the early stages of the disease mistakes are also liable to be made. Thus biliary and renal colic and various neuralgias have been diagnosed in cases of vertebral tumour. In gouty subjects pains from neuritis of the spinal roots may be severe, and may be aggravated by movement, but they are not accompanied nor followed by spinal deformity or by signs of compression of the cord.

Symptoms due to caries of the spine may present a close similarity to those occurring in tumour. But as a rule the root symptoms are much less severe, and there is a greater tendency to sharp curvatures of the spine. Definite skiagraphic appearances may also be detected. Moreover there may be signs of tubercle in the lungs or in other organs.

Treatment. When there is unmistakable evidence of a vertebral tumour, which causes extreme pain, surgical interference should be considered in relation to the removal of pressure on the nerves. In cases of primary sarcoma Horsley has suggested the injection of Coley's fluid in order to induce necrosis, and thus relieve pressure.

In many cases, however, pain may be relieved by attention to the posture of the patient and by the administration of morphia and other anodynes; precautions should also be taken for the prevention of bed-sores.

TUMOURS OF THE SPINAL MEMBRANES.

Meningeal tumours, although rare, are more common than intra-medullary tumours. According to their

situation in relation to the dura mater they are divided into two groups:—(1) *Extra-dural tumours*, which originate in the dura mater, in the tissues between this membrane and the bone, or in the periosteum of the vertebræ. Sarcomata and hydatid cysts occur most frequently. Lipomata are also met with: fibromata, myxomata, endotheliomata and mixed forms of tumour more rarely. A sarcomatous growth may penetrate the dura mater and invade the cord. Extra-dural cancer is rare, and is always secondary; it seldom breaks through the dura mater.

(2) *Intra-dural tumours* occur more frequently than the extra-dural. They spring from the inner layers of the dura mater, from the arachnoid, the spinal roots or the pia mater. Sarcoma is a common variety: it may be primary or secondary, diffuse or localised. The diffuse form has a tendency to surround considerable portions of the cord; it may spread over its whole length, including the cauda equina, and even penetrate into the cranium. It may injure the cord by direct pressure or by interfering with its blood supply.

Other varieties of tumour growth that are occasionally met with are:—tubercle, gumma, myxoma, fibroma, psammoma, endothelioma, fibro-sarcoma, and myxo-sarcoma. Multiple growths—neuromata or sarcomata—may be found on the roots and in the membranes of the brain and spinal cord.

Symptoms. In the majority of cases symptoms indicating implication of the spinal roots precede those due to compression of the cord. Pain is an early and usually a prominent symptom. At first it may be slight in degree and unilateral in distribution, but it tends to increase in severity and to affect both sides of the body. It is important to remember that the pain produced by an intra-vertebral neoplasm is rarely so intense and agonising as in the case of a vertebral growth, and that occasionally the development of paraplegia is almost or even quite painless. The position of the pain depends upon the site of the lesion.

Thus when the growth is in the dorsal region dull aching pains may be referred to the breast or the shoulder; when the roots of the cervical or the lumbar nerves are involved shooting pains may be experienced in the arms or the legs. Sometimes pain can be referred to the distribution of a particular root or roots, and there may be an associated hyperæsthesia of the skin.

Irritation of the anterior roots is occasionally expressed by rigidity of the spinal muscles, or by spasms in the limbs. After a time signs of irritation of the spinal roots are followed by signs of impaired function, namely, anæsthesia, paralysis and muscular atrophy in the parts supplied by the affected nerves.

Sooner or later the *functions of the cord* become impaired, usually as a result of its compression, but occasionally owing to the development of a myelitis. There is paraplegia, which is combined with weakness of the trunk or of the arm-muscles according to the height of the lesion. As a rule the paralysis is gradual in onset and slow in development, but sometimes it develops with great rapidity: the former indicates a slow compression of the cord, the latter inflammatory changes at the level of the lesion.

The condition of the paralysed muscles and of the reflexes, varies with the position of the tumour. In tumours of the lumbo-sacral region the paralysed muscles undergo a progressive atrophy and the knee-jerks are abolished; whereas in tumours situated at a higher level, paraplegia is of the spastic type, the knee-jerks are aggravated, ankle-clonus is present, and the plantar reflex is extensor in type. In tumours of the cervical region, spastic paraplegia is often associated with an atrophic paralysis of certain muscles in the arms, according to the roots involved.

Loss of sensation from implication of the sensory tracts in the cord is usually late in onset. It varies in degree and in extent: as a rule all forms of sensation are impaired or lost below the level of the lesion, a distinction from tumours within the spinal cord, in

which dissociated anæsthesia is more common. The position of a meningeal tumour may be definitely localised by marking the extreme upper limit of the *slightest* change in the cutaneous sensibility.

During the earlier periods of the disease one-half of the cord is frequently more compressed than the other, when there will be a want of correspondence in the distribution of the motor and sensory symptoms, paralysis being most marked on the side of the tumour, and anæsthesia on the other side. But as the damage to the cord increases, loss of power and of sensation gradually become more equal in degree on the two sides.

Loss of control over the sphincters is often an early and a prominent symptom when the tumour presses upon the sacral segments of the cord; retention of urine with intermittent or overflow incontinence when the lesion is at a higher level. In the early stages of meningeal tumour vaso-motor disturbances sometimes occur; in the later stages bed-sores are common and they may be severe and intractable.

The presence of multiple growth is suggested by the successive development of symptoms which, though similar in character, indicate by their distribution that various regions of the cord are involved.

Diagnosis. The presence of a tumour involving the spinal membranes is strongly suggested by the order in which the symptoms occur, by their unilateral commencement, their combination and their progressive character. It is to be specially noted that root symptoms may be present for a long time before the onset of paraplegia, which begins insidiously, one leg being affected before the other.

In the early period of the disease when root symptoms are alone present the diagnosis has to be made from other affections involving the roots, for example, aortic aneurysm, vertebral cancer and neuritis. At a later period when signs of compression or of inflammation of the cord have developed, pachymeningitis and caries of the spine are the chief diseases to be considered.

The symptoms of radicular neuritis in the cervical region may closely simulate those of a meningeal tumour, and it may be necessary to reserve opinion for a time; the presence of a tumour would be unlikely if root symptoms had existed for several months without any signs of cord disease appearing. Cancer of the vertebræ would be excluded by the absence of any local signs of growth or of spinal deformity: further, the pains in this disease are more markedly intensified by movement than they are in meningeal growths.

In hypertrophic cervical pachymeningitis severe pains may be followed by signs of compression myelitis, but as a rule there is a tendency for the symptoms to be limited to those of root or of meningeal origin.

In caries of the spine the root-symptoms are less prominent than in extra-medullary tumours and may be absent altogether. When there is no obvious spinal deformity, the existence of sight irregularities of the vertebral spines should be carefully looked for: the presence of deep-seated tenderness and of rigidity of the back on movement are also significant of early caries.

With regard to the situation of the tumour it must be remembered that this is always at a higher level than the paralytic symptoms caused by it; for example, the highest level of anæsthesia produced by a growth in the dorsal region is often two or three inches below the position of the growth. But if account be taken of the most minute changes in sensibility, the area innervated by the compressed spinal segment can be more accurately determined.

In considering the probable nature of the growth, it is important to obtain a complete history of the case, to study the rate at which the symptoms have developed and to look for signs of tumour growth in other parts of the body.

Prognosis. The prognosis in a case of meningeal tumour depends chiefly on the possibility of its successful removal by a surgical operation. Benign growths which are definitely localised may be extirpated and

the patient may make a complete recovery. Unfortunately the common variety of growth is malignant in character; in such a case the outlook is most unfavourable and the issue is almost invariably fatal.

Treatment. When symptoms of an extra-medullary tumour occur in a patient who present unmistakable evidence of syphilis, it is always desirable to give anti-syphilitic remedies a fair trial. But if after a month's treatment no improvement is observed the question of laminectomy requires consideration.

In all non-syphilitic cases this operation ought to be performed as soon as possible, for it is very difficult in any other way, than by exposure of the tumour, to be absolutely certain as to its nature, its size or the possibility of its removal. If the growth is found to be benign and localised, its complete removal may cure the patient. If, however, owing to its malignant nature, the surgeon decides against an attempt to extirpate it, he may still consider the advisability of removing a portion of the growth, in order to relieve pressure symptoms; or of dividing the posterior roots for the relief of agonising pain.

In cases which are inoperable the treatment consists mainly in the alleviation of pain by morphia and other remedies, and in the prevention of complications, such as pneumonia and cystitis and other sources of sepsis.

SECTION VIII.

Disorders of Equilibrium.

In discussing the physiology of muscular co-ordination it was pointed out that the cerebellum is the great centre for the co-ordination of muscular movements, and that lesions of it or of its afferent tracts or of its connections with the cerebrum are apt to cause ataxia, with disturbance of the equilibrium of the body.

The chief conditions in which co-ordination of movement occurs are:—

1. Diseases of the cerebellum. *Acute lesions*:—Encephalitis, softening from vascular occlusion, hæmorrhage. *Chronic lesions*: Primary parenchymatous degeneration, olivo-ponto-cerebellar atrophy, progressive cerebellar disease due to vascular or interstitial lesions; tumours, abscess. [Classification based on that of Gordon Holmes.]

2. Diseases implicating certain parts of the cerebrum, as tumours of the prefrontal region or of the corpora quadrigemina; presumably because they interfere with impulses passing between the cerebrum and cerebellum. In this group may be mentioned the acute ataxia of Leyden, produced by disseminated inflammatory patches in the medulla, pons and crura (see p. 278).

3. Diseases which interrupt the connection of the cerebellum with the periphery of the body. (1) The most important are those which involve the posterior columns or the cerebellar tracts of the spinal cord, viz.: Tabes dorsalis; Friedreich's disease; ataxic paraplegia; subacute combined degeneration of the spinal cord; some forms of spinal syphilis and of disseminated sclerosis; (2) Menière's disease. (3) Tumours or other lesions involving the posterior spinal roots. (4) Rare varieties of peripheral neuritis.

Many of the above conditions are considered in other sections of this book. In the present section the effects of lesions involving the afferent tracts of the cerebellum will be mainly considered. It is convenient to begin with tabes which, being one of the commonest chronic diseases of the nervous system, affords frequent opportunities for the study of inco-ordination of movement.

CHAPTER I.

TABES DORSALIS. LOCOMOTOR ATAXY.

Tabes is a chronic progressive disease of the sensory neurons, the fibres of the posterior roots being specially affected; the disease is manifested clinically by pains, paræsthesia, anæsthesia, diminution of muscular tone, loss of the deep reflexes, inco-ordination, together with visceral and trophic disturbances. It has a close association with the general paralysis of the insane.

Etiology. The most important factor in the production of tabes is syphilis. In about eighty per cent. of the cases the history of a hard sore or of secondary symptoms can be obtained, and in some of the remaining cases it may be difficult, owing to a history of gonorrhœa or for other reasons, to exclude the possibility of syphilitic infection. Such infection, however, in about ten per cent. of the cases cannot be proved nor even reasonably suspected; we have then to consider the influence of other antecedents, such as injury, overstrain, exposure to cold and wet, alcoholic and sexual excesses. One or other of these antecedents may also act as an exciting cause of tabes in persons who have suffered from syphilis.

That other causes than syphilis must play a part in determining the incidence of the syphilitic virus on the brain or the spinal cord or on both is suggested by the small percentage (one to five) of persons who have contracted syphilis who subsequently suffer from tabes or from general paralysis. A few

authorities, for example, Ferrier and Mott, do not believe that these diseases can develop apart from previous syphilitic infection, and go so far as to say 'no syphilis, no tabes.' In support of this opinion it has been frequently stated that a cytological examination of the cerebro-spinal fluid in tabes and general paralysis gives the same marked lymphocytosis in cases in which there is no evidence of previous venereal infection as in cases in which such evidence is irrefutable. The correctness of this statement is rendered doubtful by the results of an investigation undertaken by the author and A. Ramsbottom. A careful cytological examination was made of thirty-four cases of tabes and general paralysis: in twenty-nine of these cases (about 85 per cent.) there was a decided lymphocytosis, while in the remaining five (15 per cent.) there was no lymphocytosis, nor could any evidence of antecedent syphilis be obtained. It is therefore justifiable to conclude that lymphocytosis is not a constant phenomenon in tabes and in general paralysis and that syphilis, although the most important factor in the production of these diseases is not an absolutely essential one.

Stress, produced by over-fatigue or over-exertion, occasionally appears to be an exciting cause of tabes, and sometimes a connection can be traced between the site of the strain and that of the first symptoms. Thus ataxia has begun in the arms when these limbs have been subject to over-strain, or an early development of optic atrophy has seemed to depend on prolonged overuse of the eyes. Furthermore, it is to be noted that degeneration of the posterior columns has been experimentally induced in animals by excessive traction on the limbs.

Many articles have been written on the subject of *traumatic tabes*, and it seems likely that occasionally a concussion of the spine may initiate the degenerative changes in the posterior columns which characterise the disease. It is rare, however, for injury to be the sole

factor, and frequently it will be found that the tabetic patient was not quite free from symptoms before the accident. Of injury, of sexual excess and of other antecedents, except syphilis, it may be said that generally they only tend to light up disease which was pre-existent or for which the ground was already prepared. In considering the influence of excessive venery it must be remembered that sexual excitement is often a symptom of the disease.

Tabes is a disease of middle life, most cases beginning between the ages of thirty and fifty; it occurs more commonly in men than in women. A hereditary neuropathic taint can be traced in only a few cases; direct inheritance of the disease can scarcely be said to occur, for when the child of a tabetic parent develops tabes it is owing to the transmission of syphilis and not to the inheritance of tabes. Juvenile tabes is almost invariably the result of inherited syphilis; exceptionally the syphilis has been acquired, as by infection from a nurse.

In a few cases both husband and wife have suffered from the disease in consequence of syphilis, one having infected the other, or both having acquired syphilis independently; this has been called conjugal tabes.

Symptoms. There is scarcely any other disease of the nervous system which presents such variations in its clinical manifestations as tabes. This applies not only to the character of its symptoms, but to their arrangement, order of development and course. In many cases three stages may be distinguished; namely, a pre-ataxic, an ataxic and a so-called paralytic stage, which is really a stage of profound ataxia. These divisions are quite arbitrary; there is no definite line of demarcation between them, the symptoms of one stage pass insensibly into those of the other. It is therefore desirable to begin with a description of the individual symptoms of the disease and afterwards to consider variations in their grouping.

Sensory symptoms. The earliest and most constant symptoms of tabes are *pains* of a peculiar and distressing

character; they may precede other symptoms by months or years, and may last throughout the whole course of the disease. Two varieties may be distinguished—the momentary and the prolonged. *Momentary pains* are the most characteristic. They are sudden, darting in nature and of brief duration: from their resemblance to electric shocks they are often called 'lightning pains.' At first not very urgent, they tend to increase in severity until there may be paroxysms of almost continuous pain. Such paroxysms, which are apt to come on at night, may recur every few weeks, or in the severest cases almost every few minutes. These lancinating pains are referred to the muscles and bones rather than to the joints; sometimes they are referred to the skin, which for a time may be left tender to the slightest touch; in some cases even the contact of the bed-clothes is unendurable. Although usually affecting various parts of the lower extremities, they may be experienced in the arms, especially along the inner side of the forearm; in the trunk, or even in the head and face. As a rule their localisation corresponds to a territory supplied by a spinal segment; but sometimes the pains are limited to the area supplied by a particular nerve, as the sciatic.

The *prolonged pains* are often severe and are described as 'boring,' 'burning' or 'dragging': they may continue in the same place for hours, days or even weeks. Dull, aching pains similar to those of rheumatism are sometimes present: they tend to become worse in cold damp weather. The patient may also suffer from aching pains along the spine, or from a feeling of constriction round the body. The latter, known as the 'girdle sensation,' is an almost constant feature, and is often present at an early period of the disease; it is usually situated on a level with the upper abdominal region. A similar tight sensation may be felt around the foot or the leg. Rarely a paroxysm of severe pain may be followed by erythema or a patch of herpes.

It is to be noted that lightning pains, although usually frequent and severe, may be quite insignificant or even absent, and that they bear no proportion to the other symptoms of the disease: sometimes at a late period they abate or cease altogether owing to complete destruction of the posterior roots.

In addition to pains the patient may complain of various *paræsthesiæ*, such as numbness and tingling, or formication; the soles often feel as if covered with wool or other soft substance; rarely the face feels numb and stiff; numbness and tingling may be felt in the region supplied by the ulnar nerve, or along the whole of the inner side of the hand and arm, a region which is supplied by the eighth cervical and first dorsal roots. In many cases, as pointed out by Biernacki, no pain or tingling is felt when strong pressure is applied to the ulnar nerve near the inner condyle of the humerus. Sometimes sensations of intense cold or heat are felt in the extremities.

Hyperæsthesia, especially hyperalgesia, of the skin at the level of the girdle sensation is common; sometimes it occurs in patches on the lower limbs. A patch of hyperæsthesia may be found either within an anæsthetic area or in its immediate neighbourhood.

In most cases of tabes some form of *anæsthesia* is present. In the early stages it is met with on the trunk where it occurs in the form of bands or zones which correspond to the distribution of spinal roots, especially to those of the fourth and fifth dorsal segments; sometimes the anæsthesia occupies a wider area, extending from the chest along the inner aspects of the arms and hands. This trunk anæsthesia is rarely complete and may consist of a loss of sensation to light tactile impressions only. The patient is often quite unaware of its existence; it is therefore of great importance to examine for these bands, especially as they are very common and are very helpful in diagnosis; the distribution is usually bilateral, but it may be unilateral.

Sooner or later the cutaneous sensibility of the legs and feet is diminished, sensation to pain being usually affected before that to touch. A pin-prick is felt as a light touch, and the patient is unable to distinguish between the point and the head of a pin. The sense of temperature, in respect to extremes of heat and cold, is usually impaired in the same degree, and over the same areas as cutaneous pain; it is rarely affected alone. In some cases the loss is restricted to heat or to cold; in others, loss of sensations to heat is associated with hyperæsthesia to cold. Defects in sensibility to moderate degrees of temperature are

Fig. 119.—Distribution of sensory loss in an early case of tabes. The shading on the thorax and down the arms represents loss of sensation to cotton wool and very slight diminution to pin-prick; the temperature sense was perverted, hot objects being called cold. In the legs cotton wool was felt; sensation to pain and temperature was greatly impaired.

generally found in association with defects to light touches. As a rule these defects are not present until a late period of the disease, in this respect presenting a marked contrast to the loss of sensibility to pain and to more extreme degrees of temperature, which are often present at an early period.

Delay in the conduction of pain is frequently observed, a pin-prick being felt only after an interval of a few seconds; the patient may feel the contact of the pin at once, but not the pain of the prick for some time, and the maximum intensity of the pain may be perceived

still later. Less commonly the perception of tactile and temperature sensations is delayed. The power of localising a sensation may be perverted, for example, a prick on the toe may be referred to the foot, on one leg to the other leg—allocheiria. In some cases a single prick is perceived as several pricks—polyæsthesia.

Anæsthesia, although a very common symptom, is not always present; the cutaneous sensibility may be normal even when pains and ataxia are prominent. As a rule anæsthesia is most marked in the feet and the lower part of the legs, one leg being more affected than the other. It may extend to the thighs, the perineum, the genitals, the trunk and even to the face and head: the skin of the face may feel to the patient as if stretched or compressed by a mask. In a few cases anæsthesia affects the whole of the cutaneous surface below the level of the second rib.

The vibrating sensation is often lost in the bones of the legs and feet at an early period of the disease, and sometimes when the cutaneous sensibility is normal.

Impairment of sensation in muscles, tendons and joints, which also occurs, is shown not only by the absence of pain on squeezing these parts, but by the patient's inability to tell the exact position of his limbs when his eyes are closed, or to recognise that passive movement of a limb is being made. The loss of this sense of position is chiefly observed in the lower limbs. It must not be confused with ataxia; it is a common experience to be unable to discover in a patient, who presents decided ataxia in standing and walking, any lack of knowledge regarding the position of any part of his leg when passively moved as he lies in bed or sits in a chair with his eyes closed. The pressure sense, as tested by pressing with the finger on a part, may be preserved when stroking the part with cotton wool is not perceived; conversely tactile sensibility may be preserved when the sense of pressure is entirely lost.

When anæsthesia affects the upper limbs, its distribution is similar to that in the lower limbs, the distal

portions being mainly affected; it may be limited to the palms, just as sometimes it is limited to the soles.

Ataxia. Inco-ordination of movement usually develops very gradually, and nearly always begins in the lower extremities. At first it is only noticed when the guiding influence of vision is removed. Thus the patient walks unsteadily in the dark, or he sways when standing with his eyes shut. Inability to stand steadily with the feet close together and the eyes shut is known as Romberg's symptom. A severer test is for the patient to stand on one leg, first with his eyes open and then closed. In the early stages of the disease he may walk without much difficulty, although it may be noticed that he staggers a little when he turns abruptly round, or when he gets up suddenly from a chair, or when he assumes the erect posture after stooping, his eyes being closed at the time. The difficulties mentioned are always greater when the patient's feet are bare; then also the slightest degree of inco-ordination in standing is evinced by the irregular tightening of the tendons on the dorsum of the foot.

Associated with the unsteadiness in standing and walking, errors may be noticed in attempts to touch prescribed spots, or in the performance of other movements. For example, the patient when seated may be unable to touch accurately the knee of one leg with the heel of the other; or he may exhibit inco-ordination in crossing his legs, or in trying with his foot to describe a circle in the air. Such movements, however, just as the sense of position, are often quite normal, even though there is obvious swaying of the body when the patient stands with his feet close together.

As the disease advances, standing and walking without support becomes increasingly difficult and finally impossible. At this stage a patient standing with the aid of two sticks presents a characteristic attitude. The feet are wide apart and the legs are drawn backwards so as to form an obtuse angle with the feet; the thighs are extended on the legs, the

buttocks project backwards, while the body is inclined forwards in order to keep the line of gravity in front of the arches of the feet.

The *gait* now becomes typically ataxic; it is characterised by a quick and high elevation of the advancing foot, which is suddenly and forcibly projected forwards and outwards and then brought down again with a stamp. In advanced cases walking becomes impossible, and even when the patient is well supported, the legs are thrown hither and thither in the greatest disorder, their movements being comparable to those of a marionette.

The movements of the trunk may also be affected, and sometimes to such an extent that the patient is unable to sit steadily in a chair and will at once fall off if he closes his eyes. In time, too, the movements of the arms become disorderly: at first there is inability to execute delicate movements, such as writing, or the patient is unable, when his eyes are shut, to touch a prescribed spot, such as the tip of his nose. At a later period of the disease the fingers fumble in buttoning and unbuttoning the clothes; an object is grasped in an uncertain and spasmodic manner, while at a still more advanced stage the patient may be quite unable to dress himself, or to convey food to his mouth. In cervical tabes the arms are early and chiefly affected.

Hypotonia and Atonia. A diminution or a loss of tone in the muscles can be frequently demonstrated in cases of tabes: the change is often present in the pre-ataxic stage, and is therefore of much value in diagnosis. It is shown by flaccidity of the muscles and, when the impairment of tone is considerable, by an abnormal degree of movement at the various joints. Thus a patient lying on a couch may be able to raise the leg with the knee extended to an angle of 90°, 100° or more above the horizontal level, instead of to only an angle of 50° or 60° as in health. He may also be able to flex the knee and the hip to such an extent that the heel is brought into contact with the buttock. Hypo-

tonus of the trunk muscles may be indicated by undue flexibility of the spine, so that in some cases the patient can bend down far enough for his face to touch the couch.

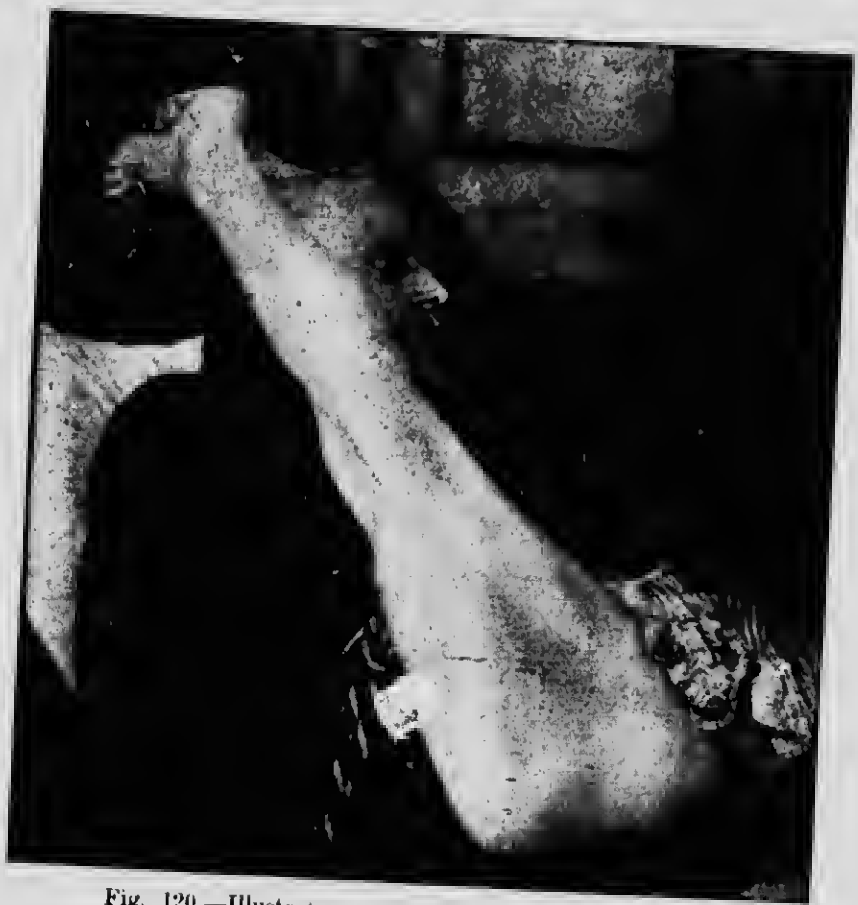


Fig. 120.—Illustrates an extreme degree of hypotonia.

Paralysis. A common early symptom is transient weakness of an external ocular muscle, the patient complaining of double vision, of squint or of a drooping upper lid. The weakness may last a few weeks or it may become permanent. In the latter case one or more muscles are involved: there may be ptosis, unilateral or bilateral, or paralysis of all the muscles supplied by the third nerve; sometimes there is paralysis of the external rectus only; occasionally partial or complete

paralysis of all the muscles of both eyes gradually develops, leading to the condition known as total ophthalmoplegia.

It is usually stated that the strength of the limb muscles is well maintained. In many cases of tabes, however, a careful examination will reveal definite weakness of the flexors of the hip or of the dorsi-flexors of the ankle; such weakness may persist and become marked, sometimes it is observed at an early period of the disease. Very rarely unilateral paralysis of the muscles supplied by the musculo-spiral nerve, or by the peroneal is met with. Transient weakness of the

Fig. 121.—Showing upward movement of the eyeballs during convergence; all other movements of the eyes greatly impaired; from a case of early tabes.

lower limbs comparable to that of the eye muscles also occurs; sometimes this is momentary, for example, a patient when walking may fall to the ground without any warning, owing to the sudden giving way of his legs. In rare cases, and generally at a late period of the disease, muscular atrophy is associated with tabes: the peroneal muscles are most frequently affected.

The Reflexes. The condition of the *superficial reflexes* varies to some extent with that of the cutaneous sensibility; they are usually present, and are often increased during the early stages of the disease.

Exaggeration of the abdominal reflexes, especially of the epigastric reflex, is a noticeable feature in many cases of early tabes; frequently it is associated with hyperæsthetic zones of skin. It is interesting to contrast this exaggeration of the abdominal reflexes with their early loss in disseminated sclerosis.

Loss of the knee-jerk is one of the earliest and most constant symptoms of tabes; it may precede by many years the development of ataxia. Very rarely the knee-jerk may be obtained for a time, although typical signs of tabes are present; or it may be present on one side and absent on the other. In such exceptional cases an appeal should be made to the tendo-Achilles reflex, the loss of which usually precedes that of the knee-jerk. The wrist and elbow-jerks are preserved, unless the cervical segments of the cord are implicated.

The muscular irritability, as tested by tapping, is always present; that of the quadriceps femoris may be increased although the knee-jerk is lost.

The Bladder and Rectum. Some disturbance of micturition is common. At first the patient may complain that he has to wait a little, and to strain before he can pass water, or that the act takes longer than formerly; a variety of such early disturbance is known as 'stammering micturition,' by which is meant an urgent call to pass water, with a temporary inability to do so, followed by precipitate and uncontrollable micturition. Frequently the bladder is not completely emptied; retention, however slight, is a source of grave danger, for residual urine is apt to decompose and set up cystitis and pyelonephritis. Retention is often associated with an intermittent overflow of urine: it is rarely absolute except in an advanced stage of the disease. Paralytic incontinence of urine is rare, though there is often a loss of reflex tonus in the sphincter vesicæ, so that coughing or laughter may cause the escape of a little urine.

Constipation is common, and probably depends on

weakness of the muscular wall of the colon. Occasionally the sphincter ani becomes weak, when there is a difficulty in retaining a loose stool.

Ocular Symptoms. Of equal importance to the lightning pains and the loss of the knee-jerk in the diagnosis of commencing tabes is the Argyll-Robertson phenomenon which is found in about seventy per cent. of the cases. It is characterised by loss of the reaction of the pupil to light, with preservation of its contraction during accommodation. In a few cases the reaction to accommodation is also lost, and very rarely this loss is observed when the reaction to light is preserved. Loss of the light-reaction may be unilateral or bilateral, complete or incomplete, and it is to be specially noted that a normal condition of the pupils is sometimes present in tabes, and therefore does not exclude this disease. In some cases a feeble contraction of the pupil to light is followed by a rapid dilatation, or the latter only occurs. The pupils are often very small, occasionally they are dilated; they may be equal in size, but as a rule some inequality can be detected. They are rarely circular, some irregularity in outline being usually present.

One of the earliest symptoms for which a patient may seek advice is impairment of sight, which is found to depend on optic atrophy. The loss of sight is associated with a peripheral contraction of the visual fields for the perception of colours and the recognition of objects. Central scotomata are very rare. The optic atrophy is primary and progressive; it generally leads to total blindness in three or four years. In some cases it may cease to progress, and may even partially improve. The occurrence of optic atrophy appears to be followed by delayed onset of ataxia, or to arrest of its progress if already present. Occasionally it happens in tabes that sight fails rather suddenly without any ophthalmoscopic changes.

Affections of other cranial nerves. Sometimes the sense of smell is lost as a result of degenerative changes

in the olfactory nerve; loss of taste may also occur. Transient or persistent deafness is met with, in consequence either of disease of the labyrinth, or of atrophy of the auditory nerve; attacks of vertigo may be associated with the deafness.

The functions of the fifth nerve are sometimes impaired. As already mentioned, pains and anæsthesia are met with in the region supplied by this nerve; very rarely other signs of its implication are observed, namely, paralysis and atrophy of the muscles of mastication, neuro-paralytic ophthalmia, ulceration of the buccal mucous membrane and herpes zoster on the face.

In some cases the vocal cords become paralysed: the abductor muscles the posterior crico-arytænoids are most frequently affected. Unilateral atrophy of the tongue sometimes occurs; this is frequently associated with paralysis of the vocal cord and the palate on the same side.

Visceral crises. This term denotes various forms of paroxysmal disturbance of function in which pain is generally a marked feature. The *gastric crisis* is the most frequent. This consists of attacks of severe epigastric pain, accompanied or followed by distressing vomiting; the vomited material at first contains food, afterwards it consists of a watery mucus, which ultimately becomes mixed with bile and occasionally with blood. Associated with the pain there is often a band of anæsthesia in the mid-dorsal region, together with hyperæsthesia of the skin over the epigastrium. During the attack the patient may suffer from palpitation, from vertigo, or from agonising pains in the limbs. In some cases the attack is represented by pain or by vomiting only, and very rarely by nausea without vomiting. Its duration varies from a few minutes to several days. Attacks often recur every few weeks, but however severe and exhausting are the symptoms, it is remarkable how quickly the patient recovers when the attack is over. Gastric crises occur during the early

stages of the disease; they may be its first noticeable manifestations.

Intestinal crises sometimes occur, consisting of attacks of paroxysmal diarrhœa. Occasionally there are *rectal crises*, in which the patient, owing to the sensation of a foreign body in the rectum, has an urgent desire to go to stool, and he may suffer from paroxysms of pain accompanied by severe tenesmus.

Renal crises, resembling attacks of renal colic, and *bladder* and *urethral* crises are also present in some cases. In the latter varieties there is an urgent desire to pass water, and severe pain may be referred to the neck of the bladder or to the urethra; sometimes the attack is ended by the passing of a little blood.

Other crises are *cardiac*, resembling attacks of angina pectoris; *nasal* or *bronchial*, consisting of attacks of sneezing or coughing; and *laryngeal*. A laryngeal crisis is characterised by noisy respiration, by cough and by dyspnœa; in some cases the symptoms resemble those of laryngismus stridulus. When the attack is severe there may be much pain, and stridor together with a feeling of suffocation: the lips become blue owing to the obstruction to inspiration, which is also shown by the indrawing of the chest wall.

Trophic changes. Of these the most striking and important are perforating ulcers and arthropathies. Other trophic disturbances that are occasionally observed are: —Local sweating, as of the palms or soles, or of one side of the face; an altered growth or a falling off of the hair or nails; a painless loss of teeth owing to changes in the alveolar processes; ecchymoses in the skin, thickening of the epidermis, patches of herpes, œdema, and spontaneous rupture of tendons.

The general nutrition of the patient also is much impaired; in many cases emaciation becomes extreme and even when an adequate amount of nourishment is being taken.

Perforating ulcers are found on the soles of the feet, usually at the base of the great or the little toe, and

PLATE VI.

Skiagram of a case of tabes, showing old fracture of bones of forearm, thinning of these bones, and disease of the carpus.

occasionally on the heel. The ulcer generally begins as a corn; it has sharp irregular edges, a circular outline and exudes a serous or a sero-purulent discharge; it may penetrate deeply and may set up disease in the subjacent bone or joint.

Arthropathies. The peculiar changes in the joints which occur in some cases of tabes were carefully described by Charcot—hence the name Charcot's joint disease. The condition is characterised by a painless and rapid swelling of the joint; the adjacent parts and even the whole limb may participate in the swelling, which has often the character of a hard and tense oedema. The skin is pale and there are no signs of inflammation. In mild cases the effusion gradually subsides and the joint resumes its normal size and shape; relapses, however, are apt to occur, or other joints may become affected. In severe cases the enlargement is followed by erosion of the cartilages, wasting of the ends of the bones and relaxation of the ligaments which may become calcified; sometimes there is much periarticular new bony formation, but usually osteophytic deposits are less conspicuous than in osteo-arthritis. Relaxation of the ligaments of the joint with or without atrophy of the ends of the bones leads to increased freedom of movement and sometimes to partial or complete dislocation.

As a rule the affection is limited to one joint; the larger joints are the most commonly attacked, and in the following order of frequency: the knee, hip, shoulder, ankle and elbow. Arthropathies may develop at any period of the disease; they are not uncommon in the pre-ataxic stage.

In some cases of tabes the bones of the limbs, especially of the lower limbs, become rarefied and brittle; they are therefore liable to fractures which may occur without obvious cause, or in consequence of a trivial injury. In the process of union, which is often long delayed, an abnormal amount of callus is thrown out.

The '*tabetic foot*' is the result partly of relaxation, and of changes in the ligaments, and partly of disease of the bones and their articulations. At first the dorsum of the foot becomes prominent, subsequently the arch of the foot sinks, the sole becomes flattened, and its inner border swollen; the toes are extremely flexed and the whole foot is shortened.

Complications. There is a close alliance between tabes and general paralysis of the insane. The two diseases may be combined in the same patient, the symptoms of the one disease or of the other preponderating; thus many tabetics present mental symptoms—often slight in degree—which characterise general paralysis, whilst many cases beginning as general paralysis develop symptoms of tabes.

In rare cases tabes is complicated by true syphilitic disease of the brain or the cord—gummata, meningitis and the various forms of cerebro-spinal syphilis; or by myelitis; or by the development of general muscular atrophy.

Occasionally in tabes there are signs of aortic valvular disease, or of aneurysm; both conditions being probably the indirect results of syphilitic infection. Tachycardia and glycosuria are other rare complications.

The course of tabes is extremely variable. A patient may remain in the pre-ataxic stage for many years, indeed he may never suffer from inco-ordination of movement; this is apt to be the case when optic atrophy develops at an early period. In other cases there is no pre-ataxic stage, inco-ordination and loss of the knee-jerk being the earliest symptoms.

The sequence of symptoms also presents every variation, and almost any one of the symptoms may be the first to appear. In the pre-ataxic stage, in addition to shooting pains, loss of the knee-jerk and the Argyll-Robertson pupil, we may meet with anæsthesia, muscular hypotonus, paralysis of ocular muscles, optic atrophy, crises and joint affections, the order of their development varying in almost every case.

PLATE VII.

Skiagram, showing fracture of patella, with wide separation of fragments: the same case as Plate VI.

Although frequently steadily progressive, the course of tabes may be characterised by long periods in which the symptoms are stationary, or even show a tendency to improve. An arrest in their development may occur at any stage of the disease; sometimes it is permanent. In some cases periods of arrest alternate with periods in which the symptoms become gradually or rapidly worse. An intercurrent malady, such as influenza, or the over-indulgence in alcohol, is apt to increase the degree of ataxia or the severity of other symptoms.

Tabes is not directly fatal, but the suffering and the general impairment of health which attend the malady, by lowering vitality, tend indirectly to shorten life; the average duration of life is from twelve to fifteen years. Death is usually the result of some complication, kidney disease caused by sepsis from residual urine being probably the commonest.

Rare forms of the disease. In addition to the ordinary forms of tabes, including the form in which optic atrophy is one of the earliest symptoms, the following varieties are occasionally met with.

(1) *Cervical tabes.* In this variety, which is very rare, the arms are first affected; pains, anæsthesia and ataxia may be limited to them; their deep reflexes are lost. The knee-jerk is present for a time, but ultimately disappears. Gastric crises are apt to occur early. The cranial nerves are more liable to be involved than in the ordinary form of tabes.

(2) *The neuralgic type.* This variety is characterised by the presence of severe and long-continued lancinating pains; at first there is no ataxia and the knee-jerk is present. The diagnosis is based on the lightning-like character of the pains and on their association with, or the subsequent development of, the other early signs of the disease.

(3) *The juvenile type* has been already mentioned. It is exceedingly rare; it occurs more frequently in girls than in boys, a distinction from tabes in the adult. Optic atrophy is generally an early and a prominent

feature. In many cases some form of bladder disturbance is observed at an early period.

Morbid Anatomy and Pathology. In *advanced* cases of tabes the chief observable changes are atrophy of the posterior nerve roots, thickening of the pia-arachnoid over the dorsal surface of the cord and sclerosis of the posterior columns. To the naked eye the posterior columns have a grey translucent and shrunken appearance. Under the microscope they are seen to consist of degenerated nerve fibres and an increased amount of neuroglia connective tissue. As a rule these changes are most marked in the lumbo-sacral region where they occupy nearly the whole of Burdach's and Goll's columns. At a higher level the degeneration becomes gradually more limited to the inner portions of the posterior columns until in the cervical region it affects the columns of Goll only. In cervical tabes the degeneration begins in the cervical roots, and then is found in the postero-external columns, but not in the postero-internal. In late stages of tabes other fibres may be affected, namely, the fine nerve fibres in Clarke's column, the fibres passing from the posterior column into the posterior grey horns, and the reflex collateral fibres in the median grey matter and in the anterior horns; very rarely some of the fibres in the lateral columns show degenerative changes.

To understand the distribution of the changes in the spinal cord in *early* cases of tabes it is necessary to be acquainted with the origin and arrangement of the fibres in the posterior columns. These columns are composed of exogenous fibres which originate in the ganglia on the posterior roots, and of endogenous fibres which arise from cells within the spinal cord, and are probably commissural in function between different portions of the grey matter. The exogenous fibres, which constitute the posterior roots, chiefly suffer in tabes; the endogenous are affected only in very advanced cases.

Each posterior root as it penetrates the cord divides

into two parts, an external and an internal part. The former consists of fine fibres which pass into the apex of the posterior horn or zone of Lissauer. The latter is composed of three distinct sets of fibres:—(1) Short fibres which form the inner part of the posterior root and end in the cells of the adjoining posterior horn. (2) Fibres of medium length, which after passing up some distance in the middle portion of the posterior column are distributed amongst the cells of Clarke's column and the cells of the anterior horn. (3) Long fibres which do not end in the cord but ascend in the posterior column to the medulla. As the posterior root

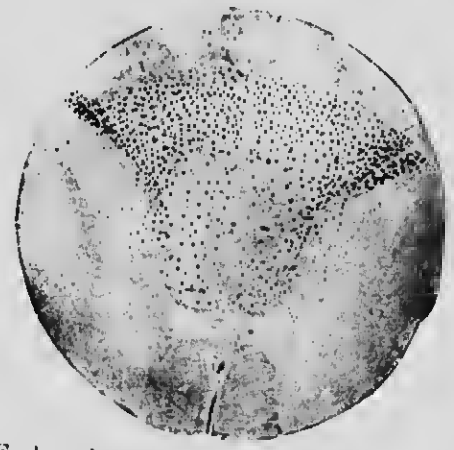


Fig. 122.—Early tabes dorsalis. Marchi's stain. Degenerated fibres = black dots. Note degenerated fibres most numerous close to posterior horn of grey matter. (Williamson.)

fibres of every spinal nerve enter the cord between the posterior horn and the nerve fibres which have entered lower down, the fibres of each root are pushed more and more towards the middle line. The consequence is that in the cervical region Goll's column is composed mainly of the long fibres from the lumbo-sacral and lower dorsal roots, whilst Burdach's column is composed of fibres from the upper dorsal and cervical roots. The former fibres terminate in the nucleus gracilis, the latter in the nucleus cuneatus.

The medium fibres are the first to degenerate, and if the lumbar region of the cord, where the disease usually

begins, is examined, symmetrical areas of degeneration will be found in Burdach's columns extending from the middle thirds of the posterior horns towards the middle

Fig. 123.—Section of spinal cord: early tabes. Weigert's stain. Pale areas in posterior columns, degenerated parts. The highest figure, lower cervical region; middle figure, upper lumbar region; lowest figure, lower sacral region. (Williamson.)

line. In some cases of early tabes the fine fibres of Lissauer's zone are also implicated. As the disease progresses, the degeneration invades the long and the short fibres of the posterior roots and with still further

progress the whole of the posterior columns becomes involved with the exception of two portions, namely, the comma-commissural zone—the part of the posterior columns that is adjacent to the grey commissure—and the oval fields of Flechsig which lie close to the mesial septum. These areas are composed of endogenous fibres which undergo degeneration only at an advanced stage of the disease.

In early cases of tabes the posterior root fibres outside the cord may show little or no changes although those within the cord may be markedly degenerated. In very advanced cases the fibres outside the cord become degenerated and the changes may extend into the ganglia on the posterior roots. The cells of these ganglia are usually normal; sometimes they show slight changes and occasionally they are atrophied. It is probable that the degenerative changes in these cells are simply secondary to those in the posterior roots.

Changes are sometimes found in the peripheral nerves; as a rule they are limited to the small sensory fibres to the skin. In cases of tabes associated with muscular atrophy, degeneration of the motor nerve fibres, or of the cells in the anterior horn may be present; the changes in the cells are probably secondary to those in the fibres.

Of the cranial nerves, the optic are most frequently affected. The degeneration begins in the peripheral portions of the nerves and extends towards the brain. In some cases it is associated with changes in the ganglion cells of the retina; in others these cells are normal, although blindness from optic atrophy has existed for many years. At their entrance into the eyeball the optic fibres lose their medullary sheaths; probably at this point they are most likely to be affected by toxins circulating in the blood. On the other hand, Léri has adduced evidence in favour of the optic atrophy being secondary to an interstitial neuritis, which he believes to be the result of syphilitic disease of the vessels and meninges. Occasionally other cranial

nerves are involved. Degeneration has been found in the peripheral fibres or in the nuclei of the trigeminal, the vagus, the spinal accessory, the hypoglossal and the auditory nerves.

The *essential lesion* in tabes is a primary and progressive degeneration of the nerve fibres in the intramedullary portion of the posterior roots. Their myelin sheaths break up, become granular, and finally disappear; ultimately the fibres themselves are destroyed.

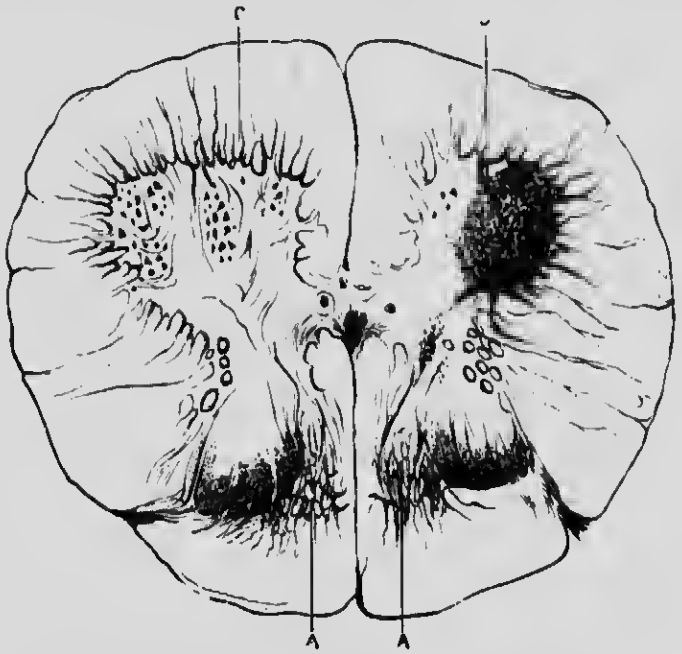


Fig. 124.—Sections of the lumbar region of the spinal cord, from a case of tabes, complicated with muscular atrophy. A, A, sclerosis of the posterior root-zone; C, left anterior horn, healthy; D, right anterior horn in state of atrophy. (Charcot and Pierret).

Proliferation of the neuroglia tissue, which occurs, is a secondary effect, and is not the result of chronic inflammation. The degeneration of the posterior root fibres begins at the surface of the cord close to the pia mater, at a point where the fibres lose their neurilemma sheaths, and where therefore they are the most likely to be attacked by toxins.

Some authorities believe that the initial changes are

the result of a syphilitic meningitis, others that they are due to a specific inflammation of the posterior system of spinal lymphatics. A more probable view is that syphilis acts indirectly by lowering the vitality of the nerve fibres; in this way it renders them more vulnerable to the action of toxins, the nature of which may vary in different cases. One variety of toxin may arise from a bacillus closely resembling that of diphtheria, which Ford Robertson has found in the alimentary canal and in other parts of the body. He believes that this toxin is the essential cause of the nerve degeneration which occurs in tabes and also in general paralysis.

Diagnosis. As a rule the diagnosis of tabes is easy; occasionally it is difficult, especially in the pre-ataxic stage. At this period the only symptom to attract attention may be diplopia, failure of vision, a painless joint swelling or slight disturbance of micturition. These symptoms should always excite the suspicion of tabes and should lead to an examination of the knee-jerk, and the pupils, and to enquiries regarding the occurrence of shooting pains. Furthermore, the trunk should be examined for areas of impaired sensation, and the lower limbs for muscular hypotonus, and for sensory disturbances, either cutaneous or osseous, as tested by the tuning fork. Visceral crises are sometimes very early symptoms, and may be easily mistaken for signs of primary visceral diseases; thus a renal crisis may suggest renal calculus, a gastric crisis disease of the stomach. In such cases an examination of the knee-jerk and the pupil will usually decide the question.

From the ordinary type of *multiple neuritis* tabes is easily distinguished, but the symptoms of the ataxic type may strongly suggest the presence of true tabes. In both cases the knee-jerk is lost and there is obvious ataxia. In pseudo-tabes, however, the pains are rarely sharp and stabbing, muscular tenderness is often prominent, the pupils act normally and optic atrophy does not occur.

In diphtheritic paralysis the presence of ataxia with loss of the knee-jerk may lead to a suspicion of tabes, especially in the absence of a definite history of sore throat. But as a rule there is evidence of a preceding paralysis of accommodation or of the soft palate. Moreover, lightning pains are absent and the course of the disease is more rapid than that of tabes.

Severe cases of *diabetes mellitus* are sometimes complicated by the presence of severe pains in the legs, by loss of the knee-jerk and by a perforating ulcer on the foot. The pains, however, are aching rather than stabbing in character: the pupils react normally, and optic atrophy does not occur. The symptoms referred to generally depend on peripheral neuritis, but in a few cases, as indicated by pathological examinations, they may be due to degeneration of the intra-medullary fibres of the posterior nerve roots.

There are cases of *cerebellar tumour* in which ataxia is associated with loss of the knee-jerks, but in this disease there are no sensory disturbances in the legs, whilst severe headache and optic neuritis are usually present.

Prognosis. Owing to the variable course of tabes it is difficult to formulate any general guides to prognosis. The symptoms and their tendency to progress must be studied in each case before an opinion can be formed as to the future course of the disease. Periods of arrest are common, and many cases under careful treatment improve to a considerable degree. Ataxia severe enough to prevent walking may disappear to such an extent that the gait becomes almost normal. The chief danger from ataxia is due to the risk of injury from falling.

It is rare for all the symptoms of the disease to disappear; the knee-jerk rarely returns and the attacks of pain are very liable to continue. Gastric crises, whatever their severity, rarely prove fatal, but they, as well as violent limb pains, lead to prostration and often to emaciation; hence the patient's resistance becomes lowered to attacks of other diseases.

Treatment. Although there is no known specific for tabes the course of the malady and its individual symptoms may be favourably influenced by appropriate treatment, the nature of which will necessarily vary in different cases.

The first question to be decided is whether anti-syphilitic remedies should be employed or not. The answer is that they are of distinct value in many cases, and are especially likely to be of service: (1) when the patient has not received adequate treatment for the original syphilis; (2) when symptoms of syphilitic lesions are still present, and (3) when tabetic symptoms have developed rapidly, or within five years, after infection by syphilis.

On the whole, experience indicates the desirability of giving anti-syphilitic treatment a trial in nearly every case of tabes, unless the disease is of long standing or is associated with much general debility. The best results are obtained by the method of mercurial inunction which is so thoroughly carried out at Aachen and other spas; in some cases inunction is used alone, in others it is supplemented by intra-muscular injections of one of the soluble salts of mercury. The outline of the treatment, the details of which will be found in works on syphilis, is as follows:— One rubbing of mercurial ointment should be given daily for six or eight weeks, and the course should be repeated every six months for a period of three years. This allows of an interval of about four months between each series of rubbings, during which it is generally advisable for the patient to take one of the iodides, together with strychnine and other tonics. An alternative plan is to give an intra-muscular injection of mercury once a week, and the rubbings on the remaining six days.

Considerable improvement often follows such a course of treatment; the patient gains weight, his colour improves and his pains and ataxia become less marked. In some cases even the knee-jerk returns and the reaction of the pupil to light is restored.

General treatment. Rest for both mind and body is of great importance in the treatment of tabes, and in all acute or severe cases the patient should be kept in bed for several weeks. Excesses of all kinds must be avoided; alcoholic and sexual excesses are particularly injurious. Exposure to cold and wet tends to increase the pains and the inco-ordination, so that residence in a sunny, dry and uniform climate is to be recommended. The tendency to emaciation and general weakness may be counteracted to some extent by living in the open air as much as possible, and by a careful attention to diet; cream and cod liver oil are often productive of great benefit. General massage is also of service: galvanism is strongly recommended by Oppenheim.

Symptomatic treatment. Attacks of severe pain are often difficult to relieve. Local applications, in the form of warm fomentations, lint soaked in chloroform, or repeated sinapisms are sometimes beneficial. A warm bath or counter irritation to the spine may also afford relief. Internally one or other of the following drugs, either singly or in combination, may alleviate the pain:—Antipyrin, phenacetin, antifebrin, exalgin, aspirin, pyramidon and aluminium chloride. A combination of aspirin and pyramidon, or one of bromide with phenacetin is often efficacious.

Superficial pains are temporarily relieved by the hypodermic injection of cocaine. Severe paroxysms of lightning pains may only yield to morphia; but neither this remedy nor cocaine should be used except as a last resort, owing to the danger of a habit being acquired. The above remedies are also of service during a gastric crisis; oxalate of cerium, and drop doses of tincture of iodine in water frequently repeated are also recommended. As a rule, however in severe attacks no drug other than morphia will give relief. In some cases the application of a blister, an ice-bag, a mustard plaster, or of the galvanic current to the epigastrium gives good results. Should gastric crises persist for several days it may be necessary to feed the patient per rectum.

In very severe and obstinate cases Forster's heroic treatment by division of the posterior dorsal roots from the seventh to the tenth seems worthy of consideration. A laryngeal crisis may be quickly relieved by the inhalation of nitrite of amyl, or of chloroform.

The condition of the bladder should be constantly investigated owing to the risk attending the presence of residual urine; whenever this is suspected and in all cases of retention, the catheter must be used. When there is evidence of cystitis it is necessary to wash out the bladder with a solution of boracic acid: urotropin also should be prescribed.

A Charcot's joint may require support by means of some mechanical apparatus. To obtain ankylosis of the joint, which is often desirable, arthodesis or excision should be performed. When the limb is quite useless the question of amputation has to be considered.

Ataxia. The best method of treating this symptom is that which was first introduced by Fraenkel. It consists in a course of systematic exercises for the re-education of the ataxic limbs. The consideration underlying the treatment is that in normal conditions the functions of the sensory tracts are seldom developed to their fullest extent, and that when the functions of one sense are impaired those of the other senses may become proportionately increased; this is strikingly illustrated by the delicate perception of tactile and of auditory impressions which is acquired by the blind. Applying this consideration to the sense of position which depends on the integrity of sensory nerve fibres from the muscles, ligaments and joints, and assuming that in most cases of tabes these fibres are not completely degenerated, it will be understood that if means be taken to increase the sensibility of still living fibres, the results will be an improvement in the perception of the position of the affected limbs, and a restoration, if only partial, of the power to co-ordinate their movements.

It is obvious that if a tabetic patient wishes to attain precision in the execution of the various movements pre-

scribed for him, he must devote his whole attention to them. No benefit can ensue if the exercises produce undue fatigue, for then the patient's attention will flag, and as a consequence his attempts to perform any particular movement will fail in accuracy. In tabes the feeling of fatigue after exertion is often impaired; the patient's feelings therefore cannot be taken as a guide to the time which may be given to the exercises. Their duration is best regulated by the condition of the patient's pulse—the quicker the pulse-rate produced by an exercise, the shorter the time to be given to it, and further movements should not be undertaken until the pulse beats with normal frequency. At first as a rule the exercises may be carried out for about ten minutes twice daily; later on, if the patient becomes stronger, the duration and frequency of the sittings may be increased. The treatment should be continued for a year or longer according to the degree of progress made in each case.

Various exercises for the legs may be arranged for the patient when he is lying down, sitting, standing or walking. Thus when he is in bed he may practise touching a prescribed spot on one leg with the heel of the other, or placing the heels into the grooves or the holes of a vertical board placed at the foot of the bed. Similar exercises may be carried out in the sitting posture. Standing and walking exercises should be performed very slowly and carefully; they comprise standing on markings on the floor, and walking along certain patterns on the carpet first with short steps and then with long ones, or on stripes—broad, narrow and zig-zag—painted on the floor.

The hands and arms may be trained by exercises in writing and in copying simple diagrams; by the use of perforated boards into the holes of which the patient tries to put the tip of a finger or to insert pegs provided for the purpose; or by means of halma and draught boards.

It has been pointed out by Stuart-Low that weighted boots tend to steady the gait of an ataxic patient. He inserts a piece of sheet lead, weighing two or three

pounds, in the sole between the layers of leather; and finds that a patient wearing a pair of such weighted boots can walk without support although unable to do so when wearing ordinary boots. The purpose of the boots is to increase the tension of the muscles, and thus to improve their muscular tone: and to increase the sensory impressions from the muscles to the central nervous system.

CHAPTER II.

FAMILIAL OR HEREDITARY ATAXIA.

As pointed out by Gordon Holmes there is a difficulty in classifying the different varieties of cases which come under the above heading. Definite types of familial or hereditary ataxia are met with, of which the type known as Friedreich's Disease is one of the best defined, but there are many intermediate varieties, the pathological grouping of which at the present time must necessarily be imperfect. Provisionally they may be classified as follows:—

1. Friedreich's disease, a combined system-degeneration of the spinal cord.

2. Spino-cerebellar ataxia, dependent on a primary degeneration of the spino-cerebellar tracts, frequently associated with degeneration of the posterior columns.

3. Cerebellar ataxia, the result of a primary degeneration of the cerebellar cortex.

The essential anatomical feature of the cases coming under these headings is a primary degeneration either of the cerebellum or of some of its afferent tracts, the degeneration occurring in consequence of an inherent lack of vitality in these parts.

FRIEDREICH'S DISEASE.

This is a progressive form of spinal disease which was first described by Professor Friedreich of Heidelberg.

Etiology. It is common to find more than one case in the same family; for example, two brothers, or a brother and a sister, the other children being unaffected. That some hereditary influence is at work is shown not only by this family tendency, but by the occasional presence of the disease in the parents, or in more distant relatives. In other cases, when no direct transmission of the disease can be traced, there may be consanguinity of the parents, or a history of alcoholism, epilepsy, insanity or of some other neurosis in the ancestors. An isolated case in a family is not unfrequently met with, and sometimes in a family in which no neuropathic tendencies are apparent.

The two sexes are affected nearly equally, males slightly preponderating. The age of onset is usually about the seventh or the eighth year; sometimes the symptoms do not appear till puberty or even later.

Symptoms. As a rule the first striking symptom is unsteadiness in walking, with a liability to stumble and fall. Slight at first, the inco-ordination gradually increases until standing and walking are only possible when the feet are kept widely separated: sooner or later these acts cannot be performed without assistance, and eventually the patient becomes bedridden. In a typical case the gait is reeling and staggering and, in the swaying of the body from side to side, resembles that of a drunkard, or of a patient with cerebellar disease. The feet are not lifted so high as in tabes, nor are they advanced so rapidly and jerkily. In standing, the patient oscillates from side to side; in many cases, though not in all, this static ataxia is increased when the eyes are closed.

The arms also become affected, very often soon after the legs; their inco-ordination is shown by a disability to execute fine voluntary movements, such as those needed for buttoning the clothes, or for picking up a pin. Lack of precision is also seen in attempts to touch the nose with the tip of the finger. Inco-ordination in the movements of the arms is constantly asso-

ciated with a fine tremor, or with irregular choreic-like movements. Frequently the muscles of the head, neck and trunk become involved, so that tremor or jerky shakings of the head, and slight swaying movements of the trunk are noticeable. Irregular and purposeless contractions of the facial muscles may be also observed.

At first the muscles preserve their bulk and strength, but as the disease advances the limbs gradually become weak. Paralysis of a group or of groups of muscles is generally a late phenomenon, and the same may be said of muscular atrophy and of contractures. In a few cases motor weakness is observed at an early period; the flexors of the hip and the dorsi-flexors of the ankle being most commonly affected. The electro-muscular contractility is usually normal; in two of my own cases the partial reaction of degeneration was obtained. In the early stages of the disease the limbs are flaccid and hypotonic; in the later stages they may become spastic.

Sensory symptoms are never conspicuous and are often completely absent. The patient may suffer from aching pains in the limbs, but only rarely from shooting or from severe pains. In most cases the cutaneous sensibility is normal; in a few, I have detected slight anæsthesia in the distal portions of the limbs. The sense of position is occasionally impaired, but its preservation in association with marked ataxia is often a striking feature.

The knee-jerk is lost at an early period; its absence may be noticed for some time before the other symptoms of the disease develop. Occasionally the knee-jerk is present and persists. The plantar reflex is of the extensor type. The superficial abdominal reflexes and the cremasteric reflex normal at first gradually diminish as the disease advances. The functions of the bladder and the rectum are rarely affected.

Nystagmus is a common and sometimes an early symptom. It is often limited to, or is chiefly evoked by, conjugate lateral movements of the eyeballs. Paralysis of the ocular muscles, loss of the pupillary

reflex and optic atrophy have been observed but only very rarely, and it is doubtful whether they should be regarded as part of the disease.

Another striking feature is a peculiar disturbance of speech, due to inco-ordination of the articulatory and respiratory muscles. As a rule it does not occur until after the development of ataxia in the limbs. The defect does not present very definable features; words are slowly drawled out, their syllables are separated or elided, and the utterance, indistinct, is of an explosive character.

Characteristic deformities of the feet and back are frequently present; the former are the first to develop.

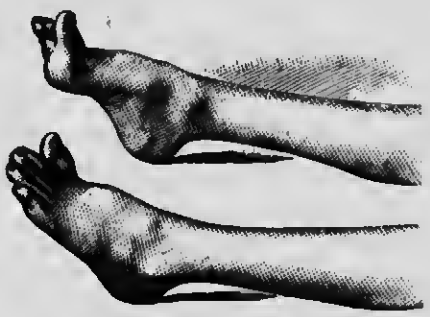


Fig. 125.—Photographs showing the attitude of the feet in Friedreich's disease.

The foot is shortened, its dorsal surface is very prominent and the sole is unduly hollowed. Its position is that of talipes equinus, or of equino-varus. The proximal joints of the toes are hyperextended, whilst the distal ones are flexed; the great toe being especially affected. The tendon of the extensor proprius pollicis stands out prominently; this prominence, or the loss of the knee-jerk, is sometimes the earliest sign of the disease; one or both of these signs may be observed in a child whose brother or sister suffers from pronounced ataxia. Lateral curvature of the spine is common in advanced cases; occasionally there is kyphosis, or kyphoscoliosis. Both the spinal and the foot deformities are probably the result of muscular weakness.

The mental condition is often lacking in acuteness and activity; it is marked by apathy or torpor rather than by actual deficiency, for example, when the patient is told to look at an object or to put his head back he will do so and will maintain the required position for a long time. The following symptoms have been

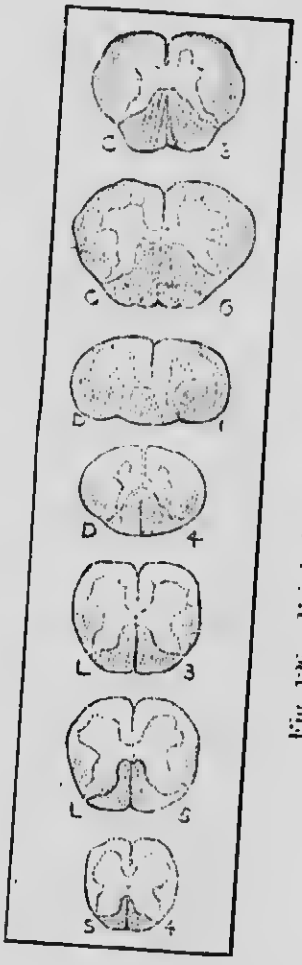


Fig. 126. Friedreich's disease; distribution of the degeneration in the white columns, indicated by the dotted shading. (Gowers, after Friedreich.)

occasionally observed:— polyuria, salivation, vertigo; cyanosis and œdema of the feet, together with other signs of cardiac muscle failure.

The march of the disease is very slow, occupying from eight to thirty years; it is fatally progressive, although long stationary periods may interrupt its

downward course. Death is generally the result of some intercurrent malady.

Pathology. Morbid changes are mainly limited to the spinal cord. In a few cases slight changes are found in the medulla and pons, namely, degeneration of the motor tracts and of the restiform bodies, and sometimes of the hypoglossal and facial nuclei. Very rarely atrophy of Purkinje cells in the cerebellum is found, but when this occurs the case is probably a type of hereditary ataxia other than the one under consideration.

The spinal cord is usually smaller and thinner than natural. The pia mater is sometimes thickened especially over the posterior columns. There is extensive sclerosis of the cord involving the posterior, the lateral and often the anterior columns. The degeneration of the posterior columns both as regards distribution and intensity resembles that of tabes; the posterior root zone in the cervical region, however, is more constantly affected. The degeneration of the lateral columns involves not only the pyramidal tracts but extends outwards and forwards into the spino-cerebellar tracts and into the anterior pyramidal tract. Associated with the sclerosis of the direct cerebellar tract, there are always degenerative changes in the column of Clarke.

Newton Pitt, who has drawn attention to disease of the vascular system, suggests that the cord degeneration is probably dependent on an inherited tendency towards general early vascular deterioration. Williamson has pointed out that the degeneration is most intense in the peripheral parts of the cord, which are less freely supplied with blood than the central areas. The most probable explanation of the disease is that of Gowers who believes that there is a congenital tendency towards early death of the nerve elements of the cord which are affected, an abiotrophy.

Diagnosis. A case of Friedreich's disease could hardly be mistaken for one of tabes, for with the exception of ataxia and loss of the knee-jerk there are scarcely

any other symptoms common to the two diseases. In Friedreich's disease we meet with nystagmus, imperfect articulation, oscillations of the head and trunk and deformities of the foot and spine. In tabes these symptoms are absent, whilst lancinating pains, crises and various disturbances of sensation are prominent features of the disease. Juvenile tabes is generally the result of congenital syphilis: the presence of any manifestation of the latter disease would be much against the diagnosis of Friedreich's disease.

Disseminated sclerosis is characterised by spastic paraplegia, exaggerated knee-jerks, and ankle-clonus: ataxia is rarely a prominent feature. Nystagmus and impaired articulation are the only points of resemblance between disseminated sclerosis and Friedreich's disease. The age of onset is different; very rarely, however, disseminated sclerosis occurs in children, and then anomalous forms of the disease may present difficulties in diagnosis.

Treatment. The disease being incurable, the treatment is limited to the alleviation of symptoms. Massage, passage movements and Fraenkel's exercises should be persevered with, especially during the early stages of the disease.

SPINO-CEREBELLAR ATAXIA.

This is a chronic slowly progressive disease which has a tendency to attack more than one member of a family, and which frequently can be traced through several generations. The onset of the symptoms generally takes place between the ages of sixteen and thirty-five. The characteristic clinical feature is inco-ordination of movement which usually affects the legs first and then gradually attacks the arms, and subsequently the muscles of the head and face and those of articulation; the gait is of the reeling cerebellar character. Failure of vision from optic atrophy occurs, and sometimes at an early period. Ptosis, diplopia and other signs of ocular palsies may be present; nystagmus also occurs in some cases. The tendon-reflexes are generally exaggerated. Weakness,

rigidity and contractures of the legs are met with in the later stages of the disease; apart from the contractures no deformities are met with.

This malady is closely allied to Friedreich's disease with which it is connected by intermediate types. It is distinguished from it by its onset at a later age, by a greater tendency to hereditary transmission, by the preservation or the exaggeration of the knee-jerk, by the absence of spinal and foot deformities, and by the frequent occurrence of optic atrophy and of oculo-motor paralysis.

The most prominent pathological change is degeneration of the dorso-cerebellar tract: the ventro-cerebellar tract or Gowers tract is affected to a less degree. The posterior columns are partly degenerated but much less so than in Friedreich's disease. The pyramidal tracts are usually spared. Occasionally the brain-stem and the cerebellum look smaller than natural; no degenerative changes are found in the cerebellar cortex.

PRIMARY PROGRESSIVE CEREBELLAR DEGENERATION.

This is a familial disease, which occurs in adults between the ages of thirty and forty and progresses slowly till death, which may result from some intercurrent affection at an advanced age. The chief symptoms are:—A staggering or reeling gait, and defective equilibrium in standing, which are not affected by deprivation of vision; ataxia of the upper limbs; tremors and choreiform movements of the limbs and head; nystagmoid jerkings of the eyes on movement, and a hesitating, scanning and explosive utterance. The cutaneous sensibility and the reflexes are not affected. The pupillary reflexes remain normal, and oculo-motor palsies do not occur.

The symptoms depend on a primary and progressive degeneration of the cortex of the cerebellum which leads to atrophy of the cells of Purkinje and of the fibres which connect them with the central nuclei. The

afferent and the efferent cerebellar tracts show no changes. A complementary neuroglial sclerosis is found in the subcortical cerebellar white matter.

Gordon Holmes believes that the selective disease of the cerebellar cortex is "primarily due to an hereditary defective vital endurance, or a tendency to degeneration of this tissue, which begun to fail from the middle of adult life." As he points out the degeneration is limited to the receptive part of the cerebellum, namely the cortex, and the fibres which arise in it.

There are two other rare forms of chronic cerebellar degeneration which may be briefly mentioned; in neither variety has a familial or a hereditary tendency been definitely traced.

Olivo-ponto-cerebellary atrophy. This is a special form of primary cerebellar disease which was first described by Thomas, who summarises its main features as follows:—"A type characterised *anatomically* by atrophy of the cerebellar cortex, of the bulbar olives and of the grey matter of the pons; by total degeneration of the middle cerebellar peduncles, by partial degeneration of the corpora restiformia, and by relative integrity of the central nuclei of the cerebellum; *clinically* by the cerebellar syndrome. It is neither hereditary, familial, nor congenital. It comes on at an advanced age and progresses slowly. It falls into the group of primary cell atrophies."

The most prominent symptoms are defective equilibration in standing and walking; ataxia and tremor of the arms, and a slow scanning articulation. Nystagmus also is usually present. In some cases, occurring in younger persons, and sometimes showing a familial tendency, slight weakness and spasticity of the legs, together with exaggeration of the knee-jerk are observed.

Progressive cerebellar disease due to vascular or interstitial lesions. Only a few cases have been recorded. In one case described by Schultze the onset occurred at the age of thirty-nine; the gait was cerebellar in type; articulation was slow and indistinct, and there was

intention-tremor of the arms, and slight nystagmus. The symptoms progressed rapidly, and death took place at the age of forty-three. There was considerable atrophy and sclerosis of the cerebellum and the brain-stem. The Purkinje cells of the cortex were scanty, and those which remained were atrophied; the molecular and granular layers were less affected. The lesions were diffuse, and there was much secondary sclerosis of the neuroglia. The central nuclei were atrophic; the superior peduncles were degenerated. There was also degeneration, probably secondary, of the olives and the transverse fibres of the pons. The spinal cord was normal except for pallor of the pyramidal tracts.

CHAPTER III.

ATAXIC PARAPLEGIA.

Combined disease of the posterior and lateral columns of the spinal cord, giving rise to an ataxic paraplegia, occurs in several spinal diseases, for example in Friedreich's disease, in subacute combined degeneration, in Erb's syphilitic spinal paralysis, and in rare cases of tabes and of general paralysis of the insane. In these diseases other symptoms are present, but cases do occur in which ataxic paralysis is the only clinical manifestation, or the only one for a time. In the year 1886 Gowers described the condition as a new clinical entity: subsequent investigations, however, have shown that many of the cases upon which he based the morbid anatomy of the condition are instances of subacute combined degeneration, whilst others eventually prove to be instances of disseminated sclerosis. For the few remaining cases the title may be provisionally retained.

The chief antecedents which may have acted as exciting causes of 'ataxic paraplegia' are, exposure to cold, severe muscular exertion and concussion of the spine. A history of syphilis is rare.

The symptoms develop gradually. At first the patient

complains of unsteadiness in walking, or of easily induced fatigue and stiffness in the legs. On examination there is unsteadiness in standing and walking, which is increased by closure of the eyes; weakness of the flexors of the hip and knee, and of the dorsi-flexors of the ankle are also usually present. The knee-jerk is exaggerated, ankle clonus can be obtained, and the plantar reflex is of the extensor type. Sensory symptoms are never prominent, and are usually absent. Sooner or later the arms become affected with weakness, inco-ordination and exaggeration of their reflexes.

The pupils react to light: nystagmus and optic atrophy do not occur or only very rarely. The bladder and rectum are not involved, except occasionally at a late period of the disease, when the sphincters may be partially paralysed.

As the disease progresses, the spastic weakness increases and gradually overshadows the ataxia, so that the case more and more resembles one of spastic paraplegia. Variations from the above type are met with, some cases approaching more closely to tabes, others to disseminated sclerosis, and still others to a dorsal myelitis. The course of the disease is very chronic, and apart from complications there is but little tendency to a fatal ending.

The post-mortem lesion is sclerosis of the posterior and the lateral columns, the degree and extent of the sclerosis varying considerably in different cases. The situation of the degeneration in the posterior columns differs from that in tabes in two respects: it is usually more intense in the dorsal than in the lumbar region, and the root zone is not conspicuously affected. In many cases the degeneration is limited to the middle portions of the posterior columns, and does not extend to the posterior surface of the cord. In the lateral columns the pyramidal tracts are mainly implicated; sometimes degeneration may be traced into the cerebellar tracts and into other portions of the white matter.

CHAPTER IV.

SUBACUTE COMBINED DEGENERATION OF THE SPINAL CORD.

The peculiar clinical features of subacute combined degeneration—as shown by the distribution and character of the nervous symptoms, by their tendency to successive changes in type and by the presence of anæmia, along with its characteristic morbid anatomy, namely a diffuse and progressive degeneration of the white matter of the spinal cord, were first pointed out and placed upon a satisfactory basis by Risien Russell, Batten and Collier. Prior to their investigations on the subject many cases of the disease had been described under a different heading, and usually under one which implied that anæmia was the essential cause of the disease. There is ample evidence that anæmia may lead to degenerative changes in the spinal cord which may be expressed during life by various symptoms, and it is possible that anæmia alone may give rise to the condition known as subacute combined degeneration. But there are strong reasons for believing that in most cases of the disease, the anæmia which is generally present, is a secondary phenomenon and that both the anæmia and the changes in the nervous system are concomitant results of the same cause, whatever that may be. Subacute combined degeneration is to be regarded as a separate entity, a malady quite distinct from a number of ill-defined conditions of the nervous system which occasionally occur as a direct result of a severe anæmic state of the blood.

Etiology. Persons between forty and sixty years of age are most often affected; women are rather more liable than men to be attacked. The exact cause is unknown, but there is much to suggest a toxic origin; thus the disease has followed influenza, prolonged suppuration or diarrhœa; it has occurred in women during pregnancy and parturition; occasionally the

patients have had syphilis or have been addicted to alcohol. In many cases the health was quite good till shortly before the onset of the symptoms.

The most common associated condition is anæmia but it is not constantly present, nor does it always precede the onset of spinal symptoms, hence it cannot be regarded as the essential cause. It seems more probable, as already suggested, that some toxic agent is responsible for the anæmia and the cachexia which occur in this disease as well as for the spinal degenerations.

Morbid Anatomy. There is a diffuse degeneration of the white matter of the spinal cord which is most marked in the mid-dorsal region. There, nearly the whole of the white matter may be destroyed; the grey matter and a narrow zone of white matter immediately surrounding it are not affected. Above and below the mid-dorsal region the destructive process gradually diminishes, degeneration being most marked in the posterior columns above the thoracic region and in the lateral columns below it. In the cervical region the columns of Goll are more affected than those of Burdach; the cerebellar tracts and the crossed pyramidal tracts are also involved, and scattered areas of degeneration are found in other parts of the cord, especially in the anterior columns. In the lumbar region also the degeneration is widespread, and although most pronounced in the lateral columns it is found in the posterior columns, and in scattered areas outside both these tracts.

Thus there appear to be two distinct processes—a focal destructive lesion in the dorsal region and a systemic degeneration of the long tracts, above and below this region. In these respects the morbid anatomy resembles that of a transverse lesion of the cord; a marked distinction, however, is the absence of limitation of degeneration to the long afferent and efferent tracts, for fibres outside these tracts are also affected. The grey matter, the anterior and posterior roots and the peripheral nerves are usually quite normal.

Fig. 127. Sections of the medulla: cervical, thoracic and lumbar regions of the spinal cord stained by the Weigert-Pal method; showing degeneration in the fasciculus gracilis (fg.c.), the fasciculus cuneatus (fc.c.), the direct cerebellar tracts, the posterior columns and in the crossed and direct pyramidal tracts. (F. E. Batten. "The Spec. Pathology and Diagnosis of Nervous Diseases.")

Retrograde lesions, however, are found in the cells of Clarke's columns and in the pyramidal cells of Betz in the ascending frontal convolution; the former are the result of interruption of the spino-cerebellar tracts and the latter of interruption of the spinal pyramidal tracts.

The walls of the blood vessels may show fatty changes and occasionally sclerosis; it is probable that such changes are secondary, for they are only marked in areas where the destructive process is advanced.

Symptoms. In many cases the clinical course may be separated into three stages; the transition from the one stage to the other is often rapid, and is indicated by a marked change in the character of the symptoms.

The three stages are:—(1) A stage in which the symptoms are those of a slight ataxic-paraplegia. This stage is the longest, lasting usually several months and occupying from a half to three-quarters of the whole illness. (2) A stage of spastic paraplegia with anæsthesia of the legs and trunk, lasting only a few weeks. (3) A stage of complete flaccid paraplegia, with absolute anæsthesia, absent knee-jerk, incontinence of urine and fæces and œdema of the legs and trunk. The average duration of this stage is also a few weeks.

In some cases the symptoms develop rapidly and are ushered in by headache, vomiting and pyrexia, but in most cases the onset is slow and insidious. As a rule the earliest manifestations of the disease are subjective sensations of numbness and tingling in the peripheral parts of the limbs. At first these sensations have a "stocking and glove" distribution, the feet being earlier and more severely affected than the hands; subsequently they spread towards the trunk. They are often associated with a slight diminution of the cutaneous sensibility.

The patient now begins to have a feeling of stiffness and weakness in the legs and sometimes in the arms; he may also show signs of anæmia. The sensory symptoms are soon followed by an obvious spastic weakness of the legs associated with ataxia; in some cases the

Fig. 121. Sections of the medulla; cervical, thoracic and lumbar regions of the spinal cord stained by the Weigert-Pal method; showing degeneration in the fasciculus gracilis (f.g.), the fasciculus cuneatus (f.c.), the direct cerebellar tracts, the posterior columns and in the crossed and direct pyramidal tracts. (F. E. Batten. "The Sec. Journ." vol xxxvi.)

spastic paraplegia, in others the ataxia is the more marked. The knee-jerks are exaggerated, ankle clonus may be obtained and the plantar reflex is extensor in type. A clumsiness and a lack of co-ordination in the movements of the hands are sometimes observed.

The transition to the second stage is usually rapid and the patient may become unable to stand and walk in the course of a few hours or days; this disability is due more often to inco-ordination of movement than to complete paralysis. Definite anæsthesia now rapidly develops; it begins in the distal parts of the limbs and gradually extends to the trunk, where its upper limit is sharply defined, and is frequently associated with a girdle sensation. Severe darting pains in the legs are often present, and sometimes there is much intercostal pain, or an aching pain in the hepatic region. In rare cases the intercostal pain is accompanied by herpes zoster and occasionally by subcutaneous hæmorrhage.

During this stage the rigidity of the legs increases and may become extreme; paralysis, however, is rarely absolute; the muscles react normally to electricity. Both the superficial and the deep reflexes are exaggerated. Exceptionally slight nystagmus, weakness of an ocular muscle, or retinal hæmorrhages are observed.

The general condition of the patient is often good, but there is a tendency to irregular rises of temperature and to attacks of diarrhœa; the pyrexia occurs both in patients who are anæmic and in those who show no signs of anæmia.

The transition to the third stage is marked by a rise of temperature and, soon after, the spastic paraplegia is replaced by a complete flaccid paralysis; the muscles show marked hypotonus and begin to waste rapidly. The muscular excitability to the Faradic and galvanic currents is much reduced, and to the former more than to the latter; as a rule no definite reaction of degeneration is obtained. During this period œdema of the legs and trunk is not uncommon; it probably depends on the anæmia and on the impaired innervation of the affected parts.

The knee-jerks are now lost, ankle clonus disappears, but Babinski's reflex is usually persistent. Anæsthesia of the legs and lower part of the trunk is absolute and may extend as high as the first dorsal segment. The sphincters become paralysed and cystitis often occurs and may lead to pyonephrosis. Œdema of the legs and trunk sometimes appears at this period. At a later period the small muscles of the hand and subsequently some of the arm-muscles undergo a progressive atrophy; there may be a band of anæsthesia along the inner aspect of the arms. The patient gradually gets weaker and thinner; bed sores form over the sacrum and death results from exhaustion, from respiratory paralysis, or from renal complications. During the terminal period, mental weakness and nocturnal delirium are common, and sometimes the patient is subject to attacks of clonic convulsions without loss of consciousness.

Such a definite course as that described is not always observed; in some cases a flaccid paralysis, with loss of the tendon reflexes, and ataxia are present at an early period; in others the flaccid stage is never reached, the limbs remaining spastic throughout the whole course of the disease.

Anæmia is a common symptom, the blood usually having the characters of a secondary anæmia, sometimes of the pernicious type. Anæmia may precede, or attend, the onset of the nervous manifestations, or it may appear at any subsequent time; occasionally it is completely absent.

Diagnosis. It is in the earlier stages of the affection that difficulties in diagnosis chiefly arise, and especially as regards disseminated sclerosis and tabes. In *disseminated sclerosis* the nystagmus is usually more marked, optic atrophy is common and there is a tendency to emotional disturbance and to remission of symptoms; the disease too occurs at an earlier age. Then certain symptoms, namely lancinating pains, marked anæsthesia, anæmia and cachexia which occur in combined

degeneration are quite exceptional in disseminated sclerosis.

Tabes is suggested by ataxia and girdle pains: it is excluded by the spasticity of the legs, the increased knee-jerks and the normal reactions of the pupils. In the third stage of combined degeneration the flaccid condition of the legs, the loss of the knee-jerk, the anæsthesia and the paralysis of the sphincters might also suggest tabes, but against this disease is the marked paralysis, together with the absence of the Argyll-Robertson pupil phenomenon and the presence of Babinski's reflex.

Tumours pressing on the cord sometimes cause a spastic paraplegia followed by flaccid paralysis, but as a rule the paralysis is preceded by severe root pains which are not conspicuous in combined degeneration. In the latter disease the hemisection phenomena are never present, whereas they are common when the cord is compressed.

Certain cases of *acute myelitis* may closely resemble those of combined degeneration; in favour of the latter are the history of a gradual onset, the definite stages, and the involvement of the arms. It is probable that many cases described as instances of chronic myelitis were really examples of the disease under consideration.

Prognosis. Although it is impossible in any given case to foretell the duration of life the prognosis is particularly unfavourable when the anæmia is severe and when the onset and the development of the nervous symptoms have been rapid. In such cases death usually occurs within six months. In other cases life may be prolonged for several years, but as a rule the fatal issue is reached before the end of the fourth year.

Treatment. The anæmia often shows marked improvement for a time under the influence of arsenic and iron, but no drugs as yet tried appear to have benefited the paralytic symptoms or to have materially affected the general course of the illness. Careful attention should be given to the condition of the mouth, the naso-

pharynx, the stomach and the bowels, in order that no possible source of infection may be overlooked, for it seems probable that some cases of subacute combined degeneration may owe their origin to absorption of toxins from a chronic infection of the alimentary tract.

CHAPTER V.

AUDITORY VERTIGO.

A study of the pathology of the spino-cerebellar tracts, lesions of which constitute the chief features of many of the diseases described in this section, shows how important for the maintenance of equilibrium are the impulses derived from the periphery of the body. Probably of still greater importance in relation to equilibration are the impulses which reach the cerebellum by the vestibular tract. The fibres of this tract originate in the ganglion vestibulare, the peripheral filaments of which supply the lining membrane of the semicircular canals, and combine to form the vestibular root of the right nerve; this root enters the pons and ends in close relation to certain of its nuclei, which are intimately connected with the cortex of the cerebellum. (See figs. 11, 12 and 14.)

A lesion of the semicircular canals, or of the vestibular nerve produces a sensation of giddiness which may or may not be associated with an involuntary movement of the body. The term vertigo is used to include both these factors; it implies a sense of defective equilibrium as well as actual manifestations of its presence.

The exciting causes of vertigo are very numerous. It occurs in disorders of the alimentary canal, of the kidneys and other organs; in connexion with defects in ocular innervation; in consequence of alcoholism, of excessive smoking and of other toxæmic conditions,

whilst it is a common symptom in diseases of the brain especially in those involving the cerebellum.

In most cases of vertigo there is satisfactory evidence based partly on the condition of the auditory functions – such as the presence of tinnitus and slight deafness—and partly on a consideration of the complicated anatomical relations of the vestibular tract, that apart from organic brain disease and epilepsy the symptom vertigo is to be regarded as the direct or indirect consequence of a morbid condition of the labyrinth or of its connexions with the cerebellum.

Certainly so far as diseases of the ear are concerned there can be no doubt that local conditions which alter the pressure of the endolymph such as suppuration in the middle ear, or the accumulation of wax in the external auditory canal—or conditions which give rise to irritability of the nerve-terminals in the semicircular canals, will severally upset the sense of equilibrium and thus cause vertigo. A distinction should be drawn between *auditory vertigo* due to primary disease of the labyrinth and *aural vertigo* produced by affections of the external and middle ear.

A further distinction is required in relation to the use of the term Ménière's disease. This should be restricted in order to identify and to give a definite entity to the condition originally described by Ménière—to acute primary affections of the labyrinth; whilst it is convenient to apply the term Ménière's symptom-complex to chronic and to secondary affections of the labyrinth. The former affections are rare, the latter are common.

Acute Labyrinthitis—Ménière's Disease.

The pathological condition which gives rise to the symptoms of this rare malady is an acute destruction of the labyrinth, the result either of hæmorrhage or of acute inflammation. The clinical picture of a typical case is as follows:—A man, past middle age, without any

signs of ear disease, and apparently in good health, is suddenly seized with intense vertigo which throws him to the ground, where he lies for a minute or two in a state of partial or even of complete unconsciousness. On recovering consciousness he suffers from nausea and vomiting and becomes pale and collapsed.

It is now found that his hearing on one side is impaired, and that he suffers from severe tinnitus. His eyes show well-marked nystagmoid movements towards the unaffected side. In a few days the vomiting and vertigo diminish in intensity, and the former soon ceases altogether; the latter is apt to be aggravated by any movement of the head. The nystagmus may continue for some time. Eventually all the symptoms pass away with the exception of tinnitus and unilateral deafness which may persist during the remainder of the patient's life.

Chronic Labyrinthitis: Ménière's Symptom-Complex.

As a rule this occurs as a complication of old-standing disease of the middle ear, especially chronic adhesive and sclerotic processes; sometimes there is a direct extension of suppuration from the middle ear to the labyrinth. Less commonly chronic labyrinthitis occurs as a primary affection in consequence of syphilis, gout or senile degeneration.

The chief **symptoms** of the disease are tinnitus, deafness and vertigo, which develop gradually in the order given; tinnitus and deafness have usually been present for some time before the patient suffers from his first attack of vertigo.

Tinnitus, which is generally the earliest manifestation of the malady, is often peculiarly distressing owing to the intensity of the sounds and their persistency. The sounds vary in character; they may be ringing, hissing, roaring, pulsating or humming. Sooner or later the hearing becomes impaired and ultimately may be lost. Both the deafness and the tinnitus are usually unilateral, but the opposite side may also become affected.

The attacks of vertigo, which are repeated at irregular intervals and which occur with great suddenness, vary much in severity. Sometimes the attack is limited to a sensation of giddiness causing the patient to feel as if he were rotating, or as if surrounding objects were in motion. Sometimes the attack is severe enough to cause the patient to fall to the ground from which for a time he may be unable to rise owing to the intensity of the vertigo or possibly to a temporary loss of consciousness. The act of falling may be immediately preceded by the impression that a blow has been struck on the head, which compels the patient to fall in a particular direction. Frequently an attack of giddiness is combined with instability in walking, giving rise to a staggering gait.

The vertiginous seizures are often accompanied or followed by nausea and vomiting. They vary in duration from a few minutes to several hours, and may recur every few weeks or months. There may be no vertigo between the paroxysms but more commonly there is continuous vertigo, although of slighter degree than that in the attacks.

The **course** of chronic labyrinthitis varies much in different cases; as a rule the tendency to giddiness gradually lessens and may pass away altogether. When, however, the morbid process is steadily progressive the symptoms may only cease when the hearing power is completely destroyed.

The **diagnosis** of labyrinthine vertigo is based on the association of attacks of giddiness with tinnitus and deafness not due merely to impairment of the sound-conducting apparatus. The greatest diagnostic difficulty is in the distinction between the symptoms of slight labyrinthine disturbance and those of minor epilepsy. In both cases the vertigo may occur suddenly and be of extremely short duration: it may be attended by a transient loss of consciousness, and it may occur during sleep. The distinction can only be made by a careful observation of the patient for some time; progressive

deafness and tinnitus would point to disease of the labyrinth, slight convulsive seizures and mental changes to epilepsy. It must not be forgotten that occasionally labyrinthine vertigo is associated with attacks of epilepsy.

The **treatment** of auditory vertigo depends to some extent on its cause. If it is secondary to middle ear disease this must be adequately dealt with, surgical aid being often required. If an underlying morbid state, such as gout or syphilis, can be determined, suitable treatment for these conditions may materially lessen the severity of the attacks. When no cause can be discovered we are limited to remedies for the labyrinthine symptoms, and for the cerebral instability caused by them. For the former symptoms the most effective treatment is counter-irritation; the application of a blister or of a seton behind the ear will often give considerable relief. The administration of salicylate of soda is also beneficial. For the central disturbance, bromide of potassium is the most efficacious medicine; a dose of fifteen or twenty grains should be given thrice daily for a long time. In many cases the administration of this drug either alone or combined with tincture of belladonna will reduce the number and the severity of the seizures, and may arrest them altogether.

In severe, intractable and progressive cases, especially when the symptoms are unilateral, the radical operation for removal of the semicircular canals may be advisable; as it usually involves destruction of the cochlea, complete deafness will necessarily ensue.

The general treatment of the patient must not be overlooked; the bowels must be kept acting regularly and tonics must be given from time to time. It is also important to treat any morbid condition that may be present, especially dyspepsia, which may exaggerate the vertigo or may co-operate with other influences to induce an attack.

SECTION IX.

Diseases characterised for the most part by the presence of some variety of Muscular Spasm.

The term spasm is generally applied to muscular contraction, which is in excess of that occurring in conditions of health. Such excess is a component part of the convulsions of epilepsy or of those produced by a coarse lesion of the motor cortex. It is the main feature of spasmodic torticollis, of tetany, of the involuntary movements of chorea, and of many varieties of tic. The regional pathology of these cases is not always the same, and there appears to be a growing tendency to vary the clinical terminology of excessive muscular contraction according to the position of the lesion which produces it, to give the term spasm a pathological rather than a clinical significance. Thus we are told that the term must be restricted to the motor reaction which is consecutive to pathological stimulation of some point in a spinal or a bulbo-spinal reflex arc, a striking instance of which is the twitching of the facial muscles produced by irritation of the sensory fibres of the fifth cranial nerve. According to this view the twitchings of the facial muscles which are due to cortical instability, whether this be the result of psychical disturbance or of a tumour or other coarse lesion, are not true spasms: they are tics or local convulsions as the case may be.

Without entering into a criticism of this view it seems desirable to briefly review our knowledge of the regional pathology of spasm, using the word as a designation for a morbid excess of muscular contraction.

When paralysis is associated with spasm, as in hemiplegia from a lesion of the internal capsule, or in paraplegia fro a lesion of the pyramidal tracts in

the spinal cord, it is clear that the rigidity of the affected limbs is due, directly or indirectly, to interference with the transmission of motor impulses along the upper neurons. The generally accepted explanation is that, owing to the spinal anterior horns being cut off from the influence of cerebral inhibition their cells react excessively on the muscles which they innervate; in other words, the spinal cells are deprived of certain healthy impulses, and are therefore abnormally prone to react to irritation, whether of reflex or of other origin; for example, the limb contracture in hemiplegia may be suddenly exaggerated by a comparatively slight peripheral injury. In a sense then the tonic spasm caused by a lesion of the upper neurons may be regarded as partly of reflex origin. But partly only, for another influence has to be taken into account, namely, that of the cerebellum. As first pointed out by Hughlings Jackson, the cerebellum exerts a tonic influence over the muscles of the body; in this respect its action is antagonistic to that of the cerebrum, which, as we have seen, is inhibitory to muscular tonus. A cutting off of cerebral impulses tends to exaggeration of the deep reflexes and to increased muscular tonus, whereas a cutting off of cerebellar impulses tends to a diminution of these reflexes and to hypotonus. Broadly speaking, it may be said that the maintenance of ordinary cerebral and cerebellar relations to the spinal cord is essential for normal tonus and for normal reflexes, and that any material change in such relationship leads to changes in the reflexes, and in the tonus of the muscles.

It seems probable therefore that the over-action of the spinal centres which results from removal of cerebral inhibition and which leads to rigidity of the paralysed muscles is partly the result of the unopposed action of the cerebellum. This hypothesis is strongly supported by the fact that a *total* transverse lesion of the spinal cord above the lumbar enlargement, which excludes both cerebral and cerebellar influences from

the spinal centres, leads to a complete flaccid paralysis, together with abolition of all the reflexes of parts situated below the level of the lesion.

Now, although a lesion of the upper motor neurons almost invariably attends the presence of a spastic paralysis, this is not always the case when muscular spasm *alone* is present: frequently indeed it is impossible to be certain which part of the motor tract is at fault. The spasms which make up the convulsions of epilepsy can be assigned to irritation of cortical cells, and so also can the local convulsions of coarse cortical lesions: in these cases the upper neurons are implicated. Other spasms appear to be related to irritation of the lower neurons, thus compression of the seventh nerve at the base of the skull may cause spasm of the facial muscles, and meningitis involving the third nerve may lead to a spasmodic squint: such spasms, however, are not necessarily due to direct irritation of these nerves, they may be reflex in origin. It is indeed noteworthy how few spasmodic affections can be traced with certainty to lesions of the lower neurons: for example, spasm is not a symptom either of acute or of chronic disease of the spinal anterior horns, and even in traumatic lesions of the motor fibres of the peripheral nerves muscular spasm is a rare event. It is much more frequently produced by irritation of sensory nerve fibres; thus carious teeth occasionally start a facial spasm, corneal ulcers may cause a tonic contraction of the orbicularis palpebrarum, and inflammation of a joint may excite spasm of its muscles. These are examples of reflex spasm.

There are other spasmodic disorders of which the regional pathology has not yet been accurately determined. It is still uncertain where the lesion is situated which gives rise to the muscular rigidity of paralysis agitans, to the tonic spasms of tetany, to the clonic spasms of torticollis or to the shock-like contractions of paramyoclonus multiplex.

It will gathered from a consideration of the above

statements that the pathology of spasm is often very complex; in every case we have to take into account the influence of the cerebrum as well as that of the cerebellum upon the spinal grey matter. We have also to consider disturbances of the grey matter from reflex causes, and, further, the possibility that spasm may be produced by direct irritation either of motor nerve fibres or of the contractile tissue of the affected muscles. On the whole then it seems desirable to use the term spasm in a clinical sense rather than to give it a definite association with any particular pathological condition. From this point of view there is adequate justification for grouping together, as in the present section, various spasmodic affections which appear to be generated by different pathological conditions.

CHAPTER I.

SPASM OF THE MUSCLES SUPPLIED BY THE CRANIAL NERVES.

FACIAL SPASM.

This may depend on many causes. It occurs in chorea, in athetosis, in tetanus and occasionally in tetany: it forms part of the convulsions of epilepsy, of hysteria or of those produced by an irritative lesion in the cerebral cortex. Should such a lesion involve only the lowest part of the ascending frontal convolution it occasionally happens that spasms are limited to the facial muscles. Persistent facial spasm has been started by compression of the facial nerve at the base of the brain, by a tumour, an aneurysm or by meningeal thickening; in rare cases transient facial spasm has been set up by a lesion of the facial nucleus in the pons. Sometimes facial paralysis of long standing becomes complicated by contractions of the affected muscles.

Facial spasm is also met with as an *idiopathic affection*, that is, apart from any detectable morbid

change which could account for it. This variety usually begins between the ages of forty-five and sixty; it affects women more frequently than men. In many cases no cause can be discovered. In others the affection has been preceded by some emotional state, such as that produced by a sudden shock or by prolonged mental anxiety; some of these cases may be instances of tic, rather than of true facial spasm. Another antecedent is peripheral irritation, such as that caused by carious teeth, or by painful affections of the conjunctiva and cornea. Occasionally the spasm appears to have developed out of some frequently repeated movement of the face.

In these cases it is difficult to say what part of the facial motor path is disturbed; there may be molecular changes in the cortical centre which start the discharges causing the spasm, but the evidence is in favour of the facial nucleus being at fault, the affection being a bulbo-reflex spasm, of which the afferent channel is probably the fifth nerve and the efferent the seventh nerve.

Symptoms. As a rule the spasm is limited to one side of the face, and is of the clonic variety. At first it is slight, and only occurs occasionally. Subsequently the clonic contractions tend to increase in severity: they are repeated more and more rapidly, and at the height of the paroxysm may pass into a tonic contraction. The paroxysms vary greatly in duration and in the frequency with which they recur. In the intervals between them there is partial or complete cessation of the spasm.

All the muscles supplied by the facial nerve, including the platysma and even the stapedius, may be involved in the spasm; the orbicularis palpebrarum and zygomatici are the most frequently and the most markedly affected; the orbicularis oris and the frontalis often escape. It is obvious that the facial contortions will vary in different cases; usually they comprise twitching and winking of the eyelids, twitchings and

distortions of the naso-labial fold and of the angle of the mouth.

In a severe attack the spasm may spread to the other side of the face, affecting at first the eyelids or the angle of the mouth, and later possibly nearly all the muscles of both sides. Occasionally it extends to the tongue, to the masseters, to the sternomastoids, and even to the muscles of the shoulder. Sometimes the spasm is very limited in distribution; it may involve the eyelids only—*blepharospasm*. In this affection there is contraction of the orbicularis palpebrarum on both sides, the spasm being either of the clonic or the tonic variety; the former manifests itself by rapid winking movements; the latter by persistent closure of the eyelids for a few minutes or possibly longer. The clonic form may occur without obvious cause; it is also seen in hysteria or as a symptom of tic. The tonic form is associated with photophobia, and is generally caused by irritation of the ocular branches of the fifth nerve.

All varieties of facial spasm are liable to be evoked or to be aggravated by emotion, by a bright light and by volitional movements of the face, as in speaking or laughing; they tend to abate or to cease altogether when the patient is at rest and in a dark room.

Sensory symptoms are rarely present, and voluntary movements of the face are unimpaired. The electrical reactions of the affected muscles are normal.

Diagnosis. The persistency and the paroxysmal character of the spasm, together with the total absence of paralysis, even when the affection has lasted for years, distinguish this variety from all other varieties of facial spasm. When spasm is produced by pressure on the facial nerve, or by a lesion in the cortex, paralysis, either transient or permanent, generally supervenes. When it follows or complicates an old facial paralysis there is always a history or the presence of muscular weakness on the affected side as well as some permanent contracture of the muscles. Pain or the presence of any tender spots in the course of the fifth nerve suggests

that the spasm is of reflex origin, and is due to irritation of the fifth nerve.

Course and Prognosis. The course is irregular, the severity of the attacks varying much from time to time. Sometimes they cease altogether, either spontaneously or as a result of treatment, but there is a great proneness to relapse. In a few cases complete recovery takes place; but as a rule the affection is of long duration and may persist to the end of life. Although not serious the facial contortions give rise to much distress and sometimes lead to great mental depression.

Treatment. In every case of facial spasm our first duty is to try and ascertain the cause, and especially to seek for any source of reflex irritation, such as carious teeth or disease of the nose, pharynx, ears or eyes. The removal of any local disease will often ameliorate or cure the facial spasm. If, however, the attacks still persist, and also in cases in which no cause can be discovered we must rely mainly on treatment calculated to improve the general health, and to strengthen the neuropathic constitution. To this end an outdoor life, regular hours and suitable food are of the first importance. The patient should be protected as far as possible from all mental excitement; he should also avoid exposure to draughts of cold air.

No drug appears to have a curative effect. Bromides may relieve the spasms for a time; gelsemium, conium and morphia are also beneficial in some cases. When the patient is anæmic, iron and arsenic should be administered. In the early stages of the affection hot applications to the face, or counter-irritants, such as a blister placed behind the ear, are sometimes of service.

The value of electricity is doubtful; the application of a weak constant current appears to have done good in some cases; the cathode should be placed on the nape of the neck, or behind the ear and the anode passed over the affected side, the current being allowed to pass continuously for a few minutes. Massage of the face and general gymnastic exercises are to be recommended:

the latter not only improve the general health, but tend to strengthen voluntary control over the muscles.

Stretching of the facial nerve has been successful in some cases, but the operation should only be recommended when the attacks are particularly severe and of long standing; moreover, the patient must be warned of the probable replacement of spasm by paralysis.

In obstinate cases another operative method is worthy of trial, namely, the injection of absolute alcohol into the sheath of the nerve. The needle of the syringe should be thrust in towards the stylo-mastoid foramen and the alcohol very slowly injected until distinct paralysis of the facial muscles is produced. This paralysis passes away rapidly; the spasms may cease and never return; their recurrence, however, after a few months is the most common result.

SPASM OF THE OCULAR MUSCLES.

Spasms, both tonic and clonic, occasionally affect the extrinsic as well as the intrinsic muscles of the eye. Spasm of a single muscle is rarer than spasm of muscles associated in a particular movement; in convulsive attacks the eyes with the head are turned towards the side most convulsed, that is, away from the hemisphere which is most irritated. In hysteria the eyeballs may be spasmodically converged, or turned upwards. Convergent strabismus occurs also in basal meningitis, and in cases of "head-nodding"; its most common cause is hypermetropia owing to prolonged overaction of the internal recti. Transient diplopia is sometimes experienced by patients suffering from chorea: it is probably the result of irregularly distributed spasms of the ocular muscles. Tonic contraction of the levator palpebræ is a feature of Graves disease; very rarely it is caused by irritation of the fifth nerve.

Nystagmus. This term is applied to rapid involuntary rhythmical movements of the eyes, which are generally bilateral and symmetrical. The movements may be constant during waking hours, or may occur only when

the eyes are moved in a particular direction. In the latter case the nystagmus may be the result of muscular weakness; for example, partial paralysis of the right external rectus would be suggested if twitching movements of the right eye were observed when the patient looked as far as possible towards the right side.

The direction of the movement is most frequently lateral; sometimes it is rotatory or vertical. Occasionally the oscillations may be detected during the use of the ophthalmoscope when they are invisible to the naked eye.

Nystagmus occurs in many diseases of the nervous system. It is a distinctive feature of disseminated sclerosis, of Friedreich's disease, and is of common occurrence in tumours of the cerebellum, of the corpora quadrigemina and of the optic thalamus. It occurs as a temporary symptom in cerebral hæmorrhage, meningitis and sinus thrombosis. Disorders of the semi-circular canals also produce nystagmus. It is often found in association with conditions which cause defects of sight, such as opacities of the cornea, optic atrophy, errors of refraction and albinism. In children it occurs also in cases of "head-nodding." Sometimes it develops in adult life apart from other evidence of disease, as in coal-miners, who work in a cramped position with poor illumination.

MASTICATORY SPASM.

Spasm of the muscles of mastication may be either tonic or clonic, the former variety being more frequent. Tonic spasms keeps the jaws firmly closed—'trismus' or 'lock-jaw'—so that the teeth cannot be separated, or only slightly; the taking of nourishment is therefore difficult, and speaking is very indistinct. Trismus is a striking and an early symptom in tetanus: it may also occur in severe cases of tetany. It occurs during the tonic stage of an epileptic fit, and occasionally in connection with hysteria. Sometimes it depends on central lesions in the pons, which irritate

the motor nucleus of the fifth nerve; in other cases it is caused by irritation of the nerve itself, as from a basal meningitis. Interference of movement by spasm must be distinguished from that due to disease involving the ramus or the joint of the lower jaw.

Clonic spasms of the muscles, causing the lower jaw to make rapid vertical movements and bringing the teeth forcibly together form part of a rigor or of a general convulsion. Such contractions are also observed in some cases of paralysis agitans. A single sudden contraction occurring at long intervals is occasionally met with in chorea.

GLOSSAL SPASM.

Spasm of the tongue, either tonic or clonic, is rarely met with as an independent affection. It may be produced reflexly, in consequence of peripheral irritation, as from inflammation of the buccal mucous membrane, or from carious teeth; or possibly in some cases by direct irritation of the hypoglossal nerve or of its nucleus. But as a rule the spasm forms a part of a general convulsion, such as that of epilepsy or of hysteria; it may also participate in the spasms of chorea. Both sides of the tongue are usually involved: a unilateral spasm is sometimes observed in cases of hysterical hemiplegia, which causes the tongue to be protruded obliquely.

CHAPTER II.

SPASMS AFFECTING THE MUSCLES OF THE TRUNK AND THE LIMBS.

Apart from the spasms which occur in tetanus, in tetany, in the occupation neuroses, and in the convulsive attacks of cortical disease, of epilepsy and hysteria, and excluding also the spastic paralyses, localised spasms are occasionally met with which do not form a part of any known disease, and which cannot always be traced to a definite cause.

A single muscle may be attacked, as the levator anguli scapulæ, the latissimus dorsi, the deltoid or one of the rhomboids, or a group of muscles supplied by the same nerve or by the same spinal root. Thus clonic spasms have been observed in the muscles supplied by the fifth cervical root, namely, the deltoid, biceps, brachialis anticus and supinator longus.

Of the muscles of the lower limb the calf muscles are especially liable to be the seat of painful tonic contractions; such cramps are common in certain varieties of multiple neuritis, and, apart from definite neuritis, may be caused by errors in metabolism. Tonic spasms may

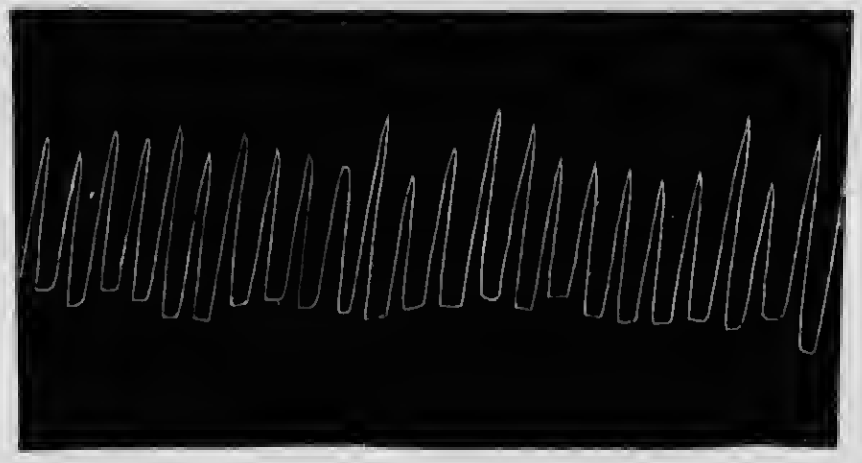


Fig. 128.

also affect the extensors of the toes and feet, the muscles of the sole of the foot, and indeed almost any muscle in the lower limbs.

In a boy, aged ten, under my care, the sartorius and the quadriceps of the left thigh were the seat of marked clonic spasms: these were regular in rhythm and numbered about one hundred a minute (see tracing, fig. 128). When the boy's knee was flexed clonic spasms quickly appeared in the hamstring muscles. There were no other symptoms beyond slight cutaneous hyperæsthesia of the affected thigh; no cause for the spasm could be discovered. In this case there were no signs of a neurosis, but in most

patients who suffer from localised clonic spasms there is evidence of hysterical or of other neurotic tendencies. Such a predisposition is less common in cases of tonic spasm, as they are more often due to over-exertion, to local irritation, or to the presence of a poison in the blood.

In the etiology of both varieties of spasm injury plays an important part: it may act directly, or indirectly, as a result of the sensory irritation set up by a wound or by a scar. In one of my cases clonic spasms of the left pectoralis major were associated with the presence of an old cicatrix at the lower edge of the muscle; removal of the scar tissue was followed by a cessation of the spasms.

The general treatment of all varieties of local spasm should be based on the lines which we have already indicated as suitable for the treatment of facial spasms.

CHAPTER III.

PARAMYOCLONUS MULTIPLEX.

This is a rare affection characterised by the repeated occurrence of sudden, shock-like contractions of single muscles, of portions of muscles or even occasionally of groups of muscles. The contractions affect the muscles of the limbs more commonly than those of the trunk, and the latter more frequently than those of the face. The proximal muscles of a limb are more prone to be involved than the distal ones; thus in the upper limb the clonic spasms predominate in the deltoid, the pectoralis major, the biceps, the triceps and the supinator longus; in the lower limb in the extensor and adductor muscles of the thigh. The muscles of the trunk, that are most commonly affected, are the abdominal recti, the obliques and the erector spinæ. The clonic spasms may be symmetrically distributed on the two sides of the body: sometimes they are isochronous, but as a rule there is a short interval between the spasm on one side and that of the corresponding

muscles on the other side. Voluntary movements are not interfered with; in some cases they inhibit the spasms, in others they aggravate or even evoke them in parts previously quiescent. The spasms disappear during sleep; they are increased by emotion and by physical fatigue. They may not be observed until the patient's clothes are removed, or they may be severe enough to move the affected limb.

Apart from the myoclonic spasms the motor functions are normal. There is no loss of power, and the affected muscles do not atrophy nor show any change in their electrical excitability. The superficial reflexes and the tendon-jerks are generally increased. The sphincters are unaffected. There is no disturbance of sensation. Any mental disturbance is quite exceptional.

Nothing definite is known regarding either the etiology or the pathology of paramyoclonus. The disease has started after an injury, after a severe mental shock, and during the weakness following an acute illness. A combination of the disease with epilepsy sometimes occurs; this suggests that possibly the cerebral cortex may be the seat of the lesion. Against such a hypothesis is the fact that the spasms may affect only one muscle, or only one portion of a muscle. A more probable view is that the spasms depend on some disturbance of function in the lower motor neurons, and, judging from the symmetrical distribution and the occasional isochronicity of the spasms, it may be that the cells of the spinal anterior-horns are the seat of the disturbance.

The prognosis of paramyoclonus as regards cessation or remission of the spasms is most unfavourable. Instances of recovery have been recorded, but it is doubtful whether they were genuine cases of the disease or merely cases of hysteria which simulated it.

CHAPTER IV.

THE TICS: HABIT SPASMS.

The term "tic," meaning a "twitch" or "jerk," is applied to a psychical disorder which is represented by a co-ordinated purposive act, performed for the fulfilment of a definite object. In many cases of tic the purposive act is one that involves the visible twitching of certain groups of muscles, as, for example, those of the face; the observed movement may then closely simulate that of a clonic spasm, for in both conditions there are rapid muscular contractions. In other cases the movement, though obviously purposive, is more deliberate and quite unlike that of a spasm; for example, the rotatory movements of the head, originally started by a frayed collar, which are continued long after the painful stimulus has been removed.

The essential difference, however, between a tic and a spasm is pathological rather than clinical, the etiology of the respective movements and their relation to various influences being often quite distinctive. The muscular twitching of a tic is a psychical stigma, a manifestation of cortical instability, which may be controlled or aborted by the exercise of the will. A spasm, on the other hand, is the result of irritation, usually either of the motor cortex or of some point in a spinal or a bulbo-spinal reflex arc; it does not depend on consciousness and cannot be produced or controlled by the will.

It must be admitted that, as already suggested at the beginning of this section, these distinctions often fail in the analysis of particular instances of the movements referred to. No better proof of such failure could be adduced than the difference of opinion which is shown by recent writers in respect to the classification of many of the spasmodic disorders we have considered, especially as regards facial spasm and spasmodic torticollis. Some authors include both of those conditions under the tics, others only spasmodic torticollis.

There is a universal agreement that a tic may be started by peripheral irritation, and that the reflexly induced movement only leads to involuntary and automatic repetition when the psychical condition is deficient or unstable; in other words, when certain nerve cells have less than normal control. A consideration of this pathology raises the question:—Can a spasm be reflexly produced if the nerve cells involved are perfectly healthy and of normal stability? Or, to put the question in another way: Can stimuli applied to afferent nerves produce muscular spasms if the centres are quite normal? Without attempting to answer this question it is obvious that the line to be drawn between some varieties of tic and of spasm is a very fine one. Further, it must not be forgotten that it is often impossible to discover any adequate cause for many so-called reflex spasms, and that a spasm, whatsoever its origin, may be transformed into a tic by the perpetuation of a morbid habit.

Etiology. The subjects of ti although frequently possessing more than average a' ty, usually betray in some way a want of mental stability. They are often members of a neuropathic stock, as shown by the presence in their parents or in other relations of hysteria, alcoholism, epilepsy, chorea, tics, neurasthenia, or even of organic disease of the nervous system.

The malady affects the sexes about equally. It may become manifest at any age after early childhood; most commonly it begins at the time of puberty. Its development is favoured by a lowered condition of the general health as a result of acute illness, of growing too fast or of other cause; and also by anything which tends to depress the nervous system, such as fright, worry or prolonged anxiety. Some cases appear to owe their origin to mimicry. Very rarely I have seen a tic develop from an attack of chorea; thus originating from an involuntary movement, and not, as is usually the case, from a voluntary one.

In many cases ties seem to be started by peripheral

irritation, as blepharospasm from conjunctivitis, twitching of the facial muscles from adenoids, or rotation of the head from a frayed collar; but perhaps it is more correct to say that a true tic is suggested rather than caused by a local discomfort.

Symptoms. Tics are classified under the headings of simple, convulsive, co-ordinated and psychical.

Fig. 129.—Photograph of a case of unilateral facial tic.

Simple tic. In this variety the abnormal movements are generally limited to a small group of muscles. The face is the part most commonly affected; there may be blinking movements of the eyelids, alternate elevation and lowering of the eyebrows, or twitching of the zygomatic muscles, with drawing of the angle of the mouth first to one side, and then to the other. Another

common movement is a sudden rotation of the head, or a lateral, a forward, or a backward jerk. The platysma is often affected, either alone or with other muscles of the neck.

Ties of the upper limb are manifested by a rapid shrugging of the shoulder, by flexion and extension at different joints, or by rotation of the limb. The lower limbs are less frequently involved; there may be a sudden stamping of the foot, or rapid kicking movements. Ties of the respiratory muscles sometimes occur; they are represented by sniffing, sobbing, coughing, and hiccoughing, and may be associated with twitching of the facial, trunk or limb muscles.

The characteristic movements of tic, which are well seen in the facial variety, occur suddenly and are of lightning-like rapidity; occasionally the movements are slower and quite voluntary in appearance, as seen in some of the head ties. As a rule the movements cease during sleep; they are increased by any excitement and if the patient is under observation, although they can usually be controlled by a great voluntary effort. The manifestations of the disorder are generally preceded by a desire to make the particular movement: this impulse may be restrained for a time, but in the end the patient is compelled to give way; he then suffers from a sense of depression and misery, because he has yielded to the impulse which he had determined to quell. In cases of long-standing the movements may take place unconsciously.

Co-ordinate tics. In all true ties the movements are co-ordinated, but in the variety under consideration they are more complex than in simple tic. In every respect the movements are identical with those of every-day life, except as regards their frequent repetition without obvious cause. A patient when walking may suddenly make a half-turn as if looking for something: another may perform stooping movements, whilst a third may repeatedly stop during his walk to rub the calf of one leg with the toes of the other foot.

Feeble-minded children are apt to repeat certain co-ordinated movements, and sometimes in a rhythmical manner; thus they may go through the movements of balancing or of jumping; they may nod or rotate the head in a vigorous manner or repeatedly hit it with the hand. Movements similar to those of a performing dervish have been observed, namely, sweeping rotations followed by rapid revolutions of the body.

In the affection known as *saltatory spasm*, alternate contractions of the muscles of the lower limbs occur when the soles of the feet are placed upon the ground, which may be powerful enough to throw the patient into the air. When the spasms are less severe the patient may hop or jump on the floor, and may be quite unable to stand still for an instant; sometimes the heels only are drawn up, the toes remaining on the floor. The contractions continue so long as the erect posture is maintained, and usually cease when the patient sits or lies down; but in some cases they can be made to reappear by pressing on the soles of the feet. Although generally limited to the legs, the spasms may extend to the muscles of the trunk and face; the arms are rarely affected.

Convulsive tics. This is a rare variety of tic characterised by four groups of symptoms, some or all of which are combined in the same case. The symptoms are:—Sudden, lightning-like muscular contractions, either localised or generalised, which are repeated without the slightest rhythm; uncontrollable utterances; impulsions to mimicry; and imperative ideas and obsessions.

The spasmodic movements resemble those of simple tic both in character and distribution. They tend however to be more grotesque and generalised, in many cases affecting nearly every muscle of the body. In the face exaggerated contortions may occur; and the head, the trunk, the limbs or any of their segments may be jerked and twisted in every conceivable direction.

The movements are not constant, they occur in attacks,

in each of which the contractions of one group of muscles is followed by that of another group, or both groups are in action simultaneously. The attacks are induced or aggravated by excitement, emotion and when the patient is under observation. Although often painfully conscious of his grimaces and contortions, the patient has but little voluntary control over them. Ordinary voluntary movements are not necessarily interfered with; they may prevent or arrest the spasmodic movements. Many patients are able to perform acts requiring the most delicate muscular co-ordination, such as writing, sewing or playing the piano.

Explosive utterances may accompany or precede the automatic movements. They consist in the emission of sounds, words or sentences which are entirely irrelevant to anything that is being said, or done at the time. In some cases the exclamation is one of blasphemous or of obscene words (coprolalia), in others there is an irresistible impulse to repeat like an echo (echolalia) or to imitate gestures (echokinesis) even when such mimicry may distress or vex those who witness it. Whatever the form of the uncontrollable impulse the patient has little or no power to prevent it, although sometimes an explosive utterance may be warded off by voluntary speech. Imperative ideas and obsessions may also form a part of the clinical picture; sometimes they occur without motor equivalents when they are called mental or psychical ties.

Psychical tics are characterised by an irresistible impulse to utter a certain word, or to count a certain number before performing a certain action. The patient may loathe or dread the utterance of some word, and may strenuously attempt to evade it, but it haunts him so persistently that he is compelled to emit it; the struggle against the impulse is often followed by depression and exhaustion. Some patients are unable to perform a simple action, such as unlocking a door or getting into bed without counting one hundred or some other number. Others feel compelled when walking to

touch certain objects, or to step on certain portions of the pavement.

A closely-allied condition is the insanity of doubt, in which the victim has the feeling that he has omitted to do something, or has done it inaccurately. Between the above-mentioned mental peculiarities and actual insanity there is no sharp boundary line, transitional cases bridging over the gap between imperative ideas—recognised as absurd—and real delusions.

Diagnosis. The movements of tic may be mistaken for those of chorea, of hysteria or of reflex spasms. In chorea the movements are inco-ordinate and erratic and are not suggestive of purpose. They are liable to exacerbations, but do not completely intermit. In tic there is a constant repetition of similar movements which occur in definite attacks with intervals of complete freedom. Moreover chorea tends to recovery, whereas tic is frequently incurable.

The spasmodic movements which occur in some cases of hysteria may be identical with those of tic, and for the differential diagnosis we may have to rely on the presence or the absence of hysterical stigmata, such as globus, anæsthesia, and contraction of the visual fields; the history and the general aspects of the case will also require a careful consideration.

We have already commented on the difficulty that may be experienced in the differentiation of a tic from a localised spasm; a consideration of the following points may be helpful in some cases. A reflex spasm is confined to some definite nerve area; it is uninfluenced by volition or by the patient's attention being drawn to it. The subject of tic, on the contrary, frequently presents some anomaly of volition. The performance of a tic is generally preceded by a desire to make the movement and by a feeling that if it is carried out mental satisfaction will ensue. The reverse, however, is the case, the movement is followed by dissatisfaction and misery. Distress also occurs if the patient is able to prevent the movements by an effort

of will. None of these sensations are experienced by sufferers from reflex spasms.

Prognosis. The outlook in a case of tic depends mainly on the age and the mental condition of the patient, and also on the duration of the affection. The longer the tic has lasted, the more likely it is to persist. Many of the simple tics of childhood pass away under the influence of good hygienic and educational treatment. The prognosis of tics which start in adult life is less favourable, and is particularly bad when there is much volitional debility.

Occasionally a tic of some standing has been temporarily, or even permanently arrested by a sudden and severe emotional shock, whether this entailed great joy or profound grief. The disorder does not involve any danger of life except as an indirect result of the pronounced mental symptoms that are sometimes present, which may lead the patient to commit suicide in order to obtain release from his sufferings.

Treatment. Although any causal relationship between peripheral irritation and a true tic is usually insignificant or impossible to determine, a careful search should always be made for the existence of a morbid local condition which, if it did not start the tic, might tend to increase the frequency and the intensity of the attacks. Thus the correction of an error of refraction, or the removal of adenoids may lessen the severity of a facial tic, even if it does not dispel it.

The chief defect, however, to be dealt with is the mental instability which underlies the manifestations of the disorder, and every method should be employed that will help to lessen such instability. Discipline, education and gymnastic exercises, together with attention to every other means that is conducive to a good condition of physical and mental health, are all of primary importance in the treatment of tic, for they tend to improve the power of self-control and thus serve as useful adjuncts to the carrying out of a special

method of treatment which has proved to be of considerable benefit in many cases.

The essential principle of this method consists in encouraging the patient to try to remain absolutely motionless like a statue, at first for a few seconds and later, as his voluntary power increases, for longer periods; and in teaching him exercises which will bring into action the muscular groups that are affected, as well as their antagonists. The séances, each of which, including both the period of immobilisation and that of the prescribed movements, must not last longer than twenty or thirty minutes, should be gone through several times daily. They should take place in front of a mirror so that the patient may be able to judge of the degree of immobility obtained, and to see and correct any mistakes and irregularities in the performance of the muscular exercises. This method should be regularly and perseveringly carried out for a long period, and must be continued for some time after the tic has apparently subsided.

In severe and rebellious cases, especially if the patient's mental state precludes the employment of systematic exercises, isolation in a nursing home for a few weeks combined with a course of Weir-Mitchell treatment is sometimes advisable.

Of medicinal remedies, arsenic given in gradually increasing doses is probably the most valuable. Aggravated cases of tic may be temporarily improved by a sedative line of treatment, consisting in the administration of the bromides, of valerian or of belladonna; a mixture containing the bromides and conium is sometimes beneficial. Contemporaneously with such sedatives nerve tonics may be given with advantage.

CHAPTER V.

SPASMODIC TORTICOLLIS.

This is a condition in which active spasm of some of the muscles of the neck causes an unnatural position of the head.

Etiology. Nothing very definite is known regarding the causation of this disease. It is most frequently met with in adult life, and more commonly in females than in males. The onset of the symptoms is often preceded by feeble health, depressing emotions, or exposure to cold; or by some injury, such as a fall, in which the neck muscles were subjected to a strain. Sometimes the affection seems to develop from a habit spasm; sometimes from excessive use of the muscles in particular occupations. Another exciting cause is some disorder of vision which leads to a strained position of the head. In many cases no adequate reason can be given for its development, nor can any neurotic tendencies be observed in the patient's relatives. In most cases, however, there is evidence of a neuropathic heredity, such as a family history of epilepsy, insanity or hysteria, or some trait of nervous instability in the patient himself.

Symptoms. The chief and usually the earliest symptom is an altered position of the head due to spasmodic contraction of certain muscles. Occasionally the spasm is preceded by pain, by uneasiness or by other subjective sensations. At first slight in degree, the spasm gradually develops in intensity, the maximum amount of which varies in different cases and in the same case at different times, being greater, for example, during fatigue and mental excitement than during rest and mental tranquility. Obviously the position of the head will vary according to the muscle or muscles involved; while the degree of head movement will depend on the intensity of the spasm and on its nature, whether tonic, clonic or tonico-clonic. The sterno-mastoid is most frequently affected, and it may be the

only muscle involved, but generally its contraction is associated with that of other muscles, especially the splenius and the upper part of the trapezius. The complexus, the scaleni, the recti and obliqui are also sometimes implicated, while contractions of the platysma, and the omo-hyoid may be associated with the spasmodic movements. The muscular combinations are numerous, and, inasmuch as the muscles involved contract in various degrees, the resulting movements of the head present considerable variations in different cases.

Fig. 130.—Spasm of the splenius (Duchenne).

Contraction of the sterno-mastoid turns the head to the opposite side, slightly raising the chin and drawing down the mastoid process towards the shoulder: contraction of the upper part of the trapezius causes slight rotation of the head towards the opposite side, while it draws the head backwards and depresses it towards its own side. Thus the action of the two muscles is similar and their association on the right side would cause marked inclination of the head towards the right shoulder, but the rotation of the head to the left would

not be greater than that produced by the action of the right sterno-mastoid alone.

The splenius inclines the head backwards and slightly rotates the face towards its own side. Hence spasms involving the right sterno-mastoid and the left splenius would cause extreme rotation of the head to the left, the face looking towards the left shoulder; whilst if the right splenius was also affected the head would be strongly retracted as well as rotated towards the left side. This retraction of the head, which is also produced by bilateral spasm of the deep muscles of the neck, is known as retrocollic spasm, and is always accompanied by raising of the eyebrows and transverse wrinkling of the skin of the forehead owing to the associated action of the occipito-frontalis muscles.

In most cases there is a tendency for the spasm to spread to muscles of the neck other than those in which it commenced, and sometimes it may involve the muscles of the arm, the face or even the lower jaw. When the arm is affected after the neck it is usually on the side opposite to the affected sterno-mastoid.

As already stated, the spasm may be either tonic or clonic, and frequently both kinds of spasm occur together. In unilateral clonic spasm of the sterno-mastoid the head is rotated by a succession of jerks owing to the alternating contractions and relaxations of the muscle. At first the contractions are separated by considerable intervals, but as the disease progresses the intervals become shortened until the contractions may number twenty or thirty in a minute.

Torticollis often gives rise to much general discomfort and sometimes to actual pain which may be severe and neuralgic in character. The affected muscles never waste; on the contrary, they may become hypertrophied; their electrical excitability is normal or in some cases increased. Atrophy of one side of the face is occasionally present. Many cases present features which characterise the tics. Thus the spasms are preceded by a feeling that they must be made in order to obtain comfort, whereas

they are followed by a feeling of remorse, and yet their repression, which, in the early stages of the affection, can be accomplished by an effort of the will, is accompanied by exhaustion and distress.

The course of the disease is usually chronic; it is characterised by exacerbations and remissions in the intensity of the spasms which often persist for many years or for the remainder of the patient's life. Some cases recover after a longer or shorter time. The disease has no tendency to shorten life nor to lead to any serious disorder. Sufferers from torticollis occasionally exhibit some form of mental disturbance, which may precede the onset of spasm.

Pathology. We have no knowledge regarding either the seat or the nature of the lesion. When a single muscle is involved the spasm may depend on an abnormal condition of the spinal or bulbar grey matter from which the nerves proceed, or possibly on irritation of the nerve fibres themselves. But when the spasm affects several muscles, which are physiologically associated in the production of a given movement, it seems likely that the cortical centres are implicated which preside over such movements; their cells may be congenitally weak and prone to discharge, and hence as the ætiology of the affection suggests, a very slight cause may be adequate to start the spasm. Such considerations have induced many authorities to regard the malady as a variety of tic.

Diagnosis. Spasmodic torticollis must be distinguished from fixed positions of the head, and from hysterical spasm; as a rule the diagnosis is easy. Of the causes leading to temporary or permanent fixation of the head may be mentioned;—permanent shortening of one sterno-mastoid by changes occurring in the muscle at or about the time of birth; pain and tenderness of the cervical muscles as in a so-called rheumatic stiff neck, and in cervical caries, in which disease the sterno-mastoid is often unduly stretched. The local signs of these conditions are usually conspicuous; moreover, the

muscles are tense, contracted or shortened on the side towards which the face is turned, whereas in true torticollis they are contracted on the opposite side.

Spasm of the neck muscles which is sometimes met with in hysteria is usually associated with spasm of the trunk muscles, and with other hysterical manifestations. Occasionally the diagnosis is difficult and may not be cleared up until the patient has been under observation for some time.

A lateral displacement of the head has been found to depend on a unilateral dislocation of the cervical vertebræ; in such a case the sterno-mastoid on the affected side would be flaccid rather than contracted.

Treatment. The directions previously given for the relief of local spasms and the ties are equally applicable to the treatment of spasmodic torticollis. If any cause for this affection can be found, its influence should be removed or counteracted. So far as possible the patient should lead a quiet life free from excitement, worry and undue fatigue.

Systematic regulated movements of the head, combined with massage of the affected muscles, are often beneficial. The patient should be encouraged to perform the various movements of the head several times a day; he should move the head forwards, backwards, laterally and rotate it first to one side and then to the other. Each of these movements should be executed a certain number of times, the number varying inversely with the degree of fatigue produced. This treatment may be supplemented by the application of a weak constant current, one pole being placed on the nape of the neck, the other on each of the affected muscles.

Sedative drugs, such as the bromides, conium, Indian hemp or morphia, will afford temporary relief. In a few cases the prolonged administration of a sedative has been followed by permanent improvement: in several cases, Bastian obtained this result by keeping the patient continuously asleep for a period of three or four weeks, by means of chloral. Colman mentions a long-

standing case of torticollis in which recovery followed deep etherisation repeated on several occasions.

Certain surgical measures have occasionally been attended by temporary or even permanent success, but on the whole the results are not encouraging. Kocher recommends the systematic division of the contracting muscles. In different cases he has divided the sterno-mastoid and the trapezius near the mastoid process, the complexus, the splenius and the inferior oblique, and in some of his cases the result was satisfactory. A more promising operation appears to be that of excision of a portion of the spinal accessory nerve on one side accompanied by section of the posterior primary division of the upper cervical nerves on the opposite side.

CHAPTER VI.

CHOREA.

Chorea, a common disease of childhood, is characterised by involuntary, irregular spasmodic movements, and often by muscular weakness and by psychical changes. It has a close relation to acute rheumatism and to endocarditis.

Etiology. Chorea is a disease of the middle and later periods of childhood, nearly eighty per cent. of the cases occurring between the ages of five and sixteen. It is rare under five and after twenty, except when associated with pregnancy; a special form is occasionally met with in older persons. Girls are attacked nearly three times as often as boys, and between the ages of twenty and thirty chorea is almost limited to females. Hereditary influences are prominent in a special group of cases called Huntington's chorea (see p. 427), but they are not common in ordinary chorea; occasionally the parents have suffered from chorea, or there is a family history of epilepsy or of insanity. But a family history of rheumatism is commoner than one of a neurosis. It is probable that chorea is particularly apt to occur in

families which show a disposition towards both rheumatic and nervous affections.

The *relation of chorea to rheumatism* is very close; indeed there are good reasons for believing that it is almost invariably a rheumatic manifestation. Thus chorea is a very common sequel of acute rheumatism in childhood, and even when there is no history of a previous rheumatic attack there is frequently evidence of some rheumatic taint, such as the presence or the pre-existence of subcutaneous nodules; of erythematous eruptions often, with slight pyrexia; of tonsillitis; or of slight pains in the limbs, sometimes in association with effusion into one joint, or into tendinous sheaths near a joint. Chorea may also immediately precede acute rheumatism, and if the subsequent history of choreic patients is ascertained it will be found that they are more prone than other children to suffer from rheumatic attacks. Chorea occurs also during the course of acute rheumatism in young adults as well as in children, and especially at the onset of pericarditis. The frequency with which signs of endocarditis are met with in cases of chorea is another important link connecting this disease with rheumatism. Lastly, it may be noticed that when, as occasionally happen, chorea occurs during convalescence from scarlet fever there is nearly always an associated joint affection of rheumatic type.

Relation to emotion. An attack of chorea is sometimes preceded by some form of emotional disturbance, especially fright. The interval between the sudden alarm and the outbreak of chorea may be a few days, or only a few seconds, as in a case mentioned by Gowers, in which the symptoms of chorea began immediately after an unexpected pistol shot close to the ear. In such cases, however, it will often be found that some signs of chorea had been previously noticed.

Relation to pregnancy. Pregnancy is a prominent factor in the chorea of young women, especially when the pregnancy is associated with much emotional

disturbance Chorea occurs most commonly during a first pregnancy; the third month is the period at which it usually begins. It may recur during a second and even during a third pregnancy. Very rarely chorea has developed shortly after an abortion, or after parturition at the full time.

Relation to hysteria. Spasmodic movements closely resembling those of chorea sometimes occur in hysteria, and hysterical phenomena may co-exist with symptoms of chorea. It is probable that most of the "epidemics of chorea" are really made up of hysterical cases, and it is doubtful whether true chorea ever spreads by imitation.

Symptoms. The characteristic movements of chorea may be the first signs of the disease, or they may be preceded by an obvious change in the character and disposition of the child, who frequently becomes inattentive and apathetic, or fretful and discontented. In some cases a dragging of one leg, or a tendency to let objects fall from the hand, is the first indication of the disorder; such symptoms are due partly to irregularities in the contractions of muscles and partly to actual muscular weakness. The muscular twitching generally begins in the face, or in the hand, and subsequently may spread to the trunk and legs; in severe cases all the voluntary muscles may be implicated.

The clinical picture of a well-marked case is as follows:—The face exhibits every variety of contortion. The brow is knit and immediately expanded; at one moment the eyebrows are raised, at the next they are lowered; the eye is alternately opened and closed; the eyeballs are rotated; the angles of the mouth are drawn outwards and as quickly retracted, or the mouth is quickly opened and then closed. The result of these rapidly alternating movements is a quick succession of different facial expressions, such as those of delight, surprise, vexation and anger. The tongue is jerked out of the mouth and quickly drawn in, and is curled and twisted in every direction. The head is jerked suddenly

from one side to the other, or is thrown back. The gesticular agitation of the upper limbs is also remarkable; the shoulder is shrugged, or the whole arm is jerked forwards or backwards, and every possible movement at the elbow and shoulder joints may be performed; the hand is alternately supinated and pronated; at one moment the fingers are irregularly extended, at the next they are flexed. The disorderly contractions of the trunk muscles produce writhing movements of the body and sudden lateral and antero-posterior deviations of the spine; these movements may be violent enough to throw the patient from a chair, or out of bed. Jerky movements of the respiratory muscles also occur, one inspiration may be excessively deep, while another is shallow or abortive, or alternations may be observed between abdominal and thoracic respiration. When the lower limbs are affected the momentary contraction of groups of muscles causes various movements at the different joints, such as eversion and inversion of the feet and various contortions of the toes; in walking the legs are thrown hither and thither, they twist and rotate and cause the body to be jerked first to one side and then to the other.

Speech is often impaired; this is due mainly to disturbance of the movements concerned in articulation, and partly, in some cases, to irregularity in the movements of the vocal cords, or to the disturbance of respiration. Words are uttered quickly, or jerked out in separated parts, while a sentence may be interrupted by a hasty inspiration. An attempt to speak is often accompanied by an increase in the convulsive action of the facial muscles; in severe cases the speech becomes so disordered as to be almost if not quite unintelligible: very rarely there is a true aphasia. Spasmodic contractions may affect the muscles of mastication and deglutition and cause these functions to be imperfectly performed; the patient may be quite unable to feed herself or to swallow the food that is put into her mouth.

The successful execution of a voluntary action by a

choreic patient is not necessarily interfered with; by an effort of will the patient may be able to keep the outstretched hand quite steady, to pick up a small object from the floor, or to write a few words with normal precision. In many cases, however, a voluntary movement cannot be accurately performed. As a rule the inaccuracy is due to a lack of power to adequately inhibit the choreic movements; sometimes it appears to depend upon a disability to co-ordinate the muscles necessary for the movement, or upon actual weakness of some of the muscles. It must be admitted that it is not always easy to distinguish between the effects of spasm, of incoordination, and of paralysis. When the spasmodic movements are very slight, a marked delay in the relaxation of a set of muscles necessary for the completion of some action may be easily recognised; sometimes the relaxation occurs suddenly, as when an object is involuntarily dropped from the hand. In the latter case the sudden relaxation of the muscles may be equally well put down to transient paralysis as to incoordination; certainly some degree of muscular weakness is very common in chorea. As a rule the weakness is most obvious in the limb that is affected by the spontaneous movements, but it may be detected in a limb that is free from them. It may precede the onset of the movements, or may develop after they have subsided; thus the sole expression of the disease may be slight paralysis of a limb, and very rarely it is neither preceded, attended nor followed by any choreic movements, or they are so slight as to escape notice. In fact no constant relation can be traced between the severity of the spontaneous movements and the degree of muscular weakness; the former may be marked when the latter is slight, or *vice versâ*.

Paralysis may be monoplegic, hemiplegic or paraplegic in distribution; most frequently it is limited to one arm. It is of the flaccid type; the affected limb is limp, its muscular tonus is diminished; atrophy of the weakened muscles rarely occurs. As a rule the paralysis is slight, occasionally it is considerable, and

very rarely it is complete; but in whatever degree it is rarely, if ever, permanent. In one case of chorea reported by the author paralysis of the dorsi-flexors of one ankle with consequent foot-drop was still in evidence four years after the onset of a well-marked paraplegia. In this case the affected muscles reacted sluggishly to both galvanism and faradism. In many cases of chorea, however, the electrical irritability of the muscles and nerves is increased; an altered qualitative response to galvanism has also been observed, ACC being equal to instead of less than KCC, as in health.

Sensory symptoms. Choreic patients often suffer from headache, especially in the early stage of the disease, when they may also complain of pains in the limbs and joints. Anæsthesia does not occur, except as a result of peripheral neuritis or of hysteria, either of which conditions may occasionally be present.

The reflexes. There is a need for more accurate knowledge regarding the condition of the reflexes in chorea. In my own experience the deep reflexes are normal or diminished, rather than increased; they may be abolished when paralysis is prominent. Oddo, who examined the knee-jerk in one hundred and forty-seven cases of chorea, found it normal in thirteen per cent., increased either on one or both sides in twenty-five per cent., and similarly diminished in sixty-two per cent. of the cases.

In studying the reflexes in chorea it is important, as Oddo points out, to observe that choreic movements are often excited by percussion of a muscle or a tendon and whether the knee-jerk is absent or increased. After percussion there may be a few seconds delay before a movement of the knee, either flexion or extension, is produced, and the contraction may spread to other muscles and sometimes to the other limb. If the movement thus produced is flexor it may counteract a true knee-jerk, if extensor it may simulate a knee-jerk which is really absent. Thus a pseudo-reflex may take the place of a true one, it may mask it, it may also follow it.

These pseudo-reflexes or involuntary movements excited by percussion of a tendon may explain the phenomenon known as the *sustained knee-jerk*, in which the leg instead of dropping immediately after its brisk forward movement is held suspended in the air for a second or two.

The condition of the superficial reflexes is also variable; frequently they are brisk, sometimes they are diminished. In cases of hemichorea diminution or loss of the abdominal reflexes on the affected side may be found in association with an extensor plantar response.

Mental condition. Some variety of psychical disturbance is frequently observed during the course of the malady, but its degree bears no relation to the severity of the other symptoms. The disturbance may take the form of mental dulness which, usually slight or moderate in degree, may amount to actual dementia. In some cases failure in the power of attention is a marked feature. In others mental irritability passing into wild delirium or even mania, is observed, the patient singing and shouting incoherently; this condition is sometimes called "chorea insaniens."

Heart disease is very common in chorea. It may be due to a previous attack of acute rheumatism, or it may develop during the course of the chorea. Many patient present no signs of cardiac disease, but if their subsequent history is followed a certain number of them become affected with it. Moreover, in about ninety per cent. of fatal cases of chorea there is post-mortem evidence of valvular disease. As a rule the mitral orifice is affected; it may be stenosed, or enlarged in consequence of weakness of the muscular wall of the ventricle. In some cases this weakness is due to anæmia; in others to muscle failure through inability to cope with the effects of the mitral stenosis. Pericarditis may accompany the endocarditis, and is especially common in fatal cases.

Temperature. In the ordinary type of chorea, even when the movements are severe, the temperature

generally remains normal. The occurrence of pyrexia is usually due to the presence of arthritis or of endocarditis. Very rarely hyperpyrexia occurs in association with endocarditis and delirium.

Varieties. *Chorea gravis.* In most cases of chorea the twitching movements are slight or moderate in degree. In some cases they are very violent and seem to involve every muscle of the body, when it may be quite impossible for the patient to effect any purposive movement. In this severe type the patient has to be fastened in bed, where she incessantly writhes and tosses about: the expression is wild and restless, the speech is unintelligible, herpes may form on the lips, while the constant friction of the limbs and trunk against the bed leads to erythematous patches, to abrasions and even to ulceration of the skin over bony prominences.

Hemichorea. In some instances the movements are exclusively limited to the limbs on one side: the muscles of the face and the trunk, however, are always bilaterally affected.

Paralytic chorea. As already mentioned, loss of power in a limb may be the only prominent symptom of chorea, the arm being more frequently paralysed than the leg. The weakness usually comes on gradually: there is hypotonia, and the deep reflexes are generally abolished.

Pathology. Slight changes in the nerve cells, dilatation of vessels, with exudation or hæmorrhage around them, and small spots of softening in association with embolism have been found in different cases, but none of these changes are peculiar to chorea. The most constant morbid condition is endocarditis, even when no signs of rheumatism, nor audible murmur, were present during life: pericarditis also may be present.

It is clear then that in order to form an opinion regarding the *situation* of the lesion in chorea we must rely mainly on a consideration of its clinical manifestations. Such a consideration leads to the conclusion that

the main seat of the disturbance is in the cerebral cortex. This is indicated by the frequent excitation of an attack of chorea by fright or by some other form of emotion, by the mental condition of the patient during an attack, by the cessation of the movements during sleep, by the effect on the movements of attention and of volition, and by the phenomena of hemichorea and of hemiparesis. The spasmodic movements—the essential features of chorea—indicate that the motor centres of the cortex are mainly implicated. That the cerebellum may be also affected is suggested by the muscular

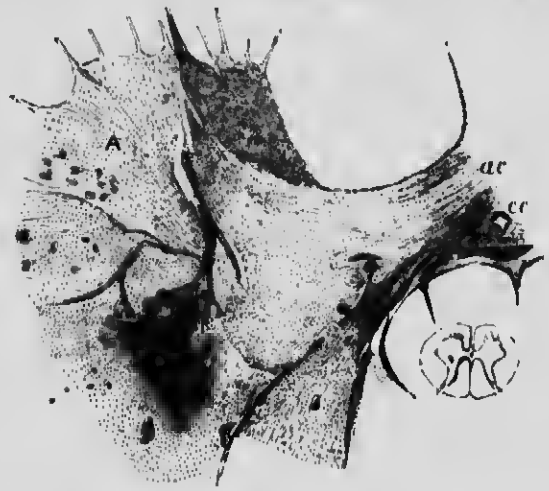


Fig. 131. Showing a small haemorrhage into the cervical enlargement of the spinal cord in a case of chorea that died on the fourth day of scarlet fever; cc, central canal; ac, anterior commissure; A, anterior horn.

hypotonus and the inco-ordination of movement which are frequently present.

As to the *nature* of the lesion, the close association of chorea with rheumatism and endocarditis, and its occasional development during pregnancy, suggest that the changes in the brain are the result of some poison circulating in the blood.

Fresh light has been thrown on the pathology of chorea by the investigations of Poynton, Holmes and Paine. These observers found vascular and inflammatory changes in the brain and its membranes, and also

in the nerve tissue itself, consisting of destructive lesions secondary to the vascular changes, and of alterations in the morphological character of the nerve cells; such changes were universal throughout the brain. The authors believe that they are due to the action of bacterial toxins, for diplococci, identical with those found in rheumatism, were found in the meninges and in the walls of the vessels. These organisms have also

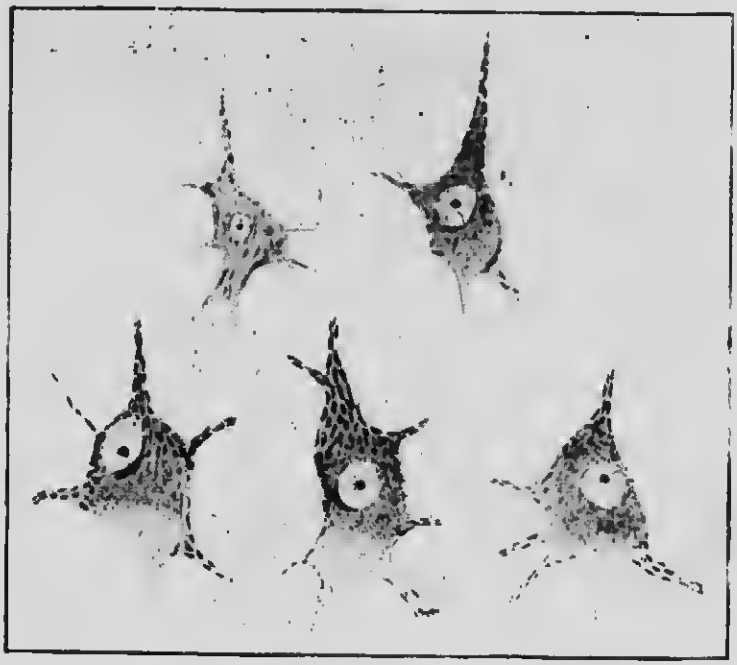


Fig. 132.—Two large pyramidal cells and three Betz cells from the precentral convolution, showing various degrees of chromatolysis. (Poynton and Gordon Holmes.)

been obtained during life from the cerebro-spinal fluid in cases of chorea.

There appears then to be every probability that the phenomena of chorea depend on multiple small cortical lesions which are set up by an organism that is also responsible for the arthritis and the endocarditis of acute rheumatism; and it seems reasonable to infer that the involuntary movements and the paresis which are

almost invariably present are due to the action of the bacterial toxins on the cortical motor cells, and that the variations in the condition of the deep reflexes may depend on the varying effects of those toxins on different parts of the nervous system. Inco-ordination of movement and hypotonia may be due to changes in the cerebellum: whilst the rare occurrence of paraplegia, or of paralysis in association with slight anæsthesia limited to the distal portion of a limb may be assigned to

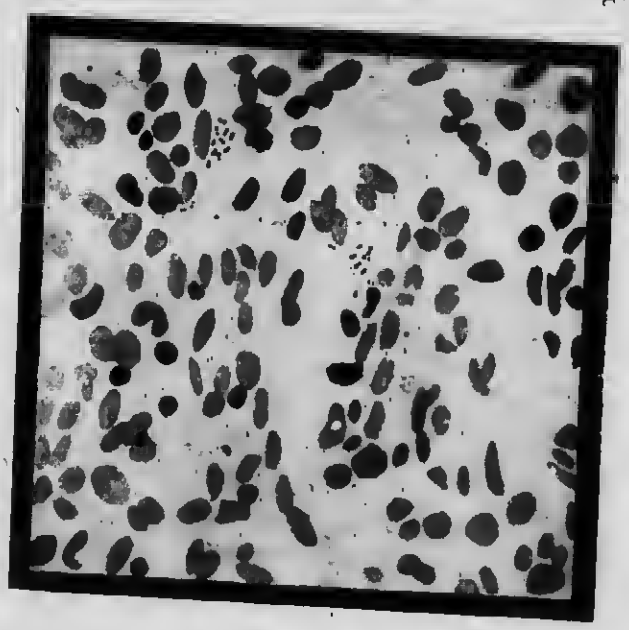


Fig. 133.—A film of pia mater showing the presence of two groups of diplococci. (Poynton and Gordon Holmes.)

changes in the spinal cord or to a peripheral neuritis respectively.

Diagnosis. This as a rule presents no difficulty. Sometimes similar movements to those of chorea occur in hysteria, but there are usually other indications of this disorder or a history of imitation. The movements in hysteria tend to be purposive in aim, those of chorea to be purposeless: the former are increased when the patient is asked to control them, the latter may be diminished or arrested by an effort of will.

When the muscular spasm is slight, and the mental symptoms assume the form of maniacal delirium, chorea may be overlooked, and the case may consequently be regarded as one of acute mania: this disease, however, usually attacks older subjects, and is characterised by more continuous gibbering, than occurs in the delirium of chorea.

In paralytic chorea involuntary movements may be only slight or even absent, but the usual limitation of the paralysis to one arm and its gradual onset in a child is strongly suggestive of chorea. In all cases of paralysis when, owing to the absence of spontaneous movements, the diagnosis of chorea is doubtful, the patient must be carefully watched for the appearance of any such movements, and especially when the arms are held above the head or straight out in front of the body; it is important to remember that paralysis, either limited in distribution or widespread, may exist for some time, even several weeks, before the movements make their appearance.

In infantile hemiplegia the paralysed hand may be affected with slight involuntary movements closely resembling those of chorea; but as a rule the limbs are spastic, and possibly smaller than their fellows.

Prognosis. The duration of chorea is uncertain, but it is common for the symptoms to disappear within two months. In children, even in severe cases, complete recovery is the rule. There is a tendency, however, for the disease to recur, and the patient may have four or five attacks during as many years; occasionally the movements never entirely cease in the intervals. The greater the number of attacks of chorea the more likely is endocarditis to occur.

The prognosis is serious in the chorea of pregnancy, especially in cases of abortion. It is also grave when there is pronounced psychical disturbance, and, although the patient may recover from the chorea, there is some risk that mental deterioration will ensue.

Apart from endocarditis, pericarditis, embolism and

hyperpyrexia, the chief danger to life is exhaustion from the incessant and violent character of the movements, and the resulting inability to take nourishment and to sleep.

Treatment. Except in the very mildest cases, rest in bed is essential, and even when the choreic movements are slight it is advisable for the child to stop in bed till noon, and to lie on a couch for an hour during the afternoon. Rest tends not only to shorten the duration of the attack, but also to prevent the development of endocarditis, or to cure it if it is already present. The patient should not be allowed to make any mental effort and should be protected from all sources of mental excitement; in many cases complete isolation is desirable, the patient being placed under the charge of a nurse, who, besides being thoroughly capable, is experienced in the management of children. Of equal importance to mental and physical rest is an abundant supply of nutritious and easily-digested food: cod-liver oil is also to be recommended. A daily tepid bath followed by gentle massage is often very beneficial.

As to drugs, there is every reason to believe that arsenic is of value; a dose of three minims of liquor arsenicalis should be taken thrice daily after food, and the amount may be gradually increased every few days until a dose of fifteen minims three times a day is reached. During the administration of the drug the patient must be carefully watched for any signs of poisoning, such as gastric irritation, pigmentation of the skin and peripheral neuritis.

In cases in which the symptoms of chorea have been preceded or are attended by definite rheumatic manifestations it is advisable to administer the salicylates or aspirin before putting the patient on a course of arsenic. It is useful to combine sodium bicarbonate with salicylate of sodium, as recommended by D. B. Lees. Aspirin, which is sometimes more efficacious than the salicylates may be given in ten to fifteen grain doses four times a day, the amount being gradually reduced as the

rheumatic symptoms abate in severity. Good results appear to have followed the use of other drugs, as, for example, large doses of quinine, phenacetin, antipyrin, chloretone, the sulphate or the bromo-valerianate of zinc, and the liquid extract of ergot.

In severe cases of chorea it is important to protect the body and the limbs from injury. As a rule it is desirable to place the patient on a water-bed and to bandage the limbs in flannel; the legs should be bound together and the arms bandaged to the body, while the trunk and hips may be steadied by placing a blanket across them and fastening it to the sides of the bed. A warm wet pack has often a soothing effect; the child is stripped, and then wrapped in a sheet wrung out of warm water, the sheet being surrounded by a blanket and a mackintosh. After lying in this pack for from thirty to sixty minutes, the child should be quickly dried and then wrapped in warm flannel. Liquid and pultaceous nutritive food should be frequently given in small quantities at a time; a little whisky in milk may be required if there are signs of exhaustion. When the movements of the head are very violent it may be impossible to administer food except by means of a feeding-bottle, or of a tube passed through the nose.

The drug treatment of the severest cases of chorea is not very encouraging. On the whole a mixture containing the bromides and chloral in sufficient doses to promote sleep seems to answer the best; or chloral may be given by itself. These drugs must be at once suspended should there be any signs of cardiac depression. The subcutaneous injection of hydrobromate of hyoscine, or of hydrobromide of scopolamine is sometimes useful.

During convalescence cod-liver oil, iron and strychnine are beneficial. Regulated exercises, baths followed by massage, and fresh air and sunshine are also of great value. For a long time frequent lying down on the couch must be enforced.

CHAPTER VII.

HUNTINGTON'S CHOREA: HEREDITARY CHOREA.

The first comprehensive description of this rare disease was given by Huntington in 1872. The affection is characterised by the appearance in middle life of involuntary purposeless movements, in conjunction with a progressive mental deterioration.

As a rule the symptoms are first manifest between the ages of thirty and forty, but they may develop at a later period. There is a tendency for several members of the same family, including one of the parents, to be affected. The disease has been traced back through four or five generations of ancestors; should it miss a generation it is unlikely to manifest itself in a subsequent one.

The spasmodic movements resemble those of ordinary chorea, although as a rule they are slower and less jerky in character, and are associated with more incoordination. At first they are slight, and may be limited to the hand or the face; gradually they become severe and may involve almost every part of the body; even the ocular muscles may be implicated, causing rolling movements of the eyeballs. The face shows involuntary grimaces, the tongue is contorted, whilst the limbs may be thrown about in every possible direction. The speech becomes hesitating and slurred; words and syllables are badly pronounced and are liable to be interrupted by inspiratory and expiratory noises.

In the early stages of the disease the involuntary movements may be suppressed by an effort of will or by the execution of a voluntary movement, but in the advanced stages the movements are usually uncontrollable and may cease only during sleep. Incoordination is often a prominent feature as shown by the attitude and the gait. The patient stands and walks unsteadily, and in walking may lurch from side to side like a drunken man; after the disease has lasted several

years walking may become impossible. Ultimately the legs may show slight spasticity, with exaggeration of the knee-jerks, probably the result of disease of the pyramidal tracts.

Associated with the motor disturbances there is a gradual deterioration of the mental faculties. At first there may be depression or irritability with outbreaks of excitement, together with a tendency to suicide, but sooner or later complete dementia supervenes.

The malady does not necessarily shorten life, but death may be hastened by the exhausting effects of the movements and by the indirect influence of the psychical condition.

The pathology of hereditary chorea is obscure; definite changes have been found in the brain, but none of them can be regarded as pathognomonic. Perhaps the most constant morbid condition is a chronic diffuse encephalitis frequently occurring in the form of a scattered miliary sclerosis and often associated with a chronic pia-arachnitis. In other instances there appears to be a primary parenchymatous degeneration of the cortical cells. The former condition may be the result of vascular disease; in some cases thickening of the vessels has been observed. The latter may be explained on the assumption that the ganglionic cells are genetically weak and are therefore prone to early degeneration. It is impossible to say which of these two main pathological findings represents the essential part of the disease.

SENILE CHOREA.

The irregular spasmodic movements which occur in this affection resemble those of other forms of chorea, but they first appear in old age, being rarely met with before the age of forty-five. Sometimes they come on after a period of mental anxiety, or after a sudden fright. As a rule the movements continue till the patient's death; the disease shows no tendency to shorten life. Senile chorea differs from the juvenile variety in the absence of any relation to rheumatism or

to endocarditis; it is distinguished from Huntington's chorea by greater freedom from mental changes, by the occasional termination in recovery within a year or two, and by the absence of any hereditary or familial tendencies.

The morbid anatomy of the disorder is similar to that of hereditary chorea, a sclerosis of the cerebral cortex being found in association with wasting of the cortical neurons.

The treatment of these two rare varieties of chorea corresponds to that of the juvenile form, arsenic appearing to be of value in some cases, whilst the bromides and other sedatives often mitigate the severity of the incessant movements.

CHAPTER VIII.

TETANY.

The characteristic feature of this disease is a peculiar tonic spasm of the extremities, which lasts for a variable time and is generally symmetrical. The wrists are slightly flexed, while the attitude of the hand resembles that of the accoucheur, the fingers being approximated and flexed at the metacarpo-phalangeal joints and extended at the other joints; the thumb is extended and strongly adducted, its tip being applied to the radial side of the forefinger; the palm is made still more hollow by the approximation of its inner and outer borders. The ankles are extended, and the feet are arched and inverted; there is extreme flexion of the toes which sometimes overlap one another; the fore-part of the sole is often contracted so as to become distinctly concave, with a median furrow due to an approximation of its outer and inner margins. The hands and feet are usually painful, tender and swollen, and their dorsal surfaces may be red and shining.

The rigidity may spread to other parts of the limbs and, very rarely, to nearly all the muscles of the body. The elbows may be rigidly flexed, the knees extended, and the arms and thighs strongly adducted; the

abdominal muscles may be tense and rigid, whilst spasm of the diaphragm and of the thoracic muscles may produce dyspnœa and cyanosis. The sterno-mastoids, the masseters, the facial and the ocular muscles are also occasionally implicated. Articulation may be impeded owing to stiffness of the tongue, and swallowing may be difficult from spasm of the pharynx. The spasms occur in paroxysms which after lasting from a few minutes to several hours gradually pass off, often

Fig. 134.—Attitude of the hands in a case of tetany.

incompletely, to recur after a variable interval of hours or days. In some cases and especially in young children a certain amount of spasm continues between the paroxysms.

The tonic spasms begin suddenly though the attack is usually preceded and is sometimes followed by tingling and numbness, or by burning sensations in the extremities. As a rule no objective disturbance of sensation can be detected; occasionally sensibility to touch and pain is impaired. The reflexes are generally normal; in some cases the knee-jerk is exaggerated, in a few cases it is diminished or even abolished. Very rarely slight

paralysis follows the spasm: atrophy of the small muscles of the hand has also been observed, whilst in rare cases general muscular atrophy has supervened.

In the intervals between the spasmodic attacks a remarkable increase of the mechanical and the electrical irritability of the motor nerves and muscles can be usually demonstrated. The mechanical irritability may be observed in the limbs, but is most frequently seen in the face, where a single tap over the trunk or the branches of the facial nerve, or even stroking the side of the face, will produce a momentary contraction of the facial muscles.

Fig. 135.—Attitude of the feet in tetany, the same case as fig. 134.

The electrical irritability of the nerves is also greatly increased to both the faradic and the constant current, and especially to the latter; frequently also a more ready response obtained is with the positive than with the negative pole. Moreover, a prolonged muscular contraction—tetanus—is usually obtained with either pole, and a feature peculiar to tetany is an anodal opening tetanus. The ulnar nerve is peculiarly liable to show this exaggerated irritability, while, curiously enough, the facial nerve, which shows exalted irritability

to mechanical stimulation rarely does so to electrical stimulation. Compression or percussion of a sensory nerve will also often induce a spasm.

During the course of the malady the pulse is often accelerated, and in severe cases the temperature may be raised.

Pathology. In the few cases of tetany that have been submitted to post-mortem examination, no definite lesions have been found in the nervous system. In rare instances, capillary hæmorrhages, collections of lymphoid cells around the blood-vessels and small foci of myelitis have been detected; changes in the ganglion-cells of the spinal cord have also been met with. But in other cases the most careful microscopical examination has failed to discover anything abnormal in the central nervous system, in the peripheral nerves or in the muscles. At present an opinion as to the probable seat and nature of the lesion can only be formed from a consideration of the causes and the clinical features of the disease.

While tetany may occur at any age it is most common in infancy and during early adult life. At the former period it occurs most frequently in males; at the latter in females, probably owing to the association of the disease with pregnancy, and with prolonged exhausting labours. When present in infancy tetany, which is often associated with laryngismus stridulus, is nearly always found in cases of rickets, and mainly in those cases where there are offensive stools or other signs of impaired digestion. In adults also it is apt to follow prolonged gastro-enteritis, and is frequently associated with dilatation of the stomach, especially when this depends upon the cicatrisation of a pyloric or a duodenal ulcer. Bouveret and Devic attribute the association of gastric ulcer and tetany to an excess of hydrochloric acid in the gastric juice, but cases have been recorded in which there was no such excess. It seems more probable that the poison exciting the tetany is derived from the decomposing contents of the enlarged stomach.

Tetany has developed during the course, or at the termination, of one of the specific fevers, especially enteric and cholera; epidemics of tetany have been observed in connexion with enteric fever. The disease has also occurred in cases of poisoning by chloroform, lead and ergot of rye. It has been met with in association with atrophy of the thyroid gland and with myxœdema; and has followed removal of the thyroid both in man and in animals, but only when the removal included the parathyroid glands as well. It is a recognised fact that when the thyroid is removed without the parathyroids tetany does not result, whereas the complete extirpation of the parathyroids alone is followed by tetany and death. Some authorities therefore believe that the tetanic spasms depend on insufficiency of the parathyroid glands which prevents an adequate neutralisation of poisons which have a tendency to injure the nervous system. It is stated that these glands have an influence on the metabolism of calcium, for the excretion of calcium is increased, and the amount of this body in the blood and the brain is diminished in cases of tetany produced by removal of the parathyroids. Further, it has been shown that a deficiency in calcium increases the excitability of the nervous system.

From a consideration of the above facts relating to the etiology and the experimental pathology of tetany it appears reasonable to infer that the disease is due to irritation of some part of the nervous system by the presence in the blood of a poisonous substance which arises under different conditions, and may vary in its nature in different cases. Moreover, there are good reasons for believing that the presence of the poison, which produces the symptoms of the disease, is in some way related to defects in the secretion from the parathyroid glands, defects which may be due either to a congenital or to an acquired insufficiency of these bodies.

As to the part of the nervous system which is primarily responsible for the peripheral spasms, there is much difference of opinion, the cerebral cortex, the

cerebellum and the spinal anterior horns each having its advocates. Against the cerebral origin of tetany is the frequent persistence of the spasms during sleep and chloroform narcosis. Then if the spasms depended on cortical disturbance we should have to admit that such disturbance, in many cases at least, is limited to the centres for the hand and the foot. It is also to be noted that the convulsions which sometimes occur during an attack of tetany do not appear to have any influence over it, the features of tetany persisting during the convulsive seizure; this apparent independence of the two conditions suggests that different portions of the nervous system are implicated.

There is a good deal to be said in favour of Hughlings Jackson's suggestion that derangement of the cerebellum may be the chief pathological factor in tetany, and as we have indicated at the beginning of this section, the effect of cerebellar impulses must be always taken into account when considering the pathology of any variety of muscular spasm. We have also drawn attention to the absence of any definite relation between spasm and disease of the spinal anterior horns. It is constantly stated that the spasms which are produced by poisonous doses of strychnine are due to the direct action of the poison on the spinal motor cells, but it is doubtful whether this has been satisfactorily proved. Houghton and Muirhead believe that in strychnine tetanus some resistance is removed to the passage of impulses between the posterior horns and the terminal fibres of the sensory nerve roots which surround the motor cells, but they consider it improbable that the poison acts either on the terminal fibres or on the motor cells.

In the year 1892 (see Treatise on Peripheral Neuritis) I made the suggestion that the phenomena of tetany may depend on an abnormal condition of the peripheral nerves. In support of this hypothesis it may be noted:— (1) That the distribution of the tetanic spasms is similar to that of the paralysis in multiple neuritis; the distortions of the hand, and feet in tetany are caused by

spasm, those in multiple neuritis by paralysis. Further, in the early stages of multiple neuritis I have seen a *spasmodic* attitude of the hands, identical with that of tetany. (2) That sometimes in tetany there is evidence that sensory fibres are implicated: this is shown by the presence of paræsthesia, of slight anæsthesia, and of an exaggerated excitability of the sensory nerves to mechanical and to electrical stimulation. (3) That the motor fibres also show increased irritability, and that occasionally in tetany fibrillary tremors and wasting of the muscles, together with paresis are observed.

But if nerve fibres rather than nerve cells are mainly selected by the toxin of tetany, it is clear that its action is not limited to the peripheral nerves for symptoms indicating disturbance of the central nervous system are also not infrequently present.

Diagnosis. It is only in aberrent cases that difficulties in diagnosis are likely to arise. Sometimes the typical spasms are very slight, and are apt to be overlooked, owing to the prominence of sensory symptoms, but a careful enquiry will usually elicit the previous occurrence of an attack of more marked spasm.

Hysterical contracture may closely simulate the spasm of tetany; the former, however, is generally unilateral in distribution, whilst the latter is bilateral. Increased electrical irritability of nerves, a characteristic feature of tetany, is rarely present in hysteria.

In a severe attack of tetany the spasms are sometimes widespread, involving the masseters and the muscles of the face, neck and trunk as well as those of the extremities, when the case may be mistaken for one of tetanus. The order, however, in which the muscles are attacked is different in the two diseases; thus trismus is generally the earliest manifestation of tetanus, whereas it is a late phenomenon in severe cases of tetany. In tetanus the fingers and hands escape; in tetany they are early and markedly affected.

Prognosis. Uncomplicated cases of tetany usually end in recovery. The duration of the disease varies

from a few hours to several weeks or months; the duration of the intermittent form tends to be longer than that of the continuous one. The tetany of pregnancy generally continues until delivery, that of lactation until the child is weaned.

Fatal results often occur in cases due to excision of the thyroid, or to dilatation of the stomach, although it is stated that the danger attending gastric tetany may be averted by the operation of gastro-enterostomy. The prognosis is grave in young children who are weakened by constant diarrhœa, and also in cases in which the spasms are severe and wide-spread, involving the glottis, or the diaphragm and other respiratory muscles.

Tetany is prone to recur if the subject is again exposed to one of its exciting causes. Occasionally the disease is associated with epileptic attacks, and very rarely with mental disturbances; in a few cases it has been immediately followed by chorea.

Treatment. The treatment of tetany consists in the employment of remedies for the relief of the contractures and for the removal or mitigation of any associated morbid condition which may be the exciting cause of the disease. The patient should be kept in bed in an equable temperature, all unnecessary movements being avoided; he should be fed with nutritious and easily assimilated food. Warm baths are often beneficial and they may be combined with the cold spinal douche.

When diarrhœa is present it must be treated with astringents or intestinal antiseptics, according to the nature of the case; it is often desirable to precede the administration of these remedies by a dose of castor oil or of calomel. Washing out the bowel with an enema of boiled water containing some antiseptic is also efficacious.

Where there are signs of gastric dilatation an emetic should be given; lavage also may be required, but this requires care, for the passage of the tube may excite a paroxysm. In severe cases of gastric tetany which do not respond to lavage, or in which the general condition

of the patient is alarming, the advisability of performing gastro-enterostomy calls for consideration: in several cases the operation has been followed by a satisfactory result.

When tetany follows excision of the thyroid the administration of an extract of this gland, or of the parathyroids is worthy of trial, although the results of such treatment have not been very successful. Moreover, if we admit that parathyroid insufficiency is the chief cause of tetany it is desirable to give parathyroid preparations in other varieties of tetany, after dealing with any gastro-intestinal disturbance or other ascertainable exciting cause. The relation of calcium salts to the excitability of nerve tissue is also suggestive. It is stated that the absence of the parathyroid secretion deprives the tissues of an adequate amount of calcium with the consequent development of hyperexcitability of nerve-tissue. If this be true the administration of some preparation of calcium, as the chloride or the hypophosphite, seems advisable, together with articles of food rich in calcium, such as milk, eggs and whey.

For the immediate relief of severe spasms full doses of the bromides may be required; chloral, and morphia have also been successful; in children chloral is best given by an enema. If there is spasm of the larynx, or of the respiratory muscles causing asphyxia, the inhalation of chloroform is indicated; when the stage of complete narcosis is reached the spasms generally relax.

During the intervals between the attacks of tetany, attention must be paid to the general condition of the patient, tonic treatment being usually required for some time; any tendency to gastro-intestinal disturbance must be carefully watched, and rickets must be combated by various hygienic measures, by suitable feeding and by the administration of lacto-phosphate of lime and cod-liver oil.

CHAPTER IX.

PARALYSIS AGITANS.

Paralysis agitans is a disease of advanced life which often appears to be started by severe emotional disturbance. Its chief characteristics are tremor, a peculiar attitude and gait, difficulty of movement, and certain subjectory sensory symptoms; its exact pathology is still unknown.

Ethology. Somewhat commoner in men than in women, the disease usually begins between the ages of forty and sixty, especially between fifty and sixty: occasionally the age of onset is under forty, or over sixty. Hereditary tendencies can rarely be traced. A brother and sister recently under the author's care were typical cases of the disease, and Erb has observed the disease in three sisters, but such instances are quite exceptional. In a few cases a simple tremor has been noticed in other members of the family.

The most important immediate antecedent is some form of emotional disturbance. Thus the general tremor which sometimes follows a sudden fright or alarm, and which usually subsides after the shock, may persist, or after subsiding for a time may return, the other symptoms of paralysis agitans subsequently developing. This sequence has been observed in persons who have witnessed a serious accident, such as a child being run over, or a fellow workman being killed by machinery; and in others who have been suddenly alarmed by hearing bad news, such as the loss of all their money, or the burning down of their houses or other property. No doubt in many of these cases pathological changes are already present in the nervous system which are stirred into activity rather than actually started by the effects of the sudden shock.

A similar remark may be made with regard to the influence of mechanical injuries or of overstrain, which occasionally appear to set up the disease; the sequence, however, is rare, and this alone suggests that some other

factor has already been acting on the nervous system. In a few cases the tremor begins in the injured part, and then something more than a coincidence may be inferred. Thus, in a painter tremor began in the left hand in which he had held his palette for many hours a day. In such cases a feeling of tiredness or of actual pain is often experienced in the overworked limb for some time before it becomes affected with tremor; frequently, also, the patient has passed through a period of much mental anxiety or has been subjected to a sudden mental shock.

In a few cases paralysis agitans has developed after an attack of acute illness; in others after exposure to severe cold and wet. Neither syphilis nor alcohol appear to have any influence in producing the disease.

Pathology. The changes found in the nervous system, examined by the most recent methods, are few and do not throw much light on the pathology of the disease. They comprise thickening of the walls of the small blood-vessels, perivascular sclerosis and a diffuse proliferation of the neuroglia connective tissue; such changes are usually more conspicuous in the cord than in the brain. They are also met with in the central nervous system of old people, though as a rule to a less marked degree than in paralysis agitans; hence some authorities regard this disease as a form of senility. The changes, however, are not constant; they may be completely absent both in cases of paralysis agitans and of senility; if inconstant they are not essential to the production of the disease. The symptoms are chiefly motor, and therefore must depend on changes in some part of the motor path. The presence of rigidity rather than of flaccidity of muscular tissue is much in favour of the upper neurons being the seat of the morbid process; whilst many of the clinical features indicate that the cerebral, and not the spinal portion of these neurons, is involved. Thus for a long time the symptoms are often unilateral or hemiplegic in distribution; the tremor ceases during sleep, and may be temporarily arrested by an attack of cerebral hæmorrhage.

If we assume that slight changes in different parts of the upper neurons may produce almost identical symptoms, it is possible that in some cases of paralysis agitans the dendrites of the motor cells of the cortex are implicated, in other cases the terminal branches of the pyramidal fibres in the spinal grey matter, or even the dendrites of the cells of the anterior horns. The possibility also that the influence of the cerebellum may be a factor in the production of the muscular rigidity must not be overlooked. The resemblance of some cases of paralysis agitans to the irritative variety of peripheral neuritis is striking, and suggests the desirability of making a careful examination of the peripheral nerves in all cases of paralysis agitans, in which a necropsy is obtained.

Symptoms. It is convenient to divide the clinical history of paralysis agitans into three periods: the period of invasion, the stationary period and the terminal period.

The period of invasion. In most cases tremor is the earliest symptom; it develops gradually, and at first may be only noticeable when the patient is excited; later it becomes constant. In other cases the onset of tremor is preceded by dull aching pains in the limbs, by transitory weakness or stiffness of a limb, by attacks of restlessness, or by unusual feelings of fatigue. It is especially common for rigidity of the thumb and forefinger to precede the appearance of tremor. In a few cases the onset of the disease is sudden, as when tremor immediately follows a mental shock and is persistent.

The order in which the tremor invades different groups of muscles is variable; most frequently it begins in one hand, especially the left, and then after a few weeks attacks the leg on the same side; at a later period the tremor extends to the limbs on the opposite side, the arm being affected before the leg. This early unilateral distribution is a characteristic feature of the disease. Occasionally the tremor begins in the leg or in the shoulder muscles.

The stationary period. In from one to three years the disease is fully developed, and the tremor becomes almost incessant although it varies in intensity. At first it is fine in quality, at a later period it tends to become coarse, and as the oscillations increase in range they lessen in frequency; an average rate for the movements, which are regular and rhythmical, is from five to seven a second.

The parts most conspicuously and constantly affected are the fingers and thumb; their oscillations produce movements which often closely resemble those necessary for the rolling of a pill or of a cigarette. Slight flexion and extension movements at the wrist, together with alternating supination and pronation of the hand, are often associated with the movements of the digits. Tremor of the shoulder muscles is occasionally present; it is prone to occur when tremor begins, or is most marked in the leg. Slight to-and-fro movements of the head are sometimes seen, but usually only when the disease is well advanced. Very rarely the eyelids, the facial muscles, the tongue, and even the palate, epiglottis and vocal cords are affected by tremor. The ocular muscles always escape. In one of my patients the lower lip is in constant up and down movements, and the masseters also show rapid contractions.

The tremor is often most marked when the affected part is supported. It may be checked by the exercise of the will, and is usually steadied by a voluntary movement. Thus the hand, trembling much when resting on the knee, becomes steady when a cup is grasped and conveyed to the mouth, or when a pen is held for writing. For a long time the writing itself shows no tremor, and even when the disease is advanced a lens may be required to detect any irregularities in the strokes; when wavy irregularities are apparent to the naked eye, they are finer than in other varieties of tremor. Writing, like all volitional movements in paralysis agitans, is slower than normal. In rare cases the tremor is seen only during the performance of a voluntary

movement; occasionally it is completely absent. The tremor is increased by emotional disturbance and by warmth: it is diminished by cold, by riding in a carriage, by a quiet life free from anxiety and excitement and it usually ceases during sleep, and during chloroform narcosis. The tremor in one limb may be exaggerated by arresting the movements in another limb.

Difficulty in movement, which as a rule develops gradually some time after tremor has been established, is due mainly to rigidity of the muscles and to the slow conduction of motor impulses, and sometimes to actual muscular weakness. Absolute paralysis does not occur, but diminished power to passive resistance is common, its distribution varying in different cases; thus the grasp may be weak or the flexor movements of one leg may be weaker than those of its fellow. As a rule weakness is most prominent in parts that are most tremulous; frequently it is more apparent than real, the difficulty in a movement depending rather on the slowness of its execution than on muscular weakness. There appears to be delay in the transmission of voluntary impulses, and the transmission seems to entail considerable effort and to engross the whole of the patient's will-power. Slowness of muscular action is well illustrated by the duration of the wrinkles produced in the forehead either voluntarily or by looking upwards. In a healthy person the wrinkles which result from looking up quickly disappear on looking down, but in paralysis agitans they often persist for some time. Another example of this delay in movement is sometimes seen in advanced cases of the disease when the patient tries to look at an object situated to his extreme right or left side: he turns his eyes quickly, but his head only very slowly.

Movements are still further hindered by the muscular rigidity which is one of the most characteristic features of the disease. To this rigidity may also be ascribed the typical attitude of the body, the peculiar features of

which are as follows:—The head and the upper part of the body are strongly bent forwards, the back being curved and rigid, making it difficult for the patient to raise the head or to turn it from side to side. The elbows are carried somewhat away from the trunk, and

Fig. 136.—Attitude in paralysis agitans.

are slightly flexed. The hands are held in front of the abdomen; the wrists are usually extended, but exceptionally they are flexed.

The position of the fingers and thumb varies; in the most frequent type it resembles that of tetany, the fingers being flexed at the metacarpo-phalangeal and

extended at the other joints, but the thumb, although extended, is not as in tetany pressed firmly against the index finger, but is opposed to it, making the attitude similar to that adopted in holding a pen or in rolling a cigarette. After a time the interossei may become permanently contracted and shortened, when it will be impossible to forcibly extend the metacarpo-phalangeal joints. The thumb may be over-extended at the phalangeal joint; sometimes its palmar surface becomes much flattened. In other cases the fingers are slightly bent at all their joints and are deflected towards the ulnar side; occasionally they are alternately flexed and extended at their several articulations so as to resemble the distortions of rheumatoid arthritis, but there is no grating on movement nor thickening of the bones, unless the two conditions are associated which is sometimes the case. The posture of the lower limbs is often quite normal; occasionally there is slight flexion at the hip and the knee, with adduction of the thighs. In advanced cases the feet may assume the position of talipes equino-varus, and the toes may be curled down towards the sole, with the exception of the great toe, which is either normal in position or is hyper-extended.

The muscles of the face, apart from trembling of the lower lip which is present in some cases, are motionless. The face has a mask-like appearance; its expression is a fixed one, being mournful, anxious or vacant, and is but little varied by any feeling of emotion. Sometimes the fixed expression resembles that of astonishment, the mouth being open, the eyebrows raised and the forehead marked by deep transverse wrinkles. The imitation is imperfect owing to the dull look of the eyes and the meaningless aspect of the lower part of the face. In other cases the forehead instead of being wrinkled is unusually smooth.

Very rarely other types of attitude are met with; for example, the trunk and limbs may be extended, the head being either bent forward or retracted.

The gait may remain normal for many years, but

sooner or later it becomes altered, and in many cases presents the following characteristic features: —The patient rises slowly and with difficulty from his seat and hesitates for a few moments before starting to walk. His body appears to move in one piece as if the joints were soldered together. Once started, his steps at first are short and are taken slowly, but the pace rapidly increases till the walk becomes a shuffling run and the patient is compelled to run forward in order to maintain his balance; this variety of gait is called propulsion or paralysis festinans. Some patients, if suddenly gently pulled from behind show a tendency to run or to fall backwards, although their bodies are inclined forwards; this phenomenon, called retropulsion, may occur spontaneously.

The voice is often affected; it is monotonous, high-pitched, and without modulation; sometimes it is weak and tremulous. Frequently there is some hesitation before beginning to speak, just as there is before beginning to walk, but when once started the utterance tends to be hasty and rapid as if the patient wished to finish speaking as soon as possible. The saliva often dribbles from the half-open mouth: the swallowing is unaffected except at a late period of the disease.

The reflexes. The superficial reflexes are unaffected and the plantar reflex is of the normal flexor type. The knee-jerks are said to be normal; in the author's experience they are more frequently exaggerated than normal; ankle-clonus is rarely present. The nutrition of the muscles may remain good for many years, but as the disease advances there is often a tendency to wasting; the electrical reactions of the muscles are unaltered or only slightly diminished.

The skin. Thickening with loss of elasticity of the skin is common in advanced cases of the disease; it may be impossible to pinch up a fold of the thickened skin, and when a fold can be raised its subsidence is much slower than normal.

Sensory and vaso-motor disturbances. As a rule the

cutaneous sensibility is perfectly normal; rarely hyperalgesia and analgesia are observed. Subjective sensations, however, are very common and often give the patient much distress. Pains of a rheumatic or a neuralgic character, or a tired, aching sensation may be complained of. In many cases cramps and a feeling of tension in the muscles, or other disagreeable sensations lead to much muscular restlessness which cause the patient to seek frequent changes of posture and often rob him of sleep.

Another very troublesome sensation is that of excessive heat. In order to obtain relief the patient has the windows thrown open even in cold weather; he wears the lightest garments in the daytime and throws off the bed-clothes at night. This sensation of heat is liable to remissions and exacerbations; an exacerbation frequently occurs after a paroxysm of trembling, and is often accompanied by profuse sweating and by flushing of the face. During one of these heat attacks the peripheral temperature may be raised one to two degrees, but the central temperature remains normal. In rare cases abnormal sensations of cold are experienced, or these alternate with sensations of heat.

Apart from attacks of depression, the mental faculties remain good throughout. The sphincters are unaffected and the urine is quite normal.

Terminal period. The duration of the disease varies much in different cases; as a rule it is from eight to twelve years, but in some cases it is twenty or thirty years. When the tremor and the muscular rigidity become intense the patient is unable to stand, and sooner or later he is compelled to keep his bed where he lies absolutely helpless. He becomes very thin and weak; bedsores may form over the sacrum, and he eventually succumbs either to general exhaustion or to some intercurrent disease, such as pneumonia. Even up to the last the intellect remains quite clear; occasionally, however it becomes weakened and there may be delusions and delirium.

Diagnosis. When the typical attitude, the rigidity and the tremor are present the diagnosis is easy. But when the symptoms develop in an irregular manner or when some of them are absent the diagnosis for a time may be difficult or impossible. Very rarely tremor is absent even at an advanced period of the disease, and then the diagnosis is based on a study of the expression, the attitude, the gait and the movements of the limbs. In other cases tremor remains unilateral for many months or years, while in a third series of cases tremor is alone present. In the last-mentioned variety we must rely on the peculiar features of the tremor, namely, its continuance during repose of the limb, its control by the will, and the pill-rolling movements of the fingers and thumb.

In some cases of paralysis agitans the earliest symptoms are aching and stiffness of the muscles with a feeling of weakness in one leg. Such symptoms might suggest involvement of the pyramidal tracts as from disseminated sclerosis; if, however, this were the case we should expect to find an exaggerated knee-jerk, an extensor plantar reflex, and slight though definite weakness of the flexors of the hip, or the dorsi-flexors of the ankle.

The tremor of old age is very similar to that of paralysis agitans, but the following differences may be observed:—The former is generally bilateral from the first, whereas the latter affects one side of the body before the other. The head and the muscles of the face are almost invariably affected by senile tremor and at an early period; they frequently escape in paralysis agitans, and when they are involved it is usually at a later period than the hand.

Prognosis. The very slow downward progress of the disease is often arrested for a time, and sometimes there are periods in which the symptoms distinctly improve, but actual recovery rarely if ever occurs. There is no special danger to life, but after a few years the patient becomes bedridden and then may sink from exhaustion or in consequence of some intercurrent malady.

Treatment. Although unable to cure the disease we may do much to relieve the tremor, the constant restlessness and the other distressing symptoms. The patient should lead a quiet life and be spared from mental excitement, anxiety and much physical exertion. Gentle exercise, however, such as a short walk or a game of croquet is often beneficial; even writing a letter or doing needlework may temporarily relieve the tremor. Passive movements, massage and warm baths are frequently of great service in maintaining the nutrition and in giving relief to many of the symptoms. The patient should live in the open air as much as possible, and in a climate which although bright and sunny tends to be cool and bracing, for the symptoms are often worse during very hot weather. When walking becomes difficult care should be taken to avoid the risk of a fall.

Of medicinal remedies the most efficacious drug is hyoscine hydrobromate which may be given either by the mouth or hypodermically, in doses varying from one-hundredth to one-fiftieth of a grain. A useful prescription is the one recommended by Williamson, namely, one-eighth of a grain of hydrobromate of hyoscine in six ounces of chloroform water. The patient should begin with a dose of two teaspoonfuls a day, which is gradually increased to six teaspoonfuls (one-sixty-fourth of a grain) providing no toxic symptoms are produced. One dose may be taken after breakfast and another at bedtime in order to abate any restlessness during the night. A few minims of liquor strychnine may be advantageously combined with the hyoscine. Sulphate of duboisin and sulphate of hyoscyamine are also recommended.

In some cases sleeplessness is a troublesome symptom, and it may be necessary to prescribe veronal, sulphonal or trional: the patient should lie on a firm mattress, and should be lightly covered. Restlessness and troublesome pains in the limbs are often relieved by small doses of antipyrin or of phenacetin.

CHAPTER X.

THOMSEN'S DISEASE: MYOTONIA CONGENITA.

This is a very rare condition to which attention was first drawn by Dr. Thomsen's description of the disease as it existed in himself.

In most of the recorded cases the disorder affected several relatives of the patient, and in some cases it appeared in successive generations. As a rule the symptoms were first noticed in childhood, and they gradually progressed during the period of muscular development. The disease is much commoner in males than in females.

The characteristic symptom is a peculiar stiffness of the muscles which comes on when they are put into action after a period of rest. Voluntary muscular contraction is slower than normal, and the ensuing muscular relaxation is also slow, hence the muscles whose action is opposed to that of the affected ones, are unable to overcome the contraction.

The rigidity limits the range of, or if severe prevents, any intended movement. The longer the muscles have rested the stronger their contraction at the beginning of a voluntary movement. Repetitions of the movement tend to lessen the intensity of the spasm, and the movement gradually becomes freer; sometimes indeed after the body has become warm by exercise the actions of the patient may seem to be as free as those of a healthy person. The spasm is but little influenced by cold, alcohol, or emotional disturbances; it tends, however, to be worse when the patient is much fatigued. The muscles are well developed or even hypertrophied; sometimes they appear to be unnaturally strong, sometimes weaker than normal.

The spasm is usually most noticeable in the movements of the legs, the stiffness and slowness of movement being well seen when the patient goes upstairs. The muscles of the arms, the neck and the trunk are also frequently

affected. Thus in grasping, the thumb and fingers are stiffer than natural, and their subsequent relaxation is difficult; if the patient, after lying on his back for a few minutes, tries to get up, his movements are noticed to be awkward and stiff, and his attitude in rising may resemble that of pseudo-hypertrophic paralysis, but if the act is repeated the patient rises better each time.

Fig. 137.

Fig. 138.

Figs. 137 and 138.—Retraction of the upper lids in a case of Thomsen's disease (Wardrop Griffith).

Fig. 137.—The patient was directed to look up forcibly and then at once to look downwards. The photograph was at once taken. It is seen that the upper lid has not come down with the eye as it normally does. The furrow at the upper part of the moveable portion of the eyelid is well marked.

Fig. 138.—He was then directed to look forcibly upwards and downwards repeatedly. At each movement of the eye downward, there was noticed a progressive improvement in the amount of the normal consensual downward movement of the upper lid. The photograph was taken after he had carried out about a dozen of these upward and downward movements.

Mastication may be interfered with by stiffness of the jaw-muscles, whilst in rare cases the tongue, the face or even the muscles of the eyeball, the larynx or those of deglutition may be involved. The orbicularis palpebrarum is tested by getting the patient to close his eyes tightly, and after half a minute to open them, when it may be noticed that they do not open so widely as in health. It is doubtful whether the acts of respiration, micturition, or defecation are ever interfered with.

No disturbance of sensation has been observed. The superficial reflexes are unaltered. The knee-jerk is often difficult to obtain at first, but repeated trials bring it out or make it more evident, and it may become unduly irritable. In many cases the affected muscles show increased irritability to mechanical and electrical stimuli and a more persistent contraction to the Faradic or the galvanic current than is obtained in health.

Specimens of excised muscle show an increased width of their fibres, which may be two or three times greater than normal. The transverse striation is feebly marked and the nuclei of the sarcolemma are increased in number. In only one case has a post-mortem examination been made; this is recorded by Dejerine and Sottas, who found the nervous system quite normal. Most writers regard Thomsen's disease as a malady essentially muscular in nature and congenital in origin. It is assumed that there is a congenital tendency for some of the muscle fibres to grow in an abnormal manner, and that this explains the abnormal muscular contractions. Similar, if not identical, contractions can be induced in animals by giving them phosphate of soda or veratria in poisonous doses. But it is at present impossible to say whether the disease is a congenital malady of the contractile tissue of muscle or whether the peculiar condition of the muscles is secondary to some derangement of the nervous system, such as a tendency to over-action on the part of nerve cells.

No treatment hitherto employed has exerted any good effect on the disease. Thomsen suggested that a life of active muscular exertion has a tendency to ameliorate the condition.

SECTION X.

Diseases of Obscure Origin Characterised by Disorders of Special Movements or of Muscular Function, which Occur Chiefly as a Result of Fatigue.

CHAPTER I.

OCCUPATION NEUROSES.

This term is applied to a group of maladies in which there is a defect in the performance of some particular action, the defect being apparently the result of an excessive use of the muscles which are required for the action. The disorder is one of a special movement and not of the individual muscles, the combined action of which produces the movement; for, when the same muscles are used for other movements, no disability in their action can be detected, except in very severe types of the disease.

In accordance with the generally accepted view that spasm is the most frequent factor in the observed disorder, it is customary to append the word cramp to each variety of occupation in which the disability occurs: thus we speak of writers' cramp, of pianists' cramp, of telegraphists' cramp and so on. In many cases, however, of occupation neuroses, weakness rather than spasm of muscles appears to account for the disability, whilst in others there is no obvious spasm or weakness of the muscles employed, and it may be difficult or impossible to give a satisfactory explanation of the muscular irregularities displayed during an attempt to perform the intended action. It is therefore more appropriate to use the term neurosis than that of cramp when speaking of the disabilities which attend the frequently repeated and prolonged use of certain groups of muscles.

WRITERS' NEUROSIS.

The facts relating to this disorder, which is a common one, are applicable, *mutatis mutandis*, to the other less frequent varieties of occupation neurosis.

Etiology. The disorder is one of the prime of life, its symptoms usually appearing between the ages of twenty and forty-five. It is much commoner in men than in women, solely because excessive writing falls more to the lot of the former than the latter. It is apt to occur in persons who have shown a lowered resistance to the effects of prolonged work or of much anxiety, or who have suffered or are still suffering from neurasthenia, migraine, neuralgia, or from other manifestations of a nervous disposition. Local disease or injury sometimes acts as a predisposing cause. In some cases there is no evidence of any neuropathic tendency nor of any cause except excessive writing the disorder arising in a person otherwise in perfect health. The neurosis is particularly common in clerks, and especially in those who are accustomed to write in a strained position. The mode of writing most prone to induce the disorder is that in which the pen is moved up and down by the muscles of the thumb and the first two fingers, the little finger or the wrist being the fixed point of support. On the other hand, the disorder is less likely to occur if the pen is held lightly and if writing is performed by a free movement of the wrist and forearm along the table, the arm and shoulder also participating in the action: the explanation being that fatigue is less readily produced when the muscles used for carrying out such a complex movement as that of writing are large than when they are small in size. The frequency of the disorder among clerks whose style of writing is usually very cramped may be contrasted with its rarity amongst shorthand writers whose style is free, the movement being generally from the shoulder.

Symptoms. According to the relative prominence of the leading symptom, four forms of this neurosis have been distinguished— namely, the spastic, the tremulous,

the neuralgic and the paralytic varieties. But such simple types are rarely met with, nor has the classification any practical value.

The onset of the symptoms is nearly always gradual. At first a sense of fatigue is felt at the end of a day's work, but before long it comes on much sooner, till ultimately it begins after writing a few words, and may compel the sufferer to desist. The feeling of fatigue gradually becomes associated with other sensory and with motor symptoms.

Painful cramps in the hand when writing may be experienced, which are often accompanied by aching pains in the bones or the joints, or by neuralgic pains along the course of the nerves; at first these pains occur only during the act of writing, but later they may attend the performance of any movement by the affected limb. Sometimes there is tenderness on pressure over the nerve trunks or their branches, as over the palmar twigs of the median and ulnar nerves.

The motor disability is often difficult to explain. It is stated that the most common cause of the disability is spasm, but the presence or absence of spasm is not always easy to determine. In the author's experience a careful examination rarely reveals anything beyond the fact that the patient has imperfect control over the muscles used in writing. As a rule the patient denies the existence of any cramp-like sensations about the fingers or thumb; but he frequently describes certain slipping-away movements either of the index finger or the thumb from the pen; these suggest a lack of sustained power rather than the presence of spasm. The strength of the muscles concerned in the movement, so far as it can be estimated is, however, unimpaired. Sometimes it appears to be less than normal, but distinct paralysis is rare. Slight wasting of the muscles may supervene, but this also is rare. The electrical excitability of the nerves and muscles of the affected arm is generally normal; when altered it is more commonly diminished than increased.

In some cases of writers' neurosis, probably in a small percentage, there is evidence of spasm. The patient finds that he is grasping the pen too tightly, and the muscular effort may be accompanied by pain. As a rule the cramps affect the flexor muscles of the first finger or of the thumb, so that one or other of these digits is drawn up the pen, causing it to be driven into the paper. In other cases the spasm is most prominent in the extensor muscles, when the fingers may be extended and separated so that the pen falls from their grasp. If attempts to write are continued there is a tendency for the cramps to spread up the arm and even to involve the muscles of the shoulder. The characters of the writing become irregular in form as well as in force, and splutters of ink may be seen on the paper where it has caught the point of the pen. To ward off the spasm the patient may hold the pen between the first two fingers and : : to write by moving the arm: spasmodic flexion of the wrist, which sometimes develops, may be counteracted by the patient placing his left hand under the wrist. As a rule the spasm is tonic, occasionally it is clonic when the first finger or the thumb may be twitched off the pen. Tremor, which rarely exists alone, may accompany the spasm and render the writing still more shaky.

In early cases of writers' neurosis the only movement deranged is that of writing: other actions, however delicate, can be readily and correctly performed. Thus the patient may be able to play the piano with ease, to paint, to button his clothes, and possibly to print letters with a pen when his ordinary writing is illegible. It is, however, scarcely possible that these acts entail the use of precisely the same neuro-muscular mechanism, otherwise they would surely be imperfectly performed, and it may be noted that certain acts, such as the winding up of a watch, or the pinning of two articles together, which presumably do involve the use of the writing muscles, are occasionally deranged. In severe cases of writers' neurosis other actions requiring delicate

co-ordination of the muscles of the affected limb may be clumsily and ineffectively performed.

Pathology. Cases of occupation neurosis may be divided into two groups. The one group is constituted by cases in which certain muscles that have been subjected to overstrain become weak and wasted, and give altered reactions to electricity; the atrophic weakness may or may not be associated with changes in the cutaneous sensibility. The other group comprises cases in which disabilities are exhibited, not in the performance of all the actions of the affected part as in the first group, but only in the performance of special work; the disability may be unattended by any detectable spasm or by weakness of the muscles brought into play by the special work, and, apart from pain, there may be no disturbance of sensation.

As examples of the first group may be mentioned a case of "sawyers' cramp," described by Vivian Poore, and a case of "drummers' paralysis," recorded by Bruns. In the former case the two muscles most liable to strain in the act of sawing—namely, the supraspinator and the pectoralis major, were slightly wasted and the nerves supplying them were distinctly tender. In the latter case the flexor longus pollicis, and, to a less degree, the adductor pollicis of the left hand were paralysed and atrophied; the flexor longus pollicis could not be excited to contraction by either the galvanic or the faradic current, and, as Bruns points out, this muscle of the left hand is kept by the act of drumming in a state of constant contraction.

It seems probable that the disability observed in the above cases and in the others belonging to the first group, although perhaps partly the direct result of the fatigue induced by the prolonged over-exertion of certain muscles, is mainly caused by a local neuritis, the presence of which is indicated by pains, paræsthesia, slight anæsthesia and muscular atrophy.

But when, as in the second group of cases, the disability is limited to a purely acquired and complex

movement, such as writing, while other movements remain normal, it is impossible to resist the conviction that nerve cells and not nerve fibres are the parts where the primary derangement is located, whilst the frequent association of this variety of occupation neurosis with neurasthenia suggests the probability that the nerve cells in the cerebral cortex are affected rather than those in the spinal cord.

Course and Prognosis. Although generally insidious in onset the neurosis may develop suddenly. In either case it tends to get worse so long as writing is continued, and to get better or remain stationary when attempts to write are given up. The symptoms are likely to be severe when the original mode of writing has been bad, and also when there is a general weakness of the nervous system. If the patient learns to write with the left hand, it also may become affected, though fortunately this is not always the case. If the patient can entirely abstain from writing the symptoms subside and may disappear altogether; but even after several months of complete rest a return to work is frequently followed by a relapse of the disability.

Diagnosis. The delicate action of writing may be interfered with by many affections in which the tissues of the hand or the arm are implicated, and by several disorders of the nervous system. It is therefore of great importance to make a complete examination of every patient who complains of disabilities in writing; the condition of the nervous system must be carefully investigated, as well as that of the affected limb, in order that any disease of its muscles, tendons, joints or nerves does not escape notice.

The tremulous form of the neurosis may be mistaken for an early case of paralysis agitans; in the latter disease, however, tremor affects other movements than writing, whilst the facial expression, the attitude and the gait usually present characteristic features.

In hemiplegia of gradual onset the patient's attention may be first drawn to his clumsiness in writing, but the

observer has rarely any difficulty in making a correct diagnosis. Brachial neuralgia, arthritis, teno-synovitis of the fingers or wrist and many other local disorders may interfere with the movements of the hand; their recognition, which is usually easy, prevents the liability of any error in diagnosis.

Treatment. Complete abstinence from writing is the essential element in treatment, and when, as is frequently the case, the disorder is associated with general nervous weakness it is also desirable for the patient to take a good holiday and to adopt every method calculated to increase his physical vigour.

If the symptoms have existed for only a short time, a three months' rest may cure the disability. On resuming work the amount of writing should be reduced as much as possible and faulty methods in using the pen corrected. A large penholder is desirable, and the patient should learn to write from the elbow or wrist.

If the symptoms are of some standing a long rest is still likely to be beneficial, though very often the resumption of writing is soon attended by recurrence of the old difficulty: when this is the case it is obvious that a change of occupation is advisable. When, however, no other occupation can be obtained, and a prolonged rest has not cured the disorder, the patient should be advised to learn to write with the left hand: by so doing he may be able to resume his former work whilst he gives his right hand the best chance of recovery. Unfortunately, it frequently happens that the left hand after a time also suffers in the same way, when the patient is compelled to give up writing altogether. A type-writer may then enable the sufferer to continue to earn his living, although there is no form of this machine which will do the work of a bank clerk or that of a cashier.

During the period of prescribed rest, benefit may be derived from the application of massage and galvanism to the affected limb, and also from the employment of carefully selected exercises for the fingers, the thumb and the hand: by these means the nutrition of the

affected parts is stimulated, and the muscles become stronger, effects which are likely to diminish the disability in writing.

Various drugs may be required from time to time for the relief of symptoms. Nervine tonics, especially strychnine, help to improve the nutrition of the nervous system, and along with the other methods of treatment aid in promoting a tendency to recovery.

CHAPTER II.

MYASTHENIA GRAVIS.

In this disease some or all of the voluntary muscles become rapidly exhausted by exercise or by stimulation with the faradic current. The muscles regain their power after rest, although in some cases a variable degree of paralysis persists; those supplied by the cranial nerves are particularly prone to be affected. A fatal termination is very common; anatomical changes have been found, but their exact relation to the symptomatology of the disease is not clear.

Etiology. Males and females suffer almost equally. The disease occurs most commonly between the ages of twenty and forty, and usually at an earlier age in women than in men. In most cases the patients have been manual workers. Sometimes the symptoms have followed an infective fever or other disease capable of generating a toxic agent, such as scarlet fever, typhoid, influenza, otitis-media and diarrhœa. Occasionally menstruation, pregnancy, parturition, emotional excitement, exertion or a chill appears to have acted as an exciting cause.

Symptoms. The most characteristic feature of this disease is the readiness with which muscular fatigue is produced. A voluntary movement is at first perfectly performed, but it becomes feebler with each repetition, and ultimately becomes impossible. Complete rest for a time restores the power to perform the movement unless the disease is very advanced.

This rapid development of muscular fatigue, called the *myasthenic condition*, is usually the first noticeable feature, but in a few cases it is preceded by headache, pain in the neck and back, giddiness and photophobia. As a rule muscles supplied by some of the cranial nerves are affected before those of the trunk and limbs. Bilateral ptosis is common; in severe cases it is constant, but in slight cases it only comes on in the evening or after the patient has been looking up for a time. To counteract the ptosis, the patient throws his head back and may wrinkle his forehead, but in many cases that is impossible owing to weakness of the frontalis The

Fig. 139.—A myasthenic patient attempting to look up. Note the unequal ptosis, the strabismus, and the drooping of the lower jaw (E. Farquhar Buzzard).

orbicularis palpebrarum rarely escapes; the slightest resistance to its action usually suffices to prevent closure of the eyes. When weakness spreads to the lower facial muscles all power of expression is lost, the face is mask-like and immobile and closely resembles that seen in some types of myopathy, or in double facial paralysis. The patient may be unable to close his eyes, to pout his lips or to whistle; on attempting to smile, the upper lip is curled and the expression has a sneering character. Weakness of the external ocular muscles, an early symptom, gives rise to diplopia and, owing to the

varying degree in which the muscles are affected, the relative position of the visual images is altered at different times; the weakness may progress to complete ophthalmoplegia externa. Easily induced fatigue of the ocular muscles is indicated by difficulties in reading: the patient, after reading a few lines, finds that the letters and words are becoming blurred and indistinct. Jerky nystagmoid movements may be present. The pupils usually remain unaffected and attempts to exhaust the sphincter iridis by exposure to a bright light or to fatigue accommodation have usually failed; the action of the ciliary muscle during accommodation is also unimpaired.

One of the most constant symptoms is weakness of the jaw muscles, which may be so marked that the mouth remains open, and in order to speak or to masticate food the patient has to support the lower jaw with his hand. Weakness of the palate and pharyngeal muscles also occurs, giving rise to a nasal voice, to regurgitation of liquids through the nose and to difficulty in swallowing; if the palate be watched during the repeated phonation of the sound "ah" its movements will be seen to become less and less marked. Articulation is often defective owing to weakness of the lips and tongue. Feebleness of the voice usually depends on weakness of the respiratory muscles, for the laryngeal muscles are rarely involved; in a few cases, however, the myasthenic condition has been observed in the abductors or in the adductors. A feeling of fatigue in the tongue after eating or speaking is sometimes experienced; it is often flabby, longitudinally furrowed and tremulous and the power of protrusion may be limited. The myasthenic condition is well shown when the patient reads aloud: at first the voice is strong and the words are distinct, but after a short time the voice becomes feeble, the speech nasal, and pronunciation indistinct, and finally the patient stops for want of breath.

Inability to support the head owing to weakness of

the neck muscles is a common symptom. The trunk muscles may also suffer, sometimes so severely that the patient is unable to alter his position in bed. Involvement of the respiratory muscles leads to dyspnœa on slight exertion, and alarming attacks with cyanosis are apt to occur and may prove fatal. The muscles of the limbs, especially those nearest the trunk, are frequently affected. Patients often complain of inability to lift the arms at night when brushing the hair or undressing, and of the need for frequent rests when walking; a sudden giving way of the legs may occur. Easily produced fatigue in the muscles of the forearms and hands is shown by the handwriting, which, good at first, gradually becomes slower and more and more illegible. The intensity of the symptoms varies much at different times, and the increase of the myasthenic condition as the day advances is a very striking feature. It is also exaggerated by cold and emotional excitement.

Especially characteristic of the disease is the *myasthenic reaction*, a name given to the readexhaustion of muscular contraction by faradic stimulation. When a strong current is applied to an affected muscle a normal contraction is obtained, but if the current is continued the contraction becomes feebler and ultimately ceases; the muscles, however, after a period of repose will respond again to the current. No altered mode of response to galvanism is observed.

It has been doubted whether localised muscular atrophy ever occurs, but recent records show that it is certainly present in some cases; thus atrophy of the tongue, of the temporal muscles, or of some limb muscles has been noted in what appeared to be uncomplicated myasthenia gravis.

Apart from feelings of aching and stiffness, sensory symptoms are rare. But, as pointed out by F. Buzzard, they may be prominent; thus at the onset or during the course of the disease sharp shooting pains, together with numbness and tingling in the fingers or toes, may be experienced; occasionally definite anæsthesia has been

observed. In one case recorded by Buzzard relative anæsthesia and analgesia was found along the ulnar borders of the arm and around the lower portion of the thorax, areas of skin which are so frequently affected in tabes; in this case there was no evidence, either clinical or microscopic, that early tabes was a complicating factor.

The knee jerks, normal or slightly exaggerated, share in the tendency to exhaustion, being often abolished by repeated tapping. The cutaneous reflexes are not affected: the functions of the bladder and rectum are normally performed. The mental state is almost

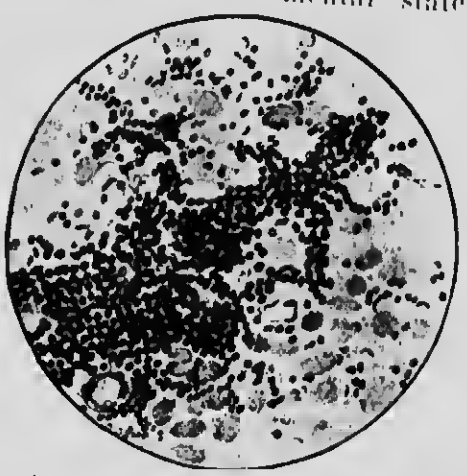


Fig. 140.—Section of ocular muscle, showing a large lymphorrhage in the midst of healthy muscle- and nerve-fibres (E. Farquhar Buzzard).

invariably normal, but very rarely mental abnormalities have been observed.

Pathology. From a careful study of five cases of myasthenia Buzzard believes (1) that the disease "has a definite and constant morbid anatomy constituted by the presence of widely distributed cellular, and sometimes serous, exudations in the tissues of the body." To these exudations he gives the name 'lymphorrhages,' for they consist mainly of clumps of cells which in appearance are identical with the ordinary lymphocytes of the blood; in one case he

observed small collections of lymphocytes between the nerve cells of the bulbar nuclei. The lymphocytes do not invade the muscle fibres, but lie scattered in the spaces between them; just as in cases of capillary hæmorrhage the tissues are infiltrated by red blood corpuscles. (2) That slight muscle-fibre changes, indicating an early stage of degeneration, are frequent and afford an explanation of the rare occurrence of muscular atrophy. (3) That proliferation and degenerative changes in the thymus gland are frequently but not constantly met with; this inconstancy indicates that the lymphocytic deposits are not secondary to the changes. (4) That the symptoms of myasthenia are best explained by assuming the presence of some toxic, possibly autotoxic, agent, which has a special influence on the protoplasmic constituents of voluntary muscle, and a less specialised influence on the functions of other tissues. Buzzard sees no reason for believing that any microbe is the causative agent of the disease; other writers, however, from a consideration of its etiology, suggest that the toxin is probably of microbic origin, and that it acts selectively upon the lower motor neuron, so as to modify its functional activity. (5) That the relation of the toxin to the incidence of lymphorrhages and to thymic alterations is not clear.

It is interesting to note that myasthenia is occasionally combined with symptoms of Graves disease and that hypertrophy of the thymus occurs in both diseases. Sometimes congenital anomalies, such as the presence of rudimentary digits, are found in patients suffering from myasthenia.

Diagnosis. In a well-marked case of myasthenia the character and the grouping of the symptoms render the diagnosis easy. Thus, in addition to a peculiar facial expression, ptosis and a nasal speech, we have weakness and ready fatigue of the muscles on exertion, the myasthenic reaction, together with absence of muscular atrophy and of anæsthesia. But it must be remembered that the myasthenic reaction is not always present, and

may occur in other diseases as in hemiplegia and neurasthenia; and also that, as already mentioned, muscular atrophy and anaesthesia are present in exceptional cases of myasthenia.

Mild forms of the disease may resemble cases of hysteria or of neurasthenia, in which also variations in the intensity of the symptoms—so characteristic of myasthenia—are a common feature. Hysteria is recognised by its emotional manifestations, by the usually marked characters of its sensory phenomena and by the absence of the myasthenic condition and the myasthenic reaction. In neurasthenia, although easily induced fatigue is complained of, there is no persistent muscular weakness. When muscular atrophy is present in myasthenia it may be difficult to eliminate one of the myopathies, and it is possible that a close relation exists between the two diseases. At the same time, a careful consideration of the history and the grouping of symptoms usually leads to a correct diagnosis.

In obscure cases of ocular paralysis where syphilis and tabes can be excluded it is important to examine the deltoid and other muscles for indications of the myasthenic reaction. From bulbar palsy the disease is distinguished by the great variability in the degree of muscular weakness from time to time, by the prominent affection of the ocular muscles, and by the absence of marked muscular atrophy.

A temporary difficulty may occur in distinguishing between the early symptoms of myasthenia and those of an occupation neurosis, for in both cases the local incidence of the muscular disability may be determined by the occupation; thus the commencement of myasthenia in a clerk may be declared by easily produced fatigue of the hand and fingers, in a teacher by impairment of speech or of mastication.

Prognosis. The malady is a grave and dangerous one; death is the usual termination and generally takes place within two years, most frequently in consequence of failure of the respiratory muscles. It is stated that

very rarely complete recovery may occur. Many cases show great improvement for a time and then relapse, and the symptoms of the disease are liable to marked fluctuations, so that it is difficult to form an opinion regarding the duration of the disease.

Treatment. The patient should avoid excitement, cold and fatigue. Except in the mildest cases absolute rest in bed is essential. If there is any difficulty in mastication or in deglutition the food should be minced, and it is desirable for the patient to take as much nourishment as possible during the early part of the day, that is, before the myasthenic state has reached its maximum. If sufficient nourishment cannot be swallowed the patient should be fed per rectum, as the introduction of a stomach tube may bring on serious symptoms. Gentle massage and galvanism may be tried, although their use has not proved of much benefit. Many drugs have been given, but without any decided beneficial effects; probably strychnine, either by the mouth or hypodermically, is the most useful.

Quite recently Pemberton, bearing in mind the close relation of calcium to the functions of muscular activity, has advocated its administration in the treatment of myasthenia. In a carefully investigated case in which he found a diminished output of creatinine and an excessive excretion of calcium, he gave the patient calcium lactate with beneficial results.

The administration of various glandular extracts has been recommended; in a few cases thyroid extract appeared to do good. Attacks of dyspnœa may sometimes be relieved or cut short by pulling forward the tongue, by performing artificial respiration and by the inhalation of oxygen.

SECTION XI.

Intermittent and Paroxysmal Neuroses.

The majority of these neuroses will be described in the following chapters: some of them, as tetany, chorea and Menière's disease, have been dealt with in other sections. They are characterised by the occurrence in definite attacks, of pain, unconsciousness, vasomotor disturbance, muscular spasm or paralysis: with a more or less normal condition of the patient between the attacks.

CHAPTER I.

NEURALGIA.

The term neuralgia should be restricted to paroxysmal pain in the course of a nerve when there is no evidence of disease in any part of the nervous system or in any of the viscera.

The character of the pain may be the same whether there is actual disease or not: thus the lightning pains of tabes, or the piercing pains through the chest produced by a disordered stomach, are identical in character with those of an uncomplicated neuralgia. The above definition is based not on the absence of disease but on the absence of *detectable* disease. It is therefore tentative and somewhat arbitrary: for the absence of symptoms or of visible pathological changes does not necessarily exclude the existence of disease. Moreover, with the progress of knowledge, it is reasonable to believe that the limitations of the term neuralgia will become more and more restricted, and, as Head suggests, that in all probability most neuralgias will cease to be regarded as diseases and will be recognised as symptoms either of lesions in the nervous system, or as reactions to visceral irritation. Meanwhile, these causes of pain should be sought for and should not be omitted from consideration

until their absence has been proved by careful examination. With these considerations in mind we may proceed to give a brief description of that form of neuralgia which appears to be unassociated with disease in any part of the body.

Etiology. Primary neuralgia is rare before puberty and is most common during the middle period of life. Weak, nervous and anæmic persons, especially those in whom gouty or rheumatic tendencies can be traced, are more prone to be attacked than are the robust. It is, however, a striking fact that the first attack of neuralgia often starts without any obvious cause, the patient being in good health and free from any suspicion of neurotic or of other morbid tendencies. Subsequent attacks may be brought about by peripheral irritation, by exposure to cold and damp, by over-work, prolonged lactation, mental worry or distress, or by any circumstances or conditions which tend to a deterioration of the general health. Of acute diseases which depress the nervous system, influenza is most commonly followed by neuralgia. The disorder is also induced by malaria, diabetes and gout, and by alcohol and lead. In such cases, however, it is often difficult to exclude an early neuritis.

Symptoms. The pain varies greatly in character and intensity. It may be boring, tearing, burning or sharp and shooting. It comes on in paroxysms which vary in duration from a few seconds to several minutes, and in frequency from two or three to several hundreds a day. During the intervals there may be no pain, or there may be a dull aching in the affected part. The pain is usually unilateral; it is referred to the deep parts rather than to the skin. It is most intense in the course of a particular nerve; during the height of a paroxysm it often overflows into adjacent nerve territories. Associated with the spontaneous pain there may be superficial tenderness of the neuralgic area. This is usually localised to certain points in the course of the nerve or its branches, such tender points generally corresponding to places where

cutaneous branches emerge from beneath bone or fascia. A feeling of numbness or deadness in the affected area may accompany the pain, and transient anæsthesia may follow it. In some cases irregular patches of erythema develop over the neuralgic area; transient small subcutaneous swellings also occur, whilst severe attacks of pain may be associated with œdema of angio-neurotic origin. A characteristic feature of all neuralgias is their tendency to relapse; sometimes, as in the malarial and influenzal varieties, the relapses may occur at a definite time of the day. Occasionally a precise periodicity is observed in cases of neuralgia produced by local disease; thus in a case of suppuration in the antrum the chief paroxysm of pain occurred between seven and eight o'clock every morning.

Varieties. Neuralgia may occur in almost any part of the head, trunk, or limbs. The most common varieties are as follows:

Trigeminal neuralgia, Tic douloureux or Epileptiform neuralgia is characterised by paroxysms of pain in the distribution of one or more branches of the fifth nerve. No pathological lesion has been found to account for the malady. In several cases the Gasserian ganglion has been carefully examined; in some no changes were discovered, in others there was a fibrous overgrowth of its interstitial tissue.

The first attack of pain may occur during a period of apparently good health and without obvious cause. Subsequent attacks are excited or aggravated by changes in the weather, or by abnormal physical states of the body and in severe cases by the actions of smiling, speaking or eating, or even by a breath of wind. The character and the intensity of the pain are very varied: there may be a mere sense of tingling, or paroxysms of the most agonizing burning or shooting pains. Great variations are also observed in the frequency and the degree of periodicity with which the neuralgia recurs: for example, an attack of pain may come on every

minute with precise regularity, or only once a day, either at different times or at exactly the same hour.

At first the pain may be limited to the territory of one of the three primary divisions of the nerve, but after a time it is apt to spread to the other territories of the nerve on the same side of the face. There is also a tendency for the pain to shift from time to time from one division to another, or from certain fibres of the nerve to certain other fibres. The pain starts in one or more spots which are always described as "just under the skin"; over these foci of pain there is usually much tenderness to pressure.

When the first division is affected, *ophthalmic neuralgia*, the pain radiates from the supra-orbital notch over the forehead and the anterior half of the scalp, and is often especially severe in the eyelid, in the eyeball and at the side of the nose. Tender points are found over the supra-orbital notch, over the outer part of the upper eyelid, at the lower edge of the nasal bone and sometimes over the eyeball. In neuralgia of the second division, *infra-orbital neuralgia*, the pain radiates over the cheek between the orbit and the mouth, and tender points are found over the infra-orbital foramen, over the malar bone, at the side of the nose and along the gums of the upper jaw. Neuralgia of the third division is felt in the region of the lower jaw, in the tongue, the ear, and over the parietal eminence; tender points are found over the inferior dental foramen, and over the posterior part of the temple, or just above the zygoma in front of the ear.

The neuralgia may be accompanied by various vaso-motor symptoms, such as flushing, local sweating, salivation, a discharge of thin fluid from one nostril, and lacrymation. The affected cheek has often a shiny and greasy appearance. The conjunctiva may be reddened. The hair and beard may change in colour: sometimes the hairs fall out, or they may be rubbed away by the pressure exerted by the patient to gain some ease during a paroxysm. The breath is usually offensive and the

month foul, as attempts to cleanse it are too painful. In severe cases the jaws are generally edentulous, the patient having submitted to the extraction of his sound as well as of his carious teeth, in the vain hope of getting rid of his malady.

Cervico-occipital neuralgia is more often bilateral and constant than any other variety of neuralgia. The pain is felt in the back of the head, along the course of the great occipital nerve and in the region of the neck supplied by the first four cervical nerves. It is accompanied by tenderness, which is usually most marked

141.—Herpes zoster in a case of cervico-brachial neuralgia.

about midway between the spine and the mastoid process, and between the sterno-mastoid and trapezius, where the branches of the cervical nerves are situated. The scalp is often extremely tender, so that even touching the hairs may be unbearable.

Brachial neuralgia. In this variety, pain is felt in the shoulder and down the arm to the tips of the fingers. As a rule it is most intense in the axilla and along the course of the ulnar nerve. The pain is intensified by movements of the arm, the consequence being that special

actions may be imperfectly performed. Such disability may give rise to the erroneous impression that the patient is suffering from writers' or other form of occupation neurosis. Brachial neuralgia is frequently the result of injury; many cases depend on neuritis of the brachial plexus. The chief tender points are in the axilla, at the posterior border of the deltoid, behind the elbow and in front of the wrist.

Intercostal neuralgia is characterised by pain with paroxysmal exacerbations along the anterior branches of the spinal nerves. The pain is aggravated by violent respiratory movements: it may radiate towards the back

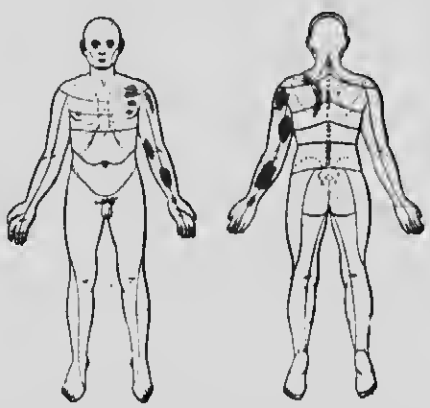


Fig. 142.—Showing painful spots in brachial neuralgia (Dana).

and arms, or into the loins. The skin in the intercostal spaces is hyperæsthetic and tender points are situated near the spine, in the mid-axillary region and near the costo-sternal articulations.

Intercostal neuralgia is a common antecedent and sequel of herpes zoster and may persist long after the eruption has passed away. The presence of the herpes indicates that the pain is a symptom of morbid changes in the posterior spinal ganglia.

Sciatica. The most common site for neuralgia is in the territory of the great sciatic nerve and its branches. Such neuralgia is usually a symptom of inflammation of the sheath of the nerve—a perineuritis, but as in many

cases there is no positive evidence that the sciatica is more than a neuralgia it is convenient to describe the affection in the present chapter.

The pain may begin suddenly or gradually: in the latter case it is often preceded by creeping sensations in the skin, or by a feeling of cold or heat, or a sense of stiffness in the limb. Slight at first and perhaps only produced by certain movements of the limb, the pain gradually increases in severity until a more or less constant gnawing sensation in the back of the thigh is associated with paroxysms of boring or darting pains along the course of the nerve. The pain is aggravated by any movement of the limb, especially if the move-

Fig. 143.—Attitude in a case of early sciatica on the left side.

ment is one which makes the nerve tense, so that slight degrees of sciatica may be detected by passively flexing the thigh with the knee extended. The patient obtains the most comfort by keeping the hip and knee flexed and the heel raised from the ground; this attitude tends to throw the weight of the body on to the other leg, and in time may lead to scoliosis, the concavity of the curve being towards the sound side. Pressure on the nerve causes pain, so that the patient when sitting rests his weight on the ischial tuberosity of the unaffected leg. There may be tenderness to deep pressure anywhere along the course of the nerve, or only in certain places. The chief tender points are over the sciatic notch, behind

the great trochanter, over the middle of the back of the thigh, in the popliteal space, behind the head of the fibula, in the peroneal region and behind the external malleolus.

In slight and recent cases there is no weakness or flabbiness of the muscles, but in severe and old-standing cases there may be weakness and atrophy of particular groups of muscles; sometimes the whole limb is wasted. Muscular cramps may also occur, which tend to be worse at night when the patient is falling asleep. Partial anæsthesia is sometimes present, consisting usually of impairment of the epicritic sensibility over the outer aspect of the leg, over the sole and outer side of the dorsum of the foot, and, if the disease is high enough to implicate the origin of the small sciatic, over the back of the thigh.

The presence of anæsthesia or of muscular atrophy, with changes in the electrical irritability of the muscles, indicates an actual neuritis. In the neuritic cases the Achilles-jerk on the affected side is generally abolished: the knee-jerk also may be diminished or absent. In the purely neuralgic cases these reflexes are normal; sometimes the knee-jerk is exaggerated.

The duration and severity of sciatica are very variable. The pain may pass away in a few weeks or may continue for many months. It may occur only on movement, or be so continuous and intense that sleep can only be obtained by the use of narcotics. Recovery is almost invariable, but the disease is prone to recur: relapses are common and may occur without any discoverable cause.

Diagnosis. An uncomplicated case of neuralgia is distinguished by the unilateral situation of the pain, by its migratory and intermittent characters and by the absence of symptoms indicating damage to the nerve fibres. If there is distinct evidence, in the presence of anæsthesia or of trophic changes in the muscles or the skin that the conducting functions of the nerve are impaired, the pain is dependent either on neuritis or

on pressure on the nerve, and the causes of these conditions must be investigated. When such evidence is lacking it may be difficult, at any rate for a time, to make a certain diagnosis. The pain of a slight neuritis may be identical with that of a neuralgia. As a rule, however, it is constant rather than paroxysmal, whilst the nerve is tender throughout the greater part of its course and not merely at certain points as is the rule in neuralgia. Exceptionally in severe cases of neuritis anæsthesia is insignificant or absent, and if some of the trunk nerves are affected, wasting of the intercostal or of the abdominal muscles may be difficult to recognise. We have then to fall back on the results of a careful search for disease of the internal viscera, the bones and the spinal cord, and on considerations relating to the character and duration of the pain. A long duration of pain without development of any other symptoms is much in favour of a pure neuralgia, but it is far from pathognomonic. For example, definite signs of an abdominal aneurysm may be preceded for many weeks by neuralgic pains in the abdomen and legs. Similarly, a tumour in the middle fossa of the skull beginning near the fifth nerve may set up shooting pains on one side of the head for a long time before definite signs of its presence can be detected; in one of my cases there was severe neuralgia in the territory of the fifth nerve for sixteen months before signs of a growth became unmistakable. Another example is furnished by the occasional long duration of the lightning pains of tabes before the other symptoms of the disease become manifest. As a rule, however, the pains are distinguished by their bilateral distribution, by their momentary duration and by their changing seat; it is rare, too, not to find impairment of the tendo-Achilles jerk or of the knee-jerk, or loss of action in the iris, or pallor of the optic disc.

In cases of pain along the sciatic nerve, the diagnosis of simple sciatica, whether a pure neuralgia or the result of neuritis, is not justified until diseases of the spine, of

the hip joint and of the pelvic organs have been excluded. In many cases it is advisable to explore the rectum and vagina for any possible source of pressure on the nerve.

The referred pains of visceral disorders tend to be aching rather than shooting in character; moreover, the pain is felt in areas which do not correspond to the distribution of peripheral nerves, and is associated with tenderness of the superficial tissues.

Treatment. The treatment of neuralgia consists in the removal of any source of irritation to the nerve, in the correction of any defect in the general health, and in the administration of remedies for the relief of the pain, or, failing these, in the performance of certain operations.

The characters of a primary or idiopathic neuralgia may be indistinguishable from those of a secondary neuralgia in which the pain is caused by disease implicating the nerve; it is therefore always necessary to make a search for any morbid local condition in order that its existence may not be overlooked. The successful treatment of nasal disease may cure an ophthalmic neuralgia, the evacuation of pus from the antrum an infra-orbital neuralgia, the removal of glands from the axilla a brachial neuralgia, the correction of indigestion severe attacks of gastralgia, and so on.

In a large number of cases of neuralgia there is some condition of debility. This association indicates the importance of a generous and nutritious diet which should include cream and other fatty foods, of adequate rest, of fresh air and of the taking of tonics, such as quinine, strychnine, and the glycero-phosphates. If anæmia is present iron and arsenic are indicated: if there is a history of syphilis, mercury and iodide of potassium should receive a fair trial: if the pain can be traced to the influence of malaria or of influenza large doses of quinine are often beneficial: whilst if neuralgia seems to be the consequence of rheumatism, of gout, or

of diabetes, the treatment suitable for these affections should be employed.

But in spite of such treatment the pain often continues; moreover, it frequently happens that no adequate cause for the neuralgia can be discovered. Hence medicinal treatment for the relief of the pain is almost invariably required. In mild forms of neuralgia one or other of the following drugs, taken either separately or in combination, will usually prove to be of service; namely, phenacetin, antipyrin, antifebrin, exalgin, pyramidon, aspirin, croton-chloral and gelsemium. Large doses of ammonium carbonate or of ammonium chloride are sometimes beneficial, and so also is a combination of nitro-glycerine with a nervine tonic. In the severest forms it is generally necessary to administer morphia; this is best given in combination with atropin hypodermically.

Local medication is often of value; ointments containing menthol, veratria, or aconite may dull the nerve endings and so reduce the degree of suffering. Temporary relief may also be obtained by freezing the painful part by means of a spray of ethyl or menthyl chloride. As soon as the acute symptoms have subsided, massage, galvanism and hydropathy are often of great service; the high frequency current is also of use, especially in cases of sciatica.

In all cases of neuralgia rest in bed is advisable; in the brachial and sciatic varieties it is essential and should be continued until the pain has disappeared or become insignificant. All movements of the affected limb must be restricted as far as possible; in brachial neuralgia this is affected by wrapping the arm in cotton wool and bandaging it in a flexed position to the side of the body, in sciatica by the application of a long splint reaching from the axilla to the foot and jointed so that slight variations in position may be obtained at the hip and knee-joints. During the acute stages of a limb neuralgia hot linseed-meal poultices should be applied along the course of the nerve. Counter-irritation, either

by the application of small blisters or of the cautery over the seats of pain, is also of great value. Acupuncture is another method which usually eases the pain of sciatica for a time; it consists in the insertion of sterilized needles into the back of the thigh to the depth of about two inches, leaving them there for a period of from fifteen to thirty minutes. Occasionally deep injections of cocaine or of morphia into the vicinity of the nerve become necessary. Morphia is the surest remedy, but cocaine in doses of from one-tenth to one-third of a grain will often abolish the pain for several hours. Recently the injection of fifty to a hundred c.c. of normal saline solution has been recommended: in seventy-three cases of sciatica treated by this method Bum obtained a complete cure in forty-two.

In many cases of trigeminal neuralgia favourable results have been obtained by the injection into the painful area of a few minims of pure chloroform, of a few minims of a two per cent. solution of osmic acid, or of absolute alcohol, either with or without eucaine or stovaine. The injection of strong alcohol into the nerve trunks as they issue from their respective foramina, namely, the supra-orbital notch, the foramen rotundum and the foramen ovale, is frequently followed by an entire cessation of the pain, sometimes lasting for several months. A practical acquaintance with the correct line and depth for the injecting needle must be obtained by experience on the articulated skull and on the dead body. With proper precautions and strict attention to asepsis there is little or no risk of any permanent morbid changes being established.

Recently Wilfred Harris has shown that it is possible to inject the Gasserian ganglion with alcohol, by passing the needle through the foramen ovale, and thus destroying the nerve-cells of the ganglion; the anæsthesia which results is permanent.

In very intractable cases of neuralgia which respond only temporarily either to internal or to external medication, certain surgical procedures must be con-

sidered. These comprise nerve stretching, neurectomy, division of the posterior nerve roots and excision of the posterior root ganglia. Extirpation of the Gasserian ganglion is the only certain cure for a severe and protracted case of trigeminal neuralgia; the operation has now been performed in several hundreds of cases and for the most part with a perfectly satisfactory result.

CHAPTER II.

MIGRAINE.

Migraine is a paroxysmal neurosis characterised by periodic attacks of headache, the pain being often preceded by some disorder of vision and accompanied or followed by nausea and vomiting.

Etiology. Direct inheritance of the disease is common. In a few cases all the members of a family are subject to some form of paroxysmal headache; in others there is a family history of neuralgia, hysteria, or of hay fever. It is commoner, however, to discover a tendency to gout in the patient's ancestors than to any nervous affection. There appears to be a slight relationship between migraine and epilepsy: thus the periodic headaches of early life may be replaced by epileptic seizures at a later period, or the parents of epileptic children may have suffered from migraine. It is rare, however, to find epilepsy in the ancestry of the subjects of migraine.

The neurosis occurs more frequently in females than in males. It usually begins in early life, the first attack occurring between the ages of five, and twenty, and only rarely after thirty. It may be difficult to discover any exciting cause for the first attack; subsequent attacks are sometimes induced by physical or mental fatigue, by worry, anxiety or mental excitement, by indigestion, eye-strain or by bad hygienic conditions. In women attacks are liable to occur at the menstrual period.

Symptoms. The headache may come on suddenly, or

it may be ushered in by certain premonitory symptoms which in some cases constitute a definite aura. These warnings comprise a feeling of heaviness in the head, numbness and tingling in the hands, face or tongue, or some perversion of the visual functions.

Visual phenomena occur in fully half the cases of migraine, and when present usually constitute the earliest symptoms of the attack. They consist of dimness of vision, of spectral appearances or of both. The loss of sight is always partial: it may affect the whole field of vision or only a small area which is usually near the centre of the field, so that the object gazed at is dim or invisible: sometimes the defect takes the form of homonymous hemianopsia. The spectra are of a bright and scintillating character and frequently appear as zigzag bands of different colours. They may develop out of the dim central area which, as it expands and clears at the centre, becomes luminous at the periphery: or they may begin as a bright spot which enlarges, its centre becoming dim and its outer boundary luminous and sometimes coloured. Within the bright ring moving luminous particles are occasionally seen. Visual appearances are not always so marked: they may be limited to a few specks or flashes of light, or to objects in constant vibration.

Sensory symptoms are sometimes experienced: they consist of tingling and of other forms of paræsthesia, which are referred to the arms, the face, lips, tongue and fauces, and exceptionally to the legs. They may occur alone or in association with the visual phenomena: in either case they generally precede the headache. Tingling often begins in the hand and passes slowly up the arm; it may be followed by slight anæsthesia or even by transient weakness of the limb. It is usually unilateral, and as it ceases the pain begins in the opposite side of the head. Gowers compares the ascending zone of tingling to the expanding luminous zigzag in the field of vision, and the subsequent impaired sensation to the dimness of sight within the zigzag.

Slight giddiness, transient mental disturbance and transient aphasia, usually of the sensory variety, are sometimes present early in the attack; the aphasia may be associated with tingling and visual disturbance on the right side.

Vasomotor symptoms are sometimes prominent, and, in one form or another, may be present throughout the seizure. The face is pale and looks pinched; the extremities are cold and the pulse tends to be feeble and slow. As the headache reaches its acme the facial pallor may be replaced by flushing and perspiration; exceptionally these symptoms are unilateral in distribution: one side of the face is pale, the eye is retracted and the pupil small, whilst as the headache subsides hyperæmia succeeds the pallor, and the pupil resumes its normal size.

The most prominent symptom of migraine is pain in the head; it may come on during the decline of some of the above-mentioned sensations, or it may constitute the whole of the attack. The pain varies in intensity; it may be so slight as not to interfere with the patient's work or so severe as to completely prostrate the sufferer. As a rule it is very distressing and of a deep and boring character; in some cases the head feels as if gripped in a vice, in others the headache is described as throbbing or pulsating. The pain is aggravated by movement, light and noise, and is worse in the erect than the recumbent position. In most cases it begins on one side, sometimes in a small spot on the temple or the forehead, or in the eyeball; in other cases it begins at the back or on the top of the head. It generally increases in intensity and spreads over the whole of one side of the head; it may either remain unilateral (*hemicrania*) or become general, and may extend to the neck or even to the arm. Sometimes it affects both sides of the head at once. In some cases it is so severe as to give rise to delirium, to stupor or to hysterical manifestations. Local tenderness of the scalp is sometimes a marked feature. In many cases the seat and

character of the headache are the same in each attack; in others they are different; for some years the pain may be frontal, and afterwards may be limited to the occipital region. The duration of the headache varies from a few hours to several days; very often it begins in the early morning and lasts all day, recovery taking place after a night's rest.

Nausea usually accompanies the headache, and interferes with the taking of food; frequently it leads to vomiting or to retching, which commonly occurs when the pain has reached its height or has begun to subside, and may bring the attack to an end. Repeated vomiting may result in the rejection of bile, leading the subjects of migraine to regard their seizures as attacks of biliousness. The temperature throughout the attack is usually normal, but in children periodic attacks of unilateral headache may be associated with considerable pyrexia.

Types of migraine. One of the commonest forms of the disease consists solely of a localised headache with sickness, sensory symptoms being absent or insignificant. The converse is less frequent, but it is important to recognise that an attack of migraine may be entirely made up of visual disturbance, or even of tingling sensations in the limbs, headache being slight or absent; sometimes a slight unilateral headache is ushered in by a sudden temporary defect of central vision on the opposite side, which is quickly followed by various visual spectra. In some cases the main feature of an attack is an inability to use the right words in speaking.

A variation in the character of the attacks in the same person is not uncommon, some being typical, others consisting mainly of visual phenomena. The character of the headache may also vary in successive attacks; one kind of headache may be attended by retching and vomiting, whilst another is not.

Duration and course. The duration of the attacks and the interval between them present many variations. An average seizure lasts from six to ten hours, a severe one

may not entirely abate for two or three days. A common interval between the seizures is three or four weeks. Sometimes the periodicity is exact, the seizures occurring at almost precisely the same time, but as a rule the periodicity is less exact, and irregularities in the duration of the intervals are often determined by the presence or the absence of exciting causes. The frequency with which attacks occur varies from two or three a week to the same number yearly. As age advances there is a tendency to a diminution in the severity and the frequency of the attacks: but this is not invariably the case, intense paroxysmal headaches sometimes occurring in women who have passed the climacteric period. The prospect of recovery or of improvement is influenced by the effects of treatment. It is best when there is some form of irritation, such as an error of refraction, or a faulty mode of living, which can be corrected or removed.

Pathology. No morbid changes have been found to account for the symptoms of migraine. The periodical derangement of function and the character of the symptoms strongly suggest that the cerebral cortex is the seat of the disturbance. The nature of the disturbance is uncertain, but it seems probable that it is analogous to that which underlies the seizures of epilepsy, although the minuter characters of the disturbance must differ in the two diseases. In migraine, as in epilepsy, the stability of the cortical cells is less than normal, and is therefore easily upset by stimulation provoked either reflexly or by irritating materials circulating in the blood. The visible vasomotor phenomena are to be regarded as secondary results of the derangement of the cortical cells.

Diagnosis. Some of the symptoms of migraine, especially the sensory warnings, may resemble those of minor epilepsy, and it must not be forgotten that occasionally one disease passes into the other and that the nature of some attacks may be difficult to determine. The following distinctions should therefore be borne in

mind:—The warning of an epileptic seizure lasts only a few seconds, that of migraine from ten to twenty minutes; in epilepsy dimness of vision is general and momentary; in migraine it is central, hemianopic or irregular in distribution, and is prolonged; in migraine the headache is usually severe and continues for several hours, whereas when headache follows an epileptic seizure it is rarely severe or prolonged, nor is it one-sided as is so frequently the case in migraine.

Certain symptoms of migraine, such as the headache and the transient paralysis or aphasia may arouse the suspicion of a tumour or of other serious disease in the brain, especially if they occur for the first time in adult life. The pain of tumour, however, is more or less constant and is usually associated with optic neuritis and other distinctive symptoms; sometimes the pain intermits for a time, but the intervals are much shorter than those of migraine. When visual disturbance is present, that of migraine presents characteristic features.

Treatment. The first and most essential step to be taken is to remove so far as possible any abnormal condition in the patient, and to correct any errors in his mode of life that are likely to excite the attacks. The diet must be carefully regulated and a daily action of the bowels secured. The patient should be placed under the most favourable hygienic conditions, over-fatigue of mind or body, late hours, hot crowded rooms and other injurious influences being carefully avoided; ocular defects should be searched for and if necessary corrected.

To prevent the recurrence of the attacks or to lessen their severity many remedies have been tried during the intervals. A measure of success is sometimes attained by the regular and continuous administration of the bromides as described under the treatment of epilepsy. They may be given alone or in combination with phenacetin, antipyrin, caffeine or cannabis indica. The extract of cannabis indica taken by itself in half-grain doses once or twice daily is often of great service. Other useful auxiliaries to the bromides are the tinctures

of gelseminm and belladonna. For cases in which the arterial tension is raised nitro-glycerine should be tried, the liquor trinitrini in doses of half to two minims being taken two or three times a day after food; in some cases it is advisable to combine the preparation with the tincture of nux vomica, with the tincture of gelsemium or with the bromides. When anæmia is present iron and arsenic are of advantage; the valerianates of zinc and iron have been strongly recommended.

During the attack itself absolute rest in the recumbent posture in a quiet and darkened room affords the most comfort to the patient and tends to shorten the duration of his illness. A thirty to forty grain dose of bromide taken at the beginning of the seizure may lessen its severity or shorten its duration. More frequently the headache is relieved by the taking of a full dose of phenacetin, antipyrin, or antifebrin, either alone or with caffein. In attacks of extreme severity in which the pain is almost unbearable and tends to be associated with delirium it may be necessary to give a subcutaneous injection of morphia. Among local agents which sometimes afford relief to the pain may be mentioned menthol, ether spray, extract of belladonna, a weak ointment of veratria, cold compresses or ice-bags to the head, and sinapisms to the back of the neck. When the attack is over it is advisable for the patient to take small doses of liquor strychninæ for a few days, or a mixture containing bromide of potassium and tincture of nux vomica.

CHAPTER III.

EPILEPSY.

Epilepsy is a disorder of the brain which is characterised pathologically by sudden brief discharges of nerve energy in the grey matter of the cerebral cortex not due to the normal stimulus, and clinically by repeated attacks in which a sudden loss or impairment of consciousness is the most constant and notable feature. Temporary unconsciousness may constitute the whole

of the attack as in the variety known as *minor epilepsy*, or it may be associated with general convulsions as in the variety called *major epilepsy*.

Etiology. In about half the cases of epilepsy there is evidence of the inheritance of a neuropathic disposition. This may be shown by the direct transmission of the disease from parent to offspring, or by a history of epilepsy, of insanity, of alcoholism or of some other neurosis in one or more members of the same family or in collateral relations. In many of the remaining cases it may be impossible to obtain any evidence indicating the inheritance of an unstable nervous system, but failure to discover this evidence does not necessarily exclude the existence of such an inheritance. It is, however, possible that toxins attacking nerve cells of normal stability may be powerful enough to set up the disease. Sometimes there is a family history of tubercle.

A notable feature of the etiology is the frequency with which the first fit occurs during certain epochs of life—namely, infancy, puberty, and the period between puberty and early manhood, when there is a normal instability of the nervous system. For although epilepsy may begin at any age, in three-fourths of the cases it begins before twenty, and its onset is especially common at the time of puberty. Many cases date from infantile convulsions; in others there is an interval of variable duration between the convulsions and the beginning of the established disease; thus fits which have occurred during retarded dentition may cease, and true epilepsy come on at puberty. In women the advent of epilepsy is often contemporaneous with irregularities in menstruation, with pregnancy or with child-bearing; in many cases these events seem to have an adverse influence upon the course of the disease, in others a favourable influence.

Of antecedents which sometimes appear to be exciting causes of epilepsy may be mentioned:—(1) Undue peripheral irritation arising from disease of the eyes, ears, nose, stomach, intestines or of the genital organs.

(2) Emotional disturbance, such as sudden fright, grief, or prolonged anxiety; and (3) infective diseases and toxic conditions; for example, scarlet fever, alcoholism, and occasionally syphilis. But it is probable that these and other alleged causes are mere coincidents, or that they would be inadequate to produce epilepsy if it were not that the nerve tissue on which they act is unstable and abnormally prone to discharge.

Symptoms. Occasionally an epileptic attack is preceded for some hours or days by premonitory symptoms such as headache, giddiness, mental confusion, irritability or depression: sometimes the patient feels unusually well, is lively and has an exceptionally good appetite.

The more immediate warnings of a fit are called *aura*. They are stated to occur in about half the cases of epilepsy, but in the author's experience they are much less common. The term *aura* is applied to any sensation or motion experienced by the patient which immediately precedes loss of consciousness; occasionally the warning constitutes the whole of the seizure. The chief varieties of aura are as follows:—

Sensory aura. These are sensations of numbness or tingling in certain parts of the hand or in the tongue; pain or a general feeling of pressure in the head, or a "rushing of blood to the head." A very frequent aura is giddiness, and this sensation may be accompanied by actual rotation of the head and eyes.

Special sense aura. Of these the visual are the commonest and the gustatory the rarest. The simple or crude sensations consist of flashes of light or of colour; hissing, whistling or crashing sounds; unpleasant smells; metallic or bitter tastes. More elaborate sensations are visions of beautiful places, or more commonly of ugly objects; and in rare cases a series of words or strains of music. Such complicated perceptions often follow the crude sensations; thus a red light may be followed by the image of a man, a confused noise by music. A dreamy state is often associated with

the olfactory sensations, an indication that the discharge has started in the uncinate gyrus.

Visceral aura. One of the most common is some sensation at the epigastrium. There may be actual pain, or an indescribable sensation which often ascends to the throat and causes a feeling of choking, similar to the globus hystericus. A sensation of nausea may accompany the epigastric aura. In other cases palpitation or vague discomfort about the heart is experienced.

Motor aura. These, which are usually brief in duration and unilateral in distribution, consist of a twitching in some part of the body, the spasm generally beginning in small muscles, which are organised for special actions, such as those of the hand or of one side of the face. Sometimes there is a sudden rotation of the head and eyes, or a sudden inability to speak or even a transient aphasia. In rare cases the epileptic attack is preceded by a complicated movement, such as running forward or backward, jumping or turning round.

Vaso-motor aura occur in the form of subjective sensations of coldness or of heat in certain parts; occasionally such parts feel cold to the touch or present spots of redness.

Psychical aura sometimes usher in a fit. Thus intense alarm may be experienced and the patient looks startled and frightened; or he passes into a dreamy state in which past events may crowd upon the memory.

Major Epilepsy or Le Haut Mal. In a typical case three stages may be distinguished—viz., (1) the tonic stage; (2) the clonic stage; and (3) the period of resolution.

The first stage. The fit, preceded or not by an aura, begins with unconsciousness, sudden falling, and often with a piercing cry or a hoarse groan. Frequently the face is very pale. The loss of consciousness is sudden and complete. The fall may be forwards, backwards or laterally; as a rule it is so instantaneous that the patient is unable to protect himself, and he may fall from a height or into fire and water; sometimes, however, he has time

to sit or to lie down. He becomes quite rigid, all the muscles of the body being in a state of tonic though unequal contraction; one side of the body is more affected than the other. The head is retracted and rotated to one side, usually to the side on which the spasm is greatest. The back is rigid and slightly arched. The legs are usually rigidly extended and rotated inwards and the feet inverted; sometimes the hip and knee-joints are slightly flexed. The arms are slightly abducted, the elbow and wrist-joints are flexed, the hands are pronated and the fingers are flexed at the metacarpal and extended at the other joints, while the thumb is pressed against the fingers or bent into the palm. The tonic spasm of the respiratory muscles fixes the chest and arrests breathing; hence the initial pallor of the face quickly passes into duskiness and often into marked cyanosis. Violent throbbing of the carotid arteries together with distension of the veins of the head and neck is also observed. The pupils are dilated and do not respond to light; they remain dilated until signs of consciousness return. During this tonic stage, which usually lasts from five to thirty seconds, the spasm may be so great as to cause fracture of the teeth or the bones, or dislocation of the shoulder or the jaw.

The second stage, which is ushered in by tremulous movements of some of the rigid muscles is characterised by the general substitution of clonic for tonic spasm. Twitching successively invades the face, the limbs and the trunk, till the whole body is thrown into violent convulsions, which usually predominate on one side of the body. The head is jerked in various directions; the facial muscles are violently twitched and the expression is rendered still more hideous by the convulsive upward rotation of the eyeballs, which hides the pupils and exposes the whites of the eyes under the blinking half-closed lids. The mouth is alternately opened and closed; the protruded tongue is often severely bitten and blood-stained mucus oozes through the clenched teeth. The limbs are powerfully extended and flexed, while the

contents of the bladder, the rectum or the vesiculæ seminales may be discharged involuntarily. The respiratory movements which are resumed at the beginning of this stage become jerky, noisy and laboured. The skin is cold and often covered with sweat. The pulse is tense and increased in frequency, while the heart beats tumultuously. Towards the end of this stage, which usually lasts two or three minutes, the clonic spasms become slower and less frequent, though not necessarily less violent, till they finally cease, and the patient for a time lies in a state of deep coma with flaccid paralysed limbs; he breathes stertorously and his pupils are widely dilated or oscillate between contraction and dilatation.

The third stage is characterised by a gradual return to consciousness and voluntary power; during the period of transition muscular twitching may here and there be noted. Sometimes recovery takes place quickly, but more commonly in a gradual manner. Sometimes the patient lapses into a profound sleep; at other times he tries to change his position, he opens his eyes and looks around him with a stupid or wild expression, and may attempt to speak. The attack is often followed by vomiting, or by the passage of a large quantity of limpid urine, which occasionally contains a trace of albumen or of sugar. Lassitude and stupor persist for an hour or more; then the patient feels bruised and exhausted, he suffers from headache and vertigo and may exhibit signs of considerable mental or emotional disturbance. The conjunctivæ are injected and petechiæ from rupture of over-distended vessels may be observed on the face and the upper part of the body. General muscular weakness is sometimes prominent, or transient hemiplegia affects the side on which the convulsions were most marked; in right-sided seizures transient aphasia occasionally occurs.

The superficial abdominal reflexes and the knee-jerks are abolished during the comatose stage; subsequently they return and the knee-jerks may be exaggerated for

a time, the exaggeration being associated with an easily elicited ankle clonus and a plantar reflex of the extensor type.

Minor Epilepsy or Le Petit Mal. In this variety there are attacks of temporary unconsciousness. This may constitute the whole of the attack or may be associated with blinking of the eyelids or other minor degrees of muscular spasm. Tasting or chewing movements are sometimes observed. A patient while speaking becomes suddenly unconscious; there is a pause, but in a few seconds he resumes the thread of his conversation as if nothing had happened. Sometimes the unconsciousness does not interrupt the action engaged in. Thus a patient, if walking, will continue to walk; he may sometimes, like the somnambulist, avoid obstacles as if the senses were still guiding him; if playing the piano, he may go on playing, and sometimes with perfect accuracy. During such attacks the pupils may be seen to dilate and the eyes to be fixed and staring; the face may become momentarily pale, while after the attack it is often more congested than natural. In other cases a feeling of giddiness or of fainting is prominent; there is mental confusion without actual loss of consciousness.

The attack may indeed be constituted by any of the auræ already described, and is not invariably attended by complete loss of consciousness. During the seizure urine is often passed involuntarily; this is especially the case in females. The period of recovery may be characterised by an outbreak of hysteroid convulsions or by the performance of various automatic actions, of which subsequently the patient has but a vague recollection. Hysteroid convulsions are most common in young women, and must be attributed to the state of brain which underlies tendencies to hysteria as well as to the preceding fit.

Automatic actions occasionally follow major seizures, but they are more common after the minor ones, and in some cases appear to constitute the whole of the attack—"masked epilepsy"; but as a rule they are preceded by

transient loss of consciousness. Of such actions, undressing is one of the most frequent; the patient may take off all his clothes and go naked into the street, or he attempts in this condition to enter a public assembly. Another patient may urinate in public, another may put in his pocket any adjacent object. In other cases anger and violence determine the nature of the action. Thus a patient helped across the street may reward the kindness by a blow on the face; a husband, apparently waking out of sleep, may beat or strangle his wife with the utmost ferocity. During this state of epileptic mania the patient may commit any kind of crime of which afterwards he has no recollection. Sometimes delirium, instead of being furious and dangerous, takes the form of unusual gaiety.

Health between the attacks. In many cases the mental and physical health remains excellent: indeed, of some epileptics it may be said that they seem all the better for a periodical cortical discharge. But in a large number of cases there is evidence of psychical degradation; the patient is self-opinionated and egotistical, his judgment is feeble, his temper is irritable, his memory is uncertain, and he shows a lowered capacity for the details of his daily work. Sometimes the mental failure is more pronounced, and gradually progresses to actual dementia. The liability to mental deterioration is greatest when the attacks begin in early life, and when they are frequently repeated, and continue for many years. But even under these adverse circumstances the mind is not always affected; on the other hand, many epileptics are naturally weak-minded. Hence it is probable that, although epilepsy *per se* often leads to mental changes, the latter may be largely due to pre-existing cerebral imperfections. Whatever the explanation, there can be no doubt that insanity and epilepsy are closely related and that many epileptics are to be found in our asylums, either in consequence of their liability to maniacal outbreaks or of their demented condition.

Course and termination. Epilepsy presents many variations in its mode of commencement, in the frequency of its attacks and in the time of day at which they occur. It may begin as the minor type which may exist for some years before convulsive seizures ensue, or as the major type which either persists or is replaced by the minor attacks; it is more common for epileptics to suffer from both varieties than from one variety only. In some cases each attack occurs at night, in the early morning, or at some other particular time; in others the attack comes on at any hour of the day or the night. Nocturnal attacks may exist for many years without their occurrence being suspected.

Some patients have only one or two seizures a year, while others have several hundreds; a common interval between the seizures is three or four weeks. Long remissions, sometimes lasting for several years, may occur; the greatest tendency to cessation of the attacks is in the period between early infancy and the second dentition. Very rarely the attacks cease altogether, either spontaneously or as a result of treatment. In at least ninety per cent. of the cases of epilepsy, however, the attacks continue throughout life, but they do not necessarily shorten its duration.

As a rule the attacks are isolated; sometimes they occur in series, each series being separated by an interval of a few weeks. In the serial variety which may complicate the major, the minor, or the combined type of epilepsy, seizures follow one another with a rapidly increasing frequency until a climax is reached, when the frequency begins to decrease until eventually the fits cease altogether for a time. Generally there is a return to consciousness between the seizures, but occasionally the succession of fits is so close that consciousness is not regained, and the patient lies in a condition of profound coma. This is called the *status epilepticus*; during its course the temperature may rise two or more degrees, sometimes to 105° or even higher, and there is a danger of death from exhaustion. Death

is rarely caused by the violence of an isolated fit, and indeed is seldom the direct result of any incident of the disease. When it occurs it is generally brought about by indirect means, such as an accident occurring during a fit, or the occurrence of some pulmonary or cardiac complication.

Pathology. There can be little doubt that the *site* of the lesions which underlie the phenomena of epilepsy is in some portion of the cortical grey matter. Evidence in favour of this is afforded by the facts of experimental physiology, and by the association of coma and general convulsions in cases where there are visible cortical lesions, as in the meningo-encephalitis of chronic alcoholism, or in new growths implicating the cortex. Another proof of this causal relationship is the cessation of convulsions on one side of the body after a lesion interrupting the motor fibres; thus if an epileptic is seized with left hemiplegia from a lesion of the right internal capsule, any subsequent convulsions do not affect the left side, whereas before the stroke the convulsions were bilateral.

Recent knowledge regarding the anatomy and functions of the nerve cells suggests that the discharge originates in the spongy grey matter rather than in the cells themselves, and that it travels from this through the dendrites along the fibrils which pass through the cells to the axis cylinders. Probably the initial discharge does not always begin in the same place. Thus a visual aura would suggest the occipital lobe as the primary seat of discharge, a gustatory or an olfactory aura the uncinate gyrus and so on; the subsequent occurrence of convulsions indicating that the discharge has spread to the motor area.

As to the *nature* of the lesion but little is known. Most of the histological changes that have been revealed by the microscope are probably not the cause but the result of the convulsions. In old-standing cases of epilepsy an increase of the neuroglial tissue is found throughout the cerebrum; the sclerosis being often most

marked in the region of the cornu ammonis, which is frequently much atrophied. Occasionally cystic tumours and angiomata are present. Multiple punctate hæmorrhages are common if death occurs during the status epilepticus. The frequent presence of general arteriosclerosis in senile epilepsy suggests that the seizures may depend on an insufficient supply of blood to the cortex. This hypothesis is held by some authorities to be the best explanation of the convulsions in all cases of epilepsy. Thus Aldren Turner suggests that local arterio-capillary thrombosis with venous stasis is a probable cause of epileptic convulsions. He bases this view partly on the pathological investigations of John Turner who found coagula of blood-plates in the veins, capillaries and arteries of the cerebrum and cerebellum; and partly from a consideration of certain circumstances in which fits are prone to occur. These are: The marked frequency of fits during the hours of deepest sleep, when, cerebral blood pressure being at its lowest, capillary stasis is most likely to ensue; and the occurrence of epileptic seizures in association with acute inflammatory disease, such as pneumonia, enteric fever and scarlet fever, and with the puerperium, in which conditions, owing to an increased coagulability of the blood, there is a tendency to venous thrombosis.

Another view is that a trifling change in the chemical constitution of the congenitally unstable grey matter will suffice to produce the first fit, and that the nerve elements are so affected by the explosion of nerve energy that similar explosions are prone to occur, and thus a habit or pathological tendency to fits is established. In order to obtain support to this hypothesis—namely, that biochemical changes act as exciting causes of epilepsy, the toxicity of the blood and the urine has been investigated by several observers, but with contradictory results. Pugh has found that the alkalinity of the blood is diminished both before and after a fit, and that the average inter-paroxysmal alkalinity is lower than the average alkalinity of the blood in health. A slight

leucocytosis is a constant feature, and the leucocytosis is much increased after a fit.

Diagnosis. This may be easy or very difficult. It is easy when convulsions are known to occur, and when they present the typical features that have been mentioned. But if attacks occur during sleep, and at no other time, they may go on for years without the patient or his friends being aware of their existence. Often, however, indications of the nocturnal attacks are present; the patient finds that his tongue is sore, or that his face is ecchymosed, or that he has wet the bed. When the convulsions occur in the daytime the disease is usually easily recognised; at the same time, even a skilled observer closely watching a single convulsive seizure may have a difficulty in deciding as to its nature. In some cases of *hysteria* the difficulty may be great, but as a rule a consideration of the following points will lead to a correct diagnosis:—In an attack of hysteria the patient rarely bites her tongue or hurts herself in falling, though she may bite her lips or hands or attempt to bite her attendants. The hysterical seizure does not follow the well-defined course which is observed in epilepsy. The patient struggles, throws her body about, kicks or fights and may become rigid, but the rigidity is not followed by clonic spasms as in epilepsy; moreover, the rigidity involves the neck and trunk rather than the limbs; sometimes there is opisthotonos which may be suddenly replaced by prosthotonos. The oscillations of the head in hysteria and the squinting movements of the eyeballs offer a marked contrast to the tonic conjugate deviation of the head and eyes in epilepsy. If the eyes can be examined, often a difficult task in violent hysteria, it will be found that the corneal reflex is usually preserved and that the pupils react to light: both these reflexes are abolished during an epileptic fit. The hysterical spasms relax suddenly and there is an almost instantaneous return to consciousness, without any signs of mental confusion or of a tendency to automatism; whereas the epileptic seizure usually

terminates in deep sleep, on awakening from which the patient looks dazed and acts in an abnormal manner. The knee-jerk, abolished during epileptic coma, becomes exaggerated during the period of recovery, when ankle-clonus and an extensor plantar response may also be elicited; such changes in the reflexes are not observed during a hysterical seizure.

Coarse brain disease. It must be remembered that occasionally the convulsions of a cortical tumour are general instead of local, and that the convulsions of epilepsy may begin locally and may be only partial in range. Hence in all cases of convulsions a careful search should be made for other symptoms of coarse lesions, and especially for optic neuritis. An examination of the motor and sensory systems and of the reflexes is also important. Headache and vomiting may occur just after an epileptic fit, but in cerebral tumour they occur quite independently of the convulsive attack.

A difficulty may arise in cases of old brain lesions, where there is little or no evidence of paralysis or of spasm. Thus in the diplegia or the hemiplegia of infancy these symptoms may be insignificant, and the convulsions, which may not come on till puberty, may be indistinguishable from those of idiopathic epilepsy; as a rule, however, slight traces of spasm or of weakness are present, or the limbs on one side are a little smaller than their fellows, or some degree of mental dulness is present.

Epileptic seizures, indistinguishable from those of idiopathic epilepsy, are also seen in association with traumatic lesions of the brain, with cerebral syphilis and general paralysis, with hydrocephalus and with vascular lesions of the brain; and in cases of alcoholism, of lead poisoning, of anæmia and in connection with pregnancy and parturition. The presence of other symptoms usually permits of a correct diagnosis being made.

The recognition of petit-mal may be difficult, and the condition is often mistaken for syncope. A cause for a

faint is usually forthcoming, as a fright or other severe emotion, a hot room, anæmia or heart disease, especially aortic regurgitation. An epileptic attack may occur when the patient is sitting quietly in a cool room; it comes on suddenly and is associated with some degree of blueness about the face. Epilepsy should be suspected in all cases of repeated "faints."

An attack of auditory vertigo may resemble one of minor epilepsy, but it is rarely attended by loss of consciousness; tinnitus and deafness are features of auditory vertigo, whereas there are no signs of ear disease in epilepsy.

Prognosis. A spontaneous cessation of epilepsy is an event of such extreme rarity that the prognosis must be chiefly based on the effects of treatment. If the patient will perseveringly carry out a well-considered plan of treatment for a long period it is justifiable to assure him that there is a great probability that his attacks will lessen both in severity and frequency, and that there is a possibility that they may cease altogether. In estimating the prognosis in the individual case the following facts should be borne in mind:—The outlook is more favourable, the shorter the duration of the disease and the less frequent the attacks; it is better if the seizures occur only at night than if they occur at other times as well; it is better in cases beginning in advanced life than in those beginning at puberty. The prospect appears to be the best in major epilepsy, when there are long intervals between the attacks.

The outlook is unfavourable when the disease begins in early childhood, or when the fits occur daily. The prognosis is particularly bad in cases of minor epilepsy, especially if the seizures are frequently repeated. It is also unfavourable in all cases of epilepsy associated with much mental impairment.

Treatment. In considering the treatment of epilepsy it must be remembered that the cortical tissue is unstable and prone to discharge. It is therefore necessary to protect it as far as possible from undue stimulation,

and to employ measures which have a tendency to lessen its instability and so increase its resisting power. These objects are best attained by the removal of all sources of peripheral irritation, by the correction of any faults in the mode of living, and by the administration of drugs which have a tendency to lessen the severity and the frequency of the attacks, if they do not abolish them altogether.

Of morbid conditions which, either directly or indirectly, may have an injurious influence on the cerebral cortex, and which therefore require to be treated, may be mentioned errors of refraction, nasal polypi, adenoids, carious teeth, pyorrhœa, gastric or intestinal disturbances and irregularities in the menstrual function; anæmia, a feeble circulation and abnormal conditions of the blood pressure.

Mode of living. The patient should lead a quiet uniform life, free from excitement and worry and overexertion. Unless his fits are very numerous, or unless he is very weak or mentally impaired, it is desirable for him to continue his schooling or his occupation, providing that all possibility of strain is carefully avoided, and that the occupation does not involve any danger to life. Work, amusement and games, if reasonable in kind and degree and judiciously proportioned to each case, are beneficial; a patient is less likely to have a fit when engaged in some engrossing mental or physical exercise than when his mind is listless and his body idle. The best occupations for epileptics are farming and gardening, the least desirable are those which entail a sedentary indoor life. The experience of epileptic colonies has taught us the great advantages to be derived from healthy occupation in the open air, combined with the systematic ordering of the habits, diet and recreation.

The food of the epileptic should be simple, plainly cooked and such as can be easily digested: it should be taken slowly and at regular hours, late suppers being avoided. In some cases a vegetable diet is beneficial, and there is reason to believe that much animal food is

harmful. In most cases, however, it is advisable to allow the patient to take a moderate amount of animal food, fish, eggs and chicken being preferable to butchers' meat. The elimination of common salt from the diet has been recommended, chiefly on the supposition that the bromide salt is then more effective, and may therefore be taken in smaller quantities. Alcohol must be strictly prohibited; water and milk are the best drinks, freshly made weak tea is better than coffee.

Epileptics should sleep on hard beds with firm pillows in order to lessen the risk of suffocation should they turn on to their faces during a nocturnal seizure.

Medicinal treatment. The bromides of potassium, sodium and ammonium, given either separately or combined, have a greater curative and palliative influence on epilepsy than any other drugs. Potassium bromide is generally regarded as the most efficient as well as the most depressing of the salts of bromine, but there is no very definite evidence in support of this view. It is stated that acne is less likely to be produced by strontium bromide than by the other bromides.

Whichever salt of bromine is selected its dose must be proportioned to the age of the patient, to his personal susceptibility to the drug and to the frequency of the attacks; in all cases the bromide should be taken regularly and continuously until two years after the last seizure, and even then it is desirable to continue the medicine, the dose being gradually diminished, for another year. If there is an interval of several months between the fits, a daily dose of twenty to thirty grains is usually sufficient to suppress them or to lessen their severity. If the fits recur every two or three weeks, or still more frequently, a similar dose should be taken two or three times a day. There appears to be no advantage in taking a larger amount than ninety or one hundred grains daily; indeed most patients who take more than a drachm a day are liable to suffer from some of the toxic symptoms included under the term 'bromism,' namely, lethargy, both mental and physical,

impaired memory, slowness of speech, tremor of the tongue and weakness and inco-ordination of the limbs. Some patients suffer also from a local dermatitis with a semi-purulent secretion, or from an eruption of acne on the face and back. The tendency to such skin affections is prevented or reduced by combining small doses of liquor arsenicalis with the bromide; after taking this combination for some years arsenical pigmentation of the skin may develop, but arsenical neuritis only very rarely, possibly because the bromide prevents the usual effects of arsenic upon the nutrition of the nerves.

When the fits occur at some particular time of the day the bromide must be taken an hour or two earlier. Thus if they occur only at night or in the early morning they may be beneficially influenced by taking a thirty grain dose at bedtime; if they occur just after the patient gets out of bed in the morning, he should take the medicine on first awakening from sleep.

The addition of a few minims of the tincture of belladonna or of digitalis to the bromide mixture is frequently of advantage: belladonna is often efficacious in minor epilepsy, digitalis in the nocturnal variety. When these remedies fail to relieve the seizures biborate of sodium is the most likely drug to be of service; it may be given in doses of five to twenty grains three times a day. Boracic acid is sometimes efficacious, and in the epilepsy of children the author is accustomed to begin the treatment with a mixture containing borax and boric acid in glycerine and water; in a few of his cases this treatment was so successful that the subsequent administration of the bromides was unnecessary.

Nitro-glycerine has been advocated for children suffering from minor seizures, and also in any cases of epilepsy in which arterial spasm is a prominent feature. The minor attacks are sometimes benefited by the salts of zinc: the lactate may be given in doses of five or six grains, the oxide in doses of one to two grains. Strychnine given either alone or in combination with

the bromides is a useful tonic and tends to combat the depression which so often attends epilepsy, and which cannot always be attributed to the action of the bromides.

Some patients can ward off a threatening attack by a powerful effort of the will, aided by muscular exertion such as clenching the teeth and grasping something tightly. When the warning of a fit is an ascending sensation from the hand or the foot, the fit in some cases may be arrested by constricting the limb by means of a ligature.

During the fit little can be done to influence its course. The patient must be laid down and protected from injury: the clothes should be loosened about the neck, and a spoon, a piece of cork or of indiarubber inserted between the teeth to prevent the tongue from being bitten. If the patient vomits he should be turned on to his side in order to facilitate the escape of the vomited matters. The post-paroxysmal sleep must not be interrupted.

When a patient falls into the status epilepticus it is advisable to inject a large dose of bromide of potassium or of chloral into the rectum. If these drugs fail the hypodermic injection of one hundredth of a grain of hyoscine hydrobromate may be successful. But as a rule the seizures can only be arrested by the inhalation of chloroform; the subsequent exhaustion may be reduced by a subcutaneous injection of strychnine.

CHAPTER IV.

RAYNAUD'S DISEASE.

This affection is characterised by temporary and recurrent attacks of retardation of the blood supply in certain parts, resulting in local pallor, in local cyanosis, and if the retardation is adequate in local gangrene. These changes which mainly affect the extremities and are generally bilateral and symmetrical, appear to be dependent on spasmodic contraction of the arterioles.

Etiology. The disorder is more common in females than in males. It may begin at any age, even in childhood or in advanced life, but most frequently the first attack occurs in early adult life. The patients are usually of a nervous disposition, and there is often a family history of other neuroses and occasionally of the manifestations of the disease itself. In some cases malaria appears to have a predisposing influence: in a few syphilis or diabetes. The chief exciting cause is exposure to cold. The attacks are more liable to come on in winter than in summer, although in severe cases they may be precipitated, even in hot weather, by a slight variation in the temperature, as by the patient passing from a warm room to a cool one. Sometimes an attack is brought on by emotional disturbance, by injury, or by gastric or intestinal disorders.

Symptoms. Three stages or forms of the disease may be distinguished:—The stage of local syncope, in which there is a pallid paroxysmal condition of the extremities; the stage of local asphyxia, in which there is a cyanotic paroxysmal condition of the extremities, and lastly the stage of local gangrene.

Local syncope. This change may represent the whole of the disease or only its first stage. It is analogous to that produced in a healthy person by exposure to extreme cold, occurring, however, in a person predisposed to Raynaud's disease on exposure to slight cold. The change consists in a rapidly developing pallor of the fingers or the toes either at different times or contemporaneously. It may be limited to the index or little finger, or involve more than one finger of each hand: occasionally the pallor extends to the wrists and ankles, and exceptionally to a higher level. The patient complains of a feeling of deadness and numbness in the affected parts, which usually show some impairment of sensation to painful impressions, and slight blunting of the tactile sensibility. Owing to these sensory defects as well as to stiffness of the fingers the patient has a difficulty in the performance of delicate movements

such as those required for sewing or the picking up of small objects. During the attack, which lasts from a few minutes to several hours, the pulse may remain normal or become small and scarcely perceptible. It is, however, in the arterioles that slowing of the blood current mainly occurs; this is shown by the absence in severe cases of bleeding when the pallid part is punctured by a needle.

In the mildest cases the attack ends without any noticeable reaction, but in the severest cases the return to normal is attended by burning pains, great intolerance of pressure, flushing and sometimes by local perspiration.

Local asphyxia. In this phase of the disorder the extremities are subject to paroxysmal attacks of cyanosis; the discoloration, which varies from a purplish-red to a bluish-black, is usually preceded by a slight and transient pallor. This cyanotic condition may be limited to one or two corresponding fingers or toes, or may gradually invade the hands and feet and extend as high as the elbows and knees. The four extremities may be affected simultaneously or successively; symmetry is generally observed, although there is often a decided inequality in the degree of implication of corresponding parts on the two sides. The discoloured extremities feel extremely cold; they are tender to pressure and often the seat of considerable pain. There may be analgesia and blunting of the tactile sense. The veins on the back of the hand are often distended; sometimes a slight degree of œdema is present. The pulse may remain unaltered during the attack, or it may feel smaller than natural, becoming full during the subsidence of the paroxysm. In some cases reddish-purple patches appear on the limbs, at a little distance from the affected parts. The helix of the ear may also suffer, either simultaneously with the hands or feet, or without these parts being involved: occasionally the nose or the zygomatic regions become discoloured.

An attack may last from a few hours to several days,

the degree of discoloration varying from time to time; during its subsidence the skin becomes red, hot and burning, and there may be a sudden outbreak of moisture over the affected part. The temperature in the mouth, usually normal during a paroxysm, may be slightly raised for a day or two after it is over. In severe cases the attacks occur in cycles, the intervals between them becoming progressively shorter until after a few months or years the condition becomes permanent when there is a risk of gangrene supervening.

Local gangrene. We have seen that the morbid process may be limited to a series of attacks either of local pallor or of local cyanosis, in which the affected parts become cold and present a dead white or a livid appearance, and are frequently the seat of severe pains resembling those of frost-bite. We have also seen that these colour changes are excited by very slight variations in the surrounding temperature, and tend to become marked and prolonged out of all proportion to the stimulus which starts them. When there is a further increase in the severity of the morbid process the state of local pallor, or more commonly that of local cyanosis, is succeeded by gangrene. This is generally of the dry variety and is remarkable for its limited and superficial distribution; very rarely it extends deeply and widely, but, although it may reach the bone, it is quite exceptional for any necrosis of bone to occur. The onset of the gangrenous process is indicated by an increase in the coldness of the affected part, and of the pains which may become almost unbearable. The end of one or more digits of the hands or feet becomes black and anæsthetic, and undergoes a gradual mummification. In some cases small bullæ form on the skin, which burst and discharge a blood-stained fluid; after their collapse the underlying skin is seen to be black, and a small sequestrum of necrosed tissue is gradually formed which is ultimately separated by the zone of ulceration which surrounds it. As a rule the loss of structure is small, and is limited to the end of a finger or a toe. Eventually

cicatrisation occurs, and if the part be examined some months later a small white scar may be the only sign of the previous necrosis.

Associated conditions. A very close relation has been observed between Raynaud's disease and paroxysmal hæmoglobinuria. If the blood drawn from a region affected with local cyanosis is examined it will be found that the corpuscles do not form rouleaux, and are much shrunken and crenated, whilst the serum contains hæmoglobin. Hence it is not surprising to find that hæmoglobinuria may be observed either during attacks of Raynaud's disease, or alternating with them. As pointed out by Barlow, in both affections the attacks are paroxysmal in character, are related to changes of temperature and may be accompanied by abdominal pain, enlargement of the spleen, slight pyrexia, a sallow complexion and a little yellowness of the conjunctiva.

The following conditions, though less frequently than hæmoglobinuria, are also found in association with Raynaud's disease:—Urticaria and generalised sclerodermia; effusion into the joints, and fibrous ankylosis of the smaller ones; thickening of the palmar fascia; cerebral symptoms, including transient paralysis, delusions and epileptic seizures; and transitory amblyopia with narrowing of the retinal arteries and pulsation of the veins.

Pathology. Raynaud sums up his view of the pathology of the disease by saying: "That in the present state of our knowledge local asphyxia of the extremities ought to be considered as a neurosis characterised by enormous exaggeration of the excito-motor energy of the grey parts of the spinal cord which control the vaso-motor innervation."

This view comprises two assumptions, viz.: (1) There is disturbed vaso-motor innervation. (2) That this depends on altered energy of the grey matter of the cord. The first assumption is strongly supported by all the evidence in our possession, and it is pretty generally admitted that whatever may be the exact pathology of

the local gangrene, the phenomena of local syncope and local cyanosis are the direct results of spasm of the small vessels. As already mentioned, spasm of the retinal arteries has been seen, and Barlow in a well-marked case of Raynaud's disease occurring in a little girl, observed contraction of some of the veins on the back of the affected hand: during one attack "they became quite moniliform, that is to say, there was an alternation of small dark swellings with narrow almost colourless intervals between them, and under observation the dark swellings gradually altered their position along the course of the veins, pointing to a varying contraction of the walls of these vessels."

But the second assumption is merely a hypothesis, and calls for discussion. Arterial spasm may be produced by stimulation of (*a*) certain parts of the cerebral grey matter; (*b*) the vaso-motor centre in the medulla; (*c*) the subordinate centres in the cord; or (*d*) the vaso-motor nerve fibres which are said to run in the peripheral nerves.

A discussion, then, of the pathology of Raynaud's disease is mainly concerned with considerations as to which part of the vaso-motor nerve tract is primarily affected, and as to the nature of the stimulant and the method in which it is brought to bear on this department of the nervous system. Information on these points is to be derived from the results of post-mortem examinations, as well as from clinical observations on the disease in question, and on other affections presenting similar symptoms.

Pathological evidence. This is very meagre, being based on the examination of only a few cases. An analysis of the post-mortem findings in seven typical cases of Raynaud's disease in which gangrene was present shows that the peripheral nerves were normal in two of the cases and definitely degenerated in the remaining five. The spinal cord, which was examined in five of the cases, presented slight changes in one case, a diffuse sclerosis in another case, and was healthy

in the remaining three cases. The arteries were found to be normal in four, and altered in two cases.

It appears, then, that as regards cases of the disease in which gangrene was present, lesions of the peripheral nerves were commoner than lesions of the cord or vessels. But that they are not essential to the disease is clearly shown by the two cases in which the peripheral nerves were quite healthy. One of these cases, recorded by Burlow, is particularly important on account of its detailed clinical history, which proves that the case was a typical instance of Raynaud's disease, and of the careful examination of the nerves and vessels. The trunks, as well as the smaller branches of the nerves going to the gangrenous foot, were found to be perfectly normal; there was, in fact, no evidence whatever of peripheral neuritis. The case is, then, sufficient in itself to upset the doctrine that Raynaud's gangrene always depends on a peripheral neuritis. The arteries in Burlow's case presented changes, but in other cases of Raynaud's gangrene they have been found unchanged, so that arterial lesions can also be excluded as essential causes of the disease.

Clinical evidence. Vaso-motor disturbance occurs in consequence of lesions of single nerves, and is met with in conditions such as alcoholic paralysis known to depend on multiple neuritis. In the former case vascular disturbance and skin lesions affect the territory supplied by the particular nerve, but these changes do not occur paroxysmally, as in Raynaud's disease, and need not be further considered. In alcoholic paralysis evidence of vaso-motor spasm is common enough. At an early period of the disease the fingers and toes become cold, dead, and white, or, at times, quite livid, while in severe cases gangrene may develop. It is highly probable that these phenomena, which are identical with those that constitute Raynaud's disease, are the result of multiple neuritis: but there is no direct proof of this, and it is possible that while the paralytic phenomena of alcoholism are due to disease of the

peripheral nerves, the vaso-motor disturbances are related to co-existing changes in the grey matter of the cord.

The ordinary symptoms of peripheral neuritis, viz., pains, anæsthesia, muscular weakness and wasting, are occasionally present in conjunction with vascular disturbance in cases of Raynaud's disease. Sometimes the initial pains are very severe and the subsequent anæsthesia may be well marked, both as regards distribution and degree. But none of the symptoms enumerated are constant in the disease; indeed they are more often absent than present.

Summary. The cardinal feature of Raynaud's disease is the occurrence of vascular disturbance in peripheral parts on exposure to cold. Paroxysmal attacks of local syncope or cyanosis, succeeded or not by local gangrene, are often met with quite apart from the ordinary symptoms of peripheral neuritis. Sometimes, however, symptoms of neuritis are unmistakably present, and sometimes marked changes in the peripheral nerves going to the affected parts are found on post-mortem examination. But the inconstancy both of the clinical and pathological evidence of peripheral neuritis points to some other cause for the explanation of the typical phenomena of Raynaud's disease.

The early paroxysmal phases of the disease are instances of reflex action developed with abnormal facility. Cold, the chief cause, irritates the morbidly active vaso-motor centres in the brain and cord. The result is spasm of the arterioles. Repeated and protracted attacks of vascular spasm may initiate actual disease of the walls of the small vessels. The peripheral neuritis which is sometimes present in Raynaud's disease, may be the direct result of arterial disease, or it may occur independently of this, the nerve degeneration being due to the presence of some toxin in the blood, to an imperfect supply of blood owing to the vascular spasm, or possibly to changes in the central cells which preside over the nutrition of the peripheral nerve endings.

But, although this appears to be the most reasonable view to be taken of the pathology, the possibility that there is more than one variety of the disease must not be overlooked. It may be that whilst in most cases the vaso-motor centres are chiefly at fault, there are other cases in which the vaso-motor nerves are mainly implicated and still others in which the whole vaso-motor tract is involved, nerve cells, as well as nerve fibres, being simultaneously or successively affected.

Diagnosis. Similar changes in the colour and nutrition of the extremities occur in other diseases, but not in recurrent attacks as they do in Raynaud's disease. The pallor of anæmia is general and persistent rather than local and transient. The blueness of congenital heart disease is also persistent and affects the nose, lips and tongue and other parts as well as the extremities; moreover, there is a marked clubbing of the ends of the fingers and toes; whereas in local cyanosis the blueness of the extremities is intermittent, the lips and tongue are not affected, and the finger-ends are tapering rather than bulbous.

Local cyanosis of a toe may closely resemble a chilblain, but the latter rarely implicates the extreme end of the digit as the cyanosis does; in chilblains also there is swelling with inflammatory exudation.

Senile gangrene may sometimes suggest the presence of Raynaud's disease, but as a rule the senile ailment is limited to the lower extremity, and tends to implicate the deep as well as the superficial tissues; generally it is associated with signs of arterio-sclerosis, whereas in cases of Raynaud's gangrene the arterial walls feel normal and there is little or no impairment of the arterial pulsations.

Treatment. Persons who are liable to Raynaud's disease should avoid fatigue and exposure to cold, and if obliged to go out in cold weather should take some nourishing food before starting and protect themselves by warm clothing, including woollen gloves and stockings. The tendency to the attacks may also be

lessened by paying attention to the state of the mouth, to digestion, and to the regulation of the bowels.

During an attack the painful spasm may often be overcome by immersing the affected part in a solution of salt and warm water in which the positive pole of a galvanic battery is placed, while the negative pole is applied to the upper part of the limb. The current should be frequently made and broken, its strength being sufficient to induce moderate contractions of the muscles.

The pain may be so intense as to require morphia; occasionally beneficial results are obtained by the taking of nitrite of amyl, nitro-glycerine and other vaso-dilators. In the intervals between the attacks massage, galvanism and physical exercises, by improving the nutrition of the limbs, are often of considerable service, especially if persevered with for a long time.

When gangrene is threatened opium is of value; the gangrene itself should be treated on surgical principles, simple protection, and antiseptic precautions being needed for the benign forms, amputation for the severest.

CHAPTER V.

ERYTHROMELALGIA.

The essential feature of this chronic and somewhat rare disease is pain, followed by redness and elevation of temperature in one or more of the extremities, the symptoms being increased by a dependent position of the limb and by muscular exertion.

Etiology. The disease is more common in males than in females, and in middle life than at other periods; it is very rare in children. It is stated to be more apt to occur in persons of Jewish descent than in others. The chief predisposing causes are exposure to changes of temperature and to physical fatigue, especially that produced by long hours of standing or by long walks. Infectious diseases, such as malaria and gonorrhœa,

have also been credited with predisposing to the malady. During the epidemic of peripheral neuritis produced by drinking beer containing arsenic many of the patients showed patches of erythromelalgia on the palms or soles. The condition has also been met with in association with disseminated sclerosis, tabes, myelitis, syringomyelia and with hysteria. In some cases a local injury has seemed to determine the onset, and the seat of the symptoms.

Symptoms. As a rule the symptoms are limited to one extremity, the foot being more frequently involved than the hand. In a typical case the initial symptom is a burning pain in the sole of one foot, which is relieved by lying down or by raising the leg to the horizontal position. This pain may constitute the whole of the attack, and may vanish altogether if the patient is able to rest for a few days. But if he is obliged to stand or walk the pain is apt to recur in attacks of increasing severity, and to be accompanied by vascular disturbance, the painful part of the sole becoming red, hot and slightly swollen; the veins are distended, the arterial pulsations are excessive, and the skin may be covered with perspiration. The surface temperature of the affected foot is higher than that of the unaffected one, and is greater when the limb hangs down than when it is elevated, which is the reverse of that which obtains in health. There is usually increased sensibility to both deep and superficial pressure. During the course of the attack the colour of the affected area gradually deepens, a rosy-red gradually changing into a dark purplish-red hue.

The symptoms are induced or intensified by a dependent position of the limb, by hot weather and by warm applications; whilst they are relieved by elevation of the limb, by cold weather and by cold applications. Walking is often very painful, and the patient may be noticed to tread on the unaffected part of the foot; in very severe cases he is obliged to get about on his hands and knees. The

attack generally lasts three or four hours, but it may be of shorter or of longer duration, in some cases lasting for a few weeks. After the disease has become established the attacks are apt to set in spontaneously, without any such determining cause as hot weather or physical fatigue.

The hand is less commonly and as a rule less severely affected than the foot; when it is very painful and congested the patient tries to obtain relief either by raising the hand above his head or by placing it across his chest. Exceptionally, patches of erythromelalgia are found on the face, the neck or on the trunk.

In consequence of the pain and the enforced disuse

Fig. 144.—Diagram showing the distribution of the reddened surface in a case of erythromelalgia (Weir Mitchell).

of the affected limb, there may be a slight general wasting of its muscles, but localised muscular atrophy does not occur, nor is the reaction of degeneration ever obtained. In long-standing cases atrophic changes in the skin and the nails are sometimes met with; very rarely gangrene has occurred. As a rule the sufferer from erythromelalgia is free from signs of any other disorder, but occasionally his psychical condition becomes changed, either temporarily or permanently, and the change may be associated with defects in speech, with hemiplegia, or with some affection of the spinal cord. The prognosis of the malady is unfavourable, the symptoms usually show a tendency to get worse and

to become permanent; very rarely they disappear spontaneously.

Pathology. The essential nature of the disease is obscure. Some authorities regard it as a variety of peripheral neuritis, others as a consequence of changes in the blood-vessels. The usual symptoms of peripheral neuritis, namely, localised anæsthesia and localised paralysis with wasting of the muscles, are absent, so that if the pain and the vascular symptoms which characterise erythromelalgia are due to neuritis, this must be limited to or must mainly affect the vaso-motor fibres of the peripheral nerves. The evidence of post-mortem examinations is much more strongly in favour of the malady being a disease of the blood-vessels. In several cases the small arteries of the affected extremity showed thickening of their walls, the middle and inner coats being chiefly involved. The small veins were also thickened, and those of the skin in two cases (Barlow) were almost occluded by changes in the intima and by thrombosis.

It may be that the primary lesion is an irritative one of the nerve centres which govern the blood-vessels, and that the ensuing vaso-motor disturbance gradually leads to permanent degenerative changes in the walls of the vessels involved. Such changes, accompanied by varying degrees of spasm and dilatation in collateral vessels would account for the alternating states of temperature and colour in the affected parts. According to this view the degeneration of nerve fibres, which is occasionally observed, is secondary to the interference with the blood supply, or possibly to pressure on the nerves from an associated overgrowth of the connective-tissue.

Diagnosis. Although there are many points of similarity between erythromelalgia and Raynaud's disease there are several important distinctions. Erythromelalgia tends to be unilateral, Raynaud's disease to be bilateral and symmetrical. Erythromelalgia, though liable to remissions and exacerbations is less paroxysmal

in character than Raynaud's disease, and is apt to persist with little change, except for the worse, for many years; its chief determining cause is physical fatigue, that of Raynaud's disease is exposure to cold. Erythromelalgia is more prevalent in males; Raynaud's disease in females. In addition to the differences presented by the two diseases as regards the colour and the temperature of the affected part it is to be noted that while permanent swelling of the extremity is common in erythromelalgia, there is a tendency to atrophy in Raynaud's disease.

Barlow suggests that the distinction which "underlies the clinical differences is that Raynaud's disease is primarily a symmetrical vaso-motor affection, although ultimately permanent disease of the vessel-walls may supervene; whilst in erythromelalgia there is primary localised disease in arterioles and venules of the skin, subcutaneous tissues and muscles, with varying accompanying vaso-motor disturbance of the collateral vessels."

Treatment. As soon as the disease is recognised or even suspected the patient should be sent to bed and kept there for some weeks in the hope that complete rest together with attention to the general nutrition and to the elimination of morbid products may lead to an arrest of the malady. If, owing to the necessity of going to work or for other reasons, the patient is unable to stop in bed, he should at all events rest as much as possible, the affected limb being kept in the horizontal position. Temporary relief to the pain is often afforded by cold applications, but in severe cases nothing but morphia will probably be of service.

In the traumatic cases the possibility of pressure on the vessels by thickened tissues will suggest the desirability of making an incision in order that the affected part may be carefully explored. The results of excision of a piece of the nerve to the part and of amputation of the limb are not encouraging. At the same time, amputation may be advisable if in a severe and protracted case there is progressive exhaustion and intense suffering which is only temporarily relieved by the administration of morphia.

CHAPTER VI.

INTERMITTENT LIMP. INTERMITTENT CLAUDICATION.

This affection, which is closely allied to erythromelalgia, is characterised by recurrent attacks of weakness, pain, paræsthesia and of a varying amount of vaso-motor disturbance usually in one leg, the attacks being brought on by muscular exercise, and subdued, at least in the early stages of the disease, by rest.

In a typical case a man in early middle life and previously healthy, begins to suffer during a walk, either suddenly or gradually, from stiffness and pain in the foot and leg, which cause him to stop. On resting, these symptoms pass away, but if he resumes his walk they return; the pain becomes more severe and the leg feels weak, cold and numb and may be the seat of painful cramps which compel him to limp and eventually to sit down. On examination the foot and leg are found to be cold and either pale, or congested and swollen: there is an absence of pulsation in the dorsalis pedis and generally also in the posterior tibial artery.

If the man can now be persuaded to take a long and complete rest and subsequently to avoid much exertion the progress of the malady may be delayed or even arrested altogether. Unfortunately, however, such a result is exceptional, the rule being that the symptoms return and persist for years with little or no sign of improvement. In a few cases gangrene of the foot has ensued. As a rule only one leg is affected, but occasionally the symptoms are experienced in both legs: very rarely the arm is involved.

The symptoms appear to depend on two factors, namely, a sclerosis of the small arteries and veins of the affected part and disturbance of the vaso-motor mechanism. A definite obliterative endarteritis is an almost constant feature, the large vessels being sometimes affected as well as the small. The muscles and

nerves are usually healthy, but a slight general wasting of the muscles and some degeneration of the nerve fibres has been occasionally observed, apparently in consequence of disease of the vasa nervorum. In some cases the vascular degeneration appears to have been brought on by alcohol, the excessive use of tobacco, gout or by syphilis; in others the interference with the circulation in the limb has been caused by the pressure of a truss or by an aneurysm of the femoral artery.

From a consideration of the above facts it may be inferred that the subsidence of the symptoms when the limb is at rest is due to the maintenance of an adequate circulation of blood in the affected part by means of healthy vessels, whilst the development of the symptoms during exertion is due to a supply of blood inadequate to the needs of the muscles brought into action, the inadequacy being the result partly of disease of some of the vessels and partly of an associated vascular spasm produced by irritation of the vaso-motor nerves or of their centres.

The treatment consists in the avoidance of alcohol, tobacco and of all other preventable causes of vascular degeneration, and in the taking of as much rest as possible; the amount of exercise to be allowed must always be less than is found to bring on the symptoms. The limb should be kept warm; gentle massage and the high frequency current are stated to be beneficial. The administration of the iodides or of the nitrites has not proved to be of much service. When gangrene supervenes amputation of the limb may be necessary.

CHAPTER VII.

RECURRENT PARALYSIS.

Temporary recurring attacks of paralysis are occasionally met with, the pathology of which is not always clear. Some persons on waking from sleep complain of weakness in the limbs or of an inability to

raise the eyelids, which may last for a few minutes or longer; such attacks are prone to occur in the subjects of neurasthenia, or of hysteria, and in those who smoke to excess. Recurring attacks of hemiparesis, of aphasia or of difficulties in articulation or in swallowing may occur in the early stages of disease of the intra-cranial arteries as a result of syphilis or atheroma: they are often followed by a permanent paralysis corresponding in distribution to the transient weakness. Very rarely a patient suffers from more than one attack of multiple neuritis; in one case there were five separate attacks. Transient paralysis in the form of monoplegia or hemiplegia may attend uræmia or follow an epileptic seizure. It is also rarely observed in cases of Raynaud's disease and of migraine.

Attacks of migraine are sometimes accompanied or followed by paralysis of the muscles supplied by the third cranial nerve. This association of recurrent oculo-motor paralysis with migraine is called *ophthalmoplegia migraine*. But it is to be noted that sometimes the recurrent paralysis occurs apart from typical symptoms of migraine, although it is usually preceded by headache, vomiting and severe pain in or over the eye. The paralysis is generally one-sided, there is ptosis with weakness or complete paralysis of some or all of the internal and external muscles of the eyeball, the fourth and sixth nerves being sometimes affected as well as the third nerve. Occasionally amblyopia is present and exceptionally paresis in the distribution of the facial nerve. Between the attacks there is either no paralysis or a persistence of the weakness which may become permanent.

There are also rare cases of facial paralysis in which, after recovery, one or more attacks of paralysis occur on the same side, or the opposite side of the face. But the most remarkable instance of recurrent paralysis is afforded by the disorder known as "family periodic paralysis," of which the following is a brief account.

Family Periodic Paralysis.

This rare affection is characterised by recurrent attacks of more or less extensive paralysis, with loss of the reflexes and of the electrical excitability of the muscles; between the attacks the condition of the patient is quite normal.

Etiology. The onset of the malady is usually about puberty. The most constant exciting cause of an attack is physical fatigue; other assigned causes are emotional disturbance, over-indulgence in rich foods, gastric or intestinal disturbances, and injury. But in many of the cases the attacks come on during a condition of perfect health. The most striking feature in the etiology is the hereditary nature of the disease; in one family eleven cases were observed in five generations, in another seventeen cases in four generations.

Symptoms. The attacks consist of a gradually extending paralysis which begins in the legs and then spreads to the arms, trunk and neck so that eventually the patient lies quite helpless, being unable to move his limbs, body or head. The proximal muscles of the limbs usually become paralysed before the distal ones. Very rarely there is ptosis, or a difficulty in speaking or in swallowing, but in the great majority of cases the muscles supplied by the cranial nerves are not affected. The paralysed muscles give no response to either faradic or galvanic stimulation. The superficial reflexes and the tendon reactions are temporarily abolished. There is no impairment of sensation, of the special senses, or of consciousness. During the attack the heart may enlarge, the dilatation being sometimes accompanied by a mitral systolic murmur. Mild attacks may be represented by a partial or a limited paralysis: sometimes only the legs or the arms are involved. The following symptoms have been occasionally observed: pallor of the face, congestion of the conjunctiva, and profuse sweating.

The attacks may begin during the daytime, but as a rule they develop at night during sleep, so that the

patient wakes up to find himself unable to move in bed. They vary in duration from a few hours to several days: they may occur daily or only every three or four months. Their disappearance, like their onset, is usually gradual, the order in which the muscles recover being the reverse of that in which they became affected: voluntary power returns and the electrical reactions become normal. Between the attacks the patient is quite well and presents no signs of disease either of the nervous or of the muscular system.

Pathology. During the attacks of paralysis it has been observed that the blood shows a moderate degree of lymphocytosis and that the urine is deficient in quantity and contains less than the normal amount of urea, uric acid and sulphates. It is also stated that the excretion of creatinine is diminished both before and during an attack, whilst it is increased after the attack. These changes, together with the temporary duration of the paralysis and its association with sweating and sometimes with gastro-intestinal disturbance, suggest that the attacks depend on the pressure of some toxic substance in the blood. That the toxic substance acts upon the muscles rather than upon the nervous system is indicated by the loss of their direct irritability to electrical and mechanical stimulation during the paralysis; and by the changes, namely, marked fissuring and vacuolation, which have been found in excised portions of an affected muscle.

It seems probable, judging from the marked hereditary character of the disease and its frequent onset in early life, that there is some congenital defect of muscular tissue which makes it abnormally susceptible to toxic influences.

Course and Prognosis. As age advances there seems to be a tendency for the attacks to become less frequent and less severe. Much dilatation of the heart is a serious sign, and, although the outlook as regards life is generally favourable, one or two cases have died in consequence of cardiac failure.

Treatment. The patient should lead a quiet life, and avoid over-exertion, exeitement and indiscretions in diet, taking every eare to keep the system in the best possible state of health. During an attack it is advisable to administer a purge together with diuretics in order to promote the elimination of toxic substances. Bromide of potassium with caffeine has also been recommended. Digitális and strychnine may be required for the cardiac weakness, and artificial respiration and oxygen for embarrassed or failing respiration.

SECTION XII.

Disorders of Speech.

As already indicated (p. 84), disorders of speech produced by disease of the nervous system may be classed into motor and sensory varieties. Motor disorders may be caused by lesions either (1) of the higher centres for the movements associated with speech and writing which are situated respectively in the posterior ends of the third and second frontal convolutions; or (2) of some portion of the motor apparatus for the expressive side of speech. This is composed of the following neurons: The centres in the lower portion of the ascending frontal convolution which preside over the movements of the lips, tongue, vocal cords and hand; the pyramidal fibres which connect these centres with the medulla and spinal cord; the bulbar and spinal nuclei and the motor nerves which proceed from them to the muscles used in articulation, in phonation and in writing.

The defects in speech present marked differences in the two varieties. When the region of the third frontal convolution—known as Broca's area—is affected there may be no defect in articulation, but the words uttered though correctly pronounced are limited in number and imperfect in arrangement; they do not form intelligible phrases or sentences, nor do they express correctly any idea the patient may wish to convey. Such a defect is called *motor aphasia*. When, however, the functions of either the upper or the lower neurons for the muscles employed in speaking are interfered with the defect is one of articulation; there is either *dysarthria*, that is, disordered articulation, or *anarthria*, in which the articulatory defects are so great that speech becomes impossible or unintelligible.

In addition to the defects produced by lesions in the two chief divisions of the motor mechanism for speech,

a third defect must be recognised, namely *aphemia*. In complete aphemia the patient is absolutely dumb, being unable to say even the simplest words, such as "yes" and "no"; in partial aphemia the patient's utterance is limited to a certain number of words; these, although they may be badly pronounced, are used appropriately, and the patient neither makes use of wrong words nor gives utterance to recurring phrases. There is thus a marked difference between the speech of a patient suffering from aphemia and that of one afflicted with motor aphasia. Aphemia occurs as the result of isolation of Broca's centre, by interruption of the paths which connect it with the primary centres for speech in the precentral gyrus and with the pyramidal fibres which proceed from those centres.

Sensory disorders of speech are almost invariably due to lesions involving the convolutions in the temporo-occipital region of the cortex; they rarely attend disease of the afferent tracts to this region. Such disorders are included under the term *sensory aphasia*, of which two main varieties may be distinguished. In the one variety called word-deafness the patient is unable to understand spoken language, and the lesion is situated in the posterior part of the first temporal convolution; in the other variety, called word-blindness, he is unable to understand written or printed language, and the lesion is situated in the angular and partly in the supra-marginal gyrus.

Aphasia then refers to defects of speech that result from destructive lesions of certain convolutions in one of the cerebral hemispheres. In right-handed persons the specialised centres subserving speech are developed in the left hemisphere, in left-handed persons in the right hemisphere. It is therefore the convolutions of the left hemisphere which are affected in most cases of aphasia. It must, however, be borne in mind that both hemispheres take part in speech processes, especially as regards the reception of speech impressions. This is particularly indicated in young children in whom

aphasia is rarely lasting, destruction of the speech centres in the left hemisphere being quickly compensated for by increased activity on the part of the right hemisphere. In the adult compensation is less striking; its degree, however, varies much in different individuals. Further, partial recovery from aphasia may depend on retrogression of the morbid process as well as on the development of compensatory action by the healthy hemisphere.

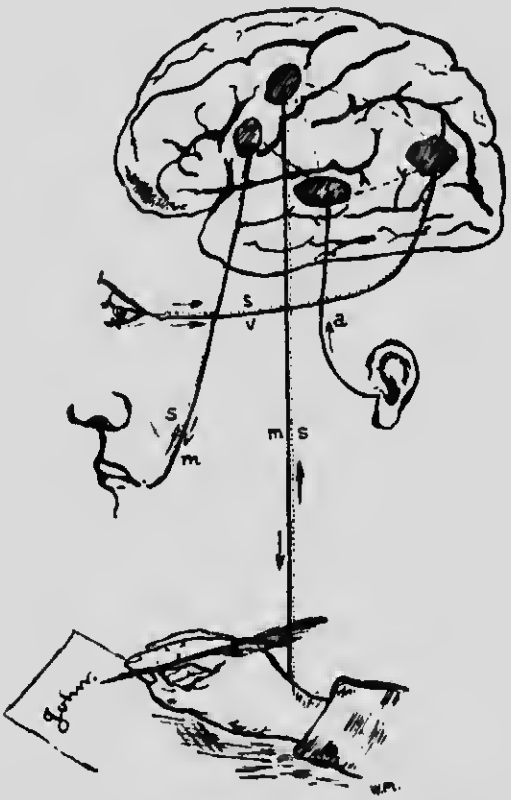


Fig. 145. The co-ordinated cortical mechanism for speech processes. From the eye and ear centripetal fibres (v and a) ascend to terminate in the angular gyrus and first temporo-sphenoidal convolutions respectively, but in reality these fibres are directly connected with a much larger area of the cortex than is here indicated. In addition to these, fibres of muscular sense (s, s, and s), indicated by dotted lines, ascend from the muscles of articulation, from those of the hand and from those of the eyeball, to reach the cortex. The centres of vocal and written expression are connected by means of centrifugal fibres, m and m, with the vocal apparatus and hand respectively.

The four main speech centres are to be regarded as specialised portions of the cortical centres for hearing, vision and for the movements concerned in speaking and in writing. They are (1) the auditory word centre situated in the posterior half or two-thirds of the first temporal convolution; (2) the visual word centre in the angular gyrus and its immediate neighbourhood; (3) the glosso-kinæsthetic centre in the posterior part of the third frontal convolution, and (4) the cheiro-kinæsthetic centre in the posterior part of the second frontal convolution. Commissural fibres connect these centres together, the connection being in all probability most intimate and complicated in highly educated persons. In these centres, word-memories are stored which in all cases are of sensory origin.

The auditory centre is the first to be developed, for the child learns to understand many words before he is able intelligently to utter them. The acquirement of auditory word-memories gradually leads to the organisation of the glosso-kinæsthetic centre, with the result that the child begins to talk, and eventually the centre becomes a store-house for memories of articulation and phonation. Later, as the child learns to read, he slowly associates the words seen with certain objects and ideas and thus acquires visual word memories which are stored in the angular gyrus. Similarly in the subsequent acquisition of writing, the cheiro-kinæsthetic centre becomes organised, and the store-house of sensory memories accompanying the movements of the hand in writing.

Motor Aphasia. In complete motor aphasia, which results from destruction of Broca's area, there is inability to express language in speech, and usually in writing; sometimes there is also impairment of the power to express thoughts by signs or gestures. The essential peculiarity is the loss of voluntary speech; the memories of words spoken and written are lost, but those of words heard and seen are retained. Thus the patient, while understanding everything that is said to him and whilst

retaining more or less his previous powers of understanding written words, is incapable of speaking spontaneously and of repeating words spoken to him. But, although speechless, he is not always wordless; under the influence of strong emotion he may give utterance to words, phrases and oaths which are of an interjectional rather than an intellectual character and which the patient cannot voluntarily repeat when asked to do so. Such ejaculations are often the same for the same patient, and have therefore been called *recurring utterances*; they are usually either the words actually spoken, or those about to be spoken when the damage to the brain occurred. Thus the recurring utterance of a librarian was "List complete" (Russell); that of a girl attacked when riding on a donkey, "Gee, gee" (Hughlings Jackson), that of a man injured in a brawl, "I want protection." Such utterances are probably produced by the action of an undamaged portion of the centre or by the intervention of the third frontal convolution on the right side.

When the aphasia is incomplete the patient is often quite aware of his errors in speaking; the utterance he has may be clear and distinct, and in singing he may be able to utter every word of a song, although unable to speak it. His powers of thinking are not materially lessened, though careful observation will often detect some impairment of the mental faculties. The degree of aphasia varies greatly in different cases and every gradation may be met with between the most severe cases in which the patient replies to questions by grunting sounds or meaningless syllables, and very slight defects, which are only indicated by a hesitating utterance and by casual mistakes in the use of words.

Owing to the close proximity of Broca's centre to the writing centre, motor aphasia is very frequently accompanied by *agraphia* (inability to write) and sometimes by *alexia* (inability to understand words seen). Agraphia may be present when there is an isolating lesion of the motor speech centre, the explanation being that in

writing the motor word representations are always revived by impulses from the percipient auditory or visual centres which pass through the motor speech centre to the writing centre. In all probability agraphia may also occur as a result of a destructive lesion limited to the writing centre in the second frontal gyrus.

Errors in writing are usually greater than errors in speaking. Their degree varies much in different cases; one patient, although his hand is not paralysed, may be unable to write a single word or form a single letter; another, while unable to write to dictation or to copy sentences, may be able to sign his own name; a third patient can write to dictation and copy from a written or printed page, but has lost the power to express his thoughts in writing; whilst a fourth patient can spontaneously write a few words and simple sentences, which, however, are rarely free from mistakes in spelling or diction.

Alexia is occasionally met with in cases of motor aphasia. Sometimes it is accounted for by a wide-spread lesion, involving the angular as well as the third frontal gyrus; but the disability may occur when the lesion is limited to Broca's area, especially in imperfectly educated persons who are unable to understand a passage without either muttering the words to themselves or making some movements of the lips, that is, without the help of motor speech processes.

Motor aphasia is frequently associated with hemiplegia, because it is common for the lesion which involves Broca's convolution to extend into the precental gyrus and its subcortical fibres.

Sensory Aphasia or Verbal Agnosia. Under this heading are included defects in speech and writing due to impaired recognition of auditory and visual word-stimuli, or to defective recall of the auditory and visual images of words; they are produced either by lesions of the convolutions situated around the posterior end of the Sylvian fissure, which, it is to be noticed, are

supplied by branches of the same artery; or by lesions of the paths by which these convolutions are united to one another and to corresponding subsidiary centres in the opposite hemisphere.

The auditory and visual word-centres may be affected either together or separately, but even in the latter case, owing to their proximity and to their intimate functional relations, a lesion occupying the one centre is very apt to interfere with the functions of the other centre. Hence the symptomatology of cases of sensory aphasia is very varied, and even with lesions similarly situated clinical differences occur in relation to different degrees of education and to the relative activity of these centres in the persons attacked. In most persons—called *auditives*—the subjective revival of words takes place in the auditory word-centre, but in others—called *visuals*—words are mainly revived in the visual word-centre, and as between auditives and visuals every transition is met with, the clinical manifestations of similar lesions are prone to be varied and complicated. For these reasons it is only possible in a short account of sensory aphasia to indicate the most common combination of symptoms in relation to the position of the lesion.

Word-deafness may result either from damage to the left auditory word-centre or to its isolation in consequence of a subcortical lesion.

(1) *Disease of the centre.* In all cases word-deafness occurs. As in other derangements of speech-processes this varies in degree according to the amount of damage to the centre and according to the amount of compensation that ensues, which appears to be largely dependent on whether the patient is a strong auditive or a strong visual.

When the centre is completely destroyed, speech may be profoundly affected on its expressive as well as on its receptive side; as a rule there is also considerable impairment of the mental faculties. With regard to the receptive side there is complete word-deafness.

The patient may hear sounds as well as formerly, but he fails to understand spoken words, which appear to him as mere meaningless sounds. An uttered request to close his eyes or to hold out his hand unless accompanied by suggestive gesture is not attended to; he hears that some one is speaking, but the words do not give birth to corresponding ideas.

Disorders on the expressive side present many variations. In strong auditives, in whom the recall of words takes place almost entirely in the auditory word-centre, voluntary speech is considerably impaired and may be represented only by unintelligible jargon. In such persons there is also much disability in regard to reading and writing, owing to the associated imperfectly developed functions of the visual word-centre. In strong visuals on the other hand the revival of words in silent thought goes on to some extent in the visual word-centre which may thus rouse into activity the glosso-kinæsthetic centre. Hence in such cases a considerable amount of voluntary speech may be preserved, in which but few errors can be detected: frequently also the ability to read and to write is retained.

When the auditory word-centre is only partially damaged the interference with speech processes is less marked. Word-deafness is incomplete and the patient may be able to repeat words he hears, though often incorrectly. Of these errors he is quite unconscious, a striking difference from cases of motor aphasia in which the patient at once recognises any errors that he makes in speech. In some cases objects are correctly named, in others the patient while unable to find a name for an object shown him may describe its nature or its use: thus instead of saying "knife," he may say "something to cut with." Wrong words are also used as "parasol" for "castor oil," whilst in bad cases all objects may be called by the same name, as in the case of a publican who called his finger or any object presented to him, a public-house. To these and other

disorders of motor speech that are met with in cases of sensory aphasia the term *paraphasia* is applied.

The term *amnesia verbalis* is specially applicable to the effects produced by minor imperfections of the auditory word-centre. The sole defect may be a loss of memory for words, which may occur as a result of defective nutrition in old persons and also during convalescence from prostrating disease. The patient forgets the name of a friend or of some familiar object, or is unable to name many of the objects by which he is surrounded, although if the name of an object is written down or uttered in his hearing he is able to associate the name with the particular object in question. The aphasia of recollection is also met with during recovery from an uncomplicated motor aphasia.

(2) *Subcortical disease.* In very rare cases the left auditory word-centre, although intact, is cut off from the central acoustic path as well as from the right auditory word-centre by a subcortical lesion. The patient retains his memories of auditory speech, and therefore can talk correctly: he can also read aloud and understands what he reads, but he is unable to comprehend what is said to him, to repeat spoken words and to write from dictation. The condition is one of uncomplicated word-deafness.

Word-blindness. Just as in the case of word-deafness, word-blindness may be produced by damage either of the left visual word-centre or of the subcortical paths by which it is brought into communication with other parts of the brain.

(1) *Disease of the left visual word-centre.* The chief defect is word-blindness, that is, an inability to read printed or written words, and in some cases to recognise a single letter. The patient sees the letters and words, and may attempt to utter them, but there is no correspondence between the actual and the spoken words. He may, however, be able to recognise portraits and simple geometrical figures, and even to recognise his own name, although unable to read any other word.

There may be little or no disturbance of voluntary speech, a striking difference from cases of destruction of the left auditory word-centre. In most persons destruction of the visual word-centre produces agraphia, and if writing is not entirely lost it is usually limited to short words written either spontaneously or to dictation, the patient often writing better when his eyes are closed than when they are open; sometimes the patient can write only his own name. In some cases of word-blindness, the patients, being highly educated and strong auditives, agraphia is absent. According to Bastian, the most probable explanation is that in such cases the visual word-centre is not needed for writing, and that the rheiro-kinaesthetic centre is brought into activity through the direct action of the auditory word-centre by means of the commissural path which connects them. The patient may, however, suffer from *paragraphia*, that is, he makes errors in the spelling of simple words, or writes wrong words, and he may be unable to complete a sentence in writing.

(2) *Subcortical disease.* Very rarely the visual word-centre is intact but is cut off from its connections with the corresponding centre in the right hemisphere and with the half-vision centres in the occipital lobes. The symptoms of this condition are almost identical with

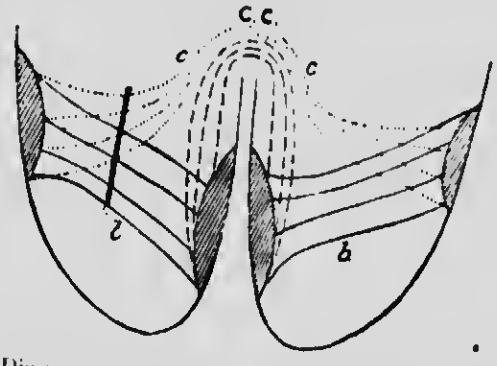


Fig. 146. Diagram representing a lesion (indicated by thick black line) which produces pure word-blindness. (Bastian.) C C, Posterior extremity of corpus callosum; c c, commissural fibres connecting the two visual word-centres; b b, fibres connecting each half-vision centre with the visual word-centre of the same side.

those produced by damage to the centre itself. The distinguishing features which are present in some cases of subcortical word-blindness are the absence of agraphia and the presence of right homonymous hemianopsia, or sometimes only of right hemiachromatopsia. The presence of paraphasia is against a subcortical and in favour of a cortical lesion.

Word-deafness and word-blindness combined. When both the left auditory and the left visual centres are severely damaged the patient is unable to comprehend either spoken or written language, and is unable to speak or to write intelligibly. His only means of communicating with others is by signs and gestures, hence it is not surprising to find that his mental condition is considerably impaired. The symptoms, however, are not always so severe, and cases are on record in which some measure of spontaneous speech was retained. The variations in the degree of speech impairment that are met with are to be explained partly by variations in the amount of damage to the centres and partly by individual differences as regards the compensatory activity of the speech centres in the right hemisphere.

Speech defects due to lesions of the two commissures between the sensory centres. The one commissure conducts impressions from the auditory to the visual word-centre, and is used in writing from dictation and sometimes in spontaneous writing. The other commissure conducts impressions from the visual to the auditory word-centre and is used in reading aloud or in naming an object seen. Destruction of both commissures produces inability to read aloud and to write spontaneously or from dictation. The patient can repeat words heard, can copy words placed before him and can understand both written and spoken language.

In opposition to the above brief description of aphasia which is based on the views so ably advocated by Bastian, Hughlings Jackson, Wernicke, Ross and other great writers on the subject, Marie has recently brought forward an entirely new hypothesis. He says that there

is only one centre for speech which is diffusely localised in the left temporo-parietal region, and that this centre is a region of intelligence specialised for language and is not a centre for the storing of sensory images: lesions of this centre are essential for the production of true aphasia. He contends that Broca's area is not the seat of articulatory memories and has no connexion with the functions of speech, and that therefore its destruction is not necessarily associated with motor aphasia. He argues that the so-called motor aphasia is a combination of aphasia with aphemia and is always the result of two lesions, the aphasia being determined by a lesion of the temporo-parietal centre, the aphemia by a subcortical lesion which interrupts the motor path by which impulses are conveyed to the muscles concerned in articulate speech.

Dysarthria and Anarthria. Allusion has already been made to the aphemia which is produced by a severance of the fibres which link together Broca's convolution and the adjacent lower part of the ascending frontal gyrus. We have now to consider the disorders of speech, comprised under the terms dysarthria and anarthria which may result from lesions of the motor neurons along which impulses are conducted from the centres in the lower part of the precentral convolution to the muscles of the tongue, lips and larynx which are concerned in articulate speech. Two main groups of dysarthrias may be recognised according as the disturbance in articulation is spastic or atrophic in character, the spastic variety being related to disease of the upper neurons, the atrophic to disease of the lower neurons.

Partial and transient forms of spastic dysarthria are common in cases of right hemiplegia as a result of acute lesions of the left internal capsule: they also occur though less frequently in cases of right hemiplegia. More marked and permanent forms can only be caused either by bilateral lesions of the lower end of the precentral gyrus or of the pyramidal tracts: or by a single lesion of the brain-stem damaging both tracts

where they are close together. In such cases of spastic dysarthria the most common history is that the patient had two separate attacks of hemiplegia, affecting first one and then the other side of the body; this condition in which dysarthria is associated with double hemiplegia is known as "pseudo-bulbar paralysis" (see p. 216).

The atrophic form of dysarthria is met with in bulbar paralysis and in other diseases of the medulla which implicate the lower motor neurons concerned in the production of articulate speech. The symptoms of the various defects in articulation are described under the diseases which produce them. There is no difficulty in distinguishing between the two varieties of dysarthria. In the spastic variety the affected muscles are stiff as well as weak; in a well-marked case the tongue is small, narrow and pointed, and may feel unduly firm. In the atrophic variety the affected muscles are flaccid and wasted and give either the partial or the complete reaction of degeneration.

In both varieties the patient, whose speech centres are intact, has no difficulty in understanding written and spoken language; he also knows what he wants to say and makes an attempt to pronounce the necessary words when it will be noticed that his utterance is defective, speech being slow and monotonous, and articulation slurred and indistinct.

A third group of dysarthrias may be distinguished in which there is ataxia of articulation due to disturbance of its co-ordinating mechanism; such ataxia may be produced by affections of the ponto-cerebellar region; it occurs in lesions of the pons, in disseminated sclerosis and in Friedreich's disease.

APRAXIA.

Closely allied to aphasia is the condition known as apraxia. This may be defined as a disorder of the cerebral functions, characterised by inability to perform

certain familiar purposive movements, in the absence of any obvious motor, sensory or mental impairment. The subject is a complex one, and has not yet been fully investigated. In the present work it is only possible to briefly allude to some of its main aspects; for further information the reader is referred to the writings of Liepmann, Maas, Kinnier Wilson and others.

Apraxia, like aphasia, may be either motor or sensory in type. It is, however, becoming customary to restrict the terms apraxia and aphasia to psycho-motor defects, and to employ the term agnosia to the psycho-sensory defects, which may affect either speech or the actions of the hands or of other parts of the body. Thus, just as word-deafness and word-blindness are varieties of verbal agnosia, so tactile agnosia, or asterognosis as it is sometimes called, is a variety of apraxic agnosia.

Apraxia may be either bilateral or unilateral. Bilateral apraxia of the tongue occurs in cases of hemiplegia: thus a hemiplegic patient who is unable to protrude his tongue when asked to do so, may be seen to lick his lips with it, or when told to put a stamp on a letter may protrude his tongue to lick the stamp. The tongue, although not paralysed, cannot be voluntarily used for a particular action. Lewandowsky mentions a case of cerebral diplegia in which the patient was unable to close the eyes voluntarily, though the lids followed the eyes downwards and were closed during sleep. Wilson refers to an aphasic patient who was unable to sniff voluntarily, but when offered a flower he sniffed it at once.

Unilateral apraxia is illustrated by defects in the use of objects, although their nature is perfectly recognised. Thus if a patient is handed a pencil which he recognises as a pencil and is even able to say what it is for, and yet when asked to use it fumbles aimlessly with it, he is suffering from apraxia. On the other hand, if the patient fails to recognise the pencil as an instrument for writing, and if when told to use it he puts it in his mouth and tries to smoke it like a cigar, he is suffering

from apraxic agnosia. The latter, or sensory variety of apraxia, was well marked in a case observed by Wilson. The patient, who was a great smoker, was handed a match, and asked what it was for. He said at once, "Oh, that's a pen." He was then requested to demonstrate its use. He replied, "Why, you clean your teeth with it," and at the same time he took it in his right hand and cleaned his finger-nails with it. In this case there was a combination of agnostic and ideational apraxia.

Apraxia, like aphasia, is associated with lesions in the left cerebral hemisphere much more frequently than with lesions in the right hemisphere. The lesions are either cortical or subcortical, being always situated above the internal capsule. They may involve the posterior ends of the first and second frontal gyri, in which the centres for the co-ordination of the complex movements of the limbs of both sides are believed to exist, just as in the third frontal gyrus there is a centre

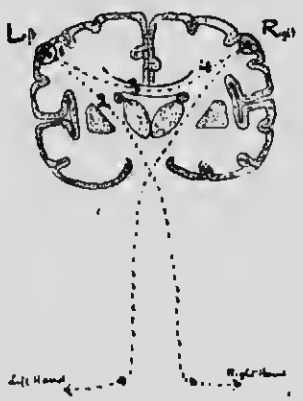


Fig. 147. Diagram illustrating the relation of the corpus callosum to apraxia. (Purves Stewart.)

for the complex movements necessary for speech; or the lesions may involve the anterior part of the corpus callosum through which the centres referred to are connected with the right hemisphere.

To understand the pathology of unilateral apraxia it

must be remembered that the left hemisphere exercises a potent influence over the movements of both sides of the body, and that many movements of the left limbs are initiated in the left hemisphere, so that a cortical lesion implicating the centre for the arm may produce not only paralysis of the right arm, but also apraxia of the non-paralysed left arm, this limb being deprived of the guidance of the centre in the left hemisphere (see 1, fig. 147). The same result may occur from a lesion interrupting the fibres immediately beneath the centre. A lesion of the callosal fibres (see 3, fig. 147) may also cause left-sided apraxia, owing to the guidance of the left arm-centre over the right being lost, but the right hand is not paralysed. Left-sided apraxia may be also produced by a lesion of the right frontal lobe which interrupts the callosal fibres passing from the left to the right arm-centre (see 4, fig. 147). A lesion, however, of the left internal capsule, which does not injure the commissural fibres, causes hemiplegia of the right side without any apraxia of the left side.

SECTION XIII.

Diseases of the Cranial Nerves.

The First or Olfactory Nerve.

Anosmia or loss of the sense of smell is usually due to disease within the nose; it is only rarely the result of nerve disease. It may arise from lesions either of the olfactory tract, or of the bulb in the anterior fossa. Thus

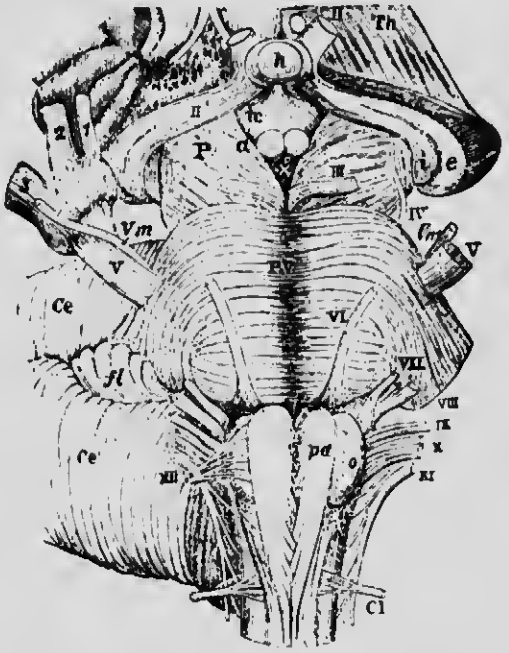


Fig. 148.—Origin of the cranial nerves (indicated by the Roman numerals). Th, Thalamus; tc, tuber cinereum; h, pituitary body; P, peduncle; PV, pons varioli; a, corpora albicantia; Ce, cerebellum; pa, anterior pyramid; o, olivary body. (Gowers, after Henle.)

these parts may be involved in a basal meningitis or in caries or fracture of adjacent bone; sometimes they are compressed by tumours in the anterior fossa, in the frontal lobe or in the pituitary region. Anosmia may

also result from injury of the delicate branches of the nerve, as by blows or falls on the head. Sometimes it occurs in association with hemianæsthesia from disease of the opposite hemisphere, either of the cortex or the posterior end of the internal capsule. Occasionally it has been observed in cases of embolism of the middle cerebral artery.

Parosmia, or perversion of the sense of smell such as the hallucinatory odour of sulphur, and *hyperosmia* or increased sensitiveness of the olfactory nerves may be present in hysteria and insanity; and sometimes as the aura of an epileptic fit, or as symptoms in cases of tumour involving the uncinate region of the temporo-sphenoidal lobe.

The Second or Optic Nerve and the Visual Path.

The chief affections of the optic nerve are neuritis and atrophy.

Optic neuritis or papillitis. Double optic neuritis, not always equally marked in the two eyes, is most commonly produced by intracranial tumours, especially when intracranial pressure is markedly increased. It is also frequently present in tubercular basal meningitis; it occurs in some cases of abscess in the brain, of sinus thrombosis, of chronic hydrocephalus, and occasionally in cases of softening or of hæmorrhage. Very rarely it has been seen in association with acute cervical myelitis, and with disseminated sclerosis. Papillitis, with or without retinitis, may also be found in profound anæmia, leukæmia, malignant endocarditis, renal disease, lead poisoning, cerebral syphilis, and in influenza, scarlet fever and other general infections.

Unilateral papillitis is usually due to disease at the back of the orbit or near the optic foramen, as from tumours, aneurysm of the ophthalmic or the internal carotid artery, or from inflammation, either limited to the nerve as in "rheumatic" cases, or spreading to it from meningitis or bone disease. Occasionally the

nerve is lacerated as a result of a blow or fall on the head.

On examination with the ophthalmoscope optic neuritis is recognised by the increased vascularity and swelling of the optic disc. Its edge appears blurred and the retinal veins are enlarged; the swelling of the disc, which may be considerable, renders it unduly prominent, and the vessels can be seen to bend down abruptly at its edges, where they are often partially concealed by the surrounding exudation. Care must be taken not to mistake haziness of the disc, often associated with hypermetropia, for true neuritis. The opacity

Fig. 149. Optic neuritis in cerebral tumour. The swelling is great, the arteries are concealed in the substance of the new tissue and the veins are tortuous. (Gowers.)

extends and appears to enlarge the disc, the veins become broader and the arteries narrower, whilst small haemorrhages may be visible on the surface or at the margins of the swollen area. These changes may subside almost entirely, but more frequently the disc gradually loses its redness, its outline becomes more defined, and it

passes into a state of "consecutive" atrophy, in which it presents a staring white colour and edges which are often blurred and may remain ragged and irregular.

Disturbance of sight may be absent even when the swelling of the disc is considerable: it is often more marked during the subsidence than during the active period of inflammation. When atrophy supervenes the acuteness of vision is always impaired and the visual field is contracted. When perception of light is lost the pupillary light reflex also disappears. In some cases, especially in cerebellar lesions, vision is lost at an early period of the disease.

Retro-bulbar neuritis most frequently depends on the excessive use of tobacco, especially when combined with intemperance in alcohol. Other causes are diabetes, gout, influenza and lead. Sometimes it is associated with rapid blindness, severe headache and pain in the eyes. As a rule the visual defect is most marked in the central part of the field, and often there is a central scotoma for red and green. The discs are slightly congested and hazy in the early stages and atrophied in the later.

Optic atrophy may be primary or it may be secondary to neuritis. Primary atrophy results either from pressure on the nerve, or from a chronic sclerosis of its fibres. The most frequent cause of such sclerosis is tabes, and after this disseminated sclerosis; probably in some cases of the latter disease the atrophy is secondary to a retro-bulbar neuritis, which did not lead to any detectable changes in the discs. Primary atrophy also occurs in Friedreich's disease, in general paralysis, and in diabetes, syphilis and other toxic conditions: it is always difficult to exclude a preceding neuritis. It is also found as a result of embolism or thrombosis of the central artery of the retina.

To the ophthalmoscope the disc appears pale in colour, with sharply-cut edges. Its central cup is not filled in as it is in secondary atrophy, and there is not much change in the size of the retinal blood-vessels. In tabes

the colour of the disc is greyish-white, and complete blindness is the ultimate result. The atrophied discs of disseminated sclerosis are usually whiter than those of tabes, and vision is rarely so completely abolished.

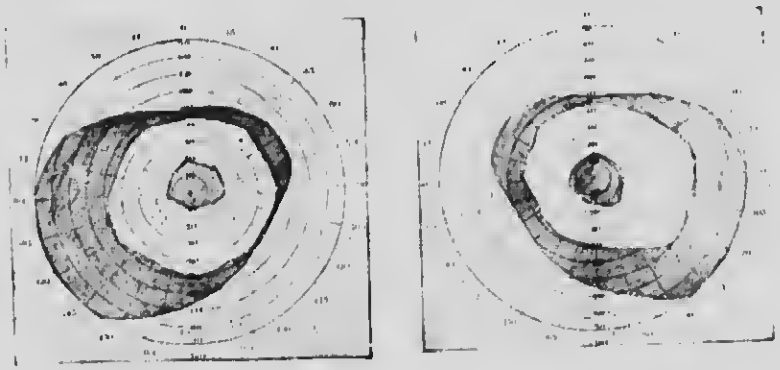


Fig. 150. Fields of vision in a case of disseminated sclerosis showing irregular restriction of the fields, and central scotomata. (Williamson.)

The **optic chiasma** may be compressed by tumours, or by syphilitic growths in the pituitary fossa, and occasionally by the distended infundibulum of the third ventricle. Rarely it is involved in adjacent inflammation.

The central portion of the chiasma being as a rule most affected, hitemporal hemianopsia is produced. In very rare cases each side of the chiasma is involved, as by pressure from calcification of both internal carotid arteries, when there may be nasal hemianopsia. If only one side of the chiasma is affected there is unilateral nasal hemianopsia, or possibly complete loss of sight on the side of the lesion. If both sides of the chiasma are destroyed there is total blindness.

The **optic tract** may be compressed by tumours at the base of the brain or springing from the temporo-sphenoidal lobe. In rare cases it is damaged by softening or hæmorrhage, or by an islet of disseminated sclerosis.

The visual defect is in the form of homonymous

hemianopsia to the opposite side. The line separating the lost from the retained half field is generally vertical, and deviates in the middle, so as not to interfere with the fixation point; it may, however, be oblique, or only a sector of the field may be lost. As a rule the hemian-

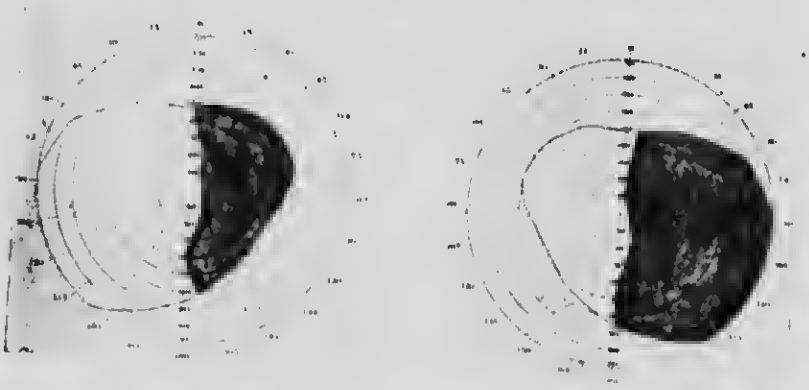


Fig. 151.—Fields of vision in a case of a vascular lesion of the left internal capsule

opsia is absolute, that is, all three visual sensations—colour, form and light—are lost.

The **intra-cerebral visual path** is sometimes damaged by tumours, softening or hæmorrhage, which may involve the optic thalamus, the posterior end of the internal capsule or the occipital lobe.

The result is homonymous heminopsia, which is distinguished from that due to disease of the optic tract (1) by the fact that when light is thrown on the blind halves of the retina, the pupil reacts normally if the lesion is situated between the corpora quadrigemina and the visual cortex, but it does not contract if the lesion involves the tract; and (2) by the hemianopsia being partial; there is blindness only of a portion of both half fields. Partial hemianopsia is unlikely to occur when a lesion affects the optic tract where the conducting fibres are close together, but is often present when a lesion affects parts where the fibres are separate and diverging from one another as in the occipital lobe.

Cortical visual centres. In lesions of the mesial side

of the occipital lobe the area of blindness may be sharply limited to a single quadrant of each field; thus it is stated that a lesion above the calcarine fissure will produce blindness of the lower quadrant, a lesion of the lower portion of the cuneus, blindness of the upper quadrant. More commonly there is homonymous hemianopsia to the opposite side, which may be unattended by any other objective symptoms, although if the lesion is a tumour, optic neuritis is usually present. The nearer the lesion is to the internal capsule the more likely are paralysis and anæsthesia on the opposite side of the body to be found in association with the hemianopsia.

Lesions of the higher visual centres in the angular gyrus and adjacent portions of the cortex, posterior to it, are apt to produce crossed amblyopia, word-blindness and possibly mind-blindness. Crossed amblyopia is a common symptom in hysteria: it is occasionally met with in local cortical disease. In mind-blindness the patient suffers from a loss of visual memory, he sees objects but is unable to recall their significance, and even well-known places and rooms appear strange to him.

Two other varieties of blindness merit a brief notice, namely, toxic amaurosis and reflex amblyopia. The former occurs in uræmia and sometimes in lead poisoning: there is sudden transient blindness without any ophthalmoscopic changes. The latter may be caused by peripheral irritation, especially by trigeminal neuralgia. Both conditions probably depend on a temporary arrest of the functions either of the nerve cells in the retina or of the cortical visual centres.

The Oculo-Motor Nerves.

These nerves may be collectively or separately affected. The type of paralysis varies according to the position of the lesion, that is, whether it implicates the individual nerves, their nuclei of origin, or their supranuclear path which connects the nuclei with some part of the

opposite precentral cortex, probably in the neighbourhood of the second frontal gyrus.

Peripheral lesions may involve the nerves at any part of their course between their nuclei and their termination in the ocular muscles. When all the muscles of one eye are paralysed the lesion is in the orbit or about the sphenoidal fissure: its chief causes are fracture of the bones or periostitis, cellulitis, pressure from a tumour, thrombosis of the cavernous sinus, and neuritis, as from syphilis, or exposure to cold. Sometimes ocular paralyses result from increased intra-cranial pressure, the sixth nerves being especially liable to be thus affected.

When the nerves are separately involved the lesion is either at the base of the brain, or within the crus in the case of the third nerve, or the pons in the case of the sixth nerve. It may be a meningitis, a new growth or gumma, a hæmorrhage or softening, or a toxic neuritis such as that produced by syphilis, diphtheria or influenza.

Paralysis of the third nerve on one side with paralysis of the face and limbs on the opposite side, indicates a

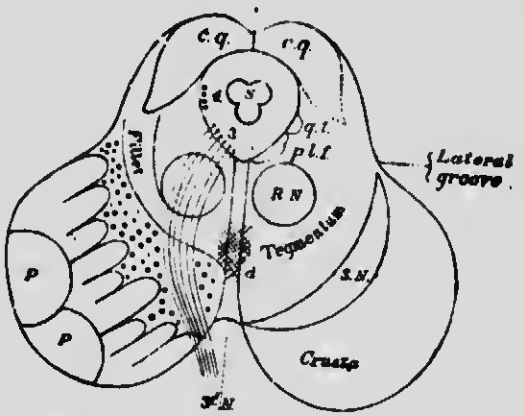


Fig. 152. A diagrammatic representation of the crura cerebri in cross-section. (Ranney.) c. q., Corpora quadrigemina; S, aqueduct of Sylvius; 4, nucleus of the fourth cranial nerve in the grey matter which surrounds the aqueduct; 3, nucleus of the third cranial nerve; q. t., root of fifth cranial nerve; p. l. f., posterior longitudinal bundle; R. N., the red nucleus; S. N., the substantia nigra; P, the portion of the "crusta" occupied by the pyramidal fibres.

lesion of the crus, which may be situated either outside the crus, as in the case of a basal meningitis, or within its substance, as in the case of a new growth. The paralysis is less complete from meningitis than from tumour; it may be represented only by ptosis and as a rule the internal muscles of the eye are spared. Very rarely both third nerves are involved by a tumour situated in the interpeduncular space. The sixth nerve may be involved by a lesion at the base—a common event or by one in the pons. In the latter position the seventh nerve also is frequently implicated.

When the *nuclei* or the *supra-nuclear paths* are diseased the general movements of the eye rather than its individual muscles are paralysed. Thus there may be loss of the upward movement of the eyes combined with drooping of the upper lids or loss of convergence, or paralysis of lateral movement, causing "conjugate deviation" of the eyes. Such deviation is the chief defect in cases of a supra-nuclear lesion, though it seems probable that the supra-nuclear path in the hemisphere is frequently involved without any detectable abnormality in the ocular movements.

The nuclei may be implicated in polio-encephalitis and in chronic degenerative processes, which are either limited to these nuclei or are associated with degeneration of the bulbar nuclei and sometimes also of the spinal anterior horns. Ocular paralysis may occur during the course of tabes, and disseminated sclerosis, and in association with myasthenia gravis, and with migraine.

Paralysis of the external ocular muscles. Certain symptoms are common to paralysis of the external muscles; they are defect of ocular movement, strabismus, erroneous projection and diplopia. *Limitation of movement* is always in the direction of action of the paralysed muscle and varies with the amount of paralysis, for example, in complete paralysis of one external rectus the eyeball cannot be moved outwards beyond the mid-position, and after a time is turned inwards by contraction

of the unopposed internal rectus. *Strabismus* or the want of correspondence between the visual axes, is called convergent when the prolonged axes of the eyeballs cross, divergent when the axes diverge from one another; thus paralysis of one external rectus produces a convergent squint, paralysis of one internal rectus a divergent squint. The term *primary deviation* is given to the deviation of the axis of the affected eye from parallelism with that of the healthy eye, whilst secondary deviation means the excessive movement of the sound eye when this is covered and an object is fixed by the paralysed eye. Paralytic strabismus is distinguished from strabismus due to muscular spasm

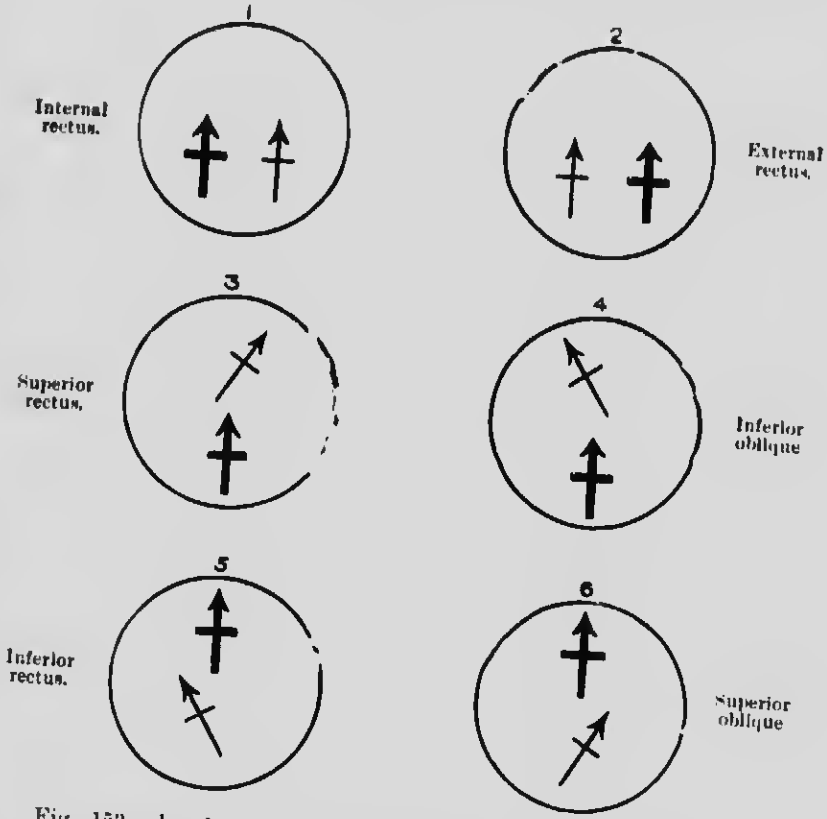


Fig. 153. In the above diagram the thick cross represents ... true image, the thin cross the false image. The left eye is suppo... to be affected in all cases. (Bristowe.)

(ordinary squint) by the fact that it is only present when the position of the object necessitates action of the affected muscles, whereas spasmodic squint is present in all positions, and there is no secondary deviation of the sound eye.

Erroneous projection and diplopia. An attempt to look at an object placed in the normal direction of vision is accompanied by increased effort, and this leads to an erroneous impression as to the position of the object, which is demonstrated by asking the patient to touch the object with his finger when it will be seen that the finger goes too far in the direction of the attempted movement. This erroneous projection which is often associated with vertigo, accounts for the perception of two images instead of one; diplopia, common in paralytic squint, is usually absent in the spasmodic variety. Of the two images of an object looked at by a patient suffering from diplopia the true one is seen by the sound eye, and is sharper in outline and more distinct than the false image, which is seen by the paralysed eye. Diplopia is

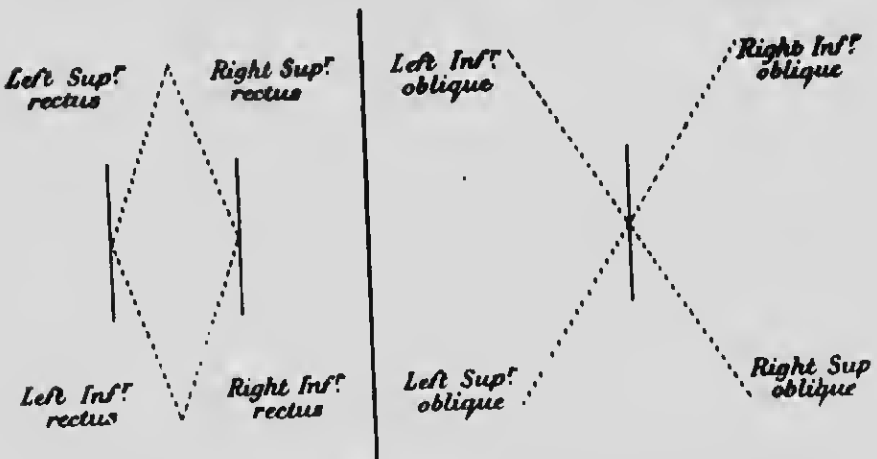


Fig. 154. Werner's "Artificial Memory" for the position of the false images in the ocular paralyses. On the left side the position of the true and false images is shown in palsy of the recti muscles: on the right side their position in palsy of the oblique muscles. The continuous line represents the true image, the broken line the false image. For example, in paralysis of the left inferior rectus, the false image is crossed, it is lower than the true image, has its upper end inclined towards it, and the diplopia occurs on downward movement of the eyes.

said to be simple or *homonymous* when the false image is displaced towards the side of the paralysed eye, *crossed* when it is displaced towards the side of the sound eye. The former occurs with convergent, the latter with divergent strabismus.

In testing for diplopia it is desirable to place a coloured glass before one of the patient's eyes while he looks at a candle flame held in different parts of the field of vision. The carrying out of this test is of the greatest value in detecting slight weakness of a muscle, for a patient may complain of double vision when there is no noticeable squint.

Affections of the *sixth nerve* or *abducens*, impair the outward movements of the eye, the patient complains of double vision on looking to the paralysed side; his

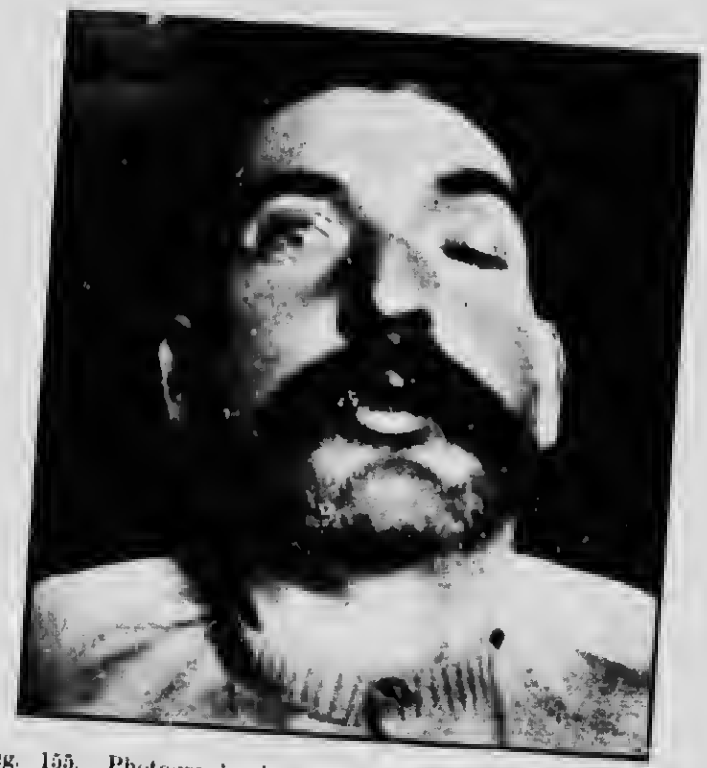


Fig. 155. Photograph showing ptosis; in this case there was paralysis of all the muscles supplied by the left third nerve, together with right hemiplegia from a syphilitic lesion of the left crus.

face is turned towards the sound side. Paralysis of the external rectus is the most frequent of all ocular palsies.

The *fourth or trochlear nerve.* Paralysis of the superior oblique leads to a difficulty in looking downwards and outwards, but the defective movement is often difficult to detect, and chief reliance in diagnosis is based on a study of the double images (see fig. 153). The head is turned downwards, the chin being inclined towards the healthy side; the nerve is seldom affected alone.

The *third nerve.* A complete interruption of motor impulses along this nerve causes paralysis of the levator palpebrarum and of all the external muscles of the eye with the exception of the superior oblique and the external rectus. The eyeball cannot be moved upwards or inwards and only slightly downwards: it is pulled outwards by the unopposed action of the external rectus. There is ptosis, the pupil is moderately dilated and does not contract to light, whilst, owing to paralysis of the ciliary muscle, the power of accommodation is lost. Such complete paralysis is rare: partial paralysis is common, and may result from damage to the nerve in any part of its course.

Paralysis of the internal muscles. The internal muscles of the eye are the ciliary muscle, the sphincter of the iris and the dilator of the iris. When they are all completely paralysed—*ophthalmoplegia interna*—the pupil is dilated, and does not react to accommodation nor to light and the power of accommodation is lost. The lesion affects either a portion of the nucleus of the third nerve or of the fibres of its root.

Cycloplegia. In paralysis of the ciliary muscle the power of accommodation is lost, so that while distant vision is good near vision is defective. Bilateral cycloplegia usually depends on a nuclear lesion. It is one of the earliest and most constant symptoms of diphtheritic paralysis: it occurs also in tabes. Unilateral cycloplegia may be due to a lesion of the nerve itself or of the ciliary ganglion.

Iridoplegia or paralysis of the iris occurs in four forms: (1) Loss of the reflex to light. In examining this reflex each eye must be tested separately, the other eye being closed; the patient should look at a distant object, while a bright light is brought suddenly in front of the eye. Loss of this reflex, with retention of pupillary contraction on convergence and accommodation, is known as the Argyll-Robertson reaction. It is usually a bilateral condition, but may be found on one side only. It is a common symptom in tabes and in general paralysis, and occurs whenever there is destruction of either the optic or the third nerve. The light reflex is stated to depend upon the integrity of a reflex arc, the afferent fibres of which pass from the retina along the optic nerve to the corpora quadrigemina, and thence to the third nerve nucleus, whilst the efferent fibres pass along the third nerve to the ciliary ganglion and thence to the sphincter iridis. In several cases, however, of Argyll-Robertson pupil, degenerative changes were found in the ciliary ganglion, and it seems probable that this ganglion, rather than the oculo-motor nucleus, is the controlling mechanism for the sphincter muscle.

(2) Accommodation iridoplegia. This means that the pupil does not contract when the patient looks from a distant to a near object. The condition is usually associated with cycloplegia, as in diphtheria, but may exist alone. (3) Cutaneous iridoplegia. In most healthy persons dilatation of the pupil may be produced by stimulation of the cervical sympathetic by pinching or faradising the skin of the neck. The reflex is lost in some cases of damage to the medulla, the cervical portion of the cord or to the cervical sympathetic. (4) Paralysis of the dilator. This is generally one-sided, and is caused by a lesion of the cervical sympathetic. The affected pupil is small, and does not dilate when shaded. The condition is associated with retraction of the eyeball, and slight narrowing of the palpebral fissure, but the movements of the upper eyelid remain normal.

Nuclear ophthalmoplegia. According to the rate of development of the symptoms, which depends on the nature of the lesion, three varieties may be distinguished. (1) The most common variety runs a chronic course, and as a rule is due to a progressive degeneration of the nuclei of the ocular nerves. All the eye muscles may be paralysed—total ophthalmoplegia, or only the external—ophthalmoplegia externa, or only the internal—ophthalmoplegia interna. Weakness of the orbicularis palpebrarum is often associated with the ocular paralysis. The degeneration may remain limited to the oculo-motor nuclei or may extend to the bulbar and spinal nuclei. Rarely bulbar or spinal paralysis precedes the ophthalmoplegia.

(2) Ophthalmoplegia sometimes develops suddenly in consequence either of hæmorrhage or of vascular occlusion, from atheroma or syphilitic arteritis. As a rule hæmorrhage quickly spreads, and death ensues within a few hours. The foci of softening from arterial obstruction are irregularly distributed, and hence, although the muscles of both eyes may be affected, the paralysis is rarely symmetrical.

(3) Ophthalmoplegia of acute but not sudden onset the paralysis reaching its maximum in a few days. As in the sudden variety the eye muscles are irregularly paralysed; in many cases the internal muscles are unaffected. Death may occur in a week or two, or the patient may survive and regain power in some of the muscles. In this variety signs of inflammation have been found in the nuclei, and the affection has been called polio-encephalitis superior. There is evidence, however, that sometimes the functions of the nerve cells are abolished by the action of some poison, for the condition has been observed in chronic alcoholism, influenza, and in diphtheria.

Recurring ocular paralysis is a rare form of transient palsy which affects some or all of the muscles supplied by the third nerve; a similar paralysis of the sixth or the fourth nerve has been described. The paralysis is

preceded or is accompanied by vomiting and by severe pain in the forehead and eye of the affected side (see p. 518). The attack may last for a few days or even a few weeks, and recur again and again after intervals of a month or longer. As the malady progresses the duration of the paralysis is increased, and it may become permanent. In such cases lesions of the nerve have been found at the post mortem examination. Transient paralyses of relapsing type affecting the third or the sixth nerve also occur in cases of cerebral syphilis.

Conjugate deviation of the eyes and head. This depends upon the associated action of certain muscles, and may be due to their paralysis or to their over-action. Thus if the head and eyes are persistently turned to the left side there is either over-action of the left external rectus, the right internal rectus and the rotators of the head to the left side; or paralysis of their opponents, namely, the right external rectus, the left internal rectus and the rotators of the head to the right side. If the movement is the result of paralysis, the head and eyes turn towards the side of the lesion if this is situated in a cerebral hemisphere, but away from it when the lesion involves one side of the pons. The reverse is the case when conjugate deviation is produced by spasm due to an irritative lesion in the pons or cerebrum. These facts will be better understood by reference to fig. 12, page 20 and by remembering that the external rectus of one side and the internal rectus of the other are innervated from the nucleus of the sixth nerve. It therefore follows that conjugate deviation will result from a lesion of this nucleus or of fibres which connect it, on the one hand, with the frontal lobe, and, on the other, with the nucleus of the third nerve. Conjugate deviation does not occur when a lesion is limited to the *fibres* of the sixth nerve, as then the external rectus alone is paralysed.

The Fifth or Trigeminal Nerve.

The fifth nerve may be affected by lesions situated in the pons, at the base of the brain, or outside the cranial

cavity. Within the pons its nuclei or roots may be implicated in tumours, softening, hæmorrhage or islets of sclerosis. In rare cases its motor nucleus is the seat of degenerative changes in bulbar paralysis, whilst the intramedullary portion of the sensory root is occasionally involved in tabes and syringomyelia.

At the base of the brain the trunk of the nerve may be damaged by tumours, meningitis or by caries of the petrous bone. In front of the Gasserian ganglion the first division of the nerve is sometimes affected by lesions about the cavernous sinus, by aneurysm of the internal carotid artery, by cellulitis or tumours within the orbit, or by fractures of the frontal bone. The second and third divisions of the nerve may be involved by tumours in or about the spheno-palatine fissure, whilst some of their branches may be damaged by injuries to the mouth or nose. A true primary neuritis of the fifth nerve is very rare, but neuritis secondary to disease of bone or membrane is not uncommon.

With regard to supra-nuclear lesions, the chief fact known to us is that anæsthesia of the region supplied by the fifth nerve, together with anæsthesia of the arm and leg, is produced by destruction of the hinder end of the internal capsule on the opposite side. Occasionally masticatory paralysis has resulted from cortical disease, but it is quite exceptional to meet with motor symptoms in affections of the upper neurons of this nerve.

Symptoms. These vary with the position and intensity of the lesion. When the sensory portion of the nerve is involved loss of sensation may be preceded by symptoms indicating irritation of the fibres, viz., numbness and tingling, or pains of a darting or burning character; there may be also hyperæsthesia of the affected parts and tender points at the bony foramina.

The area of anæsthesia is usually less than that of the anatomical distribution of the nerve to the face. The area of epicritic loss is slightly larger than that of protopathic loss: deep sensibility is also impaired. The mucous membranes are affected as well as the

skin. The cornea and conjunctiva are insensitive, the nose is no longer irritated by snuff or ammonia and the sense of smell becomes impaired owing to dryness of the mucous membrane. In the mouth, loss of sensation to a touch and prick is found on one half of the tongue as far back as the circumvallate papillæ, on the anterior pillar of the fauces and along the centre of the soft and hard palates to the upper lip. The tongue retains its deep sensibility, the fibres for which are probably conveyed in the hypoglossal nerve.

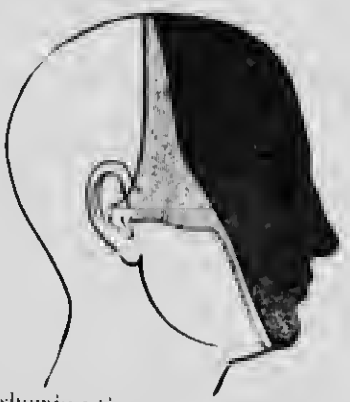


Fig. 156. Chart showing the area of anæsthesia following excision of the Gasserian ganglion. The deeply shaded portion shows the area of both epicritic and protopathic loss; the lightly shaded, the overlap of the epicritic loss. (M. Davies.)

The patient, when drinking out of a cup, feels as if it were broken. The affected half of the tongue becomes dry and covered with a thick fur. Anæsthesia of the teeth renders them liable to injury and they become loosened and ultimately drop out. In some cases the gums are swollen and ulcerated, and ulcers may form on the inside of the cheek.

Inflammation of the eyeball, called *neuro-paralytic ophthalmia*, sometimes occurs; it is probably caused by irritation of the sensory fibres of the root or of the Gasserian ganglion. The cornea becomes opaque and ulcerated, and eventually the ulcers may perforate and lead to destruction of the eyeball. Another result of such irritation is herpes zoster, especially in the area supplied by the ophthalmic division of the nerve. In old

people the eruption is often attended by severe pain which may last for a long time. In some cases the sense of taste is impaired or lost on the affected side of the tongue and palate, in other cases it is retained (see p. 26).

Involvement of the motor portion of the nerve is indicated by paralysis of the temporal, masseter and pterygoid muscles. Weakness of the two former muscles is recognised by placing the fingers over them whilst the patient brings the jaws forcibly together. The muscles feel softer than their fellows, and after a time become wasted so that the temporal fossa is hollowed and the zygoma is abnormally prominent; degenerative electrical reactions may be observed in the atrophied muscles. Paralysis of the external pterygoid is shown by an inability to move the jaw towards the sound side, and when the patient's mouth is wide open the condyle of the lower jaw is prominent on the paralysed side and the jaw itself is displaced towards this side. The tensor tympani, the tensor palati, the mylo-hyoid and the anterior belly of the digastric may also be paralysed, but the paralysis is difficult or impossible to detect.

The Seventh or Facial Nerve.

According to recent researches the seventh is a mixed nerve containing sensory and taste fibres as well as motor. The motor fibres which constitute its main portion supply the facial muscles of expression, the platysma, the stylo-hyoid, the posterior belly of the digastric and the stapedius muscles. The taste fibres are distributed by way of the chorda tympani and the lingual branch of the fifth nerve to the anterior two-thirds of the tongue; whilst the sensory emerge with the motor fibres from the stylo-mastoid foramen and are distributed to the skin of the external auditory meatus and the anterior surface of the external ear.

Etiology. The facial nerve may be affected by lesions in the pons, at the base of the brain, in its peripheral course through the temporal bone or after its exit from the stylo-mastoid foramen. Within the pons the roots

or the nucleus may be involved in tumours, in softening or in degeneration; as a rule other cranial nerves, as the sixth and the fifth, are also affected, or in nuclear cases the tenth, eleventh and twelfth nerves. At the base

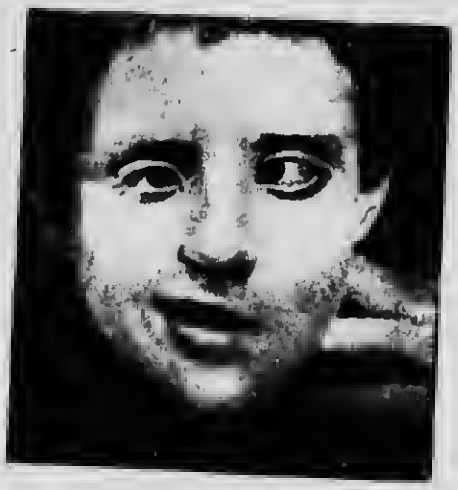


Fig. 157. Photograph showing paralysis of the left sixth and seventh nerves, in a case of glioma of the pons.

Fig. 158. A case of complete paralysis of the left side of the face; there was also deafness on the left side. The paralysis of the seventh and eighth nerves was caused by fracture of the petrous bone.

of the brain the nerve may be compressed by a tumour either in the cerebello-pontine angle or in the posterior fossa; it is also liable to be involved in a meningitis or to be injured by a fracture of the base of the skull. Within the temporal bone the nerve often suffers, especially in children, in consequence of suppurative otitis-media, or of operations upon the mastoid bone.

But by far the most frequent cause of facial paralysis is a parenchymatous neuritis, which is usually most marked in the nerve at the distal end of the Fallopian canal. In about eighty per cent. of such cases the neuritis appears to be set up by exposure to cold; this is the "rheumatic" or "refrigeration" variety. It is stated that such neuritis may be favoured by a congenital narrowing of the Fallopian canal and the stylo-mastoid foramen. In some of the cases syphilis or alcohol has been an essential or a contributory factor; in others we have to consider the possibility of microbic infection such as that which is believed to initiate acute anterior poliomyelitis. Occasionally indeed facial paralysis occurs in that disease, though then probably as the result of a nuclear lesion.

On the side of the face the nerve or its branches may be injured by wounds, blows, cellulitis or by parotid swellings. Sometimes facial paralysis is seen in newly-born infants, especially in those delivered by forceps. Rarely the facial nerve is implicated in cases of multiple neuritis and then usually on both sides. The chief causes of bilateral facial paralysis are diseases of the pons and medulla, basal meningitis and aneurysm of the basilar artery.

Symptoms. Facial paralysis of the common peripheral type usually commences rapidly or even suddenly; that due to ear disease or to the pressure of a tumour develops in a gradual manner, when it may be preceded or accompanied by pain, noises in the ear and deafness. In some cases the pain is due to involvement of the fifth nerve, in others possibly to disease of the sensory portion of the seventh nerve. Sometimes, and especially in

children, the paralysis is ushered in by fever, even when there is no local condition, such as ear disease, to account for it.

The symptoms of complete unilateral facial paralysis are characteristic, although in children whose muscles are imperfectly developed the face, when at rest, may appear normal. In the adult the affected side of the face is smooth, flaccid and expressionless, and often appears puffy. The patient cannot wrinkle his forehead, or raise or knit his eyebrow; the eye remains widely open and when he attempts to close it the eyeball is seen to roll upwards. During sleep the eye is only partially open owing to relaxation of the levator palpebræ. Having lost its protection from foreign bodies the conjunctiva often becomes inflamed, while, owing to the weakness of the lower lid, tears cannot enter the lachrymal canal and are apt to run down the cheek. The nostril falls in instead of expanding during inspiration; the naso-labial fold is obliterated, and the mouth is drawn obliquely over to the healthy side, whilst in old-standing cases the lower part of the cheek hangs down in a pouch-like manner. The lips on the paralysed side cannot be closed so that saliva is liable to escape from the corner of the mouth; whistling is impossible and articulation of the labial consonants is imperfect. Weakness of the buccinator causes the cheek to puff out and allows food to collect between the cheek and the gum. Weakness of the platysma which is sometimes present is indicated by an absence of wrinkles in the neck when the chin is forcibly pressed down. The external muscles of the ear are also paralysed, but as they are not usually under voluntary control their weakness is difficult to detect.

All the paralysed muscles waste and in severe cases may show the reaction of degeneration before the end of a fortnight; in mild cases their irritability to faradism may be alone diminished.

The sense of taste is frequently diminished or abolished over the anterior two-thirds of the tongue on the paralysed side.

Course. The ordinary type of refrigeration paralysis usually passes away in a few weeks, sometimes even within a week. Severe cases run a longer course, and the paralysis may persist for many months, and occasionally become permanent. Partial recovery of power may be complicated by secondary contraction of the affected muscles, when the mouth will be pulled towards the weak side, the naso-labial fold becoming deepened and the palpebral aperture smaller. It is now the healthy side of the face which appears to be paralysed; this impression, however, is corrected when the patient smiles or shows his teeth for then the mouth is drawn to the non-paralysed side. Spontaneous twitching movements may also be observed during recovery.

Diagnosis. Facial paralysis may result from any interruption of the nerve path, between the cerebral cortex and the muscles, and we must first determine whether the lesion involves the upper neurons—the cortical centre and the fibres leading from it to the nucleus in the pons, or the lower neurons—the facial nucleus and the peripheral fibres proceeding from it. Paralysis due to lesions of the upper neurons has the following characteristics:—(1) The upper facial muscles are relatively less paralysed than the lower ones; they do not, however, escape, for although the eye can be voluntarily closed, its orbicularis offers less than the normal resistance. (2) The muscles do not atrophy nor show altered electrical reactions. (3) The paralysis is usually associated with hemiplegia or with other symptoms of intra-cranial disease.

Paralysis due to lesions of the lower neurons is characterised by an equal degree of paralysis of both the upper and the lower facial muscles, by atrophy with altered electrical reactions and by abolition of the reflexes. The exact situation of the lesion may be determined as follows:—(1) If the lesion is situated below the point at which the chorda tympani leaves the facial nerve, taste is not affected. (2) If the lesion is situated between the points of origin of the chorda

tympani and the nerve branch to the stapedius there are, in addition to facial paralysis, loss of taste and diminution of the salivary secretion. (3) If the lesion is situated between the nerve branch to the stapedius and the geniculate ganglion, in addition to the above-mentioned symptoms there is hypersensitiveness to musical notes of low pitch. (4) If the geniculate ganglion is acutely inflamed, with implication of the adjacent facial and auditory nerves near the internal auditory meatus, facial paralysis may be associated with deafness and with herpes zoster of the external ear and sometimes in the area of the distribution of the superficial cervical plexus. (5) If the lesion is situated between the temporal bone and the pons the auditory and the facial nerves are usually simultaneously affected. The sense of taste may be impaired or lost over the whole of one side of the tongue. (6) If the lesion is in the substance of the pons, the sixth nerve, round the nucleus of which the fibres of the facial form a loop, is almost invariably affected with the seventh. Nuclear paralysis resembles cerebral paralysis of the facial muscles in the relative sparing of the upper branches of the nerve; it generally forms part of a bulbar paralysis, which may be associated with an atrophic paralysis of the muscles of the arms.

Prognosis. This is based on a consideration of the nature and severity of the lesion. When, for example, the paralysis is due to caries of the petrous bone or to a tumour in the pons, recovery can scarcely be expected. In the ordinary type of peripheral palsy a judgment may be formed regarding the intensity of the lesion by a study of the electrical reactions. If the excitability of the muscles to faradism is only slightly diminished a fortnight after the onset of the paralysis, recovery may be expected within a month. If, however, the muscles give the reaction of degeneration, and if this reaction persists over three months, ultimate complete recovery is doubtful. Between these two extremes many variations are met with.

Treatment. In all cases of the ordinary peripheral

type it is advisable for the patient to stay indoors for a time and even in bed should there be much pain or any febrile disturbance. When there is reason to believe that alcohol is an etiological factor it must be prohibited; if syphilis is suspected, mercury and the iodides should be administered.

The neuritis may be beneficially influenced by counter-irritation to the affected side by means of hot fomentations repeated every three hours for the first two or three days, and afterwards by the application of a blister or a mustard leaf to the mastoid process. A blister should not be applied in front of the ear owing to the risk of setting up cellulitis. The bowels should be kept freely open and a mixture containing salicylate of soda and iodide of potassium may be given with advantage. At a later period tonics, especially strychnine, are likely to do good.

For the restoration of voluntary power the chief reliance is to be placed on electricity and massage; and these methods should be commenced in a week or two after the onset. In applying the constant current, which is usually more useful than the faradic, two small electrodes are necessary. The negative pole should be held behind the ear near the stylo-mastoid foramen, whilst the positive pole is stroked across the forehead, around the eye, down the cheek and along the lips. The strength of the current should be just enough to cause the muscles to contract. This method can be carried out quite well by the patient with the aid of a mirror, and the application should be repeated two or three times a day for a period of ten minutes and persevered with for several months or until voluntary power begins to return, when it is well to discontinue electrical treatment in order to avoid the tendency to secondary contractures. Facial massage, which also may be done by the patient, is of still greater importance. The patient should be taught to rub the individual muscles with the tips of his fingers and to knead and compress those of the cheek and lips, between the thumb placed inside the mouth and the fingers outside.

Massage and electrical treatment should be continued for a period of six months. If at the end of that time there are no signs of improvement, the operation of nerve-anastomosis must be considered. This operation has been frequently done and in some cases with satisfactory results. It is stated that better results are obtained by grafting the facial nerve upon the hypoglossal than upon the spinal accessory. In either case there is a danger that the return of active movements in the facial muscles will be accompanied by associated movements of the tongue in the one case, or of the shoulder in the other.

The Eighth or Auditory Nerve.

The eighth nerve is composed of two parts subserving different functions, namely, the cochlear portion which is concerned with the special sense of hearing and the vestibular portion which is concerned with the function of equilibration. The anatomical relations of these two divisions of the eighth nerve are mentioned on pages 19 and 21; the affections of the vestibular part are considered in Section VIII.

Lesions of the cochlear or auditory nerve proper may cause diminution, exaltation or perversion of its functions.

Diminished function. Apart from congenital cases and from affections of the outer or the middle ear, the common cause of deafness is disease of the internal ear or labyrinth, either primary or secondary to diseases of the middle ear. These two varieties of labyrinthitis are described in the chapter on auditory vertigo. Sometimes the labyrinth of both ears is affected; this occurs in a chronic form in hereditary syphilis, and in an acute form in cerebro-spinal and in tuberculous meningitis. Temporary deafness, probably labyrinthine in origin, is sometimes produced by drugs, such as quinine and salicin, or by an abnormal condition of the blood as in anæmia.

A less common cause of deafness is a lesion of the

nerve trunk either in the internal auditory canal or at the base of the brain. The lesion may be the result of caries or of fracture of the petrous bones (see fig. 158), or of pressure from a new growth or an aneurysm. Attention may be specially drawn to tumours growing in the cerebello-pontine angle which frequently involve the auditory nerve for some time before other structures.

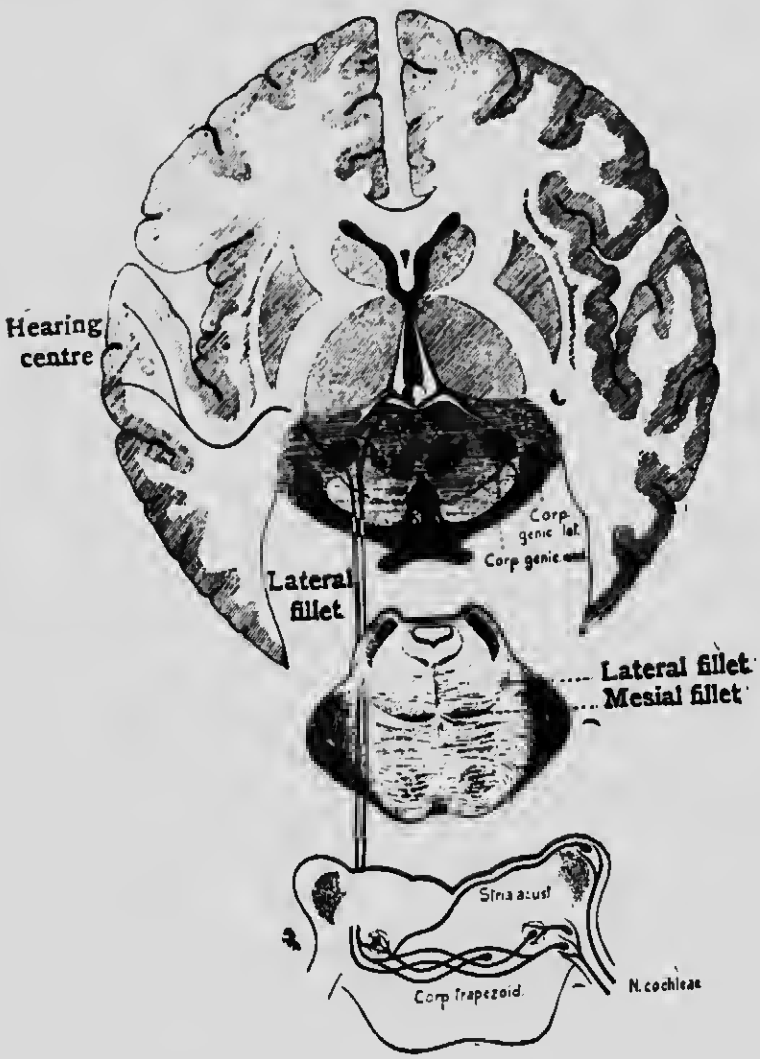


Fig. 159. Course of the auditory path. (Villiger.)

Occasionally deafness in one or both ears is caused by a tumour or other lesion involving the auditory nuclei or their roots, or some portion of the supra-nuclear path between the nuclei and the cortex. Deafness to sounds rarely arises from cortical lesions, and then only to a marked degree when both superior temporal convolutions are implicated. A unilateral cortical lesion never causes much diminution in hearing, owing to the intimate relation of the auditory nerves with both hemispheres. Word-deafness with little or no impairment of hearing for ordinary auditory impressions occurs in lesions of the left temporal lobe.

Hyperæsthesia or *hyperacousis* in which sounds are heard with undue loudness and even painful intensity is a rare condition: it occurs in hysteria and sometimes at the onset of acute changes in the auditory centres. One variety may be caused by disease of the facial nerve owing to paralysis of the stapedius muscle. *Dysacousis*, a condition in which ordinary sounds give rise to discomfort is met with in neurasthenia, and during attacks of headache, and also occasionally in disease of the brain.

Tinnitus aurium. Subjective sound sensations, such as roaring, ringing, ticking or whistling, to which the term tinnitus is applied, may be produced by morbid changes in any part of the auditory apparatus, and especially by disturbances which affect the labyrinth. The chief causes are:—(1) An altered condition of the blood as in anæmia, or of the circulation as in intra-cranial aneurysm. (2) A plug of wax pressing on the membrana tympani; various affections of the middle ear. (3) Spasmodic contraction of the tensor tympani, or the levator palati. (4) Irritation or disturbed nutrition of the auditory nerve or its centres. Tinnitus is a prominent feature in Ménière's disease: it is common in neurasthenia: it may constitute the aura of an epileptic seizure; rarely it forms part of an attack of migraine.

Even when the symptom depends on ear disease it

may be considerably modified both in character and intensity by deranged action of the auditory centre. Moreover, elaborate subjective sounds may take the place of simple forms of tinnitus owing to perversion of higher centres; such are the auditory hallucinations which are so common in cases of insanity.

The Glossopharyngeal and Vagus Nerves.

These nerves are now regarded as portions of one large mixed nerve, of which the motor fibres originate in the long nucleus ambiguus situated in the reticular formation of the medulla, whilst the sensory fibres terminate partly in the posterior vago-glossopharyngeal nucleus and partly by way of the fasciculus solitarius in the adjacent gelatinous substance. Some of the upper roots of this nerve form the glosso-pharyngeal nerve, whilst the remaining roots constitute the vagus.

The results of disease limited to the *glossopharyngeal nerve* are not definitely known. From our knowledge of its functions we should expect to find anæsthesia of the upper part of the pharynx, loss of taste over the palate and the posterior third of the tongue and difficulty in swallowing from paralysis of some of the pharyngeal muscles. Disturbances of taste which occasionally occur in affections of the middle ear may be due to involvement of branches of the nerve to the tympanic plexus.

The Vagus Nerve.

Paralysis of parts supplied by the vagus nerve may depend on lesions involving the nuclei or their nerve fibres either within the cranium, or outside it during their course through the neck and thorax. The nuclei may be implicated in diseases of the medulla, such as tumours, hæmorrhage and softening, or they may undergo degeneration as in bulbar paralysis, tabes, disseminated sclerosis and syringomyelia.

At the base of the brain the nerve is liable to be

damaged by meningitis, especially the syphilitic variety, by growths springing from the bone or membranes, and by aneurysm of the vertebral artery. In the neck the nerve may be injured by wounds or during the course of surgical operations, and both in the neck and the thorax it may be compressed by enlarged glands, tumours or aneurysms. In the thorax the recurrent laryngeal branch of the left nerve is especially liable to pressure from aortic aneurysm, whilst that of the right nerve is occasionally included in pleuritic adhesions at the apex of the lung. Sometimes the vagus is involved in cases of multiple neuritis; this has been observed in the neuritis due to alcohol, arsenic, diphtheria and influenza.

Symptoms. If the whole of one nerve is affected there is unilateral paralysis of the soft palate and larynx with anæsthesia of the larynx on the same side. Paralysis of one side of the palate does not usually cause symptoms, and at rest the palate may show no want of symmetry; the weakness is detected on movement: thus, when the patient says "ah," the base of the uvula is drawn up to the unaffected side. Speech and swallowing are but little disturbed. The vocal cord is in the 'cadaveric position,' being slightly abducted from the middle line; it is not further abducted during inspiration, nor is it adducted during phonation. The healthy cord moves freely, and on phonation may cross the middle line. There is inability to cough; the voice is low-pitched and hoarse.

If both nerves are implicated the symptoms are bilateral and sometimes extensive in distribution. Total paralysis of the palate which is caused most frequently by diphtheria and occasionally by disease of the bulbar nuclei, is shown by the absence of movement in phonation, in deep inspiration or on tickling the mucous membrane. During swallowing, liquids are apt to regurgitate through the nose; the speech has a nasal resonance. When the pharyngeal muscles are affected there is great difficulty in swallowing: food lodges in the pharynx instead of passing into the œsophagus and small particles may

enter the larynx and cause spasm and choking. In rare cases the branches of the nerves to the œsophagus are diseased when still further difficulties in swallowing will arise; the dysphagia is probably more frequently due to spasm than to paralysis.

Bilateral paralysis of the vocal cords may result from

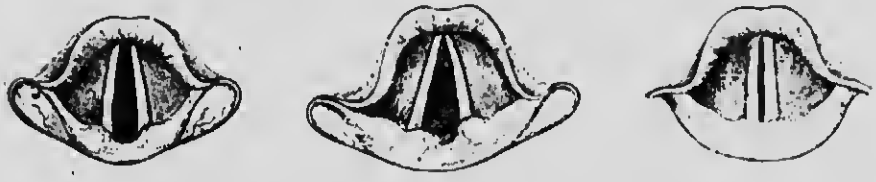


Fig. 160. Fig. 161. Fig. 162.

Fig. 160. Total palsy of both cords; cadaveric position.
Fig. 161. Palsy of left vocal cord.
Fig. 162. Bilateral abductor paralysis. (Gowers.)

disease either of the medulla, or of the trunks of the recurrent laryngeal branches of the vagi. When the paralysis is complete the cords assume the cadaveric position and are immobile; hence the patient is unable to phonate and to cough. When the paralysis is incomplete the abductors are mainly affected, these muscles being the most prone of all the laryngeal muscles to be early and severely paralysed. In such cases the cords are near together, and cannot be separated, while after a time the glottis becomes still further narrowed owing to secondary contractions of the adductors. The voice is but little affected and coughing is normally performed. Inspiration which tends to bring the cords still closer together is accompanied by loud stridor. Expiration is not impeded, and is not attended by stridor—a distinction from tracheal stenosis. With such obstruction to inspiration the condition of the patient becomes serious. The larynx is seen to move violently up and down, dyspnœa is prominent, the face becomes livid and the slightest catarrh may close the glottis, when immediate tracheotomy is called for.

In contrast to this condition is that of adductor paralysis, which is usually hysterical in origin: there is aphonia, but no stridor, nor are there other indications of respiratory embarrassment.

Various gastric, pulmonary and cardiac symptoms are occasionally met with, and more commonly in bilateral than in unilateral disease of the nerve, but they cannot be regarded as characteristic of such disease. Vomiting is sometimes present; it may be associated with vertigo owing to the close connection of the vagus with the vestibular nerve. The gastric crises of tabes are probably due to a lesion of the vagus or its nucleus. Slowness of the heart's action has been found to depend on irritation of the vagus, while tachycardia is doubtless sometimes due to paralysis of the cardio-inhibitory fibres in the upper vagal roots. In some cases of arrest of the heart's action during an attack of angina pectoris, disease of the vagus branches to the cardiac plexus has been found. Slowness and irregularity of respiration also result from bilateral disease of the vagus or its nucleus, and it is probable that Cheyne-Stokes's breathing is caused by a lowered action of the respiratory centre.

The Spinal Accessory Nerve.

The functions of this nerve may be impaired by lesions involving its nucleus or its fibres. The nucleus may be degenerated in cases of progressive muscular atrophy, disseminated sclerosis, syringomyelia, or of cervical myelitis. The intra-cranial part of the nerve is sometimes damaged near the foramen magnum by meningitis, tumours or by aneurysms. Outside the skull the nerve may be injured by wounds, or compressed by tumours, abscesses or by the results of cervical caries. In rare cases it is attacked by neuritis.

Symptoms. A lesion of the spinal accessory nerve leads to an atrophic paralysis of the sterno-mastoid and trapezius muscles, the upper portion of the trapezius being more affected than its lower portion.

The affected sterno-mastoid is wasted and less

prominent than its fellow, and its power of rotating the head to the opposite side is impaired. Paralysis of the highest part of the trapezius is indicated by a change in the contour of the neck which, instead of being straight, shows a slight curve inwards; this is especially conspicuous during a deep inspiration. Weakness of the middle portion of the muscle impairs the power to elevate the shoulder, which is dropped a little; weakness of the lowest portion leads to outward rotation of the scapula, so that its vertebral border stands out prominently and is inclined from below upwards and outwards.

If both trapezii are paralysed the head tends to fall forwards, if both sterno-mastoids, to fall backwards. This defective power to support the head sometimes occurs in children as a result of damage to both nerves from meningitis about the foramen magnum. Occasionally the spinal accessory is affected after it has perforated the sterno-mastoid, when the upper part of the trapezius is alone paralysed.

The Twelfth or Hypoglossal Nerve.

Paralysis of the tongue from disease of the upper neurons of this nerve has been already mentioned (see p. 93). The lower neurons are much more frequently affected inside than outside the cranial cavity; lesions of the nerve in the neck, as from wounds or new growths are very rare.

Intramedullary lesions of the nucleus or its root fibres, usually bilateral, form part of a bulbar paralysis and occur also in some cases of tabes, disseminated sclerosis, and syringomyelia; they may also result from softening or from the pressure of a tumour. Near the medulla the fibres of the nerve are sometimes affected by a meningitis or a new growth. Near its exit through the anterior condyloid foramen the nerve may be compressed by thickening of the bone or by the exudation produced by caries of the highest vertebræ.

Symptoms. When the upper neurons of the nerve are

involved, the muscles of the tongue do not waste. When, however, the lower neurons are involved the muscles undergo a progressive atrophy and show fibrillary tremors; the mucous membrane of the tongue is thrown into longitudinal folds with deep grooves between the folds. In some cases it is possible to obtain the reaction of degeneration.

In disease of the nucleus the paralysis is usually bilateral, and is associated with paralysis of the lips, palate and larynx. If such paralysis is complete the tongue lies motionless within the mouth and the patient is unable to protrude it or to move it in any way. Articulation is defective, and chewing and swallowing are impaired, the tongue being unable to keep the food between the teeth, or to propel it backwards towards the throat.

In infranuclear disease the paralysis is usually unilateral, the half of the tongue corresponding to the side of the lesion being weak and wasted. Within the mouth the movement is deficient towards that side, but in protrusion it is deficient towards the opposite side, and the tongue deviates towards the paralysed side.

Speech, chewing and swallowing are but little affected in unilateral disease. When this is situated near the medulla there is usually an associated paralysis of the corresponding half of the palate and of the vocal cord. When the lesion implicates the root fibres within the medulla, it frequently involves the pyramidal tract also; there is then a crossed paralysis, the limbs being paralysed on the side opposite to that of the tongue.

SECTION XIV.

Cerebral Meningitis.

Inflammation may attack either the dura mater (pachymeningitis) or the pia-arachnoid membrane (leptomeningitis). Leptomeningitis is much the commoner of the two varieties and will be alone considered in the present section. The disease which can be traced almost invariably to the action of micro-organisms may be conveniently described under the following headings: *Tuberculous meningitis* characterised by the presence of the bacillus tuberculosis. *Epidemic cerebro-spinal meningitis* and the sporadic type known as *posterior-basic meningitis*, which are characterised by the presence of the meningococcus, or diplococcus intracellularis. *Suppurative meningitis*, which occurs in two forms namely, *pyogenic meningitis* due to infection by staphylococci and streptococci, and *pneumococcic meningitis* characterised by the presence of the diplococcus pneumoniæ. *Meningitis produced by other organisms*, such as the bacillus typhosus, the bacillus influenza, the bacillus enteritidis, or the gonococcus. Syphilitic meningitis, which usually runs a subacute or a chronic course, is considered in Section XXI.

TUBERCULOUS MENINGITIS.

Etiology. Tuberculous meningitis may occur at any age, but it is most common between the ages of one and six years, and is especially frequent during the second year of life. The liability to the disease lessens as age advances, thus it is less rare between puberty and thirty than at a later period. A family history of tuberculous disease can sometimes be obtained, but perhaps not oftener than in persons who do not suffer from any form of tuberculous lesion. Occasionally two or more

members of the same family suffer from tuberculous meningitis or from some other manifestation of tubercle.

Its essential cause is invasion of the pia mater by the tubercle bacillus, and this is always secondary to a localised caseous process in some other part of the body, as in the lungs, glands, bones, joints, skin or generative organs. In a few cases the meningitis has been a direct extension from a tuberculous mass in the cerebral cortex; and very rarely, surgical interference with a tuberculous joint, a tuberculous spine or other tuberculous lesion has immediately preceded a general tuberculosis of which meningitis was a part. An acute specific disease, notably measles, is not infrequently followed by tuberculous meningitis or other form of tuberculosis.

Of predisposing factors may be mentioned, improper or insufficient food, overcrowding and a deficient supply of fresh air, together with other bad hygienic conditions. In some cases a blow on the head or excessive brain work appears to have been a determining factor in the causation of the disease.

Symptoms. The onset is more commonly insidious than abrupt, and in children the prodromal stage may last from a fortnight to two or three months. During this period, that is, before the invasion of symptoms pointing to meningitis, the child is obviously out of health; he is easily fatigued, is languid and dull, and often shows much irritability. He soon tires of his toys and is content to lie down, and cries peevishly when disturbed. He passes restless nights, often grinding his teeth and starting in his sleep, and is subject to irregular feverish attacks. His appetite is poor, and he progressively gets thinner. The bowels are usually constipated; some gastro-intestinal disturbance may be present; but without this, vomiting occasionally occurs, and especially at night. The face is pale, but often suddenly flushes. A young child may become speechless for a few days, an older child may make mistakes in speaking, whilst in adults actual aphasia may be an initial symptom.

Sometimes the disease in adults is ushered in by hysterical manifestations or by an abnormal mental condition, so that hysteria or early insanity has been diagnosed instead of tuberculous meningitis. In adults delirium is commoner and headache is usually more severe and localised than in children. Very rarely there are no symptoms to indicate the development of the disease, which is only discovered after death; in other cases the onset is suggested by a squint or by a slight paralysis of the arm or the face.

The active invasion of meningitis may be indicated by an increase in the headache, the vomiting, the constipation, or the lethargy of the prodromal stage; sometimes it is marked by convulsions. The subsequent course of the disease has been divided into three periods—namely: (1) The period of irritation, which is characterised by fever, intolerance of light and noise and by other phenomena of cerebral irritation. (2) The period of pressure during which the signs of excitement give way to somnolence and apathy, the pulse becomes slow and often irregular and the respiration sighing and irregular. The abdomen is retracted; paralysis of some of the cranial nerves and optic neuritis often develop. (3) The period of paralysis characterised by deepening coma, convulsions and a weak rapid pulse. In practice, however, it is difficult to recognise definite stages, the symptoms of one stage passing imperceptibly into those of another; hence it is more satisfactory to briefly consider the variations in the individual symptoms of the disease.

Quite early in the disease the child becomes unable to stand or to sit up; it lies in bed curled up on one side, with the legs drawn up and the arms adducted and flexed at the elbows. The child resents any disturbance; if turned on to its back it at once turns on to its side again, and if the bed-clothes are pulled down the child will at once try to pull them up. In the later stages of the disease extension of the limbs is the rule, and the child lies on its back with the legs stretched out and the hands crossed over the hypogastrium.

At first the face has a frowning expression, but subsequently this is replaced by "the far-off look" or vacant stare. Flushing of the face which is occasional in the early stage tends to become persistent, and at a late period may extend over the upper part of the body. The *tache cerebral*, although not peculiar to tuberculous meningitis, is usually present; it is a bright red flush brought out by gently scratching the skin. Dry and harsh in the early period, the skin may be bathed in sweat during the last few days. Slight stiffness of the neck and slight retraction of the head are occasionally present, but any marked head retraction is only met with in exceptional cases in which there is much tuberculous deposit at the posterior part of the base. In infants the anterior fontanelle often full or bulging early in the disease becomes flattened at a late period.

Pain in the head begins early, and is often severe and continuous. In the infant it is expressed by screaming and possibly by the sudden shrill scream known as "the hydrocephalic cry," and also by knitting of the brows or by the raising of the hands to the head, which is often bored into the pillow. Older children and adults complain of headache which tends to be localised to the frontal region. Sometimes pain is referred to the limbs, the abdomen or to other parts of the body.

The pulse, quickened at first, soon becomes abnormally slow, and is often irregular in rate, force and rhythm. In the last stage of the disease the pulse loses its irregularity and becomes gradually softer and more frequent, till it reaches two hundred or more per minute shortly before death. The temperature may be nearly normal throughout the disease, but as a rule there is an initial slight or moderate degree of pyrexia, which is often lowered at the onset of the meningitis. Subsequently the temperature may become high, and hyperpyrexia is common shortly before death; occasionally, however, at this period the temperature is subnormal. An irregular

and sighing character of respiration may be noticed at an early period, and the Cheyne-Stokes type is common at a later period. During the ascending phase of this type, the pulse quickens and the face becomes deeply flushed, whilst during the descending phase the pulse gets slower and smaller and the flushing often gives way to pallor.

Vomiting, an early symptom, often ceases after the first few days, but may return towards the end of the illness. Sometimes it is excited by the taking of food or by an ophthalmoscopic examination, but frequently it recurs without apparent cause; it is unattended by nausea or retching. Constipation, present from the onset, is usually persistent throughout the illness; occasionally it is very obstinate, but as a rule it responds to simple remedies. Rarely diarrhœa is present. The abdomen is flattened or concave.

In the earliest stages attempts at walking are often attended by incoordination and coarse tremor. In the established disease tremor, especially of the hands, is usually conspicuous. Restless purposive movements are also often present, such as a tremulous plucking at the lips or the genitals. Other movements of similar character are, grinding of the teeth, champing of the jaw and biting the lips. Convulsions may be general or partial in distribution. General convulsions occur at the onset, and more frequently during the third period. The limited variety occurs during the middle period: it corresponds to the Jacksonian type, and indicates a localised cortical lesion, either unilateral or bilateral. Sometimes attacks of unconsciousness take the place of convulsions. Rigidity of one or more limbs may persist after a convulsion: in some cases rigidity of the limbs of one side of the body is associated with a flaccid paralysis of the limbs of the other side. A monoplegic is commoner than a hemiplegic distribution. The knee-jerk present at the beginning becomes sluggish or lost towards the end of the illness, when the superficial reflexes are also impaired or lost.

During the greater part of the illness the urine and feces are apt to be passed involuntarily. Towards the end of the illness retention of urine often occurs: and the power of swallowing tends to diminish, when the patient may have to be fed by a tube.

The pupils, often contracted at first, gradually become dilated, and sometimes show a curious oscillation to light, alternately contracting and dilating. In the last stage they are markedly dilated, and lose their reaction to light. Strabismus and ptosis, varying in degree and temporary or permanent, are common both at an early and a late period in the disease. Slow or jerky movements of the eyeballs, rather than true nystagmus, are sometimes present. During the last few days of life muco-pus collects on the conjunctiva, and superficial ulceration may be seen on the lower margin of the cornea. Optic neuritis, which is usually present during the later stages, is seldom severe. Tubercles in the choroid are occasionally seen, but as a rule only in cases where tuberculosis is wide-spread: by themselves choroidal tubercles do not afford positive proof of the presence of tubercles in the pia mater. Loss of consciousness, a frequent sequel to the early drowsy irritability, gradually deepens into profound coma: sometimes [illegible] partial, and rarely it comes on suddenly [illegible] end of the illness.

The blood shows a moderate degree of leucocytosis: the cerebro-spinal fluid is found to contain tubercle bacilli and a large number of lymphocytes.

Course and Prognosis. In the secondary cases, those in which meningitis supervenes upon phthisis, hip disease or other chronic forms of tuberculosis, the duration of the disease is short and the prognosis is hopeless.

In the primary cases, those in which meningitis comes on during apparently fair health, the duration is longer, usually about three weeks, or occasionally six to seven weeks, and the prognosis, though very bad, is not so absolutely hopeless as in the secondary cases. At any

rate, cases in which characteristic signs of tuberculous meningitis were present have recovered for a time and have died subsequently either from a second attack of meningitis or from general tuberculosis; in such cases old cicatricial tuberculous deposit has been found in the pia-mater. The more localised the deposit the greater the chance of recovery; the more generalised, the less the chance.

The temporary improvements that occur during the course of the disease often give rise to false hopes. Thus a child may suddenly pass from a state of deep coma to apparent convalescence: for a few hours he may be bright and play with his toys and then relapse into fatal unconsciousness.

Pathology. In a well-marked case of tuberculous meningitis the characteristic appearances are the presence of tubercles and of inflammatory exudation at the base of the brain, of a moderate quantity of fluid in the ventricles and of softening of certain parts of the cerebral substance.

The tubercles and inflammatory exudation vary much in character and in relative proportions. The tubercles are grey or yellowish in colour, and vary in size from the finest dust to a millet seed; they are either isolated or they form confluent groups. The exudation thickens the pia-arachnoid and renders it opaque, or it takes the form of greenish or yellowish lymph. In some cases lymph is abundant and tubercles are few and difficult to find; in other cases the converse is the case. These deposits are chiefly found at the base of the brain, especially over the chiasma and in the interpeduncular space; they extend along processes of the pia mater, which dip into the brain and are conspicuous in the Sylvian fissures, the sides of which become glued together. Tubercles are also found in the choroid plexus, in the velum interpositum, over the anterior portion of the superior vermiform process of the cerebellum, and in the ependyma of the lateral ventricles.

They are absent or scanty in proeesses of pia-mater near the cortex.

Tuberculous deposits in the perivascular sheaths of arteries branches sometimes lead to thrombosis, and to areas of softening in the brain. But besides this localised softening the whole cerebrum may feel soft owing mainly to interference with its nutrition in consequence partly of the deposition of tubercles along the small vessels, and partly to actual cellular infiltration of the brain substance. This softening may occur when there is little or no hydrocephalus and appears to be the result of a wide-spread cerebritis. In other cases, when there is a considerable quantity of fluid in the ventricles—the cerebral softening may partly depend on simple maceration. The hydrocephalus, which is usually slight or moderate in degree, is probably rather the result of the inflammatory changes than a mere dropsy from mechanical obstruction; at any rate, it is difficult to find anatomical evidence of adhesions in the neighbourhood of the foramen of Majendie.

In addition to older tuberculous deposits in other parts of the body, to which the meningitis is secondary, it is common to find miliary tubercles in the spinal membranes, in the liver, spleen, lungs and pleuræ and sometimes in the choroid.

EPIDEMIC CEREBRO-SPINAL MENINGITIS: CEREBRO-SPINAL FEVER.

This is an acute infeetions disease due to the invasion of the pia-arachnoid by the diplococcus intracellularis. The disease occurs in epidemics which are rarely widespread, being usually localised to certain towns or districts. Sporadic cases are also met with, and it is probable that posterior-basic meningitis, which may also occur in epidemics, is a closely-allied, if not an identical, form of the same disease.

Etiology. Cerebro-spinal fever is most frequently met

with in countries having a temperate or a cold climate, and is especially apt to appear during the spring and winter months. Its development is favoured by overcrowding and bad sanitary conditions, hence the disease is most prevalent in the poorer quarters of a city. The degree of contagion is slight, probably not more than that of pneumonia, with which in some respects, as in the frequency of herpes and the character of the exudate, it appears to have an affinity. The disease is one of early life; for, although no period is exempt, the majority of patients are under fifteen years of age.

The meningococcus or diplococcus intracellularis is regarded as the specific cause of cerebro-spinal fever. During the first week of the disease it is present in the nasal mucus and in the secretions of the mouth and conjunctiva. It is also found within the polymorphonuclear cells, both of the cerebro-spinal fluid and of the meningeal exudation. The diplococcus is of low vitality, and is rapidly killed either by drying or by exposure to sunlight; infection by dust is therefore unlikely. In all probability the organism reaches the subdural space by means of the lymphatics from the naso-pharynx and the sphenoidal sinuses. Busse, however, draws attention to the frequency with which bronchitis and laryngitis are found in cases occurring early in an epidemic, and holds that any part of the respiratory tract may serve as the port of entry of the meningococcus. The lesions of cerebro-spinal meningitis are sometimes more advanced in the cord than at the base of the brain; in such cases it is possible that the organisms enter the blood-stream and attack the nervous system from the spinal canal.

Symptoms. Occasionally the disease is ushered in by a feeling of malaise, by shivering and pains in the head, back and limbs, but in most cases it sets in abruptly and develops rapidly. Headache is one of the earliest and most constant symptoms: at first occipital or frontal, it soon becomes general, and may occur in paroxysms of great severity. Vomiting is also common at the onset.

and sometimes persists throughout the course of the disease. Rigidity of the muscles of the neck and the back is rarely absent, and frequently causes retraction of the head and arching of the spine; in severe cases

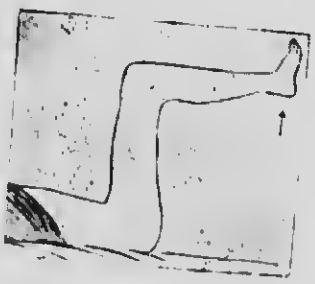


Fig. 163.—Kernig's sign in Meningitis: the leg cannot be extended on the thigh,—cannot be moved upwards in the direction of the arrow. (Williamson).

the head is bent back almost to a right angle and there is opisthotonos. Sometimes the rigidity spreads to the muscles of the jaw, causing trismus; or to the abdomen causing its retraction; or to the limbs. Kernig's sign is generally present. In infants convulsions are common at the onset of the disease and may occur during its course; usually general, they may be unilateral or local owing to irritation of certain portions of the motor cortex. Muscular tremors and twitchings of the face and limbs also occur.

Paralysis most frequently affects the eye muscles; the pupils are dilated, sometimes unequally, and they do not react to light; strabismus is an early symptom which may be permanent. Paraplegia is rare; in some cases its spinal origin is indicated by disturbance of the bladder, by girdle sensations, and by changes in the condition of the knee-jerk. As a rule the knee-jerks are increased and the abdominal reflexes are absent. Cutaneous hyperæsthesia to all forms of stimulation is a characteristic feature of the disease, and tends to be more extreme than in other forms of meningitis.

Marked intolerance to light and sound is frequently present.

During the early stage of the malady delirium of an active character is prominent; it tends to be worse at night and during a rise of temperature: sometimes it is furious and maniacal. In a few days it gives way to stupor which rapidly develops into coma.

The temperature runs an irregular and variable course, and no uniform or typical curve can be distinguished. Exceptionally there is little or no fever, but as a rule the temperature rises to 102° or 103°, and in severe cases to 105°, or even higher just before death. Three types of pyrexia have been observed: a type resembling that of enteric fever, a remittent type, and an intermittent type like that of malaria. The pulse-rate is irregular, and frequently does not correspond with the temperature; it may not be more than ninety when the temperature is 103°. Marked daily variations are common, at one time the pulse being rapid and irregular, at another slow and regular. Respiration not often increased in frequency unless pneumonia is present, is sighing in character and irregular in rhythm: it may be of the Cheyne-Stokes type.

The most characteristic skin lesions are purpuric spots and herpes. The purpuric eruption, which is common in severe cases, spreads rapidly, and may become general within a few hours. Herpes, which occurs as frequently as in pneumonia, is met with on the lips, nostrils and ears, and occasionally on the limbs. Urticaria and a diffuse erythema about the joints, abdomen or chest, also occur.

A marked polymorpho-nuclear leucocytosis is present in all acute cases, but not usually in cases which run a protracted course; the number of leucocytes varies from 25,000 to 50,000 per c.mm. The cerebro-spinal fluid is opaque, turbid or purulent; polymorpho-nuclear leucocytes are abundant and much in excess of the lymphocytes; the diplococcus is found either within or outside the polymorpho-nuclear cells.

The optic discs are frequently congested; sometimes there is definite papillitis. Purulent choroido-iritis and keratitis also occur. There may be deafness as a result either of otitis media, or of inflammation of the labyrinth.

Complications. The following complications have been met with:—Pneumonia, endocarditis, pericarditis, tonsillitis, parotitis and arthritis. The arthritis may be either serous or purulent; the knee-joints are especially likely to be affected. Sometimes there is a multiple suppurative arthritis of great intensity, when the case may closely resemble one of acute pyæmia. The most common sequela is deafness, which, if it occurs in children and is complete, may lead to permanent deaf-muntism. Other consequences of the disease are blindness, chronic hydrocephalus, mental impairment and epileptic fits.

Course and Prognosis. The course of the disease is very variable; its duration may vary from a few hours to several months. In fatal cases death usually occurs within a week, and in the malignant or fulminant type the patient may die within twenty-four hours from the onset. In favourable cases, characterised by incompleteness of the coma, and by mildness of the other symptoms, improvement often begins towards the end of the first week; convalescence is usually tedious and prolonged. Sometimes the disease persists for many weeks or months; in such protracted cases the patient becomes greatly emaciated, and may die either from exhaustion or possibly in consequence of hydrocephalus or of cerebral abscess. An abortive type is described, in which, after a day or two of headache, stiffness of the neck, pains in the limbs and fever, recovery sets in and is soon complete.

In all cases of the disease the outlook is very grave: the rate of mortality varies much in different epidemics, the average percentage of fatal cases is about sixty. In the event of recovery some degree of deafness, blindness or of dementia is likely to be left.

Morbid Anatomy. In malignant, rapidly fatal cases, there may be only extreme congestion of the membranes. In acute cases which have lasted more than a few days, the meninges of the brain and spinal cord are inflamed and often covered with a purulent exudation, which is most marked at the base of the brain, and tends to collect in the subarachnoid space. The ventricles of the brain are frequently enlarged and may contain either turbid or purulent fluid: their walls may be red and œdematous. The brain, the spinal cord and their nerve roots may also be inflamed and softened in places. In chronic cases the pia-arachnoid is thickened, and shows yellow patches where the exudation has been.

POSTERIOR BASIC MENINGITIS.

Etiology. Unlike tuberculous meningitis, posterior basic meningitis occurs most frequently during the first year of life, and attains its maximum frequency between the ages of three and six months. Probably not more than twenty per cent. of the cases are met with during the second year, and only very few at a later period of childhood. The disease presents a seasonal variation, being most common during the later winter months and in the spring. Of immediate antecedents, signs of catarrh, either nasal, bronchial or intestinal, have been observed in some cases. In other cases a fall or blow on the head appears to have been a factor in the production of the disease; its influence probably being to lower resistance to the invasion of the organism. This, as shown by Still, is a diplococcus, which differs markedly from that of pneumonia, but closely resembles the diplococcus intracellularis of cerebro-spinal meningitis, of which it is probably a modified and sporadic form.

Symptoms. The onset of the disease is usually acute: the first symptoms are vomiting and retraction of the head, or sometimes convulsive attacks.

Head retraction always occurs at an early period, and is a characteristic feature throughout the disease: in

the fatal cases it usually persists until death; in the cases that recover it continues for several weeks. In degree it varies much in different cases, and in the same case from day to day. It depends on a tonic contraction of the posterior cervical muscles, which is usually caused by irritation of the upper cervical nerves; occasionally, however, these nerves are not implicated in the meningitis when the retraction may be excited reflexly either by an otitis, or by inflammation in the sub-arachnoid space.

Fig. 164.—Showing attitude in a case of posterior basic meningitis. (Photograph taken by Slinger).

Tonic spasm may be limited to the neck, but sometimes it affects the muscles of the back and the limbs. Kernig's sign is present at an early period. Spasm of the dorsal muscles produces opisthotonos, which may be so extreme that the occiput almost touches the sacrum. Spasm of the limbs produces certain attitudes varying according to the predominance of the spasm in the flexor or the extensor muscles. Thus the joints of the upper limbs may be flexed, whilst those of the lower are extended, or *vice versâ*. As a rule, and especially when the spasm is intense, extension predominates; the arms are adducted and rigidly extended, while the hands are super-pronated with the fingers firmly clenched and

pressed into the palm. The lower limbs are usually adducted and rigidly extended; the heels are drawn up and the feet inverted.

In such marked cases there are often periodical exacerbations of the spasm in which the limbs become stiffer and the back more arched; the chest, too, may be rigidly fixed and the jaws tightly closed, the whole attitude closely resembling that of severe tetanus. This tonic spasm of the dorsal and limb muscles may exist apart from spinal meningitis, when it is probably due to irritation of the cerebellum which has a direct effect on the lower neurons of he same side. Paroxysms of tonic spasm are commoner than epileptoid convulsions, which, however, sometimes occur at an early period as well as, although more rarely, at a late period of the illness. The convulsions are probably due to irritation of the cortex owing to the presence of inflammatory changes in the membranes covering the convolutions. In some cases spasm of the neck-muscles is followed by weakness, when the patient becomes unable to hold his head erect; paralysis of other muscles is rarely seen, exceptionally it affects a limb or one side of the face.

During the early period, crying and screaming may occur, and especially if attempts are made to bring the head forward or to turn the child from its side on to its back. During the later period when hydrocephalus is present, crying usually ceases and the child lies quiet, often in a state of stupor. Sucking movements of the lips, champing of the lower jaw, grinding of the teeth and protrusion and retraction of the tongue are often present. They are probably caused by irritation of cortical centres; when the inflammatory exudation involves the tips of the temporo-sphenoidal lobes, there may be olfactory and gustatory sensations.

The tendon reactions are present, and sometimes exaggerated. In addition to the tonic spasms above mentioned, there may be strabismus of the spasmodic variety, or a marked retraction of the upper lids, giving the eyes a fixed staring look. Nystagmus is sometimes

present: the oscillations which are not dependent on forced movements in a particular direction may be vertical, lateral or rotatory. The pupils often react sluggishly to light: they tend to be contracted in the earlier period and dilated in the later period of the disease.

With regard to vision, it may be noticed that the child does not follow moving objects and does not blink when the finger is approached suddenly towards the eye. In fact some degree of blindness occurs in about one-third of the cases of basic meningitis, sometimes soon after the onset, but usually not till the third or fourth week. Probably it is of central origin, and certainly it does not depend on optic neuritis or optic atrophy, either of these conditions being very rare, even in cases when the optic commissure is surrounded by inflammatory lymph. Vision may be restored if the child survives. The hearing is generally acute; the rarity of deafness in the sporadic form of meningococcic meningitis may be contrasted with its frequency in the epidemic variety.

Undue fulness of the anterior fontanelle may be noticed during the first week: the fulness becomes more marked as the pressure from hydrocephalus increases, when the posterior and lateral fontanelles may reappear, the cranial sutures becoming separated. If the ventricles are much distended, the head gets larger and heavier, and tends to become spherical in shape. When life is prolonged the hydrocephalus is sometimes extreme, and the aspect of the head and eyes may be identical with that of congenital hydrocephalus. Stupor, a common and sometimes an early symptom in posterior basic meningitis, frequently deepens into persistent coma.

The temperature is variable; at first it is usually slightly raised, subsequently it is prone to be irregular. In protracted cases the temperature often becomes normal or sub-normal, whilst either a subnormal temperature or hyper-pyrexia may usher in a fatal

termination. The pulse, often rapid, is usually regular; it may be slow in older children. The common type of respiration in the advanced disease is an alternation of long pauses with a series of rapid deep inspirations, which are of nearly equal depth; in some cases Cheyne-Stokes type of respiration is present.

Vomiting occurs more frequently, while constipation is less frequent and obstinate than in tuberculous meningitis. Another difference between the two varieties of meningitis is the appearance of the abdomen, much retraction being very rare in the posterior basal variety. Skin eruptions are rare; a dusky erythema on the body and herpes on the lips have been occasionally observed. There is always loss of flesh; in protracted cases emaciation is marked, even when there is no diarrhœa and scarcely any vomiting, and it may last for some time after all active symptoms have subsided.

During the acute stage of the disease the blood generally shows a leucocytosis of from 15,000 to 25,000 per cmm.; the cerebro-spinal fluid is always turbid and frequently purulent, and contains an excess of polymorpho-nuclear cells. Diplococci may be found both outside and inside the cells. In the later stages the fluid becomes less turbid and contains fewer cells.

Death may occur within a few days, but the average duration of fatal cases is about seven weeks. Recovery takes place in about fifteen per cent. of the cases, but it is often very incomplete, and the child remains physically and mentally backward, being afflicted with blindness, or suffering from some degree of hydrocephalus, or of dementia. In many cases during the period of apparent cure death occurs suddenly and without obvious cause; in other cases it is the result of some complication, as diarrhœa or broncho-pneumonia.

Pathology. The chief region implicated is the posterior part of the base of the brain. Here the pia-arachnoid is thickened and is covered with exudation, which is fibrino-purulent in character, but not often markedly purulent. This exudation extends over the medulla,

the inferior surface of the cerebellum and the fold of arachnoid between them. Morbid changes may extend forwards over the pons as far as the optic chiasma and the tips of the temporo-sphenoidal lobes, and upwards along the lining membrane of the ventricles and downwards around the spinal cord. Some degree of hydrocephalus is present in nearly every case. This results either (1) from pressure of abundant lymph or from adhesions between the cerebellum and medulla, which close the foramen of Majendie and the two lateral foramina of Luschka; or (2) from adhesions which obliterate the iter and thus shut off the third and lateral ventricles from the fourth ventricle. The obstructive origin of the hydrocephalus is indicated by the transparency of the fluid which distends the ventricles. Sometimes, however, the fluid is opaque and may contain pus, when there is usually evidence of inflammation of the lining membrane of the ventricles. The path of invasion by the special micro-organisms is probably from the naso-pharynx along the Eustachian tube to the middle ear and thence to the pia-arachnoid.

The viscera are usually normal: occasionally a peri-arthritis occurs as a complication, and it is interesting to note that the same diplococcus has been found in the exudation about the joint as in that of the meninges.

SUPPURATIVE MENINGITIS.

Pyogenic meningitis. In this variety, which is induced by staphylococci and streptococci, the source of infection is usually some focus of disease in the immediate neighbourhood, such as otitis media, suppuration of the mastoid cells, abscess of the brain, or disease of the nasal sinuses, or of the cranial bones. Sometimes the meningitis occurs as a complication of septicæmia, erysipelas, scarlet fever or of some other infective disorder.

The meningitis may be limited in area as when it has been set up by local disease, but as a rule it is widespread and the meninges both of the vortex and the base

of the brain may be involved; frequently the whole surface of the brain is covered with a thick yellow or green layer of pus. The purulent exudation is seen to follow the course of the larger vessels and to dip down with the pia-mater into the sulci; sometimes it spreads into the spinal canal, the posterior surface of the cord being involved to a greater extent than the anterior. The tissue of the brain and spinal cord may appear quite normal; although sometimes the cut surface of the brain presents ecchymoses and minute abscesses.

The symptoms are those common to other forms of acute leptomeningitis, namely, headache, vomiting, pyrexia, optic neuritis, convulsions, delirium and coma. They tend to be very severe, and the course of the disease is exceedingly rapid, death often occurring within two or three days.

Pneumococcal meningitis. This may be either primary, the meninges being the first part of the body to be affected by the pneumococcus, or secondary to a pneumococcal infection of the lung or pleura. The disease occurs most frequently in cases of empyema and of ear disease; it may also be set up by malignant endocarditis. Out of twenty-three cases collected by F. Batten, seven followed empyema and purulent pericarditis, seven ear disease, whilst others followed pneumonia, abdominal infections, abscesses and joint infection.

The symptoms and morbid anatomy are similar to those of pyogenic meningitis. There is a rapid development of headache, vomiting, high temperature, localised paralysis, convulsions and coma. The course is often even more rapid than that of pyogenic meningitis, death sometimes occurring within twenty-four hours.

Meningitis produced by other organisms. Of the remaining forms of meningitis it is only necessary to make a brief allusion to the influenzal and the typhoid varieties.

Influenzal meningitis. In a well-marked case a patient with or without the usual symptoms marking the

PLATE VIII.

Cut surface of brain, showing hyperæmia of the cortex, scattered bleeding points and hæmorrhage into the white matter (from a drawing by Dr. W. E. Fothergill).

The same case as Fig. 165.

onset of influenza, gradually becomes drowsy and apathetic; he answers questions with difficulty, and in a few days becomes comatose. Recovery may occur, but a fatal termination is more common. The brain may reveal nothing abnormal, or there may be either con-

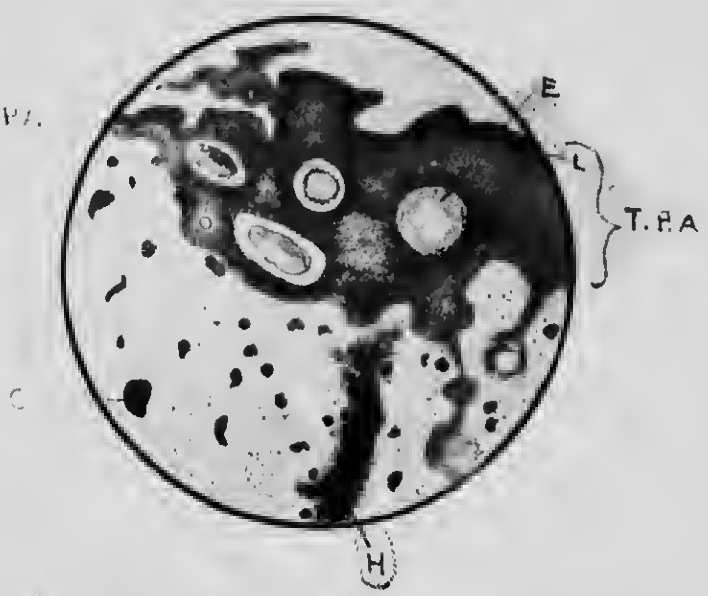


Fig. 165.—Microscopic appearances of pia arachnoid and subjacent tissues in a case of hæmorrhagic meningo-encephalitis from influenza. P.A., pia arachnoid; T.P.A., thickened pia arachnoid; E., endarteritis; L., leucocytes and fibrin; C., engorged vessel; H., hæmorrhage. (Drawn by Dr. W. E. Fothergill).

gestion of its surface or purulent meningitis with or without encephalitis, which is generally hæmorrhagic in character. In such cases Pfeiffer's bacillus has been found both in the cerebro-spinal fluid and in the fibrino-purulent exudation over the brain. Rhea has suggested that the areas of acute encephalitis in influenzal meningitis may have a bearing on the transient as well as on the permanent paralyses which follow some of the acute cerebral conditions in children. From one of his cases, in which life was prolonged for eighty-nine days, the bacillus influenza was obtained from the circulating blood, in a pure culture, on the fourth day of the illness.

The meningitis, although frequently accompanied by encephalitis, may exist alone. It affects the vertex more commonly than the base, and is usually of the suppurative variety. It may be either the primary effect of the bacilli, or it may be secondary to suppuration in the ear or in the nasal cavities, which is set up by the bacilli. No doubt in many cases of meningitis there is a mixed infection, staphylococci and streptococci being active agents as well as influenzal bacilli; the bacilli may first attack the brain and pave the way for the entry of other organisms.

Typhoid meningitis. A few cases are on record in which a patient has suffered from symptoms of meningitis without any signs of enteric fever; from the fluid withdrawn by lumbar puncture a pure culture of the typhoid bacillus has been obtained, and the necropsy has revealed purulent exudation over some part of the brain, but no lesions in the intestine.

Less rarely symptoms of cerebro-spinal meningitis have developed during the course of enteric fever, and at the necropsy either a serous or a purulent meningitis has been found in association with lesions characteristic of enteric fever.

Diagnosis. The diagnosis of meningitis is often one of the most difficult problems in clinical medicine. In the first place, the initial symptoms of meningitis may closely resemble those produced by disease outside the nervous system. In the second place, although there may be unmistakable signs of intra-cranial disease, it may be difficult to determine the situation and nature of such disease, to decide whether the symptoms are due to meningitis or to tumour, abscess or other local lesion. In the third place, the symptoms of meningitis may be quite insignificant or they may be masked by those produced by an associated morbid condition, such as hysteria. In the fourth place, there may be typical symptoms of meningitis in cases which either recover, or if they prove fatal from any cause, do not show the morbid changes of meningitis. To this condition the

name meningism is given. Lastly, even when the presence of meningitis is established, it is not always easy to come to a decision regarding its nature.

1. *Diseases outside the nervous system.* Headache, vomiting, pyrexia, drowsiness and convulsions may constitute the early symptoms of gastro-intestinal catarrh, of one of the exanthems or of pneumonia, when it may be impossible for a time to say whether or not meningitis is present. In some cases of enteric fever in which the rash is either late in appearing or is absent altogether, and in which constipation and irritability are prominent and persistent features, the difficulty in diagnosis may be great. Apart from the assistance to be derived from Widal's reaction and lumbar puncture the following points are often of service. In enteric fever there is a tendency to muscular flaccidity, in meningitis to rigidity, the tendency being often shown by the facial expression and the attitude of the body. The relaxed features of the patient with enteric are in marked contrast to the frowning and the far-off fixation expression of the patient with meningitis. The decubitus in enteric is dorsal, and the patient looks as if he were sinking down into the bed: the decubitus in meningitis is lateral, and there is a tendency to the curled-up position. The abdomen is usually distended in enteric, whereas it is often retracted in meningitis. In a case of enteric fever, when the bed-clothes are pulled down the patient takes little or no notice, but in a case of meningitis the patient immediately draws them up again. Headache may be very severe in enteric fever, but it subsides when delirium appears, whereas in meningitis it persists after the appearance of the delirium. Kernig's sign, which is frequently present in meningitis, does not occur in enteric fever.

Cerebral symptoms similar to those of tuberculous meningitis are sometimes observed in marasmic children, giving rise to the condition known as "hydrocephaloid." The fontanelle, however, is depressed, whereas it is frequently bulged in meningitis.

Retraction of the head, which is such a marked feature of the meningococcic variety, and which may occur in other varieties of meningitis, is also sometimes present in cases of otitis media, of rheumatism of the muscles of the back of the neck, of apical pneumonia, of mumps, of enlarged cervical glands, or in cases of retropharyngeal abscess. If these facts are borne in mind errors in diagnosis may generally be avoided. A careful examination of the ears is of special importance, for disease of these organs does not always give rise to obvious local signs. The acute double otitis media of young children may be attended by severe pain in the head, by vomiting, pyrexia and convulsions, symptoms which usually subside when the tympanic membrane on both sides is punctured.

2. *Other intra-cranial diseases.* In thrombosis of the cerebral sinuses there may be intense headache and vomiting, together with short repeated screams simulating the hydrocephalic cry. The presence of venous engorgement or of œdema about the eyes, the temples or the mastoid process, or of haemorrhages in the fundus of the eye would be strongly in favour of thrombosis.

The symptoms of intra-cranial tumour may resemble those of tuberculous meningitis, but as a rule they are more prolonged. In both diseases paralysis of the limbs may come on at an early period, but in tumour and also in abscess of the brain the paralysis develops more gradually than in meningitis. Further, the optic neuritis of tumour is much more intense than that of meningitis. The symptoms of an abscess supervening on a chronic empyema may closely simulate those of tuberculous meningitis supervening upon pulmonary tuberculosis. The differential diagnosis is largely based on a consideration of the history of the case and of the pathological conditions associated with the cerebral symptoms.

3. *Hysteria.* In adult life, and even in later childhood, hysterical manifestations may develop during the early stage of tuberculous meningitis: they alone may

be present or their prominence may prevent the right significance being attached to other symptoms which, though less prominent, are of greater importance. The recognition of this fact will lead to repeated careful examinations of the patient, and will thus lessen the risk of overlooking definite signs of serious disease.

4. The symptoms of *meningism* may be identical with those of meningitis: after a few days, however, they pass away rapidly, and the child regains perfect health. The diagnosis mainly rests upon the examination of the cerebro-spinal fluid obtained by lumbar puncture. In meningism this fluid is perfectly normal and contains neither cells nor organisms. Other points of distinction, according to Tylecote, are as follows: In the acute infections meningism is often early in its occurrence and of short duration: Kernig's sign is usually absent in meningism; fever is more often absent in meningism than in meningitis: the onset of cerebral symptoms for the first time during the defervescent or convalescent stage of an acute infection is much against meningism: the onset of meningism is acute, whereas in meningitis the symptoms often come on gradually; slowness of the pulse and irregular respiration are common in meningitis, rare in meningism.

In the *differentiation of the various forms of meningitis* the microscopic and cultural examination of the cerebro-spinal fluid is often of considerable value. The results of such an examination, together with a summary of some of the characteristic features of the chief varieties of meningitis are given in the following table:—

	Tuberculous Meningitis	Cerebro-Spinal Meningitis	Posterior Basic Meningitis	Suppurative Meningitis
Age at onset	Children and Young Adults		Under one year	All ages
Special features	Irregular temperature and pulse. Combination of headache, vomiting, and marked constipation.	Marked and persistent head retraction. Erythematous herpetic, and petechial rashes: arthritic swellings.	Extreme emaciation.	Convulsions, delirium, and coma.
Cerebro-spinal fluid	Clear or turbid; preponderance of mononuclear cells; tubercle bacillus.	Turbid or purulent. Preponderance of polymorpho-nuclear cells. Diplococcus intra-cellularis.		Staphylococcus, Streptococcus, Pneumococcus.
Duration	Two to eight weeks; often about *three weeks*.	A few days to several weeks or months.	One week to four months: often about *three months*.	A few days to two or three weeks: often about *three days*.
Prognosis	Almost invariably fatal.	Recovery not uncommon.	Commonly fatal.	Invariably fatal.

Treatment. In all cases of meningitis the patient should be kept on a water-bed in a quiet, darkened, cool and well-ventilated room. Perfect quiet, especially in the case of young children, is of great importance, and the nurse should be instructed to avoid making any unnecessary noise or movement, and as far as possible to leave the child alone. It is usually advisable to shave the head and to keep it cool by the application of an ice-bag. If the ice-bag does not relieve the headache

and seems to increase the child's irritability, it must be removed; sometimes the headache is definitely relieved by small doses of antipyrin or of phenacetin with caffein citrate. When there is much restlessness and moaning with pain, the cautious administration of morphia may be desirable. For a high temperature sponging the body with tepid water is often beneficial.

Abundant liquid nourishment is required; stimulants should only be given when there are signs of cardiac failure. During the earlier stages of the disease there is little or no difficulty in swallowing, but as soon as coma sets in, nasal feeding and nutrient enemata become necessary. Troublesome vomiting which does not respond to the usual remedies is sometimes relieved or removed by washing out the stomach. Constipation, which is such a marked feature in tuberculous meningitis, is best treated by calomel, or by grey powder. In the later stages of the disease an action of the bowels may be obtained by enemata or by glycerine suppositories.

There is no satisfactory evidence that any drug has much influence on the course of the disease. It is, however, generally believed that mercury, which is best given by inunction, is of some service during the acute stage; one drachm of the blue ointment should be rubbed into the scalp, axilla or groin once or twice a day, the patient being kept slightly, but definitely, under the influence of the mercury. For the later stages of meningitis the iodides appear to be useful; when not well tolerated ten to twenty minims of iodipin may be injected subcutaneously.

Lumbar puncture is advocated for all forms of meningitis, and especially for the meningococcic varieties; a few cases are recorded in which the operation was followed by recovery. But whether or not the withdrawal of cerebro-spinal fluid has a curative effect it is certainly of value for the relief of symptoms of increased intra-cranial pressure, such as severe headache, convulsions and coma. In some cases the operation may be performed daily, the amount of fluid withdrawn being

regulated by the amount of pressure, as shown by the degree of rapidity with which the fluid escapes. At the first operation the fluid may spurt out in a stream, when thirty c.c. may be safely removed even in the case of an infant. If on subsequent occasions little or no fluid escapes lumbar puncture must be discontinued.

Intra-thecal injections of various salts of silver and of anti-bacterial serums have been tried, but without definite success, except possibly in cases of cerebro-spinal meningitis. In this variety Flexner's serum appears to have been beneficial in several cases. In mild forms of the disease fifteen c.c. and in severe forms twenty c.c of the serum should be injected into the spinal canal, after the withdrawal of about forty c.c. of the cerebro-spinal fluid. It is stated that by this method of treatment the mortality of cerebro-spinal meningitis is considerably reduced. The serums known by the names of Ruppel, Dopter and of Kolle and Wassermann have also been tried and with some success, but no good results have yet been recorded in cases of posterior-basic meningitis.

The recent preparations of Koch's tuberculin (T.R. and the Tub. Bac. Emulsion) have been subcutaneously injected in cases of tuberculous meningitis, but there is as yet no satisfactory evidence regarding their effects on the disease.

SECTION XV.

Cerebral Vascular Lesions.

The most important cerebral lesions of vascular origin are hæmorrhage from rupture of an artery, softening from complete occlusion of an artery and degeneration of the nerve elements without either hæmorrhage or softening as a result of extensive arterio-sclerosis. In complete occlusion the obstructing agent is either locally formed thrombosis, or is brought from the heart or elsewhere embolism. Venous hæmorrhage and venous and sinus thrombosis may also occur. Hæmorrhage from the cortical veins is one of the causes of paralysis in young children (see p. 100). Lastly one of the larger arteries is sometimes the seat of aneurysm.

CEREBRAL HÆMORRHAGE.

Etiology. Although no period of life is exempt, cerebral hæmorrhage is rare before middle life and occurs most frequently in persons between the ages of forty and sixty: at a later age there appears to be less liability probably because a dangerous condition of arterial degeneration was reached at an earlier period. It occurs oftener in men than in women perhaps because they are more exposed to muscular strains, to syphilis, alcohol, gout and other causes which tend to promote the development of arterial degeneration. Heredity plays an important part as is shown by the frequency of cerebral hæmorrhage in some families and its rarity in others.

The condition is liable to occur in certain general blood diseases as pernicious anæmia, purpura, scurvy and especially leukæmia. It is met with in the course of infective endocarditis as an indirect result of embolism. In many cases rupture of the weakened vessel occurs at a time when the blood pressure is

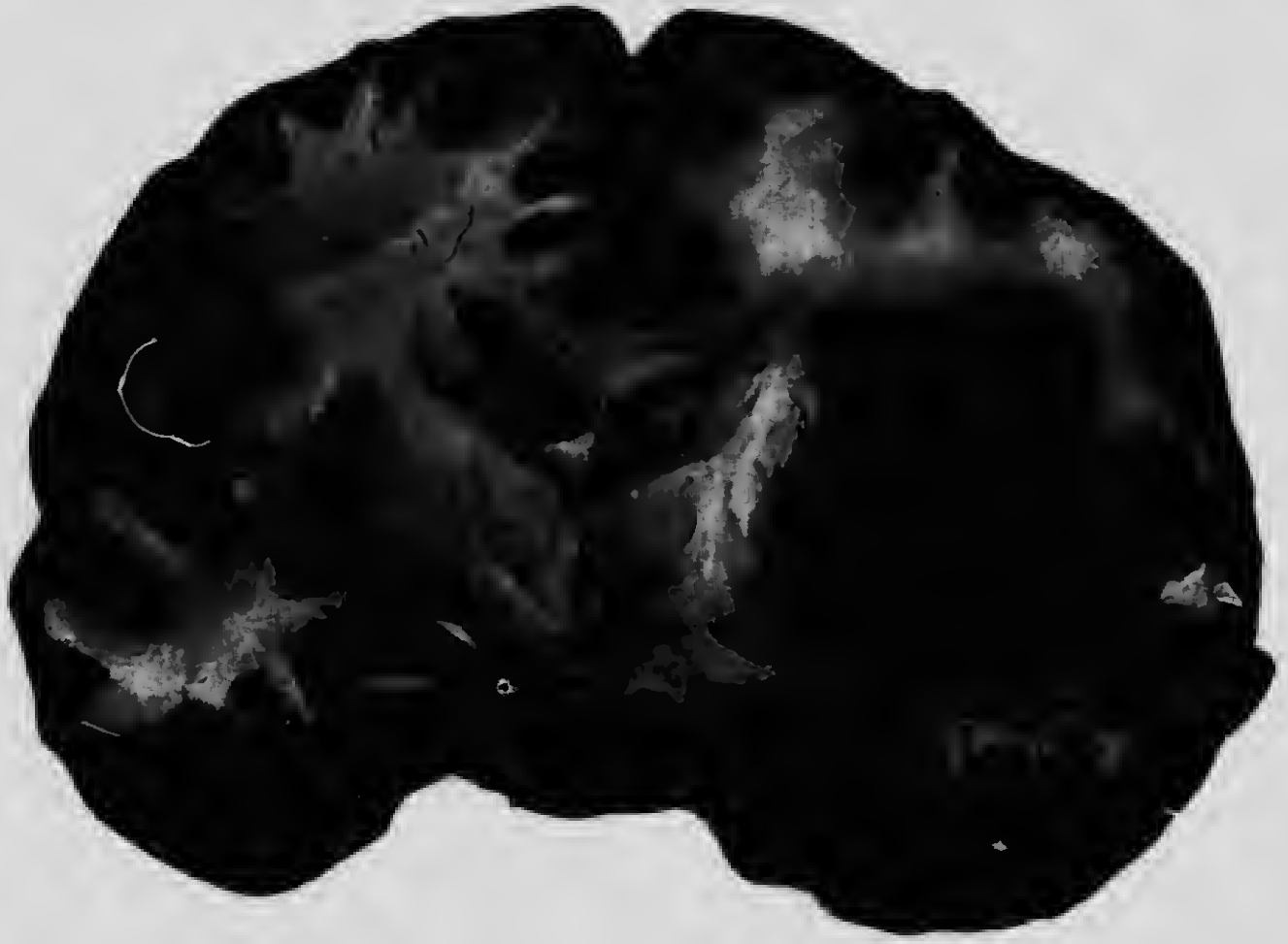
temporarily increased, as by sudden mental excitement; or by a violent muscular effort, as in lifting a heavy weight, in coughing, vomiting, or in straining at stool.

Yet in other cases the rupture takes place during sleep when the general circulation is tranquil; although at this time it is possible that there may be increased blood pressure in certain parts of the brain, as the central ganglia. The investigations of Ernest Jones seem to indicate that "rest in bed and especially sleep, protect to some extent against cerebral hæmorrhage," and that "severe exertion and time of day appear to have had too much stress laid on them in the past."

Pathology. Post mortem examinations bring out clearly the close association of cerebral hæmorrhage with granular kidneys, hypertrophy of the left ventricle and arterio-sclerosis. A consideration of this fact indicates that two causes are prominent in leading to rupture of a vessel, namely, degeneration of its walls and increased blood pressure. A third factor is also of importance, namely, the condition of the tissue which surrounds and supports the weakened vessels. If this tissue is healthy the blood pressure may be inadequate to rupture a degenerated artery, but if its resistance is impaired, owing to the changes produced by malnutrition—a result which frequently occurs in consequence of the slowing of the blood stream along the narrowed and roughened vessels—then the degenerated artery may be easily ruptured by a temporary yet abrupt increase in the blood pressure, such as that produced by physical strain or by a sudden emotion.

The character of the arterial degeneration varies in different cases. Atheroma which mainly affects the larger arteries at the base of the brain, and the diffuse form of arterio-sclerosis which affects the smaller arteries may be present either separately or in combination. The degenerative changes in the walls of the small arterioles lead to the formation of miliary aneurysms, which are found in a large number of cases of cerebral hæmarrhage. To the naked eye they look like minute

PLATE IX.



Large haemorrhage into right hemisphere.

red grains; they vary in size from that of a pin's head to that of a pea. They may be found apart from disease

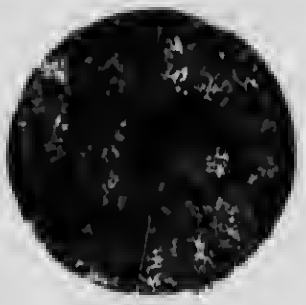


Fig. 166. A miliary aneurysm of the cerebral cortex. (Rindfleisch and Van Schaick.)

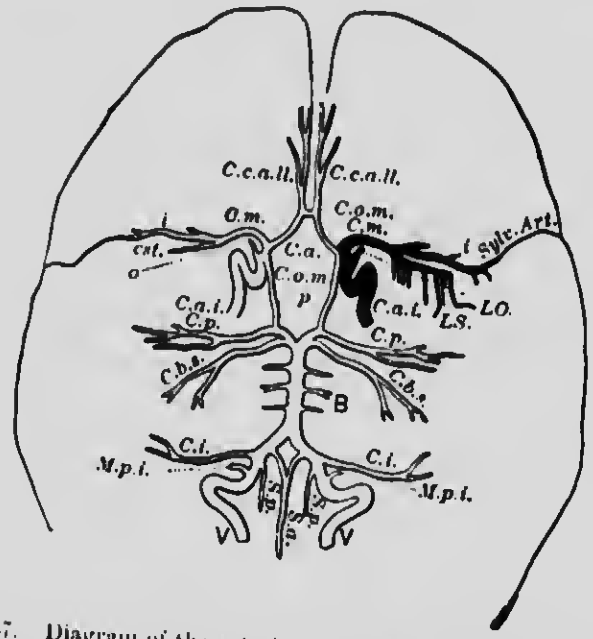


Fig. 167. Diagram of the arteries of the base of the brain, showing L.O., the lenticular-optic, and L.S., the enticular-striate set of arteries. One of the latter is called the artery of cerebral hæmorrhage. V, vertebral; S.a., spinalis ant.; S.p., spinalis post.; B, basilar with median branches; Cb.s., superior cerebellar; C.i., inferior cerebellar; C.p., posterior cerebellar; Com.p., posterior communicating; Ca.i., internal carotid; o, ophthalmic; C.m., middle cerebral; c.st., artery to corpus striatum; C.call. artery to corp. callosum. (Dercum.)

of the heart, the kidneys or the larger arteries: but their formation and their rupture are much favoured by the presence of atheroma, which by impairing the elasticity of the large arteries tends to raise the blood pressure in their branches, the walls of which receive the shock of the heart-beat with abnormal force. The relative frequency in position of miliary aneurysms corresponds to that of hæmorrhage, the most frequent region being the internal capsule, the lenticular nucleus and the optic thalamus. Hæmorrhage also takes place into the centrum ovale, especially its anterior portion, the cortex and the pons, and more rarely into the cere-

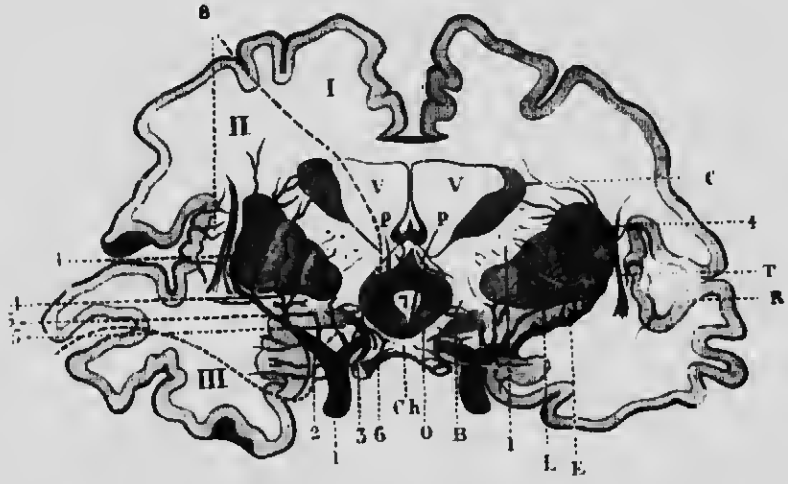


Fig. 168. Transverse section of the cerebral hemispheres, about 1 cm. behind the optic commissure. (From Duret.) Arteries of the corpus striatum: Ch, chiasma; B, section of the optic tract; L, lenticular nucleus; I, internal capsule; C, caudate nucleus; E, external capsule; T, claustrum; R, island of Reil; V, V, section of the lateral ventricle; P, P, anterior pillars of the fornix; O, grey substance of the third ventricle. Vascular areas: I, anterior cerebral artery; II, middle cerebral artery; III, posterior cerebral artery—1, internal carotid artery; 2, sylvian artery; 3, anterior cerebral artery; 4, 4, external arteries of the corpus striatum (lenticulo-striate artery); 5, 5, internal arteries of the corpus striatum (lenticular arteries). The opto-striate artery is not represented in the figure.

bellum, the temporo-sphenoidal lobe, the crus and the medulla. In explanation of the frequency of rupture of the arterioles which supply the internal capsule and

the basal ganglia may be mentioned that they come off at right angles from the middle cerebral trunk, and are terminal arteries. The lenticulo-striate branch is particularly prone to disease and rupture and hence has been called "the artery of haemorrhage." When the extravasation is a large one it may tear its way into the lateral ventricles and escape below into the sub-arachnoid space, or it may force its way through the hemisphere towards the cortex; sometimes it extends down to the crus. If the patient survives the attack, the blood coagulates and the clot gradually undergoes disintegration and absorption, until ultimately either a cicatrix or a cyst-like cavity with a fibrous wall and serous contents alone remains.

The occasional occurrence of haemorrhage in the acute specific fever, in leukaemia and other blood diseases is probably the result of an acute degeneration of the arterial walls produced in some cases by the action of toxins circulating in the blood. In infective endocarditis a cerebral artery may be blocked by an embolus consisting of fibrin loaded with microbes; as a consequence acute inflammation of the arterial wall is set up tending to the formation of an aneurysm which may rupture. Another source of haemorrhage is the escape of blood from a soft tumour, especially a glioma.

Symptoms. The usual and immediate result of haemorrhage into the brain is loss of consciousness; should consciousness be regained a common event — the most frequent permanent result is hemiplegia.

The rapidity of the onset and the degree of the coma vary much in different cases, partly owing to the position and partly owing to the amount of the extravasation. Thus a comparatively small haemorrhage into the pons or the medulla may cause profound and rapidly fatal coma, whereas one near the internal capsule may be expressed by paralysis without loss of consciousness. The escape of a large quantity of blood into the brain gives rise to sudden and deep coma; the

patient is struck down—has a stroke— and may die in a few minutes. Such a rapidly fatal result is rare; as a rule unconsciousness has a gradual development — ingravescent apoplexy—and for a short time is preceded by various symptoms which doubtless indicate the escape of a small quantity of blood. These symptoms, occurring separately, or in combination, are: a sudden pain in the head, giddiness, vomiting, convulsions, a difficulty in speaking, weakness or tingling in the extremities, or some mental disturbance. Sometimes such symptoms have occurred for days or weeks before the stroke, when they usually depend on a defective supply of blood to the brain owing to disease of its vessels; they more frequently precede thrombosis than hæmorrhage. In some cases, however, they may depend on minute hæmorrhages occurring from time to time before the apoplectic attack. Of other antecedent conditions may be mentioned epistaxis and the presence of retinal hæmorrhages or of albuminuric retinitis. The degree of unconsciousness presents every variation between slight mental dulness and profound coma: in many cases the patient can be roused sufficiently to protrude his tongue or to mumble a few words in answer to questions.

When the coma is deep the patient does not respond to sensory stimuli; his limbs are flaccid and their muscles toneless. Muscular relaxation is also seen in the puffing out of the cheeks during expiration, and in the flaccidity of the soft palate which gives rise to stertor. The veins of the face and neck are usually distended, and the face has a flushed turgid appearance; occasionally it is pale and pinched. The tendon jerks, the conjunctival, and all the superficial reflexes are abolished. The condition of the pupils varies: they may be unequal; sometimes they are widely dilated, sometimes, as in pontine hæmorrhage, they are much contracted: their response to light is completely lost. Respiration is deep, laboured, stertorous and often irregular; it may take on the Cheyne-

Stokes rhythm. The pulse as a rule is full, bounding and incompressible; often slow it may be rapid, and in grave cases is irregular. The power of swallowing is lost; urine and fæces are passed involuntarily.

Acute bedsores may form on the gluteal region of the paralysed side; hypostatic pneumonia also may occur. Both these conditions are of grave significance. The temperature usually falls immediately after an apoplectic attack reaching 96° or lower, and in the rapidly fatal cases it may remain low until death. If life is continued for twelve or more hours the initial fall is followed by a rise of temperature, either gradual or rapid. In fatal cases the secondary rise may reach 105° or even higher, but in less severe cases the temperature often oscillates for some days between 99° and 101°, and then in cases of recovery gradually sinks to normal.

This pyrexia associated with headache, anorexia and other symptoms of febrile disturbance indicates the presence of inflammatory changes about the lesion; during this stage stiffness of the paralysed limbs "early rigidity" may be detected. Frequently also there is a difference in the temperature of the two sides of the body, the paralysed limbs being redder and warmer than their fellows.

In cases of haemorrhage into the pons, the medulla or the grey matter of the central ganglia, there may be no initial fall of temperature, which rises from the first and quickly reaches 105° or 106°.

In cases such as capsular haemorrhage, in which if life is preserved the result is hemiplegia, there may be signs during the coma, if this is not too deep, pointing to the side paralysed. Thus the limbs on that side when lifted from the bed and then allowed to fall will drop like inert masses, whilst those of the non-paralysed side may offer resistance to passive movements or may be the seat of spontaneous movements. On the affected side the cheek is puffed out more during expiration, the naso-labial fold is less marked, and the angle of the mouth is lower than on the sound side. Conjugate

deviation of the eyes with rotation of the head occurs as a temporary symptom in most cases of severe hæmorrhage. When the lesion is in the hemisphere the deviation is usually away from the paralysed side; the position is as if the patient were trying to look at his lesion. Sometimes the deviation is due to spasm and then is towards the paralysed side. The abdominal and cremasteric reflexes are often absent on the paralysed side.

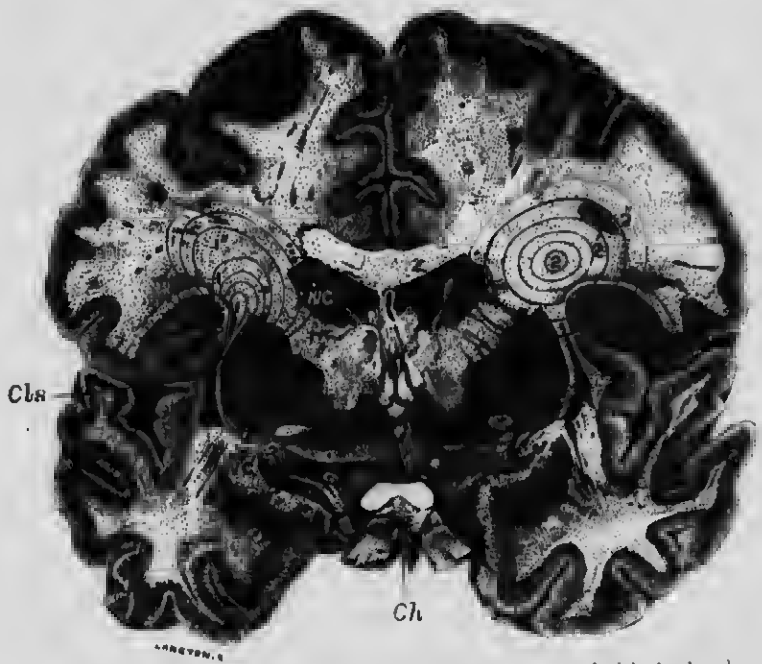


Fig. 169. Vertical section of the brain a little behind the knee of the internal capsule, showing the effects of rupture of the lenticulo-striate artery. (Modified from Charcot.) NC, head, and NC′, tail of the caudate nucleus; Ch, chiasma; NL, lenticular nucleus; IK, internal capsule; Cls, claustrum; 1, the most frequent position in which the lenticulo-striate artery is ruptured; 1′, 1″, 1‴, progressive extension of the hæmorrhage producing compression and rupture of the fibres of the pyramidal tract (hemiplegia); 2, primary focus in the internal capsule; 2′, 2″, 2‴, successive extension of the clot.

In cases of fatal coma death occurs usually in a few hours, sometimes not for some days; very rarely it occurs suddenly or in a few minutes. In some cases after a few hours the patient recovers consciousness for a short time and then passes suddenly into fatal coma

this indicates rupture of the hæmorrhage into the ventricles. When recovery takes place consciousness generally returns within twenty-four hours, but a few days elapse before the patient is free from headache, mental dulness or disturbance of speech.

Localising Symptoms. The signs due to the local effects of the lesion are now apparent: they are wider in extent at first than at a later period. Thus in cases where the persistent condition is hemiplegia only, other symptoms, as hemianæsthesia and hemianopsia, may be present during the first few days, showing that an area much greater than that destroyed by the extravasation has had its functions interfered with. The initial exten-

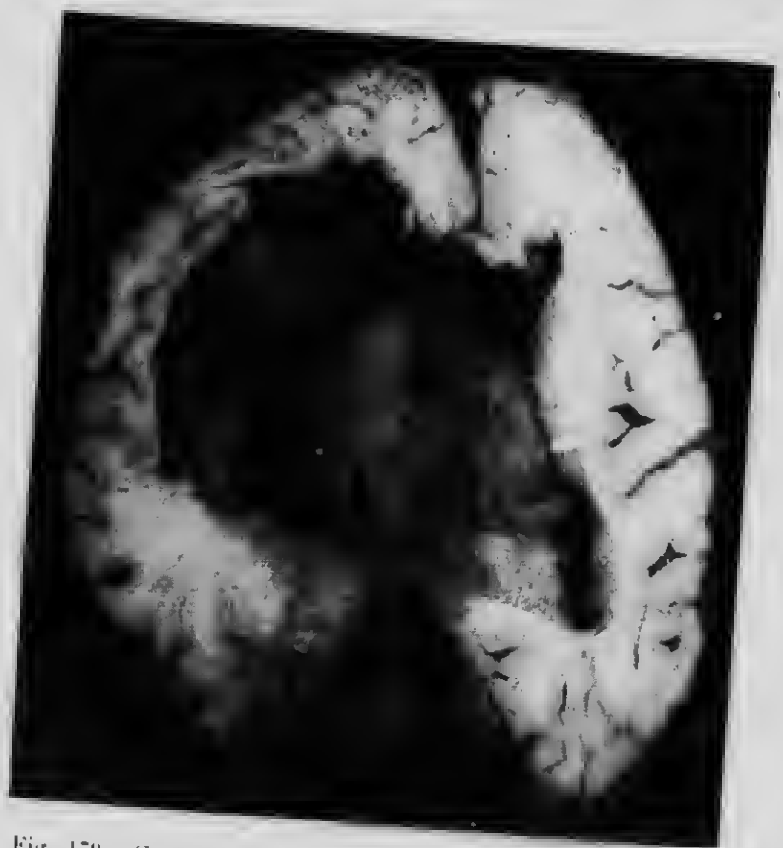


Fig. 170. Cavity formed by large hæmorrhage into left cerebral hemisphere; laceration of outer portions of caudate and lenticular nuclei. (Pathological Museum, Manchester University.)

sive disturbance of function is due partly to pressure, with its resulting anæmia, and partly to inhibition owing to irritation by the lesion.

Capsular hæmorrhage. Hæmorrhage most frequently occurs in the neighbourhood of the internal capsule: hence hemiplegia is the common enduring symptom of cerebral hæmorrhage. Its features and varieties have already been described (see p. 91).

Corona radiata. The nearer the hæmorrhage is to the cortex the more limited in distribution is the resulting paralysis; that is, a monoplegia or an aphasia

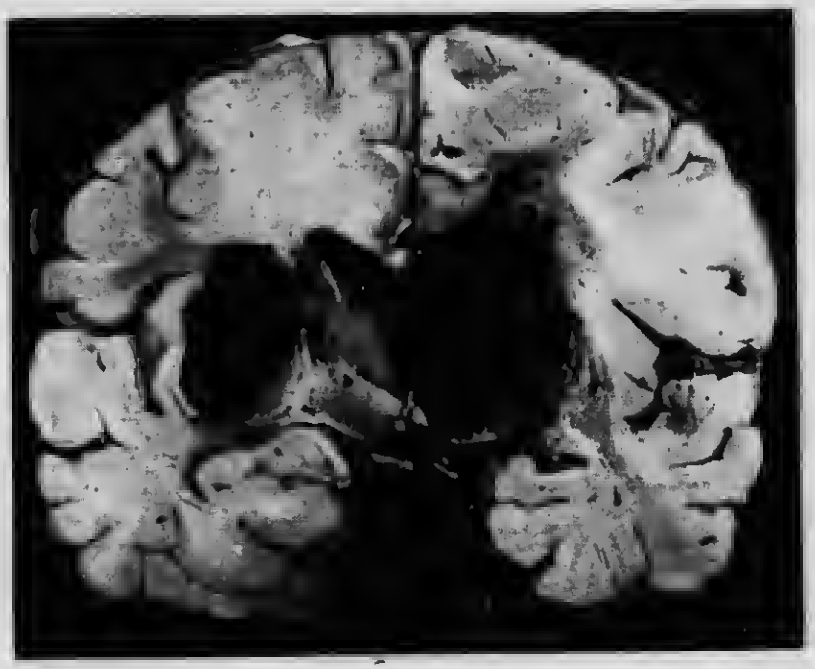


Fig. 171. Bilateral cerebral hæmorrhage; the smaller hæmorrhage on left side occurred about five weeks before the larger one on right side; atheroma of cerebral arteries, granular kidneys, in a woman aged fifty-seven. (Pathological Museum, Manchester University.)

is commoner than a hemiplegia. Hæmorrhage into the white matter of the occipital lobe may cause homonymous hemianopsia without any limb paralysis.

Cortex. Hæmorrhage into this part is very rare and

usually small in size. Convulsions may usher in or attend the apoplectic attack whilst a monoplegia or some variety of aphasia may be permanent.

Crus cerebri. When hæmorrhage is limited to this part a crossed paralysis results—the third nerve being involved on the side of the lesion and the limbs on the opposite side. More frequently, hæmorrhage is of wider extent; it may extend from the thalamic region down to the crus, when anæsthesia is common on the paralysed side.

Pons. Owing to the closeness of its texture hæmor-

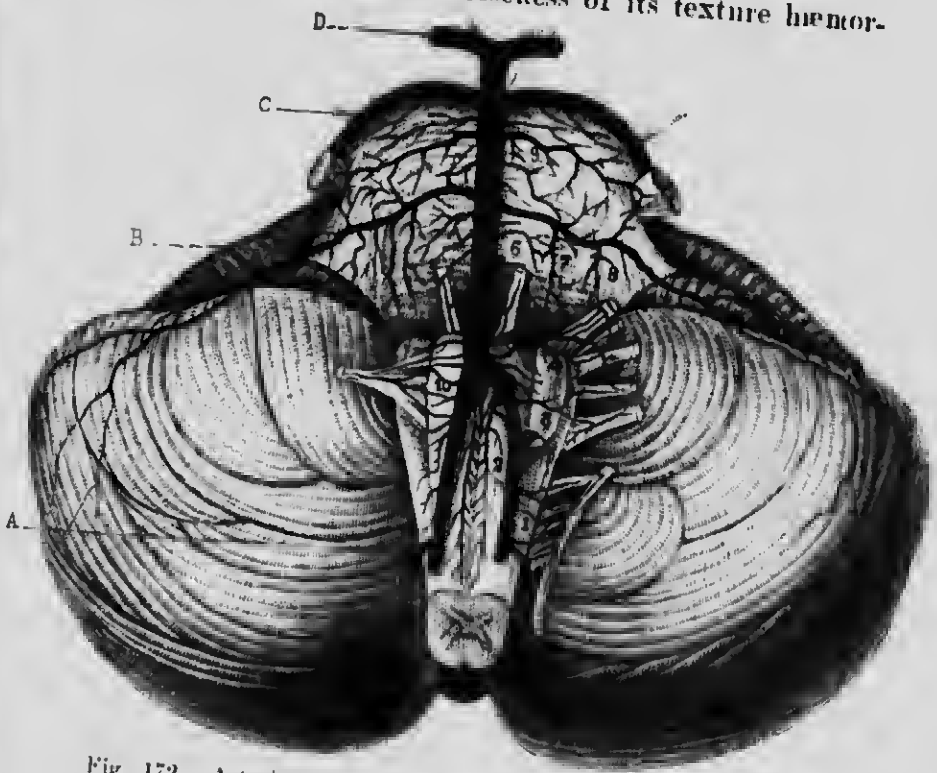


Fig. 172. Arteries of the medulla oblongata, pons, and inferior surface of the cerebellum. (After Duret.) 1, Root arteries of the spinal accessory nerve; 2, anterior spinal arteries; 3, arteries of the pneumogastric and glosso-pharyngeal nerves; 4, inferior arteries of the auditory and facial nerves (vertebral branches); 5, root arteries of the sixth nerve; 6 and 7, arteries of the sub-olivary fossa; 8, superior arteries of the auditory and facial nerves (branches of the middle cerebellar artery); 9, arteries of the trigeminal nerve; 10, arteries of the hypoglossal nerve (branches of the vertebral and anterior spinal arteries); A, inferior cerebellar artery; B, middle cerebellar artery; C, superior cerebellar artery; D, posterior cerebral artery.

rhage into the pons is usually small and circumscribed, and frequently is situated near the middle line; sometimes it is large, the pons being completely hollowed out by clot.

If the extravasation is large, coma comes on suddenly and soon becomes profound; death may occur within a few hours or even a few minutes. Convulsions, usually bilateral, are common initial symptoms;

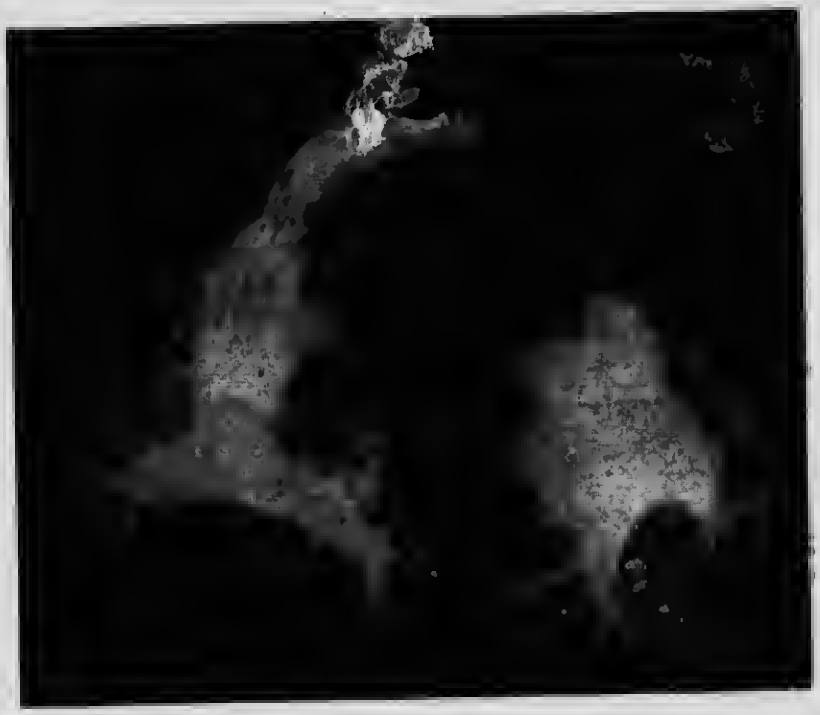


Fig. 173. Haemorrhage into pons, in the case of a man aged twenty-six; granular kidneys; numerous subcutaneous haemorrhages, also submucous haemorrhages in large intestine. (Pathological Museum, Manchester University.)

sometimes they affect the legs only. The temperature tends to rise rapidly to 106° or higher. Vomiting is a frequent symptom; interference with respiration occurs shortly after the onset. If the extravasation is small, there may be no loss of consciousness; the symptoms—paralysis of the limbs and cranial nerves—will

vary with the position of the lesion. In pontine hæmorrhage the pupils are usually much contracted, occasionally they are dilated, their varying size depending on the condition of the nuclei of the third nerves whether irritated or paralysed.

Medulla. Hæmorrhage into the medulla generally causes death very quickly. Should the patient survive the onset a rare event the persisting symptoms are similar to those of bulbar paralysis.

Fig. 174. Hæmorrhage into right lobe of cerebellum; male aged forty-four; granular kidneys, cardiac hypertrophy, signs of former hæmorrhage in right cerebral hemisphere. (Pathological Museum, Manchester University.)

Cerebellum. Hæmorrhage into the cerebellum rarely occurs and is difficult to diagnose. Most frequently it is due to rupture of a branch of the superior cerebellar artery supplying the dentate nucleus; sometimes it occurs in the posterior part of the hemisphere from rupture of a branch of the posterior cerebellar artery. Loss of consciousness varying in degree may be preceded by sudden pain in the back of the head or neck. Vomiting is more frequent and persistent than in hæmorrhage elsewhere. Skew deviation of the eyeballs may be present, and the patient may show a tendency to rotate to the side of the lesion. A large extravasation usually

bursts into the fourth ventricle or into the arachnoid space, when death speedily results. If the patient survives the attack the localising signs of a cerebellar lesion may be expected, namely, unsteadiness of movement, with or without paresis of the limbs on the side of the lesion.

Ventricles. As a rule blood escapes first into one lateral ventricle, and then passes into the other and lastly into the third and fourth ventricles. Such hæmorrhage is rarely primary, being usually secondary to hæmorrhage into the brain substance, especially in the region of the internal capsule. In primary hæmorrhage coma develops suddenly; in secondary there is either deepening of coma already present, or a return of coma after a partial or complete recovery of consciousness. Both forms are usually quickly fatal, though cases of recovery have been recorded. In primary cases the hæmorrhage depends on the same conditions that predispose to intra-cerebral hæmorrhage, and is often due to rupture of miliary aneurysms in the choroid plexus or in the velum interpositum.

Meningeal hæmorrhage. An injury to the head, which causes fracture of the skull, or laceration of the membranes without fracture is the most common cause of meningeal hæmorrhage. The blood may be effused between the dura mater and the bone—subcranial hæmorrhage; or between the dura mater and the cerebral cortex—subdural hæmorrhage.

Subdural hæmorrhage also occurs from compression of the skull during difficult labour (see p. 109): from rupture of aneurysms of the larger arteries, especially at the base; from bursting through the cortex of a hæmorrhage in the brain substance; in the course of some of the acute infectious diseases, in purpura, leucocythæmia and other blood diseases; under the same conditions that lead to cerebral hæmorrhage; and lastly in the form of hæmatoma as a part of a hæmorrhagic pachymeningitis.

The clinical course of meningeal hæmorrhage presents

many variations chiefly owing to the different causes of this condition. In severe uncomplicated cases, as from the rupture of an aneurysm, deep and rapidly fatal coma quickly ensues: sometimes owing to the escape of blood being gradual the comatous condi-

Fig. 175. Diffuse sub-arachnoid hæmorrhage, in the case of a woman aged seventy; granular kidneys, and atheroma; no history of injury. (Pathological Museum, Manchester University.)

tion is preceded by severe headache, giddiness and vomiting. Sometimes the onset is marked by epileptic convulsions.

In traumatic cases a distinction may be drawn between cases of subcranial and of subdural hæmorrhage.

In a typical case of the subcranial variety, three clinical stages may be distingnished; an initial stage in which there is partial or complete loss of consciousness as an immediate result of the blow to the head. Then consciousness is regained and the patient passes into the second stage, known as the "lucid interval," which lasts from a few hours to a couple of days. During this period the patient may feel and appear to be pretty well, and sometimes is able to resume his work. Sooner or later, however, according to the amonnt of blood that is separating the dura mater from the bone, consciousness is again lost, and stupor deepens into fatal coma. During this third stage spasms, accompanied or followed by paralysis, may be observed on the side of the body opposite to that of the blood-clot.

The subdural is distinguished from the subcranial variety of hæmorrhage by the absence of a lucid interval and by the more rapid onset and development of compression symptoms, which too are more general in distribution. To these rules, however, there are many exceptions, thus in cases of subdural hæmorrhage there may be not only a lucid interval but one of considerable duration. It is also to be noted that an injury to the head, severe enough to produce meningeal bleeding, may cause laceration, with extravasation of blood into cortical tissue; hence many symptoms such as headache, vertigo and mental disability, are due to a bruised brain rather than to a clot on its surface, the latter lesion being responsible for pronounced stupor or coma.

The situation of the lesion may be indicated by the presence of unilateral symptoms—convulsions or paralysis; and when the brain is much compressed, by dilatation of the pupil on the side of the hæmorrhage.

OCCLUSION OF CEREBRAL ARTERIES FROM THROMBOSIS AND EMBOLISM.

The two chief causes of **thrombosis** of the cerebral arteries are: Disease of the vessel and changes in the blood. The condition may also be determined by narrowing of an artery from compression, or from invasion by new growth. Slowness in the movement of blood owing to feeble cardiac action, whether produced by illness, or by prolonged anxiety or other cause, is another factor to be considered.

The common causes of arterial disease are atheroma

Fig. 176. Atheroma of vertebral and basilar arteries, also elongation and tortuosity of basilar artery; granular kidneys and cardiac hypertrophy, in a male aged sixty-one. (Pathological Museum, Manchester University.)

and syphilis. *Atheroma* beginning as a degenerative thickening of the inner coat chiefly involves the larger arteries at the base and often extends along their branches. It lessens the elasticity of the vessel, diminishes or sometimes enlarges its calibre and roughens its lining membrane. One or all of these results of atheroma tend to coagulation of the blood; the coagulation often starts at the orifice of a branch artery and may extend to or be limited to the smaller vessels. Thus thrombosis may spread from the internal carotid to the middle and anterior cerebrals and even to the ophthalmic artery. It must be borne in mind that marked atheromatous changes may be observed in the brachial and other limb arteries when the cerebral are free from change, and that the cerebral may be much diseased when the arteries elsewhere are but little affected. The frequency of cerebral softening from atheroma increases as life advances: the condition is not common under fifty, though gout, chronic Bright's disease and alcoholism tend to early degeneration.

Syphilitic disease of the cerebral arteries is met with as a result of the inherited as well as of the acquired disease: in inherited syphilis the endarteritis develops in early childhood, in acquired syphilis generally between the ages of twenty and forty-five, and usually from between two and twelve years after infection. One or more of the larger arteries are affected: their walls show irregular nodular thickenings, which cause projections and narrow their lumens, the narrowing being often greater than in atheroma. Compression from a tumour, or arteritis from adjacent inflammation are other causes of occlusion and thrombosis.

In addition to disease of the vessel an *altered blood state* may lead to coagulation. Thus thrombosis is produced by the changes in the blood which are associated with chlorosis, cancer, tuberculosis, childbirth, gout or with diabetes; occasionally it occurs after typhoid or diphtheria. Indeed any very lowering condition predisposes to thrombosis: in old persons the

influence both of arterial disease and of poor blood is often observed.

The most common cause of **embolism** is either a diseased cardiac valve or a clot in the left auricle: from the valve a piece of fibrin, from the auricle a fragment of clot is detached and carried by the blood to a cerebral vessel. The accident is less common during a first attack of endocarditis than when fresh endocarditis attacks valves already diseased. The embolus is derived from the mitral much more frequently than from the aortic valves. Mitral stenosis is the common condition, and here in addition to valvular disease there is often imperfect compensation, so that the left auricle becomes much enlarged when a clot readily forms in the auricular appendix. The left auricle is also the usual source of embolism in pregnancy and the puerperium in which conditions the state of the blood renders it apt to coagulate. In rare cases the plug is derived from an atheromatous aorta or from the clot in an aortic aneurysm or from some disease in the lungs which involves the pulmonary veins. When, as in ulcerative endocarditis, the detached particles carry infective organisms the vessel is not only blocked but its walls become inflamed and softened and yielding to pressure may form an aneurysm which subsequently bursts. Micro-organisms may also obstruct the capillaries of the brain leading to minute areas of suppuration which by coalescence form an abscess. In inflammatory processes in the pelvis, septic materials may be carried to the lungs and possibly may pass through its capillaries, but in most cases the embolic particles are derived either from a pulmonary abscess or from a septic endocarditis, which is associated with the pelvic inflammation.

The middle cerebral or one of its branches is the most frequent seat of embolism. The posterior cerebral and the vertebral are also sometimes involved; other arteries are more rarely obstructed. Cerebral embolism may occur at any age but is most commonly met with in

young persons and generally in those who have suffered from rheumatic fever, chorea or from scarlet fever.

Pathological Effects of Obstruction. The first effect is anæmia of the part supplied by the vessel. If the obstruction is not complete this local anæmia may be temporary only, the blood returning to the part either by means of collateral circulation or by the absorption of the plug. If, however, the obstruction is complete,

Fig. 177. Diagram of an embolic infarction. (Weber.) a, artery obliterated by an embolus (e); v, vein filled with a secondary thrombus (th); 1, centre of the infarction, which is becoming disintegrated; 2, area of extravasation of blood into the tissues; 3, area of collateral hyperæmia.

anæmia is succeeded by softening, the colour of which depends on the amount of blood in the softened area: thus the softening may be of the red, yellow or white variety. The affected area is surrounded by an inflammatory zone in which the nerve elements are less severely and less permanently damaged; for this reason the symptoms are wider in extent at first than at a later period.

The cortex, the corpus striatum and the optic thalamus with the closely related internal capsule are the most frequent seats of softening. The change occurs also in the pons and occasionally in the medulla and the cerebellum. The temporo-sphenoidal lobe is much more frequently the seat of thrombotic

softening than of hæmorrhage. Obstruction in the small arteries to the corpus striatum and optic thalamus is invariably followed by softening; but obstruction in the cortical arteries is not always followed by softening, and when it does occur it is usually partial either in degree or in distribution, the variations depending on the freedom of anastomosis between the superficial vessels. The effects of thrombosis are generally more permanent than the effects of embolism, for in thrombosis other vessels in the district of the one occluded are often diseased, hence the restoration of nutrition by collateral circulation is less likely to occur.

Symptoms. The cerebral symptoms caused by arterial obstruction closely resemble those of hæmorrhage and without an appeal to other circumstances it is generally impossible to determine the nature of the lesion. More especially is this the case when the seizure is attended by loss of consciousness—a less frequent event, however, than in hæmorrhage. On the other hand if the onset is marked by complete hemiplegia without loss of consciousness softening is more likely than hæmorrhage. The more sudden the obstruction and the larger the vessel occluded the more severely is consciousness affected, there being no unconsciousness either when a small vessel is obstructed or when the onset is gradual. Consciousness is often preserved in thrombosis from syphilitic disease.

The apoplectic condition itself is indistinguishable from that due to hæmorrhage, and when a large vessel is suddenly obstructed coma may be as deep and as prolonged as in severe hæmorrhage; as a rule, however, it is less intense and is shorter in duration. Initial convulsions, comparatively rare in thrombosis, are not uncommon in embolism. Sometimes the onset is indicated by sudden headache, vertigo and mental disturbance; in cases of thrombosis owing to the pre-existence of vascular disease the symptoms mentioned or others, such as impairment of memory and transient paralysis affecting either speech, face or limb, may exist

for weeks or months before the onset. Such premonitory symptoms, especially severe and paroxysmal headache, are frequent in syphilitic disease. They rarely occur in cases of embolism, in which the onset is usually quite sudden. In cases of embolism there may be signs of the same process elsewhere, as in the kidney or in the central artery of the retina. In ulcerative endocarditis the circulation of infective organisms often gives rise to rigors and pyrexia and to signs of embolism in the spleen, kidneys or other organs. In syphilitic cases the symptoms of arterial thrombosis may be complicated by others produced by a meningitis or a gumma.

With regard to the focal symptoms, those due to the situation of the lesion, hemiplegia, as in cases of hæmorrhage, is the most common form of paralysis, the *middle cerebral* being the artery most often involved. The frequency of other symptoms is different in the two cases. Thus the *cortex* being affected by softening oftener than by hæmorrhage aphasia, monoplegias, and recurring convulsions are produced more frequently by thrombosis than by hæmorrhage: athetosis and other disorders of movement are also more commonly met with.

Embolism of either the *anterior* or the *posterior cerebral artery* is rare. The anterior cerebral comes off at right angles to the internal carotid and therefore it is difficult for an embolus to enter it: softening from thrombosis of this vessel is less rare, the symptoms are those of progressive dementia. Softening from obstruction, of the posterior cerebral, is usually prevented by the freedom of its anastomoses: its occurrence is indicated by the presence of heminæsthesia, hemianopsia and sometimes of tremors of the limbs on the opposite side.

Obstruction of the whole *basilar artery* produces bilateral paralysis, all four limbs and often both sides of the face being affected. As a rule articulation, swallowing and respiration are impaired: respiration may assume the Cheyne-Stokes rhythm. Death often

quickly ensues. Obstruction of the upper part of the vessel causes much less threatening symptoms, for the circulation to the vagi nuclei is not interfered with; sometimes the nuclei of the oculomotor nerves are involved when partial or complete ophthalmoplegia results. Occlusion of the basilar artery is attended, occasionally by convulsions but more commonly by an apoplectic seizure.

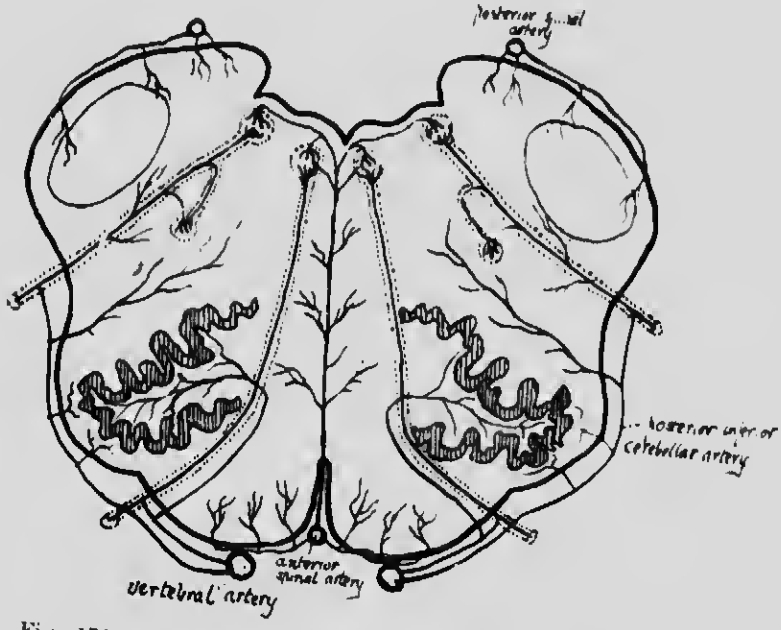


Fig. 178. Schematic representation of the arterial supply of the medulla. (Kinnier Wilson, after Van Gehuchten.)

Occlusion of one *vertebral artery* produces symptoms which tend to assume the hemiplegic form. The hemiplegia may be either on the same side as the lesion or on the opposite side, according to the situation of the obstruction. It is usually associated with paralysis of the lips, tongue, palate and larynx on the same side; but as the bulbar nuclei are partly supplied by the anterior spinal artery the paralysis is generally incomplete; for a similar reason the hemiplegia is usually transient.

Thrombosis of the *posterior inferior cerebellar* artery produces a characteristic group of symptoms. On the side of the lesion we find:– paralysis of the palate and the vocal cord, anæsthesia of the pharynx and the face, incoordination of the limbs, and sometimes loss of taste

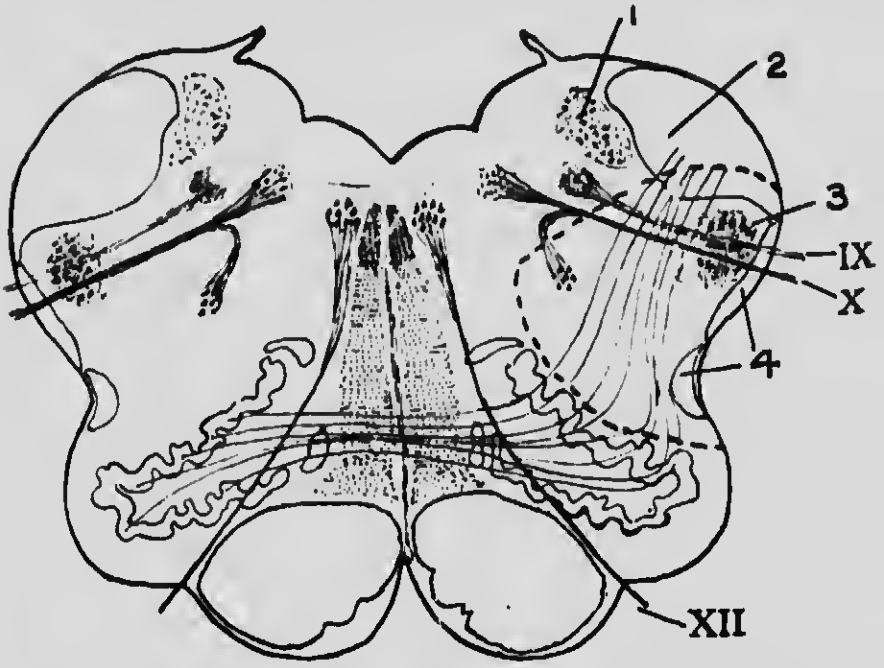


Fig. 179. Composite diagram to indicate the structure of the medulla commonly involved in the lesion—indicated by the dotted line—as found by pathological examination. (Kinnier Wilson.)
1, Descending root of 8th nerve; 2, restiform body; 3, descending root of 5th nerve; 4, spino-cerebellar tracts.

and nystagmus on deviation of the eyes to the side of the lesion. On the opposite side there is anæsthesia of the trunk and limbs and sometimes of the face. The anæsthesia is of the dissociated type, pain and temperature sensations being impaired while tactile sensibility and the muscular sense are preserved. Vasomotor disturbances corresponding in distribution to the anæsthesia may also be present. At the onset of the affection the symptoms are often more extensive; there may be a transient disturbance of the eighth nerve or paralysis of the sixth and seventh nerves on the side of the lesion.

Vomiting and difficulties in swallowing and articulation may also occur. The residual symptoms are explained by the situation of the area in the medulla which is partially or wholly destroyed. According to Kinnier Wilson the structures which are most commonly involved are the reticular formation and its nuclei, the descending root of the fifth nerve, the nucleus ambiguus, the vago-glossopharyngeal nucleus, the glossopharyngeal and vagus nerves, the spino-cerebellar tracts and other fibres passing into the inferior cerebellar peduncle (see fig. 179).

PARTIAL OCCLUSION.

Failure in the nutrition of nerve cells and nerve fibres may occur in consequence of arterial degeneration apart from the presence of either hæmorrhage or softening. The arterio-sclerosis may be either widespread or limited to certain areas. When a large number of vessels are degenerated the mental condition of the patient becomes gradually and progressively impaired; there is general muscular weakness, usually slight in degree; the limbs are spastic and the deep reflexes are exaggerated. In some of these cases the facial expression becomes fixed, like that of paralysis agitans, in others the condition of the patient resembles that of general paralysis of the insane, whilst in a third group of cases there is a resemblance to disseminated sclerosis owing to the association of tremor with the spastic weakness of the limbs.

When the degeneration affects only a few arteries the symptoms will vary according to the area of brain involved. Convulsive seizures may occur when the cortical arteries are diseased, difficulties in articulation and swallowing when the bulbar arteries are sclerosed. It is of the greatest interest to observe that symptoms of gross lesions in the brain may be closely imitated by the manifestations of local arterio-sclerosis. Such manifestations may be either temporary or permanent. Cases of temporary aphasia and paralysis are not

uncommon. The condition of the limbs in a case of transient hemiplegia may be identical with that dependent on a hæmorrhage into the internal capsule, when it will be impossible to foretell the duration of the paralysis. Should recovery take place in a few days the most probable explanation is that the paralysis depended on spasm of the sclerotic vessels. Sometimes, as a result of degeneration of the nerve elements supplied by the sclerotic vessels, the paralysis is permanent. The author has seen cases of homonymous hemianopsia and of incoordination of movement which were undoubtedly instances of occipital and cerebellar disease respectively, and which could only be accounted for by the presence of degenerative changes produced by arterio-sclerosis.

Diagnosis. The chief points of distinction between hæmorrhage, thrombosis and embolism are summarised in the accompanying table.

	Hæmorrhage	Thrombosis	Embolism
Age	40-65	In old persons from atheroma; in young persons from syphilis	Any age but chiefly young adults.
Immediate antecedents	Severe physical exertion or mental excitement	Physical or mental exhaustion	—
Prodromata	Rare	Common	Rare
Onset	Sudden	Gradual	Sudden
Loss of consciousness	Develops at onset or soon after; deep and prolonged, frequently ingravescent; slight to sleep in a few hours	Consciousness preserved or loss follows hemiplegia; presence and degree of coma vary with size of artery and with suddenness of closure; coma rare in syphilis because branches affected rather than trunk of artery.	Usually slight and transient
Convulsions	Occasionally at onset and general	May occur a few days after onset; tend to be local	Frequent
Persistent aphasia with little or no hemiplegia	Rare	Not uncommon	Not uncommon
Hemianopsia alone, or with hemianaesthesia	Rare	Not uncommon	Not uncommon
Temperature	Initial fall followed by rise	Subnormal or normal, may rise to 100° about third day	No initial fall
Heart and pulse	Hypertrophied heart and high tension pulse	Weak dilated heart and soft feeble pulse	Valvular disease common
Albuminuria	Common	May be present	Possibly from renal embolism
Eyes	Albuminuric retinitis	May be thrombosis of central artery and oedema	May be embolism of central artery, or hæmorrhages

Some other conditions which give rise to coma may now be briefly considered.

Alcoholic poisoning. The coma produced by alcohol cannot always be distinguished from cerebral apoplexy; it is therefore of the greatest importance to keep the patient under observation for a few hours when the question can usually be decided. It must be remembered that cerebral hæmorrhage from fracture of the skull often occurs during a drinking bout, and that spirits are generally given for fits and impaired consciousness, hence the smell of alcohol in the breath is of little use in diagnosis. A careful search should always be made for any unilateral symptoms, and for signs of fracture of the skull. The stomach contents and the urine should be examined for the presence of alcohol.

Opium poisoning is less difficult to diagnose. The coma develops gradually, the respirations are infrequent, the skin is moist, the breath may smell of opium, if the crude drug has been taken, and the pupils are strongly contracted. Extreme contraction of the pupils occurs in pontine hæmorrhage, but in this condition coma comes on suddenly; whilst the presence of paralysis or of other objective symptoms and a rapid rise of temperature are sufficient to exclude narcotic poisoning.

In *diabetic coma* the urine contains a large amount of sugar, a little albumin and frequently granular casts: it gives a brownish-red colour with perchloride of iron, and the breath has an acetone odour. Quite distinctive too is the methylene blue reaction with the blood, which distinguishes diabetic from other forms of coma.

Uræmia. The comatose condition presents no special characteristics; as a rule it comes on more gradually and is less profound than in cerebral hæmorrhage. Frequently it is preceded by convulsions or by sudden total blindness. The urine contains albumin and casts and is deficient in urea; albuminuric retinitis and general anasarca may also be present. Bright's disease however, is a frequent cause of cerebral hæmorrhage.

Epilepsy. The short duration of the coma, the absence of unilateral symptoms and a history of former convulsions would prevent the unconsciousness following an epileptic fit being mistaken for that of a vascular lesion ushered in by a convulsive attack. Unconsciousness may occur during an attack of *hysteria*. The history of hysterical manifestations, and the absence of any cause or of symptoms of a cerebral lesion are the chief diagnostic indications. Of other causes of coma, such as intracranial tumour or abscess, thrombosis of sinuses, general paralysis and sunstroke, a careful investigation of the history and clinical course of the case will usually enable a correct diagnosis to be made.

Prognosis. Cerebral hæmorrhage, however slight, is always a serious matter, for the arterial disease which underlies it suggests the probability of a recurrence. Life, however, may be prolonged by carefully regulating the habits of the patient, attention being especially given to the condition of the blood pressure. The following are unfavourable signs:—Coma that is deep and lasts longer than twenty-four hours; a low initial temperature or a considerable rise within a few hours; much interference with respiration, especially if it assumes the Cheyne-Stokes type and is associated with an accumulation of mucus in the lungs; the occurrence (during the inflammatory period) of delirium, high fever, and the formation of an acute bed-sore on the buttock; bilateral paralysis of the limbs indicating hæmorrhage into the ventricles, pons or medulla. In meningeal hæmorrhage when consciousness is regained the outlook as regards life is good, but the prospects of complete restoration of mental and bodily vigour are less favourable.

The prospect of recovery from paralysis depends on whether there has been actual destruction of the motor tract or merely disturbance of its functions by a lesion in its neighbourhood. In the latter case some movement may return to a paralysed limb within a few days or weeks, when the prospect of ultimate recovery

is good. If, however, at the end of a few months the paralysis is still complete, but little return of movement can be expected.

In thrombosis the prognosis varies much in different cases, being related to the severity and the duration of the initial symptoms, to the occurrence of previous attacks, to the artery occluded and to the nature of the morbid process. Deep coma is of grave significance, but prolonged coma, if slight or moderate is not so serious as in hæmorrhage. Previous attacks increase the immediate danger to life, especially when there is evidence of extensive disease of the vessels. Obstruction of the basilar, the internal carotid or of the trunks of both middle cerebrals is rarely recovered from; obstruction of the vertebral artery is more serious than that of the middle cerebral. Thrombosis from syphilitic endarteritis, and embolism are rarely fatal unless the basilar artery is involved, but vascular occlusion and softening from arterio-sclerosis is much more serious, for such degeneration tends to be progressive, and the possibility of recurrent attacks is great especially if the heart is weak and dilated. On the other hand a second attack is rare in embolism and also in syphilitic disease if this is early and properly treated.

It must be borne in mind that, although arterial thickening from syphilis may be lessened or removed by treatment, the necrotic softening which results from the vascular occlusion is as little amenable to treatment when due to syphilis as when dependent on embolism or on atheroma. The prognosis of the developed paralysis depends rather on the seat than on the nature of the disease, and also to some extent on the age of the patient, for the younger the patient the more likely is recovery or compensation to occur.

Treatment. In a case of cerebral hæmorrhage the patient should be placed in bed with his shoulders and head slightly raised; bending and constriction of the neck should be avoided. The patient should be turned on to his side in order to allow the

free escape of saliva and to avoid obstruction of the larynx by the falling back of the tongue and epiglottis. The mouth should be frequently wiped out with some mild antiseptic solution. If there is much cyanosis, if the pulse is full and incompressible, if the heart is hypertrophied and the breathing is laboured and stertorous, venesection is indicated; the withdrawal of ten oz. of blood from the arm often gives marked relief, the pulse becomes softer and consciousness is often quickly regained. Should bleeding be deemed inadvisable, owing to uncertainty of diagnosis or for other reasons, free purgation should be induced by placing two drops of croton oil in a little butter on the back of the tongue. For the relief of profound cerebral compression trephining the skull or the withdrawal of fluid by a lumbar puncture is sometimes useful. Hæmorrhage from the middle cerebral and its branches may be controlled by compression or by ligature of the carotid artery on the same side. In cases of meningeal hæmorrhage, trephining, removal of the blood and tying the ruptured artery are the only methods likely to be of service. The tendency to further hæmorrhage is lessened by the application of an ice bag to the head and of mustard to the abdomen and feet. Adrenalin given hypodermically or per rectum has also been recommended. Convulsions are controlled by chloroform or by the injection of twenty grains of chloral into the rectum.

At first nourishment is not required, but if the coma is prolonged, nasal or rectal feeding is advisable. When consciousness returns, a little cold liquid nourishment may be given from time to time during the first two or three days. Alcohol, coffee and tea must not be given unless there are signs of cardiac failure, when alcohol in small doses may be needed; a hypodermic injection of strychnine or of camphor also is sometimes beneficial. If the pulse tension is high the nitrites may be tried, and small doses of aconite often tend to subdue an over active heart. During the comatose state the bladder must be emptied by a catheter. The skin must be kept scrupu-

lously clean and still farther to obviate the risk of bed-sores the position of the patient should be frequently changed, pads of cotton wool being placed over points of pressure. During convalescence and afterwards, the necessity for a quiet life free from mental excitement and physical exertion should be enforced. The food should be light and nutritious, and alcohol should not be taken. A strict diet and carefully regulated habits will do much to prevent the recurrence of an attack and to prolong life.

If there is reason to believe that the initial apoplexy is the result of thrombosis or of embolism, the measures recommended for hæmorrhage, namely, venesection, brisk purgation and the administration of adrenalin are contra-indicated. The objects now are to lessen the tendency to coagulation, and to stimulate the circulation rather than to lower the blood pressure. During the stage of unconsciousness heat should be applied to the body and sinapisms to the neck; the inhalation of ammonia and the injection into the bowel of a small dose of brandy in hot water are sometimes efficacious. As soon as the patient can swallow, a mild aperient rather than a strong purgative is required; afterwards the patient should take beef-tea, hot milk and other light nourishment. A little alcohol may be given but this must be stopped after a few days owing to the danger of increasing the tendency to inflammatory reaction. During the inflammatory stage, ice may be applied to the head and moderate doses of antipyrin or phenacetin administered. The bromides are useful for restlessness; veronal or trional for insomnia, and digitalis or strophanthus for a weak heart.

In syphilitic cases mercurial inunction and the administration of iodide of potassium should be commenced as early as possible, for although such treatment cannot undo the mischief it may prevent its increase and the development of fresh lesions.

In cases where it is impossible to determine whether thrombosis or hæmorrhage is present it is well to avoid

active medication and to restrict the treatment to absolute rest, to instructions as to posture in bed, the application of ice to the head and the administration of simple aperients and light nourishment.

THROMBOSIS OF THE CEREBRAL SINUSES.

Two varieties of sinus thrombosis may be distinguished: a simple form the result of some general condition, and an infective form due to local infection

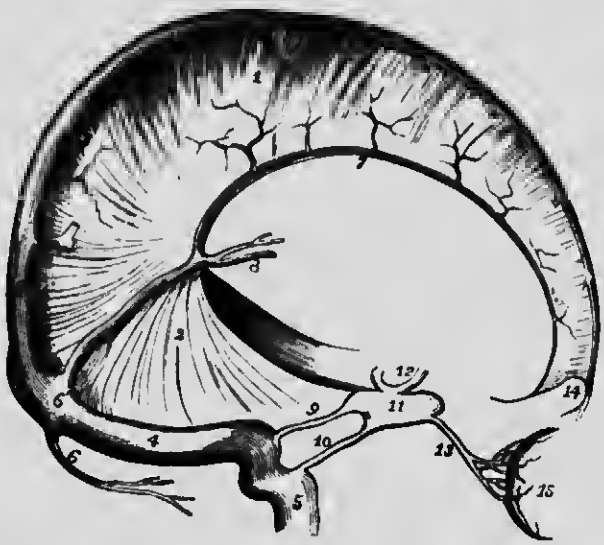


Fig. 180 (from Turner). Dura mater and cranial sinuses. 1, Falx cerebri; 2, tentorium; 3, 3, superior longitudinal sinus; 4, lateral sinus; 5, internal jugular vein; 6, occipital sinus; 7, 8, veins of Galen; 9 and 10, superior and inferior petrosal sinus; 11, cavernous sinus; 12, circular sinus, which connects the two circular sinuses together; 13, ophthalmic vein, from the eyeball; 14, crista galli of ethmoid bone.

Thrombosis in a sinus is favoured by the rigid walls and the irregular shape of the cavity as well as by the trabeculæ which cross it. Thrombosis is prone to occur either when the circulation is retarded as in consequence of a feebly acting heart, or when the blood is altered in quality as in chlorosis, or is reduced in volume as after protracted diarrhœa. An infective thrombosis may be the result of direct extension through

a vein or other tissue between the focus of disease and the sinus. Frequently the thrombus spreads into tributary veins or into an adjacent sinus: owing to the obstruction of the circulation there is intense congestion of the cerebral veins and capillaries, and minute hæmorrhages may occur in the brain as well as considerable œdema. In the infective variety of throm-

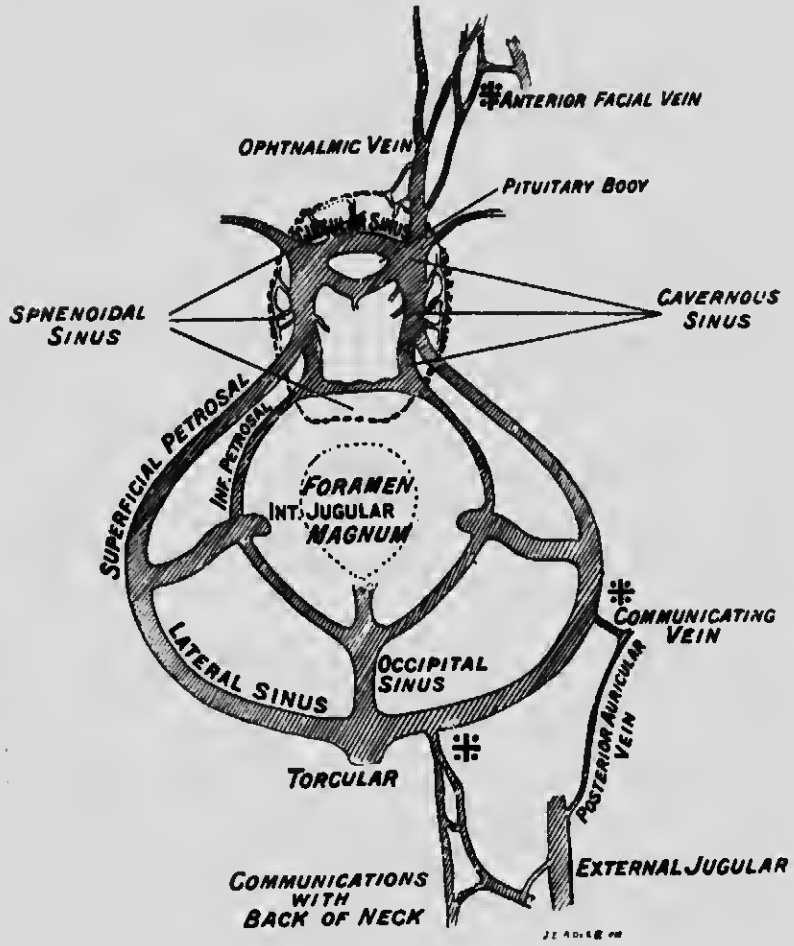


Fig. 181. Diagram to show communication of veins of cavernous sinus with venous sinuses of the cranium; showing also inter-relation of transverse and cavernous sinus with external veins. The asterisks (*) mark the anastomoses of the intra-cranial venous system with the veins of the surface. (St. Clair Thompson, after Leube.)

bosis the clot frequently becomes friable and puriform; particles detached from it may be conveyed by the blood as infective emboli to the lungs where they set up pneumonia, abscess or gangrene; sometimes a general pyæmic condition is established. Suppuration, meningitis and intra-cranial abscess are other results of the local infection.

Etiology. The *simple* or *primary* variety of thrombosis may occur at any age, but it is most commonly met with in young children, especially during the first six months of life, and in old people. In children its chief cause is marasmus in association with severe and exhausting diarrhœa, hence the condition is often called "marantic thrombosis"; its most common site is the superior longitudinal sinus.

In adults primary thrombosis occurs in the terminal stages of phthisis, during the course of cancer, enteric fever, or other exhausting malady, and sometimes in association with parturition, especially if there has been much hæmorrhage. Another cause is chlorosis, when the condition may be associated with venous thrombosis in other parts of the body.

The *infective* variety occurs much more frequently in adults than in infants and old people. Usually it results from injury or disease of parts adjacent to the sinus. It may be caused by caries or fracture of the skull, or by dental caries with periostitis; sometimes the infective focus is found in the nose, tonsils or orbit; also in erysipelas of the face or scalp, or in a carbuncle of the face or neck.

The researches of St. Clair Thompson indicate that intra-cranial complications, as a result of septic infection from the sphenoidal sinuses, occur much more frequently than has been generally admitted; he urges the importance of making a careful examination of the nose and its accessory cavities "in cases of headache and neuralgia, in intra-cranial inflammation, and in the presence of thrombosis of the ophthalmic vein or of the cavernous sinus." But by far the commonest

cause of thrombosis of the cerebral sinuses is suppurative disease of the middle ear with a cario-necrotic condition of the surrounding bone, and hence the superior petrosal or the lateral sinus is the one in which thrombosis most frequently occurs. The disease is said to spread oftener from necrosis of the posterior wall of the tympanum by way of the petroso-mastoid canal than from disease of the mastoid cells. Compression of the sinus, as by a tumour, is another cause of secondary thrombosis.

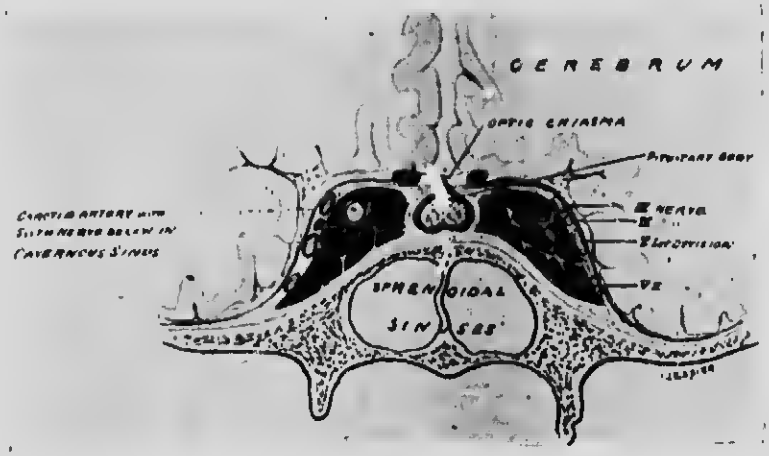


Fig. 182 (semi-diagrammatic). Coronal section, through sphenoidal sinus, to illustrate immediate relationships with cavernous sinus and its contents, optic nerve and base of brain. (St. Clair Thompson.)

Symptoms. These vary much according to the sinus affected and according to the nature of the thrombus, whether simple or infective. Thrombosis of the *longitudinal sinus*, which is usually of the simple variety, may give rise to headache, vomiting, convulsions or delirium, mental apathy, somnolence and coma. Sometimes there is rigidity of the muscles of the neck or back or even of the limbs. Strabismus and tremor of the tongue have been observed. Unilateral convulsions or paralysis indicate extension of the thrombus into the veins on one side, or its commencement in the veins.

The local signs of obstruction of the sinus are epistaxis from over distension of the nasal veins, and engorgement of the veins of the scalp with œdema about the temples and forehead. In infants, the anterior fontanelle often becomes tense and prominent. In chlorotic females phlebitis of some of the veins in the leg may be present.

Fig. 183. Thrombosis of the cavernous sinus, showing the œdema of the eyelids, proptosis, chemosis, ophthalmoplegia, and commencing ulceration of the conjunctiva. (St. Clair Thompson.)

Thrombosis of the *cavernous sinus*, which is usually bilateral, occurs as a result of infective disease in the orbit, nose or throat. The condition is indicated by engorgement of the orbital veins, giving rise in some cases to proptosis of the eyeball, œdema of the eyelids, and a transient distension of the retinal veins with

hæmorrhages, together with a slight œdema of the optic disc. Implication of the ocular nerves which run in the wall of the sinus may lead to various forms of strabismus; the first division of the fifth nerve may also be involved.

The effects of thrombosis in the *lateral* sinus are partly due to its obstruction, and partly to the septic nature of the thrombus. Simple obstruction of the circulation leads to distension of the veins over the mastoid, and to tenderness and œdema of that region. Thrombosis of the jugular vein, which is usually present, gives rise to tenderness down the side of the neck and to pain and stiffness of the movements of the head, especially in turning it to the opposite side, while the thrombosed vein may be felt as a hard cord in the neck. Interference with the cerebral circulation may be expressed by headache and optic neuritis. But local signs are not always present and if present may be overshadowed by the symptoms of pyæmia or of an associated meningitis. Pyæmia would be indicated by a high and intermittent form of pyrexia, by profuse sweating, diarrhœa and recurrent rigors, and by signs of infarctions in the lungs, liver, spleen or kidneys: meningitis by irritability, paralysis, convulsions, rigidity of limbs, delirium and final coma.

Diagnosis. In marasmic infants cerebral symptoms, such as stupor, coma or convulsions may be due to cerebral anæmia ("false-hydrocephaloid") rather than to thrombosis, which can only be diagnosed with certainty when there is external œdema and distension of the veins.

The general symptoms associated with infective thrombosis may resemble those of enteric fever—hence the importance in doubtful cases of making a careful examination of the nose, the ear and the jugular vein. In meningitis, delirium, convulsions and optic neuritis are commoner than in sinus thrombosis; headache is more severe and there may be retraction of the head.

In abscess the temperature is usually normal or

subnormal and the pulse is slow, while signs of a focal lesion or in the cerebellum or in the temporo-sphenoidal lobe are often present.

Prognosis. The constitutional state underlying a simple thrombosis is always serious, and even if patients recover they may be afflicted with a squint, with ptosis or with some lowering of the mental functions. The unfavourable prognosis of infective thrombosis is largely due to the difficulty of its early recognition, when only can surgical treatment be of much avail.

Treatment. In simple thrombosis of the longitudinal sinus the flow of blood to it should be aided by having the head and shoulders well raised; the neck should not be bent and should be free from any constricting clothing. It is also important to strengthen the heart by strychnine, digitalis, and stimulants, and to combat the tendency to exhaustion and syncope by the frequent administration of strong nourishment.

In cases of infective thrombosis of the lateral sinus we have to rely mainly on surgical measures. The sinus must be opened and its contents evacuated, the jugular vein having previously been ligatured.

Quinine, salicylate of soda and full doses of tincture of the perchloride of iron have been recommended for the septicæmic condition; in some cases the injection of anti-streptococcus serum appears to have been efficacious.

INTRACRANIAL ANEURYSMS.

The miliary aneurysms which in cases of cerebral hæmorrhage are sometimes found on the smaller arteries have been already described. We have now to consider the aneurysms of the larger arteries.

Such aneurysms are rare; as a rule they are met with between the ages of ten and sixty, being most common during mid-adult life. They occur more frequently in males than in females.

The [illegible]aining of the arterial wall which necessarily precedes the formation of an aneurysm has been caused

in some cases by injury to the head, in others by primary arterial degeneration. But much commoner causes of aneurysm than either injury or atheroma are embolism as a result of endocarditis and endarteritis from syphilis. The partial occlusion of an artery by an embolus leads to changes in its walls and so to the gradually yielding of the vessel to the pressure of the blood. The middle cerebral artery is a frequent seat of aneurysm from embolism, the basilar from syphilis. The other arteries of the brain are affected according to Gowers in the following order of frequency: internal carotid, anterior cerebral, posterior communicating, anterior communicating, vertebral, posterior cerebral, inferior cerebellar. As a rule there is only one aneurysm; occasionally there is more than one. The size varies from that of a pea to that of a pigeon's egg or even much larger.

Symptoms. In many cases the presence of an intracranial aneurysm is unattended by symptoms. In others the symptoms are either trivial or they are those common to a growth or to other lesions in the brain. There may be headache, sometimes pulsating in character, associated with singing or buzzing noises in the head, with mental dulness and irritability and occasionally with paralysis of the limbs or of some of the cranial nerves. Double optic neuritis is sometimes present. Apoplectic attacks may occur during the course of the disease as a result of repeated hæmorrhages from a small opening in the aneurysmal sac. Variations in the intensity of the symptoms sometimes occur; such intermittency is of diagnostic significance, although it is not uncommon in other forms of cerebral disease. From a careful analysis of 555 cases of aneurysm of the cerebral arteries found after death, Beadles obtained the following results:—In about forty-six per cent. of the cases the first indication of a cerebral lesion was an apoplectic seizure due to rupture of the aneurysmal sac. In thirty-seven per cent. there were signs suggestive of the presence of a tumour or

of other lesion in the brain, and in about half of these cases the symptoms were followed by fatal apoplexy. In three per cent. there were signs of compression of some of the cranial nerves, while in the remaining fourteen per cent. of the cases no symptoms whatever were observed during life. The hearing of a murmur on auscultating the skull is of little or no value for it occurred in only two of the 555 cases collected by Beadles. Moreover a murmur is sometimes heard when no aneurysm is present, as in cases of anæmia and Graves disease, or when a large vessel is compressed by a tumour.

In aneurysms of embolic origin there is a tendency for rupture of the sac to occur at an early stage of the disease, owing to the softened condition of the walls of the vessel. The larger the aneurysm the more likely is it to give rise to symptoms, although there are many cases on record in which a large aneurysm gave no indications of its presence during life. The symptoms depend more upon the situation of the aneurysm than upon its size.

In aneurysm of the cavernous portion of the *internal carotid* artery the chief symptoms are homolateral blindness from compression of the optic nerve, and paralysis of the ocular muscles, those supplied by the third nerve being first affected. Engorgement of the retinal veins may be observed, but it is usually transient owing to the free communication of the ophthalmic and the facial veins. In some cases the sensibility of the eyeball is impaired in consequence of damage to the ophthalmic branch of the fifth nerve. Sometimes the sense of smell is diminished or lost. When the aneurysm is large it may cause paralysis of the limbs from pressure on the motor tract, or if it is on the left side, aphasia from pressure on the speech centres.

An aneurysm of the *anterior cerebral* artery may cause loss of smell and of sight, by pressing upon the olfactory and the optic nerves. In aneurysm of the *posterior cerebral* there may be hemiplegia with paralysis of the third and sixth nerves on the side of the lesion.

Aneurysm of the *posterior communicating* artery is apt to compress the optic tract causing homonymous hemianopsia to the opposite side, and the third nerve causing paralysis of the muscles supplied by it.

Aneurysm of the *middle cerebral* artery, which is usually situated within the fissure of Sylvius is liable to be attended by hemiplegia and convulsions: the cranial nerves are rarely involved. There may be aphasia if the aneurysm is on the left side.

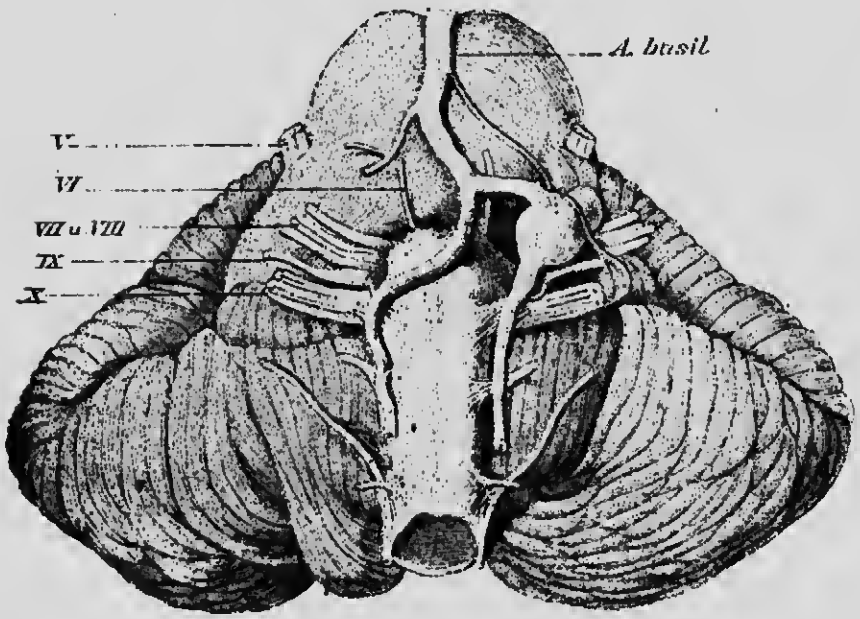


Fig. 184. Aneurysm of the left vertebral artery, compressing the facial nerve, and causing facial spasm. (Gowers, after Schultze.)

Aneurysm of the anterior portion of the *basilar artery* often exerts pressure upon the crus causing paralysis of the third nerve on the same side and hemiplegia on the opposite side. When the posterior portion of the artery is involved serious symptoms are likely to arise from compression of the pons. There may be paralysis of the limbs, unilateral or bilateral, in associa-

tion with paralysis of one or more of the cranial nerves on the side of the lesion. Difficulties in articulation, in swallowing and in breathing may also occur.

Diagnosis. About fifty years ago Gull said that "although we may from the circumstances sometimes suspect the presence of aneurysm within the cranium we have at the least no symptoms upon which to found more than a possible diagnosis." The correctness of this statement is fully confirmed by the careful investigations of Beadles, who believes that the certain diagnosis of aneurysm of any one of the cerebral arteries is practically impossible. Even when there are definite signs of an intra-cranial tumour it is difficult to distinguish those due to aneurysm from those due to a growth. Further the apoplectic symptoms which follow rupture of the aneurysmal sac are indistinguishable from those due to hæmorrhage from other causes.

The only presumptive evidence in favour of aneurysm is the presence of focal symptoms which appear to depend on a lesion in the position of one of the cerebral arteries, especially when such symptoms are intermittent in character and occur before the degenerative period in persons who have suffered from either syphilis or endocarditis.

Treatment. If there is reason to believe that an aneurysm is present it is advisable for the patient to rest in bed, with his head and shoulders well raised, and to avoid all unnecessary movements. The bowels should be kept freely open. The promotion of coagulation of the blood in the aneurysmal sac is favoured by the administration of large doses of iodide of potassium, and occasionally, as in a case described by Gowers, this method of treatment has proved efficacious. When the aneurysm can be localised with certainty the artery from which it derives its blood should if possible be ligatured, the internal carotid if the aneurysm involves this vessel or one of its main branches, and the vertebrals in cases of basilar aneurysm.

SECTION XVI.

Intracranial Tumours.

Almost every variety of tumour-growth may be met with in the cranial cavity. The growth may originate in the tissue of the bones, the meninges, the walls of the blood-vessels or in the neuroglia; thus some tumours grow from the bone or membranes and press into the surface of the brain, whilst others begin within its substance, as in the pons or central ganglia; their most frequent site, however, is the cerebellum or one of the cerebral hemispheres.

The commonest forms of tumour are tuberculous masses, gummata, gliomata and sarcomata. In children tubercle occurs most frequently, in adults glioma. Carcinomata are not common; parasitic cysts, fibromata, osteomata, endotheliomata, cholesteatoma, psammomata, angiomata, lipomata and neuromata are of rare occurrence.

Fig. 185. Tuberculous tumour of the middle lobe of the cerebellum. P, pons. (Gowers.)

Tuberculous tumours occur most frequently in the cerebellum and not uncommonly in the cortex cerebri and the pons. They are firm, rounded masses, varying in size from a marble to a small orange, and appear to spring from the lymphatic sheaths of the blood-vessels.

On section they have a cheesy aspect and they may be softened in places; sometimes they are encapsuled. Occasionally one tumour only is present, but it is common to meet with two or three or even more. As a rule the tuberculous mass is secondary to tuberculosis in some other part of the body, and it is often accompanied by tuberculous meningitis; it may remain quiescent for a long period.

Gummata are common in adult life, and occasionally occur in children as a result of hereditary syphilis. Usually small in size and irregular in shape, their cut surface has a gelatinous appearance, and sometimes presents cheesy spots separated by fibrous tissue. At a later stage they become shrunken and surrounded by a fibrous capsule, while ultimately they may be represented by a fibrous cicatrix. They develop from the peri-vascular sheaths, and whether situated on the surface or, as occasionally happens, within the substance of the brain, they are connected with the pia-mater. Their usual situation is the cortex cerebri; as a rule other syphilitic lesions are also present, especially endarteritis.

Gliomata are composed of delicate fibrous tissue cells quite similar to those of the neuroglia. They progressively infiltrate without displacing the brain tissue, and, being of much the same consistence and colour, their outline is difficult to determine; in fact the presence of such a growth may easily be overlooked. The delicate vessels in a glioma are easily ruptured, giving rise to hæmorrhage within its substance; cystic degeneration also occurs. Gliomata are found in the cerebellum, the pons and in other parts of the brain; their most frequent site is the cerebral cortex.

Sarcomata occur more frequently outside than inside the brain and commonly originate in the membranes or in the bone, especially at the base of the skull. They may perforate the cranium and appear as swellings on its surface. In contrast to the gliomata they have well-defined limits and compress and displace rather than infiltrate the brain. Sometimes primary, they are often

secondary to growths in other parts of the body. According to the variations in structure and in the degree and kind of degenerative changes which may occur in these growths, we meet with fibro-, glio-, myxo-, melanotic- or cystic-sarcomata.

Carcinomata. Carcinoma may occur as a primary growth, but usually it is secondary to cancer of the breast or of some other organ. It is often soft and very vascular, and tends to grow rapidly. The growth is most commonly found in the cortex cerebri or in the cerebellum; sometimes it arises from the choroid plexus and grows into the ventricles. Occasionally small vascular growths are found on the dura-mater; they may perforate the bones, forming "fungus hæmatodes."

Cysts in the brain are generally the result of either hæmorrhage or softening; they may also be caused by degeneration in sarcomatous and gliomatous tumours. Sometimes they are *parasitic* in origin, being either hydatid or cysticercus: the former being usually single, the latter multiple.

The rarer varieties of tumour are of little practical importance; they are described in works on pathology.

Etiology. Tumours of the brain are twice as common in males as in females, and the sexual difference is especially noticeable in the case of tubercle, glioma and cancer; the incidence of sarcoma is about equal in the two sexes. The influence of syphilis and of injuries to the head may account in some degree for the greater liability of males to be affected, but that some other explanation is required is shown by the preponderance of tumours in male children as well as in the male adult.

Nearly three-fourths of the cases occur in childhood and the active period of adult life, that is, before forty-five years of age, and owing to the frequency of tubercle in early life, about one-third during the first twenty years. The common period for glioma and sarcoma is between twenty and forty years, that for cancer between forty and sixty years, while parasitic tumours usually occur between ten and twenty years of age.

A family history of phthisis is commonly obtained in cases of tuberculous growths, and in adults there are often physical signs of chronic lung disease.

In some cases the influence of falls and blows on the head appears to be great. Sometimes symptoms of a brain tumour quickly follow an injury to the head, and occasionally the position of the growth corresponds to the seat of injury: it is possible that the molecular changes produced by concussion may initiate the development of a new growth.

Symptoms. In order to fully understand the manifold symptoms that may be associated with a brain tumour we must recognise that they are due not merely to the direct effects of the growth on the parts involved by it, but also to its indirect effects on distant parts, and indeed often on the whole intracranial contents. The cranium is a closed cavity with rigid walls, except in infants with the open fontanelle. This cavity being already filled, any addition to its contents must result either in destruction of tissue or in its compression into a smaller compass. The symptoms therefore of a growing tumour are the results partly of destruction and partly of compression of nerve elements. The cerebral vessels also may be compressed, and thus a portion of brain at some distance from the tumour may become softened and functionless, when other symptoms will arise which complicate the diagnosis.

Moreover, as the tumour increases in size, compression is not limited to adjacent parts, but it affects the brain as a whole, and increases the general intracranial pressure. A tumour in one cerebral hemisphere may interfere with the functions of the other hemisphere in spite of a partial barrier the falx between them; and although a tumour situated beneath the tentorium—a firm and complete septum—has for a time its pressure effects mainly limited to the cerebellum, pons and medulla, it will, as it increases in size, obstruct the flow of the cerebro-spinal fluid, and thus lead to distension of the lateral ventricles and so to deleterious influences on the

cerebrum. Other lesions, as meningitis, local spreading œdema, vascular disease at a distance from the tumour may also be responsible for symptoms which still further complicate the clinical picture.

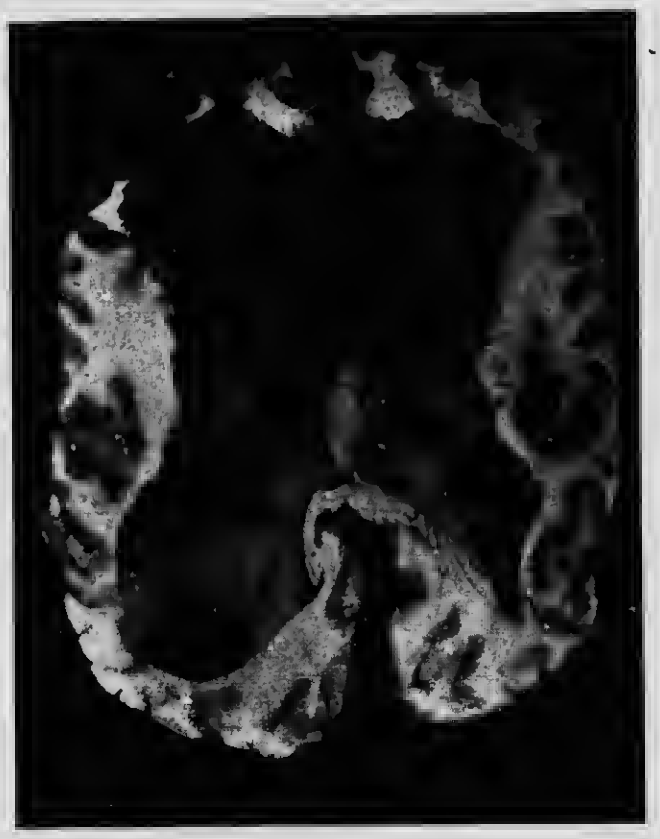


Fig. 186. Hydrocephalus, secondary to a large solitary tuberculous mass in the right lobe of the cerebellum, pressing on the veins of Galen. (Pathological Museum, Manchester University.)

Finally, it must be remembered that, very exceptionally, particles are detached from a cerebral neoplasm and being conveyed by the cerebro-spinal fluid are deposited in other parts of the nervous system, where they form secondary growths.

Degeneration of the posterior columns of the cord, not infrequently found in association with intracranial

tumours, is another factor which may be responsible for some of the symptoms present. Variability in symptoms, such as the disappearance of paralysis affecting either some of the cranial nerves or the limbs, or the transition from apathy to alertness, may be accounted for in some cases by the changing structural relations of a growing tumour, and in others by changes in the quantity and quality of the blood supplied to the brain. Certainly the post-mortem findings do not always adequately explain the varying aspects of a case observed during life.

The symptoms may be divided into general and localising or focal. The general symptoms are common to growths in any part of the brain, the focal depend on the situation of the neoplasm and are helpful in determining its regional diagnosis.

General Symptomatology.

Of the general symptoms, *headache*, *vomiting* and *optic neuritis* are the most noteworthy. *Headache* is one of the earliest and most prominent of the symptoms of intracranial growth; it mainly depends on increased intracranial pressure. The pain varies in character and in intensity; in some cases it is comparatively slight and may disappear for a time, but as a rule it is severe and constant and is liable to paroxysmal exacerbations of great intensity; it is often aggravated by light, noise and muscular exertion. In many cases the pain is of a dull aching character; in others it is boring or lancinating. Sometimes it is diffused over the head, sometimes it is localised to particular regions. A tumour in the cerebellum often causes pain in the back of the head and down the neck; one in the frontal lobe, pain in the forehead; one in the temporo-sphenoidal lobe, pain above the ear. But the situation of the pain is not a certain indication of the site of the tumour. A more reliable sign is local tenderness on percussion of the skull; this is frequently present in cortical tumours. In infants, pain in the head is expressed by outbursts of

screaming and by the putting of the hands to the head.

Vomiting is often an early and a persistent symptom, and especially when the growth is situated in the cerebellum, the medulla or in the corpora quadrigemina. It occurs more frequently in children than in adults. In its characteristic form vomiting is fitful, has no relation to the taking of food and is unattended either by nausea or by abdominal pain.

Optic neuritis occurs in about eighty per cent. of the cases; it is therefore an important sign of intracranial tumour, but its absence does not exclude the presence of a tumour. Its intensity depends more upon the rapidity of growth and upon the seat of the tumour than upon its size. At first neuritis may be unilateral, but as a rule it soon becomes bilateral; occasionally, however, it remains unilateral, or one eye is more affected than the other; such a condition would suggest that the tumour was on the same side as the more affected eye, but it is not a certain indication.

Vision is not necessarily impaired, hence the importance of the routine use of the ophthalmoscope. After a time atrophy succeeds the neuritis, when vision becomes diminished, but the diminution is not always proportionate to the apparent atrophy of the disc. As a rule optic neuritis develops more rapidly and becomes more intense in cases of intracerebellar and intraventricular growths than in cases of extracerebellar and intrapontine growths.

Opinions differ as to the mechanism by which the neuritis is produced, and it probably varies in different cases. In most cases of brain tumour the development of optic neuritis is closely related to a decided increase of intracranial pressure; in others there may be direct infection of the nerves by the products of disease, or an extension of the inflammation from an adjacent meningitis may occur.

General convulsions indistinguishable from those of idiopathic epilepsy may occur in tumours of any part of the brain. Such occurrence indicates the importance of

looking for signs of a brain tumour in every case of epilepsy. A convulsive attack may be the first prominent symptom of a tumour; in some cases it is not repeated; in other cases general convulsions occur at irregular intervals throughout the course of the disease. Sometimes they appear to indicate a rapid progress in the new growth, and as a rule are more closely related to irritation of brain tissue than to an increase of intracranial pressure. Sometimes attacks of petit-mal take the place of general convulsions.

Mental condition. Disturbance of the mental faculties is common. As a rule it is slight except in the final stages when stupor gradually deepens into fatal coma. During the course of the disease the usual condition is one of apathy and dulness with depression and impairment of memory; the mental torpor is often indicated by a dull, heavy, drowsy expression of the face. Marked irritability is sometimes noticed, especially in children. Hysterical manifestations are not uncommon and may lead to a wrong diagnosis. Occasionally mental disturbance is more pronounced, the patient suffering from hallucinations and even actual delusions. In rare cases maniacal attacks, and delusional forms of insanity have been observed.

The early development and the persistence of psychical symptoms in cases of supra-tentorial growths is in marked contrast to the insignificance or absence of such symptoms in cases of sub-tentorial growths. Mental changes are particularly apt to be prominent and to develop at an early period when the tumour is situated in one of the frontal lobes.

Vertigo is often present; it is most intense and constant when the tumour involves the cerebellum or its peduncles. Sometimes it is related to derangement of the eighth nerve, or to paralysis of an ocular muscle.

The *general nutrition* is often maintained for a long time, and some patients even put on fat. Sooner or later, however, emaciation and bodily weakness develop and steadily progress. Polyuria, albuminuria and glycosuria are occasionally observed.

Tumours of the Prefrontal Region.

The presence of a tumour in the portion of brain situated in front of the ascending frontal convolution, may be unattended by any symptoms other than headache and optic neuritis; the neuritis is frequently most intense on the side of the lesion. In a large number of cases, however, there are localising signs. A lowering of the mental faculties is particularly common, and

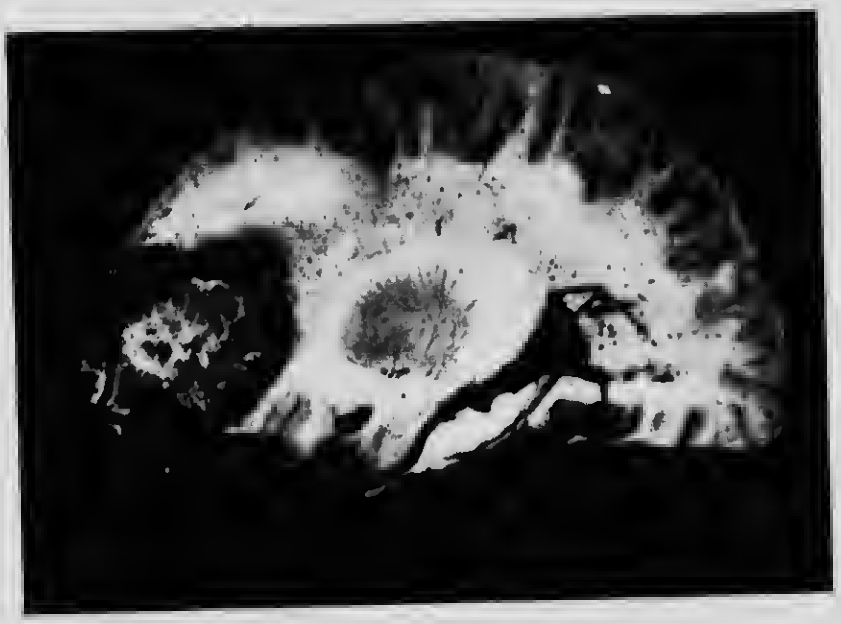


Fig. 187. Tumour in left frontal lobe, secondary to endothelioma of the cervical lymphatic glands. (Pathological Museum, Manchester University.)

frequently occurs at an early period of the disease. The character and disposition of the patient become altered; the attention is blurred, the memory fails, and there is much apathy with a tendency to somnolence; eventually delusions and dementia may supervene. Sometimes restlessness and irritability are prominent.

A tumour implicating the posterior ends of the frontal gyri may give rise to epileptic seizures, either generalised convulsions, or attacks of 'petit-mal. In some cases

there are fits beginning with localised spasms; the head and eyes are rotated to the side opposite to that of the lesion; sometimes the spasm spreads to the face and limbs, and it may be followed by transient weakness of the convulsed parts. These attacks are not preceded by a sensory aura, as is frequently the case when a growth is situated posterior to the Rolandic fissure. When the

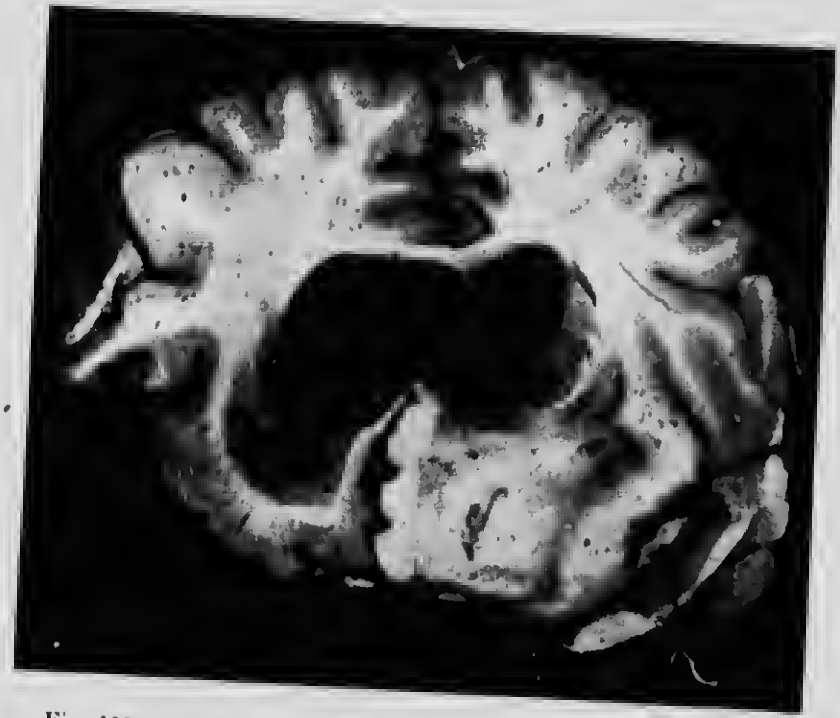


Fig. 188. Spindle-celled sarcoma in the hinder and lower part of the right frontal lobe. (Pathological Museum, Manchester University.)

growth is on the left side and the patient is right-handed the fit may be represented by transient motor aphasia followed by twitching of the right side of the face, and, if the discharge spreads to adjacent centres, by convulsive movements of the limbs. Persistent motor aphasia would indicate destruction of the third frontal convolution.

As pointed out by Grainger Stewart, a fine rapid

tremor of the limbs, especially of the upper limb, on the same side as the growth is a characteristic feature of frontal tumours; it is best seen when the patient extends his arms horizontally in front of him. Stewart has also drawn attention to the diminution or absence of the abdominal reflexes on the side opposite to that of the tumour; this phenomenon points to an affection of the pyramidal system, and frequently ushers in a hemiparesis, the weakness being caused by involvement either of the precentral convolution or of its efferent fibres. The first and the third cranial nerves may be affected by direct pressure of the growth; unilateral anosmia is a valuable indication of the side on which the tumour is situated.

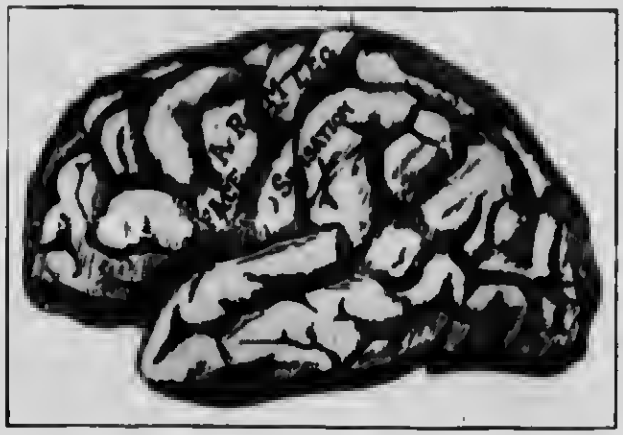


Fig. 189.

Tumours of the Precentral or Motor Area.

A tumour in the ascending frontal convolution gives rise at an early period of its growth either to spasm or to paralysis of some of the muscles on the opposite side of the body. If the tumour is situated in or near the surface of the brain, spasm precedes paralysis: if it occupies the subcortical white matter, paralysis is usually the earlier symptom. From our knowledge of the locality in the cortex in which the movements of

various parts of the body are represented, the position of the initial local spasm or paralysis enables us to determine the site of the tumour. The spasms take the form of either tonic or clonic convulsions, and tend to spread from the part first affected to other parts that are represented in portions of the cortex adjacent to the disease. Thus spasm beginning in the toes and spreading gradually up the leg, affecting successively the muscles of the ankle, knee and hip, would indicate initial irritation of the highest portion of the precentral convolution, with extension of the irritation for a short distance downwards from this part. Sooner or later in all cases of precentral tumour either a monoplegia or a hemiplegia slowly develops; the paralysis persists and is steadily progressive. In many cases psychical and sensory symptoms occur, owing to the extension of the lesion to the adjacent frontal and post-central convolutions. The frequent association of anæsthesia and paralysis when the tumour is in the subcortical white matter is explained by the close proximity of the sensory and motor tracts.

Tumours of the Parietal Region.

The parietal region includes the post-central convolution, and the superior and inferior parietal lobules. The chief focal symptom of a tumour limited to the post-central gyrus is impairment of the cutaneous sensibility and possibly of the stereognostic sense on the opposite side of the body, tactile sensibility being impaired to a greater degree than painful and thermal sensibilities; there is also impairment of the senses of active and passive movements of the limbs. Owing to the proximity of the motor area Jacksonian epilepsy is not uncommon; it is usually ushered in by a local sensory aura consisting in a sensation either of numbness and tingling, or of movement in the face, trunk or limbs on the opposite side of the body. Some degree of weakness of the opposite limbs generally ensues, and there appears to be a tendency for the paralysed muscles to waste; the

myotatic irritability, however, is always increased. Extension of the lesion backwards may lead to involvement of the higher visual centres in the angular gyrus, giving rise to word blindness and sometimes to crossed amblyopia. When the tumour is on the left side and grows downwards towards the superior temporal convolution the patient may suffer from word deafness.

Fig. 196. Glioma in the posterior parietal region of the left cerebral hemisphere. (Pathological Museum, Manchester University.)

Tumours of the Temporo-Sphenoidal Lobe.

A tumour involving the *uncinate gyrus* gives rise to subjective sensations of smell or of taste which are often of a disagreeable or perverted character; such sensations may usher in an attack of epileptic vertigo, and may be accompanied by a peculiar dreamy state in which there is a feeling either of unreality or that what is happening has been previously experienced. During the seizure movements of the lips and jaws are sometimes present; occasionally the patient suffers from a subjective feeling of excessive hunger and thirst. A destructive lesion of the uncinate lobe may cause impairment of the senses of smell and taste.

Irritation of the *superior temporal gyrus* causes subjective auditory sensations; its destruction causes incomplete deafness of the opposite ear. Word deafness may also occur when the tumour is on the left side.

Other parts of the temporal lobe appear to have no special functions and may be the seat of a large tumour without definite localising symptoms, unless the subjacent white matter is much involved, when hemiplegia and hemianæsthesia on the opposite side of the body may be present.

Tumours of the Occipital Lobe.

A tumour in the occipital lobe may be entirely latent, but as a rule it produces disturbance of vision. Irritation of the cortex may cause colour and visual auræ. Destruction of the cortex, whether on the external or the mesial aspect, is usually attended by homonymous hemianopsia to the opposite side. The sense of colour—the most special of the visual senses—is probably the earliest affected, but this is speedily followed by defect of vision for form and light when there is complete hemianopsia. The patient may be quite unaware of the defect, or he may complain of loss of sight which the observer may attribute to the optic neuritis, which in these cases is often intense; it is therefore important to test the visual fields in every case of suspected brain disease. Central vision remains unaffected and the reaction of the pupils is quite normal.

In some cases the hemianopsia depends on implication of the optic radiations rather than on the cortex itself; it may be associated with some degree of hemiplegia and hemianæsthesia from pressure on the internal capsule. Mind blindness and word blindness may be produced by the forward extension of the growth to the angular gyrus.

A large growth in the occipital lobe may exert pressure on the lateral lobe of the cerebellum giving rise to ataxia and hypotonus of the limbs on the same side.

Tumours of the Mesial Aspect of the Cerebrum.

A tumour involving the gyrus fornicatus may give rise to partial anæsthesia and paralysis on the opposite side of the body. If the growth begins in the anterior part of the falx cerebri and compresses the adjacent hemispheres mental symptoms and epileptic attacks may be prominent, the condition sometimes resembling that of general paralysis of the insane. A tumour growing

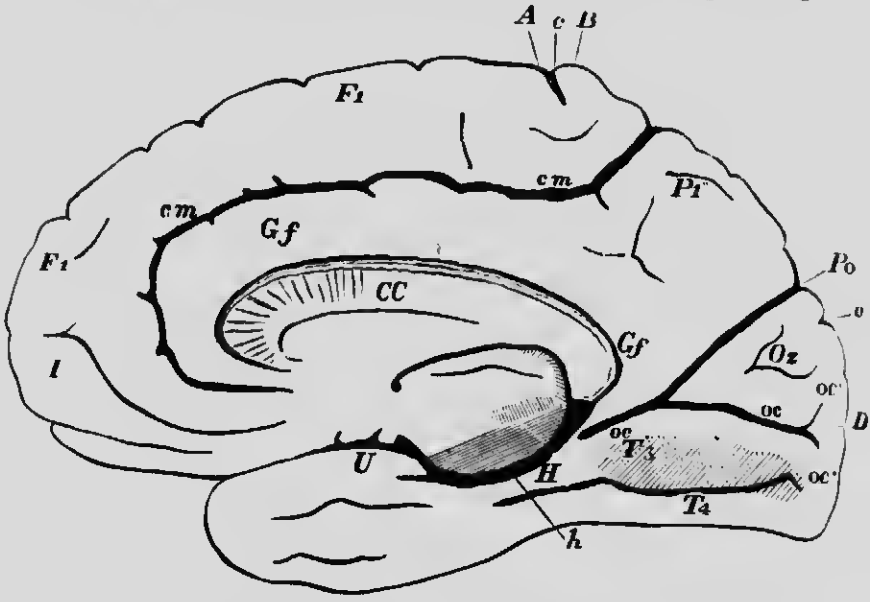


Fig. 191. The median aspect of the right hemisphere. (Ecker.) CC, corpus callosum; Gf, gyrus fornicatus; U, uncinate gyrus; F1, first frontal convolution; Oz, cuneus; Po, parieto-occipital fissure; oc, calcarine fissure.

in the posterior part of the falx is apt to give rise to Jacksonian seizures which begin in the lower limbs and are probably due to irritation of the paracentral lobule: the convulsive attacks are accompanied or followed by paralysis of one or both legs according as the growth extends into one or into both hemispheres.

Tumours of the Corpus Callosum.

To diagnose with certainty the presence of a tumour in the corpus callosum is always difficult and usually

impossible. The most characteristic symptoms are a gradually developing hemiplegia, first on one side and then on the other, and progressive mental changes ending in fatal coma. A striking feature of a growth in the anterior portion of the corpus callosum is intense optic neuritis associated with blindness, owing to direct pressure of the tumour on the optic tracts. Williamson suggests that in cases of intracranial tumour the presence of bilateral Babinski reflexes, without either ankle clonus or paresis of the limbs, is a useful indication of the median position of the growth.

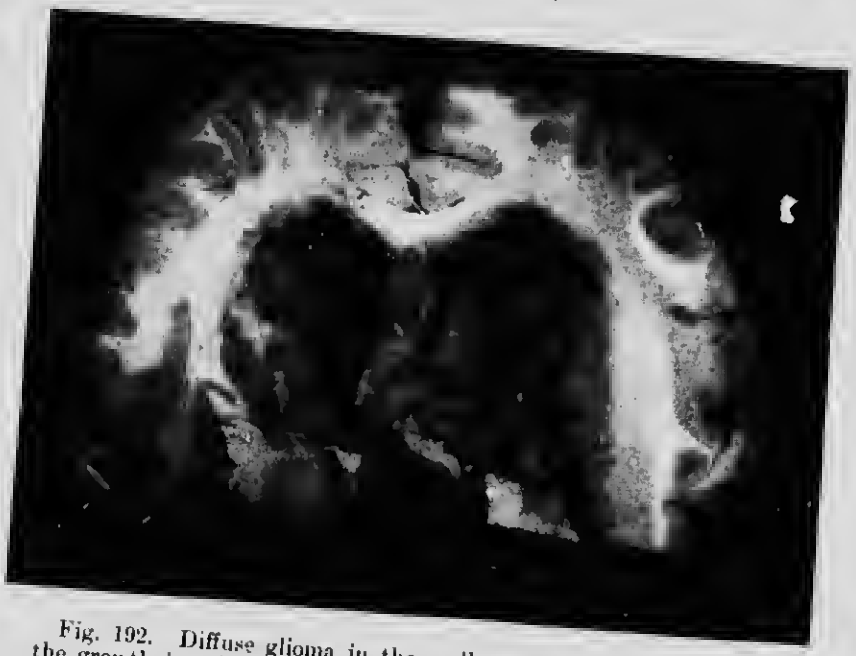


Fig. 192. Diffuse glioma in the walls of both lateral ventricles; the growth invaded the crura cerebri, and the anterior part of the pons. (Pathological Museum, Manchester University.)

Tumours of the Lateral Ventricles.

The development of a tumour in one of the lateral ventricles may be indicated by a rapid onset of headache and optic neuritis, these symptoms being associated with weakness and spasticity of the limbs and exaggeration of the deep reflexes. Mental impairment is prone to occur at an early period. In the later stages of the disease

there may be signs of compression of the parts below the tentorium, namely, vertigo and ataxia, and sometimes nystagmus and weakness of the external rectus. Death is generally caused by respiratory paralysis as a result of bulbar anæmia.

Tumours of the Basal Ganglia.

The symptoms of a tumour limited either to the corpus striatum or to the optic thalamus cannot always be distinguished with certainty from the symptoms of a tumour involving in the one case the motor division, in the other the sensory division of the internal capsule and the optic radiations.

Fig. 193. Showing displacement of the left eye outwards, and limitation of its movement upwards.

It has been pointed out by Roussy that a lesion of the optic thalamus may be diagnosed when the following symptoms are present:—(1) Marked and persistent hemianæsthesia, the loss of deep sensibility being more pronounced than the loss to touch, pain and temperature. (2) Slight hemiplegia, usually without contracture, and rapidly passing away. (3) Hemiataxia and astereognosis. (4) Severe pains in the affected side, persistent, paroxysmal and often intolerable. (5) Tremor, choreic or athetotic movements in the limbs of the affected side. Roussy believes that the sensory loss and

the pains are alone due to the lesion of the optic thalamus, whilst the other symptoms are produced by destruction of adjacent parts. The significance of this "*syndrome thalamique*" has been amply confirmed by the elaborate investigations of Head and Holmes, of which they give a full account in the Croonian lectures delivered by them in 1911. In these lectures they briefly report a

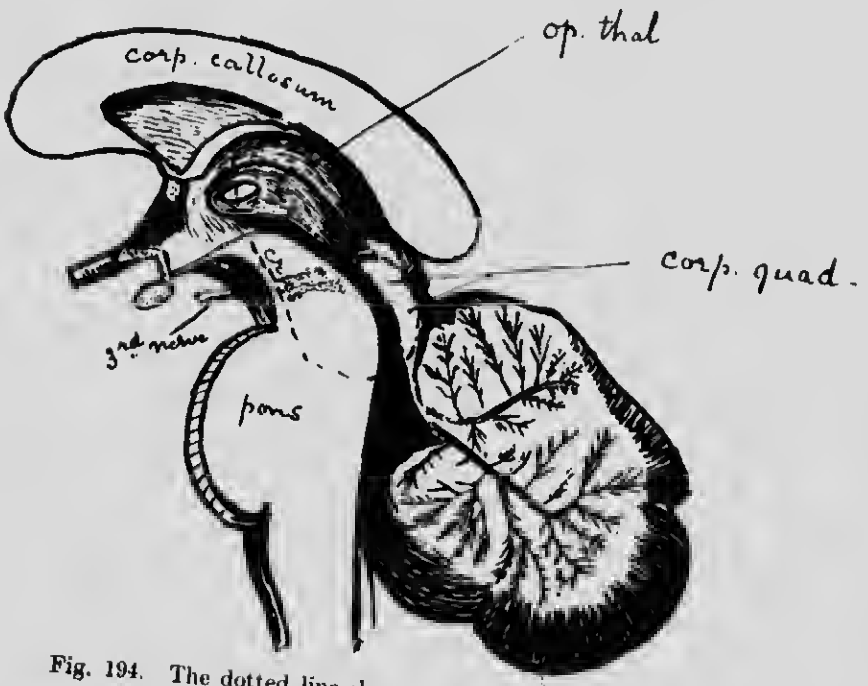


Fig. 194. The dotted line shows area occupied by new growth.

case of malignant growth involving the corpora quadrigemina and the right optic thalamus, a case which was observed by the late Dr. Beevor, and which was also under my own care; the clinical history and post-mortem appearances are fully recorded in the *Medical Chronicle* for 1909 (see figs. 193 and 194).

Here it may be noted that tumours beginning outside the internal capsule and growing towards it may produce a monoplegia before a hemiplegia—the face, the arm or the leg fibres being pressed on before the others.

Tumours of the Corpora Quadrigemina.

Tumours, although rare, are the commonest lesions of these bodies. In most cases neighbouring parts are implicated, hence it is difficult to say how far any observed symptoms are due to a lesion of the corpora quadrigemina. Their involvement, however, is strongly suggested by an unsteady, reeling gait, especially if this appears as the first symptom and is associated with impaired movement of the eyes (see fig. 193).

The gait resembles that of cerebellar disease rather than that of tabes; in advanced cases the disturbance of equilibrium is so great that the patient is unable to walk or even to stand. There is no real paralysis unless the growth involves the motor tracts. The ophthalmoplegia does not affect the ocular muscles in equal degree: a frequent combination is ptosis on one or both sides, impairment of the upward movement, with or without defective lateral movements of the eyeballs, together with paralysis of their convergence. The pupils may be dilated and unequal and may show sluggish reactions. Defective vision, which is sometimes observed, is probably related to optic neuritis or to internal hydrocephalus.

In a case reported by Williamson there was no paralysis of the third and sixth nerves. He urges the importance of examining for signs of paralysis of the fourth nerve, and believes that the presence of such paralysis, together with ataxia and a tendency to fall forwards, would suggest a lesion either in the region of the corpora quadrigemina or at the anterior part of the cerebellum and region of the superior peduncles.

When the posterior tubercle is implicated, hearing may be impaired on the opposite side, the probable explanation being that the central tracts of the cochlear nerve pass through this tubercle to reach the internal capsule. Tremor and athetosis also occur, probably as a result of extension of the growth to the superior cerebellar peduncle.

Tumours of the Third Ventricle.

Tumours growing within the third ventricle nearly always give rise to internal hydrocephalus, which is expressed by mental impairment, bilateral spastic weakness of the limbs, exaggeration of the tendon reactions and diminution of the superficial reflexes. From the standpoint of symptomatology Weisenburg, who has analysed the records of thirty cases with necropsy, divides tumours of the third ventricle into

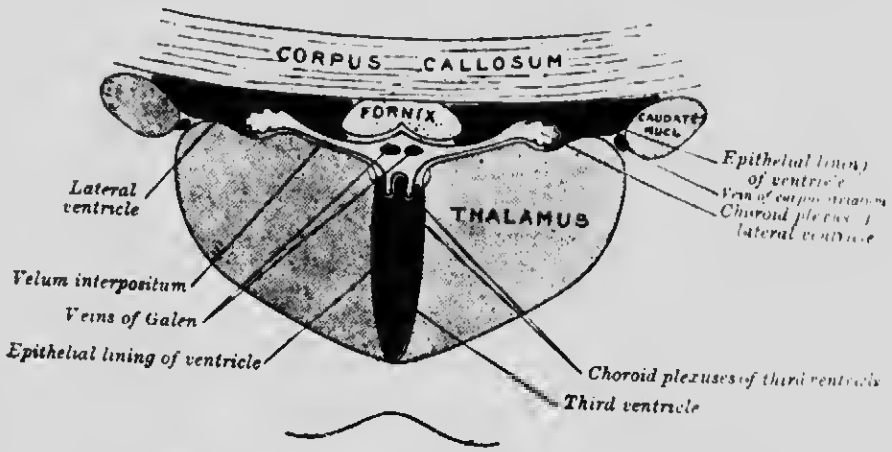


Fig. 195. Coronal section of lateral and third ventricles,—diagrammatic. (Gray's Anatomy.)

three classes:—(1) Growths situated in the floor of the ventricle which do not extend either into the foramina of Monro or into the aqueduct of Sylvius. In these cases there are no specific symptoms. (2) Growths so situated as to obstruct the foramina of Monro, causing dilatation of the lateral ventricles. The degree of obstruction varies with the position of the tumour, which may be altered by certain deviations of the patient's head; variations in the symptoms may thus be accounted for. (3) Growths which affect the structures around the ventricle, by pressing either directly upon them, or indirectly after their extension into the aqueduct of Sylvius. It is only in this group of cases that definite

localising signs are met with. These are:—impaired pupillary reactions together with paralysis of the associated ocular movements upwards, and less commonly laterally and downwards, in consequence of pressure upon the oculo-motor nuclei, especially those of the third nerves; ataxia of the cerebellar type caused by implication of the red nuclei or of the superior cerebellar peduncles; and occasionally protrusion of one or both eyeballs as a result usually of direct pressure on the cavernous sinus.

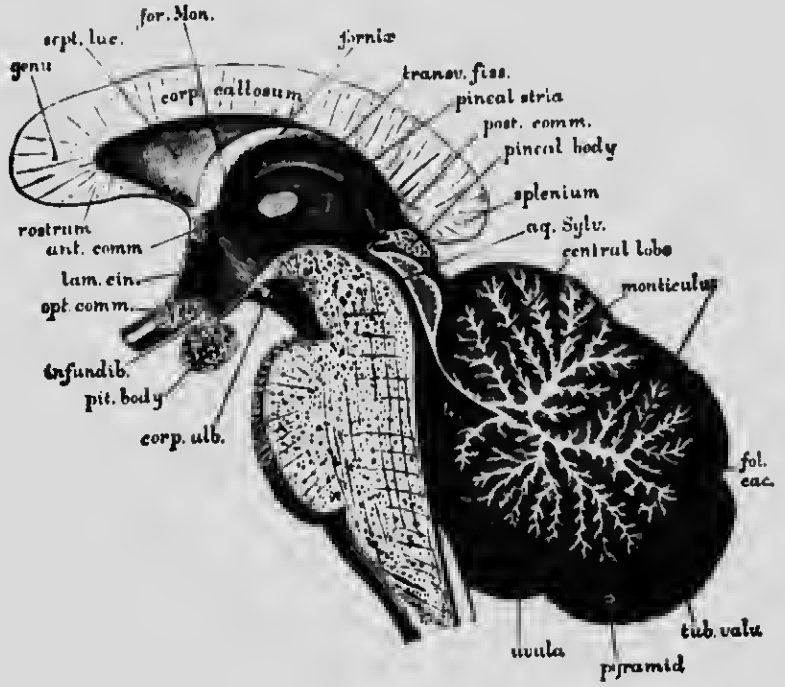


Fig. 195. (Quain).

Tumours of the Hypophysis Cerebri or Pituitary Body.

The most characteristic sign of a pituitary tumour is bitemporal hemianopsia from pressure on the optic chiasma. In some cases this visual defect is associated with acromegaly; the general symptoms of intra-cranial tumour are then usually absent. In other cases in

which acromegaly does not develop, frontal headache, optic neuritis and mental impairment are generally prominent and occur at an early period of the disease; the temporal hemianopsia is soon replaced by blindness, one eye being affected before the other, so that there may be loss of vision on one side and temporal hemianopsia on the other. The complete blindness which ensues cannot be accounted for by changes in the fundus of the eye, there being little or no atrophy of the optic disc.

Attacks of intense and paroxysmal drowsiness are often noticeable features. Amenorrhœa occurs in the female, impotence in the male. Some patients become excessively fat.

Tumours of the Crura Cerebri.

The characteristic symptom of a tumour growing in or compressing the crus is more or less simultaneous paralysis of the third nerve on the same side and of the limbs on the opposite side. Should the tegmentum be involved hemianæsthesia may accompany the hemiplegia.

In rare cases the following symptoms have occurred. Paralysis of the limbs on both sides owing to a tumour beginning in one crus and gradually encroaching on the other; tremors or ataxic movements on the hemiplegic side; hemianopsia from pressure on the optic tract; vasomotor disturbances in the paralysed limbs; acute nuclear ophthalmoplegia by a lesion in the middle line beneath the corpora quadrigemina.

Tumours of the Pons.

Pontine tumours are more common in children than in adults; the most frequent varieties are glioma and tubercle. A gliomatous tumour usually gives rise to a uniform enlargement of the pons—the so-called hypertrophy of the pons. In this variety of growth it is remarkable how frequently the cranial nerves escape; the general symptoms of intracranial tumour, namely, headache, vomiting and optic neuritis, may be entirely absent or may only occur in the terminal stages

of the disease. The want of proportion between the symptoms and the extent of the lesion is explained by the fact that the gliomatous growth slowly infiltrates the pons without destroying the axis cylinders of the nerve fibres. In cases of long standing, primary optic atrophy sometimes develops in consequence of compression of the optic tracts by a distended third ventricle, the distension being caused by occlusion of the aqueduct of Sylvius.

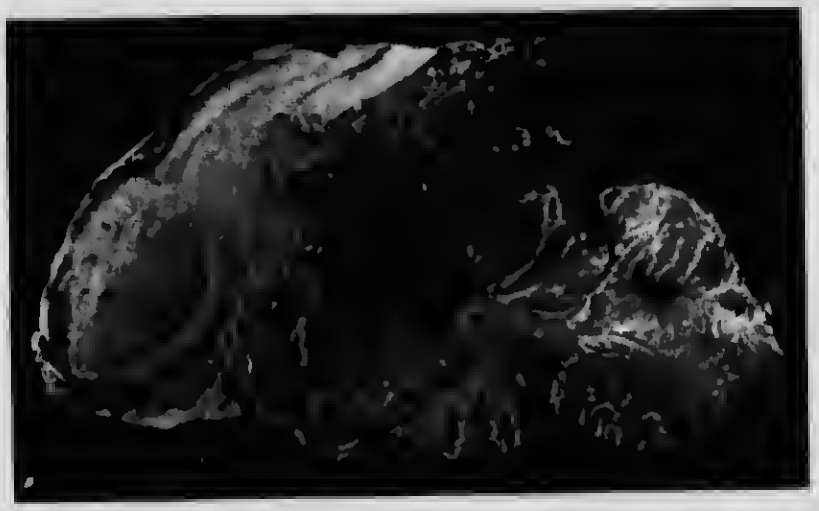


Fig. 197. Peritheloma of pons, filling up the fourth ventricle. (Pathological Museum, Manchester University.)

On the other hand the development of a tuberculous growth in the pons is often attended by headache, vomiting and febrile disturbance and sometimes by optic neuritis.

The most characteristic feature of a pontine lesion is "crossed paralysis," of which the commonest variety is paralysis of the limbs and often of the tongue on the opposite side to the lesion, and of the face on the same side; the facial palsy is of the peripheral type. In addition to the seventh, the sixth and fifth nerves are often involved, and the eighth occasionally. If the lesion extends to the medulla the eleventh and twelfth

nerves may be caught, giving rise to unilateral paralysis of the tongue, palate and vocal cord. With regard to the sixth nerve, the symptoms will vary according as its root fibres, or its nucleus is implicated. In the case of the root fibres the external rectus is alone paralysed; in the case of the nucleus the internal rectus of the other eye also suffers, when there is loss of conjugate deviation of the eyes towards the side of the lesion. If, however, the lesion is an irritative one there is spasmodic deviation of the eyes towards that side. Involvement of the fifth nerve leads to loss of sensation on one side of the face, sometimes the anesthesia is associated with weakness of the masseter and temporal muscles. Irritation of the nerve may be expressed by pains in the face and hyperæsthesia and also sometimes by trismus instead of paralysis.

In some cases there is crossed hemianæsthesia, the face being affected on one side and the limbs on the other. Neuroparalytic ophthalmia of the anæsthetic eye indicates irritation of the sensory fibres of the root, or of the Gasserian ganglion. Destruction of the nerve may cause loss of taste on one side. In cases of anæsthesia of the limbs, all four being sometimes affected, the lesion is probably in the formatio reticularis which is near the floor of the fourth ventricle. Insecurity in standing and a reeling gait which are occasionally present may depend on vertigo or on implication of the cerebellum or its peduncles. Bilateral symptoms are of frequent occurrence, a common type is spastic paresis of all four limbs with difficulties in articulation and in swallowing. Impaired articulation indicates an affection of the medio-dorsal part of the pyramidal tracts, in which the central fibres for articulation run, while dysphagia suggests that the medulla is directly or indirectly implicated.

In a few cases hemiplegia, indistinguishable from that due to disease of the internal capsule, has been observed; this indicates that the lesion is situated in the uppermost part of the pons.

In cases of tubercle death is usually preceded by symptoms of tuberculous meningitis; in cases of glioma by symptoms of bulbar paralysis.

Tumours of the Medulla.

The medulla may be invaded by tumours growing at the base of the skull, or in the cerebellum or the pons; less commonly it is the seat of a primary growth. Bilateral paralysis more or less complete, occurs; it is distinguished from that due to a pontine lesion by the

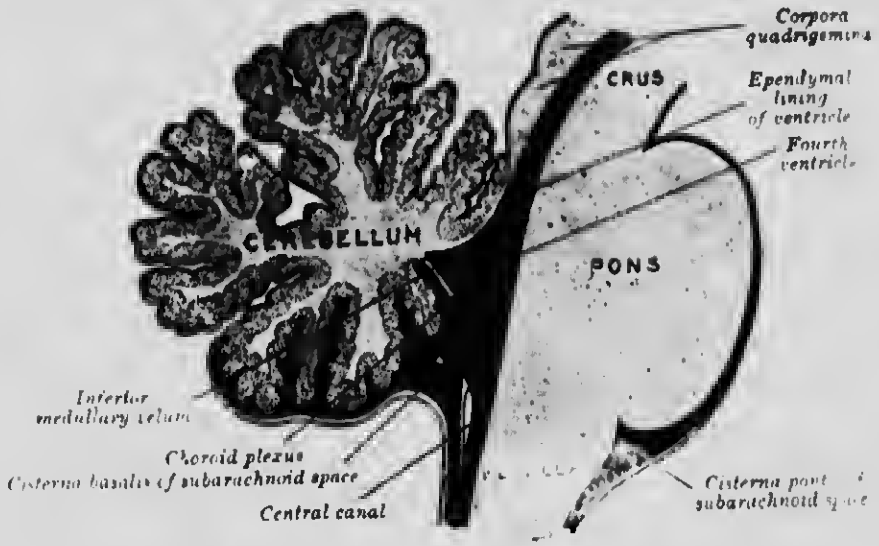


Fig. 198. Scheme of roof of fourth ventricle; the arrow is in the foramen of Majendie. (Gray's Anatomy.)

fact that it is associated with disturbance of the functions of the lower cranial nerves, rather than of the fifth, sixth and seventh nerves. Thus characteristic symptoms are loss of the reflex excitability of the palate, impairment of articulation, phonation and swallowing, with irregular respiration or intense dyspnœa and irregular action of the heart. Occasionally the seventh nerve is involved.

In some cases, tumours grow in the **fourth ventricle**: cysticercus also occurs, the vesicles either being attached

to the ependyma or floating free in the ventricle. According to Grainger Stewart, the most characteristic symptoms of new growth are: (1) The general symptoms of brain tumour: occipital headache, vomiting and giddiness develop early and are constant: optic neuritis may appear early or late: its development is always rapid. (2) Nystagmus on lateral deviation of the eyes and some weakness of both external recti and of the conjugate lateral movements of the eyes and sometimes of the lower facial muscles on both sides. (3) Slight hypertonicity of the limbs and instability of gait. It is remarkable that marked flattening or displacement of the medulla and pons may be found in cases in which during life local symptoms were quite insignificant.

The symptoms of cysticercus are somewhat different from those of new growth. The distinguishing features are stated by Bruns to be: The sudden onset of attacks of headache, vomiting and giddiness with periods of long and complete intermission; the association of such attacks with sudden movements of the head: the late appearance of optic neuritis and the tendency to sudden death.

Tumours of the Cerebellum.

Tumours of the cerebellum, which occur frequently both in children and in adults, present the characteristic general symptoms of brain tumour, namely headache, vomiting and optic neuritis, at an earlier period and in more marked degree than tumours in other parts of the brain.

The headache may be frontal or retro-ocular, but as a rule it is occipital and tends to radiate down the back of the neck: it is constant and severe. Definite tenderness on percussion of one side of the occiput is of some localising value.

Vertigo also is a frequent symptom: sometimes it is indefinite, consisting of a feeling of giddiness associated with intense nausea and a tendency to fall either backwards or towards the side of the lesion: sometimes it is

definite and both the movements of objects in front of the patient and his own feeling of rotation are from the side of the lesion to the opposite side.

Disturbance of equilibrium is a striking feature of cerebellar disease. If the tumour involves either the

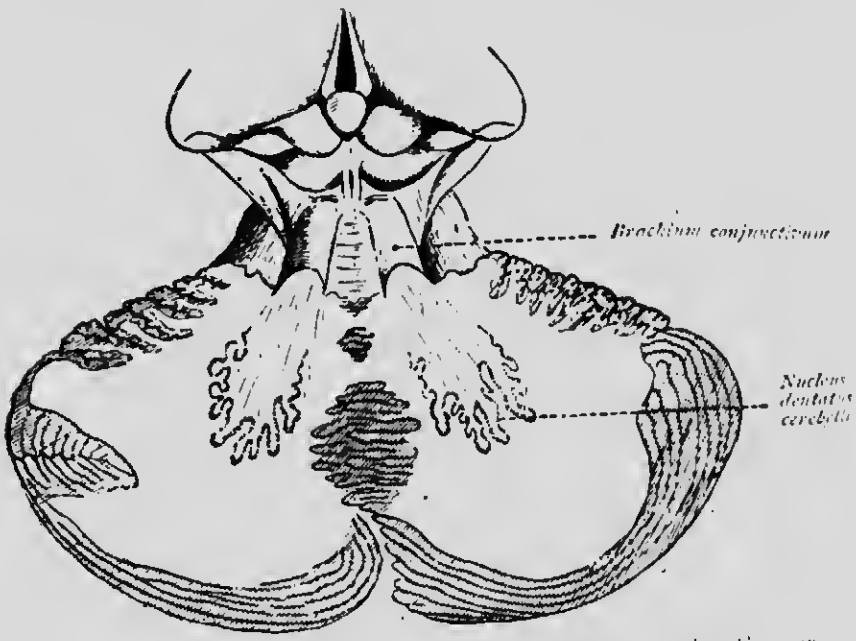


Fig. 199. The cerebellum, showing the brachia conjunctiva or crura cerebelli to the corpora quadrigemina. (Villiger.)

middle lobe, or both lateral lobes, the gait is reeling and the patient staggers from side to side like a drunken person; in severe cases he may be unable to stand. The reeling gait is largely due to irregular action of the trunk muscles. In a unilateral lesion the usual tendency is to stagger and stumble towards the same side and in walking the patient often deviates from the desired direction towards the side of the lesion. This tendency he endeavours to correct; thus a patient with a tumour in the right hemisphere who feels compelled to deviate to the right, will rotate the right shoulder forwards and try to direct his footsteps towards the left of the line of progression. It is to be noted that the

inco-ordination of movement is not related to vision as it is in tabes, and that the muscles brought into use for any particular action may co-operate in perfect harmony.

Fig. 200. Glioma of the middle lobe of the cerebellum. (Pathological Museum, Manchester University.)

Definite weakness of the limbs on the same side as a unilateral tumour is often present and may be pronounced; sometimes it is associated with weakness of the trunk muscles which is greatest on the side of the lesion. That this weakness is a direct effect of the cerebellar lesion and not due to pressure on the pyramidal tracts is shown by the absence of rigidity; indeed the weak limbs are often limp and flaccid and their muscles soft and flabby; sometimes the hypotonicity is striking.

The position of the head is of some importance in regional diagnosis. In many cases of unilateral tumour the occiput is tilted towards the shoulder on the side

of the lesion, the chin being rotated towards the opposite side (see fig. 201). When the middle lobe is the seat of a tumour the head may be retracted and the neck muscles rigid; sometimes the patient is subject to tetanic-like seizures.

Fig. 201. Showing the attitude of the head in a case of tumour of the left lobe of the cerebellum.

The reflexes are variable. A tumour in the middle lobe through pressure on the pyramidal tracts leads to exaggeration of the knee-jerks. In tumours of the lateral lobe the deep reflexes may be either increased or diminished, and may change from day to day; sometimes they are unequal; in many cases the knee-jerk is diminished or absent on the side of the lesion; in other cases it is marked on that side. The extensor plantar response is rarely met with in uncomplicated cases of a unilateral lesion.

A marked affection of the cranial nerves is against limitation of a tumour to the cerebellum. Slight unilateral paralysis of the sixth nerve sometimes occurs and is of value in localisation. When the middle peduncle is involved the fifth nerve may be affected and there may be a tendency for the trunk to rotate in one direction usually away from the side of the lesion. A "skew deviation" of the eyes has been observed in some cases; the eye on the side of the lesion looking

inwards and downwards whilst the other eye is turned outwards and slightly upwards.

Nystagmus is usually present; in tumours of the middle lobe slow nystagmoid jerkings occur when the patient looks to either side, in tumours of the lateral lobes similar movements of the eyeballs are observed when the patient looks towards the side of the lesion, but finer and more rapid movements when he looks towards the other side.

Fig. 202. Showing 'skew deviation' of the eyes from a lesion of the cerebellum. (Turner and Stewart.)

It is to be noted that some cases of cerebellar tumours are unattended by definite symptoms. This happens when the growth of the tumour has been very slow or has become stationary; the probable explanation being that the interference with one portion of the cerebellum has been compensated for by the action of its other portions and by the higher cerebral centres.

Extra-cerebellar Tumours (Tumours of the Cerebello-pontine Angle).

From a careful study of forty cases of cerebellar tumours Grainger Stewart and Gordon Holmes have been able to draw marked distinctions between the symptoms of intra-cerebellar and extra-cerebellar growths. By the latter they mean tumours "which lie in the posterior cranial fossa in the angle between the pons varolii and the cerebellum, compressing but not

directly invading either. Such growths may arise either in connection with the ventral surface of the cerebellum, when they are generally gliomata and occasionally cystic; or from the adjacent cranial nerves, especially from the sheath of the auditory nerve, when they are generally fibromyxomata. These growths are circumscribed, encapsulated tumours, and are easily separable from surrounding tissues." They press chiefly on the middle cerebellar peduncle.

The distinctive symptoms of such growths are as follows: Headache, vomiting and optic neuritis may be absent or may only occur at a late period of the disease. The eighth, seventh and sixth nerves on the side of the lesion are almost invariably and considerably affected, and usually in the order given. Nerve deafness is the earliest and most constant symptom of extra-cerebellar growths; the deafness is associated with subjective noises in the ear; sometimes the deafness is complete. Facial paralysis of the peripheral type next develops; the weakness may be slight even when the nerve is severely compressed. Paralysis of the sixth nerve also occurs; occasionally the trigeminus is implicated especially its sensory division, giving rise to tingling and partial anæsthesia in the area supplied by it.

Pressure on the lateral lobe of the cerebellum is indicated by atonia, ataxia and slight paresis of the limbs on the side of the lesion, pressure on the pyramidal fibres in the pons by spastic paralysis of the limbs on the opposite side. In the later stages of the disease when both sides of the pons are compressed, the spastic weakness may be observed on both sides of the body. Sometimes the outstretched hands show coarse tremor, which when associated with a spastic-ataxic gait makes the condition resemble that of disseminated sclerosis.

The following table, taken from the article by Stewart and Holmes, shows the chief points to be considered in the differential diagnosis of lateral cerebellar, extra-cerebellar and intrapontine tumours.

Symptoms and Signs.	Lateral Cerebellar Tumours.	Extra Cerebellar Tumours.	Intrapontine Tumours.
Optic neuritis ...	Early and intense ...	Variable	Often absent or late
Vertigo	Subjective rotation of self from the side of the lesion	Subjective rotation of self to the side of the lesion	Indefinite.
Cranial nerves V.	Rarely affected	Often affected ...	Affection of these nerves often bilateral. Paresis may be supranuclear or nuclear, and grouped according to nuclear arrangement. Paralysis of a nerve on one side and of an adjacent or distant nerve on the opposite side. Permanent paralysis of conjugate deviation of the eyes.
" " VI.	Weakness of conjugate deviation to side of lesion. Weakness of external rectus on side of lesion. Slow deliberate nystagmus to side of lesion.	Same as in unilateral cerebellar tumours	
" " VII.	Paresis slight if present	Paresis more marked	
" " VIII.	Deafness on side of lesion incomplete and variable. Tinnitus general	Deafness on side of lesion marked — generally complete. Tinnitus referred to ear on side of lesion	
" " IX.	Never affected	Occasional paresis on side of lesion	
" " X.	Ditto	Ditto	
" " XI.	Ditto	Ditto	
" " XII.	Ditto	Supranuclear paresis on contralateral side	
Motor system ...	Homolateral paresis, ataxia and atonia	Homolateral paresis and ataxia; contralateral spastic paresis common — occasionally hilateral	Paresis often hilateral, with spasticity. Ataxia general.
Sensory system ...	No change	No change	Occasionally hemianæsthesia.
Reflexes — Tendon	Variable, often diminished	Generally increased, especially on contralateral side	Increased, often unequally.
" Superficial	Normal	Often diminished on contralateral side	Diminished, often unequally.
" Plantar	Flexor	Flexor or extensor. Extensor on contralateral or both sides	Extensor on one or both sides.
Sphincters ...	Not affected	Rarely affected ...	Generally affected.

Tumours of the Base of the Skull.

Tumours which spring from the base of the skull are usually sarcomatous in nature. They may penetrate through the bone into the nasal and pharyngeal cavities, and sometimes they are palpable externally. Growths

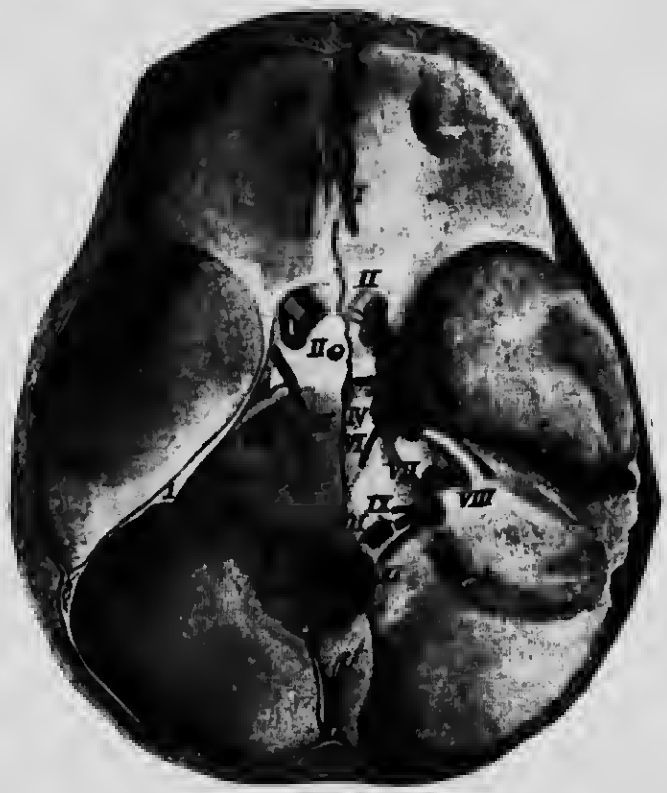


Fig. 203. The base of the skull, with the cranial nerves. (Ranney.)

in the anterior or in the middle fossa may invade the orbit and cause protrusion of the eyeball. For a long time the symptoms of basal tumours are mainly those produced by involvement of the cranial nerves, the general symptoms of intracranial tumours being absent, or only occurring at a late period of the disease.

The anterior fossa. In cases of tumour in the anterior fossa the local symptoms, which occur either separately or combined, are:—Anosmia from compression of the

olfactory nerve; unilateral blindness and temporal hemianopsia from extension backwards of the growth to the optic nerve and the chiasma respectively; and sometimes psychical changes from compression or invasion of the frontal lobes of the brain.

The middle fossa. As a rule the early symptoms of tumours of the middle fossa are referable to implication of the fifth nerve. At first there may be pain in the distribution of one or more branches of the nerve. After a time there is impaired sensation over one side of the face, while a rapid and destructive inflammation of the

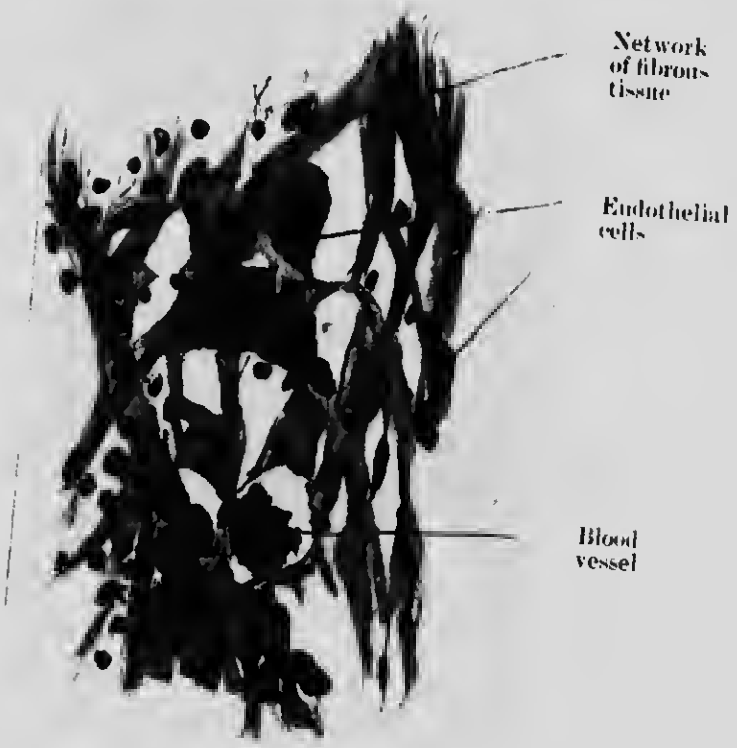


Fig. 204. Endothelioma growing from the dura mater in the right middle fossa.

eyeball is not uncommon; subsequently wasting and weakness of the masseter may occur. In some cases the seventh and eighth nerves are implicated, and when the

growth is near the sphenoidal fissure the oculo-motor nerves are liable to suffer. Slight hemiplegia from compression of the crus, and subjective sensations of smell and taste from involvement of the uncinate gyrus also occur.

The posterior fossa. The development of a tumour in the posterior fossa is generally first expressed by symptoms referrible to the eighth, seventh and sixth nerves. Subsequently there may be paralysis of the fifth nerve from forward extension of the growth, and paralysis of the ninth, tenth, eleventh and twelfth nerves from backward extension to the side of the medulla. Pressure upon the pyramidal fibres in the pons or the medulla leads to a gradually developing progressive paralysis of the contra-lateral limbs; compression of the cerebellum to paresis, ataxia and atonia of the homo-lateral limbs.

It will be noticed that the clinical picture is similar to that produced by tumours in the cerebello-pontine angle, but, as pointed out by Turner and Grainger Stewart, the cranial nerves are more rapidly involved, pain in the head and face is more frequent and persistent, deafness is less complete, nystagmus is preceded by paralysis of the external rectus muscle, and cerebellar symptoms develop at a later period of the disease.

Diagnosis of intracranial tumours. Under this heading three questions require brief consideration, namely: Is there a tumour present, and if so what is its situation, and what is its probable nature?

To the first question a correct answer can usually be given. But it must be remembered (1) that although headache, vomiting and optic neuritis are collectively strong evidence in favour of a tumour they may depend on other causes, and (2) that any one or all of these symptoms may be absent when a brain tumour is present. Severe *chlorosis* in young girls is sometimes attended by headache and papillitis, but as a rule these symptoms quickly subside under suitable treatment for the anæmia. In some of these cases headache and congestion of the

discs, simulating early papillitis, depend on hypermetropia; when this is corrected by suitable glasses the symptoms disappear. Greater difficulties in diagnosis occur when the general symptoms of brain tumour are found in young women suffering from amenorrhœa, and especially when there is little or no anæmia.

In *Bright's disease*, headache is often severe, and papillitis may exist without the characteristic degenerative retinal changes. The presence of casts in the urine, of increased arterial tension and of cardiac hypertrophy usually enables a correct diagnosis to be made.

Lead poisoning may lead to headache, vomiting, epileptiform convulsions and double optic neuritis; but almost invariably such cerebral symptoms are preceded by typical symptoms of saturnism.

Chronic cerebritis, hydrocephalus and local forms of meningitis occasionally give rise to diagnostic difficulties. The points of distinction between tumour and abscess of the brain are given in the Section on Abscess.

Great care is always needed to avoid the danger of mistaking a case of brain tumour for one of *hysteria* or of *neurasthenia*. Hysterical manifestations are easily excited by coarse brain disease, and the diagnosis of hysteria, as the sole condition present, must never be made until repeated examinations have proved the absence of all signs of serious mischief. In cases of brain tumour, as already mentioned, optic neuritis may be absent from first to last, and headache may be insignificant. Furthermore, marked neurasthenic symptoms may overshadow the importance of persistent headache which only after some months may be attended by the slow development of optic neuritis and other symptoms of a tumour, symptoms that are liable to be overlooked owing to the bias which has determined the observer's mind in favour of neurasthenia.

The situation of the tumour may usually be decided by a consideration of the localising symptoms already described, and especially of their grouping and the order of their development. But as pointed out by

James Collier the local symptoms do not always indicate the position of the tumour. They may depend on the presence of vascular lesions, meningitis, hydrocephalus, local spreading œdema of the brain, secondary deposits of new growth, and degeneration of the posterior columns of the cord. Moreover if they occur late in the course of intracranial tumours they are often of false portent, being produced by pressure on surrounding parts or by ventricular distension; for example, symptoms of a cerebellar lesion developing many months after the appearance of headache, vomiting and optic neuritis may be indirectly caused by a frontal tumour pushing the cerebellum downwards and backwards against the foramen magnum.

Of diagnostic value in some cases, is the absence of local signs during the early stage of an intracranial tumour, for this suggests that the disease is situated above the tentorium. With regard to convulsive attacks it must be borne in mind that local spasms are not always signs of a coarse lesion in the cortex. They may depend on idiopathic epilepsy, and when they occur for the first time long after the appearance of the general symptoms of an intracranial growth they have no localising value as they usually depend on a secondary hydrocephalus.

The nature of the growth. The most important indications are the age of the patient, the situation of the tumour, the course of the illness, the effects of treatment and the history or the presence of disease in other parts of the body. Such indications, however, are helpful chiefly in regard to syphilis and tubercle; other varieties of new growth are more difficult to diagnose.

A tumour is likely to be tuberculous if it occurs in an adult who is the subject of phthisis, or in a child, especially if it is situated in the cerebellum, or is complicated by symptoms of general meningitis. Tuberculous tumours often grow rapidly at first and then become stationary, and they may improve under the influence of tonic treatment.

Syphilitic growths commonly affect the surface of the brain and give rise to unilateral convulsions. Confirmatory evidence as to their nature is afforded either by a history of syphilis or by the presence of specific lesions. Moreover the effects of treatment are often remarkable, the symptoms in many cases being completely removed by the administration of mercury and iodide of potassium.

That the tumour may be a sarcoma, a cancer, or a parasitic cyst is suggested by the presence of a primary growth or of a cyst in some other part of the body. In favour of sarcoma also would be signs that it originated outside brain tissue, as at the base of the skull. A sudden apoplectic seizure during the course of the illness would in itself be suggestive that the growth was a glioma.

Course and prognosis. It is characteristic of the symptoms of an intracranial growth that they develop slowly and very gradually get worse. The downward progress may be interrupted by periods of improvement but these are often followed by periods in which the symptoms become greatly exaggerated. The rate of progress varies according to the character and position of the lesion; a soft sarcoma grows more quickly than a firm one, and a tumour in the pons ends life much sooner than one in the cerebrum. Death may occur in a few weeks or not for many years, but as a rule it takes place within two years from the first symptoms. Its chief causes are exhaustion owing mainly to the severity of the headache or the vomiting; coma which usually depends on intracranial pressure; and the development of other lesions such as meningitis or general tuberculosis in tuberculous cases, and syphilitic disease of the vessels in cases of syphiloma. Sometimes death occurs quite suddenly; this is not uncommon when the tumour involves the cerebellum or the pons.

The outlook therefore is very gloomy but it must be remembered:—(1) That the growth of almost every variety of tumour is occasionally arrested, and that very

rarely the symptoms disappear and the patient appears to make a good recovery; this has been especially observed in cases of tuberculous tumour. (2) That the effects of syphilomata may often be removed by drugs, although the persistence of some symptoms, as hemiplegia or epilepsy, indicates that nerve elements have been permanently damaged. (3) That the brilliant achievements of surgery have done much to lighten the gloom. In some cases a tumour has been removed, the symptoms have disappeared, and the patient has regained health. The success of an operation will obviously largely depend on the correctness of the diagnosis as to the position and the nature of the growth, and on its accessibility.

The presence of the following conditions indicates that the prognosis is grave, and their absence that danger is remote:—Obstinate vomiting, general convulsions, much mental dulness, apoplectic seizures and intense optic neuritis of rapid onset. Indications of improvement are usually afforded by a diminution of the optic neuritis and the severity of headache.

Treatment. Except in the case of syphiloma the removal of tumours by medicinal treatment is practically impossible, and although their growth is sometimes arrested, this cannot be anticipated in any particular case. Hence the question of surgical interference should be considered and settled as early as possible. No doubt at first it is advisable to put the patient on a course of mercury and iodide of potassium, for even when the growth is not syphilitic these remedies may be beneficial. Iodide of potassium should be given in rapidly increasing doses and if twenty grains thrice daily produce no effect, double this quantity should be tried. To this mercury may be added either in the form of corrosive sublimate or grey powder, or it may be given by inunction. How long must this treatment be persevered with? The answer is often difficult to give. When the growth is undoubtedly syphilitic, it is well to continue the treatment for five or six weeks,

but even in such a case and still more when the growth is non-syphilitic it may be desirable to decide at a much earlier period whether an operation is feasible or not; for if the symptoms are getting worse and if it seems desirable to try and remove the tumour, the sooner the operation is done the better.

An operation is feasible when the regional diagnosis of the growth can be accurately determined and when its situation is an accessible one. Thus the surgeon would expect to be able to remove a tumour situated in, or immediately beneath, the cerebral cortex, or in the lateral lobe of the cerebellum, but he would scarcely attempt to remove one situated in the pons, in the central ganglia or at the base of the skull. It must be borne in mind that although many tumours have been successfully removed, the percentage of complete recoveries after the operation is much smaller than was anticipated in the early days of brain surgery.

The operation is attended by many dangers, and these should be fairly placed before the patient and should be contrasted with the probable outlook if medicinal treatment only is relied on, in order that he may be able to come to a decision in the matter. In each case, the progress of the symptoms, the apparent character of the growth and the general condition of the patient must be carefully considered. The following are the chief risks attending the operation. Death from shock; the discovery that it is impossible to remove the tumour; the possibility that there is more than one tumour present. We must also consider the possibility that the tumour may recur, or as in the case of tubercle that meningitis may set in after the operation, and, finally, that although the tumour is successfully removed, the paralysis or the convulsions may persist, or the patient may become speechless or mentally afflicted.

If for any reason it is deemed inadvisable to try and remove the tumour then we have to fall back on palliative measures. For the relief of severe headache, antipyrine and phenacetin are often useful. Bromides, gelseminum,

butyl chloral and other analgesiacs may also be tried. Should these remedies fail and the headache become intense and agonising, hypodermic injections of morphia may be required. In some cases much comfort is derived from the application of ice bags to the head, of a mustard leaf or a blister to the back of the neck, or of leeches to the temple or behind the ear.

Spasms and convulsions are often restrained by the administration of bromides and chloral. When they depend on syphilitic deposits in the cortex large doses of iodide of potassium should be combined with bromides. When vomiting is severe and is unrelieved by bismuth or the sucking of ice, bromide of sodium or one hundredth of a grain of hydrobromate of hyoscine is sometimes of service.

Palliative trephining, that is, the removal of a piece of bone and incising the dura mater, should be employed in those cases in which the tumour cannot be removed nor its symptoms relieved by drug treatment. It has been found that this surgical procedure often relieves an agonising headache, controls urgent vomiting, leads to subsidence of optic neuritis and convulsions, and tends to prolong life. The operation, however, is not free from danger and should only be performed when the sufferings of the patient are intense, and are not relieved by the administration of drugs and other remedies.

SECTION XVII.

Abscess of the Brain.

The principal varieties of intracranial suppuration are: generalised suppurative meningitis; localised suppuration, either extra-, or intra-dural, and collections of pus within the substance of the brain. It is to the last variety that the following account mainly applies.

Etiology. The chief causes of abscess of the brain are injury, local inflammatory conditions and general infections, including infection from distant parts.

Injuries of the scalp or the cranium are common causes; even a slight blow or fall on the head may lead to intracranial suppuration. As a rule the abscess is near the seat of injury; occasionally it is on the opposite side of the brain.

Local inflammatory conditions. The commonest source of infection is suppurative inflammation of the middle ear, which usually has existed for several years; frequently the history is that the patient suffered in childhood from otitis media as a result of scarlet fever, and subsequently was subject to purulent otorrhœa, not however, constantly present; sometimes the formation of a brain abscess follows the cessation of the discharge. As a rule some part of the temporal bone is diseased and the membranes over it are thickened; sometimes these structures are normal and no direct connection can be traced between the infective focus and the abscess inside the brain, the intervening tissue being apparently healthy. In such cases it is probable that the infective agents reach the brain either by the veins or by the perivascular lymph channels. The most common situation for the abscess is the temporal lobe especially the third temporal-sphenoidal gyrus; sometimes the abscess

is in the lateral lobe of the cerebellum, being prone to develop in that part of it which is adjacent to the lateral sinus; occasionally the abscess forms in the frontal or in the occipital lobe. In some cases a relation may be observed between the part of the temporal bone that is diseased and the situation of the abscess, caries of the roof of the tympanum leading as a rule to temporo-sphenoidal abscess, disease of the mastoid cells to cerebellar abscess.

Chronic disease of the nose and its accessory cavities may also give rise to cerebral abscess. The mucous membrane may be alone involved, but as a rule the sphenoid, ethmoid or the nasal bones are also diseased, and not uncommonly as a result of syphilis. In such cases the abscess is generally situated in the frontal lobe; another though rarer source of frontal abscess is disease of the orbital bones.

It is to be borne in mind that, whereas chronic infections of the ear, nose and accessory sinuses tend to induce abscess of the brain, acute infections of these parts are more prone to give rise to suppurative meningitis and sinus thrombosis.

Some of the rare local infective sources of cerebral abscess are carbuncle in the neck; erysipelas of the scalp; osteomyelitis of the skull; tuberculous and syphilitic affections of the cranial bones; and chronic ulcerations involving some part of the face.

General infections. Multiple metastatic abscesses in the brain, in consequence of septic embolism, occur in cases of pyæmia and of ulcerative endocarditis. A single abscess, often situated in the posterior part of the cerebrum, is sometimes secondary to septic disease of the bronchi, lungs or pleura; it may also result from suppurative peritonitis, from periostitis or from osteomyelitis. In a few cases abscess of the brain has occurred as a sequel to influenza, to enteric fever or to some other specific fever.

Symptoms. The clinical history of brain abscess varies much in different cases, the variations depending

partly on its character whether acute or chronic, partly on its position and partly on its cause. Moreover the symptoms may be complicated by the presence of meningitis, or of sinus thrombosis.

It is important to remember (1) that the abscess may run either an acute or a latent course; in the latter case indications of its presence may be entirely absent or of the vaguest description, and (2) that in cases of general pyæmia the symptoms of metastatic abscesses are often overshadowed by the stupor, delirium and other symptoms of the septic condition.

The *general symptoms* of brain abscess are mainly those of increased intracranial pressure. Headache is one of the earliest and most prominent symptoms; it is, however, rarely constant; frequently dull and deep-seated the headache may be intense. In traumatic abscess the pain is generally referred to the side of the lesion; in otitis the headache is often associated with severe pain in the ear. Attacks of vertigo and purposeless vomiting are not uncommon; they are more likely to occur and to be persistent when the abscess is in the cerebellum than when it is in the cerebrum. Optic neuritis sometimes occurs, most frequently during the late stages of the disease; as a rule it is less common and less intense than in cases of tumour.

Contrary to what might be expected a normal or a subnormal temperature is the rule in cases of uncomplicated suppuration in the brain; a moderate degree of pyrexia, however, may be present in the earliest as well as in the latest stages of the disease, whilst a pyæmic type of temperature is apt to occur when the abscess is associated with sinus thrombosis. A sudden rise of temperature to 104° or 105° is an indication that the abscess has ruptured into one of the ventricles.

The pulse is usually slow and regular, its rate being often fifty or forty or even less per minute; a rise in temperature is not accompanied by a corresponding increase in the frequency of the pulse, although in the earliest as well as in the latest stages of the disease the

pulse may be both frequent and irregular. Slow respiration also is a common feature, especially when the abscess is in the cerebellum; in the last stage of the disease the respirations are often irregular and may show the Cheyne Stokes rhythm.

In most cases the mental condition becomes changed: there is an early tendency to somnolence and mental lethargy with impaired power of attention, and sooner or later the stupor deepens into unconsciousness, passing into fatal coma. Anorexia, rapid and marked emaciation, and cachexia are other features that are met with in many cases.

The *focal symptoms* are practically the same as those which have already been described under Intracranial Tumours. It is desirable, however, to give a summary of the localising symptoms of abscess in the temporo-sphenoidal lobe and in the lateral lobe of the cerebellum, these parts being the most common situations for the disease.

Temporo-sphenoidal abscess. In some cases disturbances of smell, taste and hearing occur, and should the lesion be on the left side in right-handed persons a variety of partial word-deafness may be observed, consisting in an inability to name objects, although the patient retains a knowledge of their functions. Large abscesses are prone to give rise to various symptoms in consequence of the pressure they exert on adjacent parts of the brain. Pressure downwards may implicate the third and sixth nerves at the base of the brain, and the internal capsule, causing paralysis of some of the ocular muscles on the side of the lesion and hemiplegia on the opposite side. Pressure forwards on the third frontal, and the base of the precentral convolution may produce motor aphasia, with weakness of the face, and sometimes of the arm on the opposite side. Compression of the parietal lobe is apt to produce sensory disturbances on the opposite side, while pressure on the angular gyrus and the subjacent white matter may lead to word blindness and hemianopsia.

Of considerable diagnostic significance is impairment or loss of the superficial abdominal reflexes on the side opposite to that of the abscess; the defect may occur without any signs of paralysis.

Cerebellar abscess. The presence of an abscess in the cerebellum is usually attended by severe headache, vomiting and vertigo. Optic neuritis is more frequently

Fig. 205. Abscess in the right lobe of the cerebellum, secondary to suppurative otitis media, and septic thrombosis of the right lateral sinus. (Pathological Museum, Manchester University.)

present and is more intense than in temporo-sphenoidal abscess. Rigidity of the neck and retraction of the head often occur.

The following symptoms would indicate an abscess in the right lateral lobe:—The patient lies on the left side with his limbs flexed; the eyes are directed towards the left, and are affected with a slow nystagmus towards the

right side; weakness of the right limbs, especially of the arm, the movements of which show marked incoordination; muscular spasm or rigidity on the left side; a roeling or staggering gait with a tendency to stumble and fall as well as to deviate to the right side.

Course. As already stated, the clinical course of brain abscess is very varied, and every gradation may be met with between acute cases with severe and urgent symptoms, which end fatally in two or three weeks, and chronic cases which run a latent course for many years. The acute variety is common when the cause is injury, the latent variety when the abscess depends on ear disease. Occasionally three distinct stages may be recognised: the invasiou stage, in which headache, vomiting and febrile disturbance indicate the onset of inflammation of the brain or its membranes; the stage of remission or latency, in which, although cerebral symptoms have subsided either completely or partially, the abscess slowly increases in size and becomes encapsuled; and the terminal stage, in which acute symptoms suddenly interrupt the period of latency, being caused by inflammatory œdema and softening round the abscess, or sometimes by its rupture. The abscess, especially when in the temporo-sphenoidal lobe, may rupture either into the ventricles, and give rise to symptoms similar to those of ventricular hæmorrhage, or into the subarachnoid space, and set up suppurative meningitis, which is rapidly fatal. The rupture of a cerebellar abscess may be followed by purulent inflammation about the pons and medulla, giving rise to retraction of the head, rigidity of the neck and slight opisthotonos.

Diagnosis. In cases of acute abscess following injury the symptoms are usually unmistakable, especially when they have lasted longer than a week. Cerebral symptoms in association with *ear disease* often present diagnostic difficulties. Severe pains radiating from the ear over the head, pyrexia, vomiting and attacks of rigidity, and even optic neuritis may depend on suppuration of the mastoid, apart from any brain complica-

tion; the presence, however, of optic neuritis, especially if well marked and associated with a slow pulse and with mental lethargy and somnolence would be in favour of abscess.

The symptoms of *suppurative meningitis*, which may also be set up by ear disease, are sometimes similar to those of abscess; moreover the two conditions may co-exist. Decided pyrexia would be in favour of meningitis, a subnormal or a normal temperature in favour of abscess; in suppurative meningitis the cerebro-spinal fluid escapes under increased pressure, it is purulent or semi-purulent and contains large polymorpho-nuclear cells as well as staphylococci, streptococci and pneumococci; in abscess the cerebro-spinal fluid may have a milky appearance, and may contain polymorpho-nuclear cells and micro-organisms, either both being present, or the one without the other. *Tuberculous meningitis* in children with chronic otorrhœa is distinguished from abscess by the earlier occurrence of mental apathy, by signs of irritability and by a greater tendency to convulsive seizures, to irregular palsies and to pyrexia.

In the differential diagnosis of *cerebral tumour* and cerebral abscess it is to be noted that as a rule the symptoms of tumour develop more slowly and insidiously than those of abscess, and that although remissions occur, the course of tumour is more steadily progressive whilst double optic neuritis occurs earlier and is usually more marked than in the case of abscess. Decided leucocytosis is in favour of abscess. In all doubtful cases it is of the greatest importance to make enquiries with regard to the previous occurrence of injury, of aural, nasal or more distant suppuration, or of any disease such as erysipelas or influenza, which might possibly have been a source of infection. The middle ear, the mastoid process, the nose and naso-pharynx should also be carefully examined in relation to the symptoms of intracranial disease.

Prognosis. If the presence and the situation of an

abscess can be diagnosed at an early period, and if its position is accessible to the surgeon, the prognosis in many cases is very hopeful. But if surgical interference is deemed inadvisable, the prognosis is very grave, a fatal result being almost certain. Occasionally, however, the contents of a stationary abscess become inspissated and its wall calcified; in such cases the outlook is better, and life may not be shortened, especially if the exciting cause of the abscess has been cured.

An operation may fail to save life owing to the presence of encephalitis, of sinus thrombosis or of pulmonary disease. Moreover, it may be found impossible satisfactorily to drain the abscess, or the operation, being too long delayed, exhausts the feeble resistance still remaining to the patient.

The treatment of abscess of the brain is mainly surgical. If the locality of the abscess can be accurately determined the sooner an attempt is made to evacuate the pus the better. But before trephining, especially when the diagnosis is uncertain, it is desirable to treat any local suppurative disease of the ear or of the accessory cranial sinuses. In addition to surgical measures, remedies may be required for the relief of pain and for the neutralisation, if possible, of the septic condition; careful attention must also be given to the nursing and appropriate feeding of the patient.

SECTION XVIII.

General Paralysis of the Insane: Dementia Paralytica

GENERAL PARALYSIS is characterised clinically by a series of mental and physical symptoms, which begin insidiously and steadily progress to profound mental deterioration and paralysis; and pathologically by a progressive degeneration of the cerebral cortex, the association neurons being mainly affected. The malady terminates fatally in two or three years.

Etiology. Although no period of life is exempt, the majority of the cases occur between the ages of thirty and fifty. The disease is more common in men than in women. A larger percentage of the cases is met with in the town than in the country. As a rule it is difficult to discover any tendencies to mental or to nervous diseases in the family history of the patient.

The chief etiological factor is syphilis, evidence of previous infection being obtainable in from seventy-five to eighty-five per cent. of general paralytics. Other proofs of the causal relation between the two diseases are:— (1) The marked lymphocytosis of the cerebro-spinal fluid, which obtains in the majority of cases of general paralysis. (2) The presence of Wassermann's reaction in about ninety per cent. of the cases (Mott). (3) The alleged inability to infect sufferers from the disease with syphilis. (4) The close relation between tabes and general paralysis, as shown by the co-existence of the two diseases in the same person, and by cases of conjugal tabes and conjugal general paralysis. (5) The occasional occurrence of the disease in children, when there is almost invariably either a history or clinical evidence of hereditary syphilis.

Mott believes that general paralysis occurs only in persons who have either acquired or inherited syphilis; other authorities maintain that syphilis, although a potent factor in the production of the disease is not

absolutely essential. The latter view receives some support from the observations on the cerebro-spinal fluid made by A. Ramsbottom and the author (see p. 335).

Ford Robertson and others believe that general paralysis is due to an invasion of the body by diphtheroid micro-organisms, which naturally will be specially prone to injure a nervous system, the resistance of which has been reduced by the effects of syphilis. Flashman and Latham hold that syphilis is the prime factor in the causation of general paralysis, but they are uncertain whether it alone is adequate to produce the disease, or whether it requires to be aided by the influence of non-syphilitic bacteria.

In many cases, excesses in alcohol or in venery, or prolonged mental strain and anxiety, appear to have acted as exciting causes of the disease. Very rarely, injuries to the head are followed by a train of symptoms which closely resemble those of general paralysis.

Symptoms. When the mental and physical signs are well developed the recognition of the disease is easy. Unequal pupils, tremor of the tongue and facial muscles, impaired articulation, loss of self-control, deterioration in conduct and character, and mental exaltation with progressive dementia form a combination of symptoms which cannot be misunderstood. The clinical picture, however, is not always so characteristic: both in the early and the advanced stages of the disease, difficulties in diagnosis may arise. In the early stages the symptoms may be those of neurasthenia, or of dyspepsia; sometimes the disease is ushered in by attacks of neuralgia, of migraine or of epilepsy. In the later stages the physical signs may be in abeyance, and the mental condition may simulate that of some other form of insanity. Moreover at any stage of the disease the characteristic manifestations may be complicated, or overshadowed, by the symptoms of alcoholism.

It is therefore desirable to consider (1) the individual symptoms and (2) the various types that the disease may

assume. The earliest manifestations may be restricted to disturbances either of the psychical or of the physical functions.

Psychical changes. At first the symptoms may be those of depression, indecision in action, instability of temper, and a variability in mood, together with sleeplessness, vertigo, a feeling of pressure on the top of the head and an easily produced fatigue both of mind and body. Sooner or later—it may not be for many months changes occur in the behaviour, character and intelligence of the patient. He may become unduly emotional, bursting into tears, laughing immoderately, or flying into a passion without any adequate cause. Formerly modest, courteous and moral, the patient becomes conceited, boastful and rude in his behaviour. He may give himself up to alcoholic or to sexual excesses; and drunkenness, thefts, or indecent exposure may get him into trouble and bring him to the police court. Self-control is reduced to a minimum and instinct rather than morality becomes the guide to conduct.

Periods of depression may be followed by, or may alternate with, periods of excitement, amounting in some cases to actual mania. Increased mental activity may show itself in acts of destruction, in wild speculations, or in purchasing quantities of goods for which the patient has no use.

Loss of memory and inability to fix the attention are frequently conspicuous. The patient forgets where he has put things, he neglects his business and family, fails to keep appointments and becomes unable to recall the names of his intimate friends, or even of the members of his own family. He also shows errors of judgment and defects in reasoning power, and may be incapable of doing a simple sum in arithmetic.

Delusions accompany the mental impairment, and usually are expansive in character. In hospital and private practice, they frequently relate to the physical condition; the patient has a look of self-satisfaction, and says that he never felt so well nor so strong in his

lite. Sometimes the delusion is one of grandeur as regards position, ability or wealth: the patient imagines himself to be a king or an emperor, a perfect linguist, or a champion runner; he boasts of his strength or his riches, and yet curiously enough he does not appear to be embarrassed when his statements are contradicted.

Less commonly there are delusions of persecution, or of a melancholic or a hypochondriacal character, many of which, however, are tinged with an expansive element; thus a patient may state that his debts amount to millions, or that his sufferings have lasted for hundreds of years. Sometimes he passes through a state of great anxiety and fear, during which he may have delusions as to the poisoning of his food, or may exhibit a tendency to suicide or to homicide. In other cases there is a belief that some part of the body is seriously deranged, as that the genital organs are diseased, that the abdomen is full of feces, or that the brain is too large for the skull. A striking feature of the delusions of general paralysis is that they are easily revealed by a brief conversation, and are readily modified by suggestion.

As the disease progresses dementia becomes more and more marked, or that in course of time the memory is abolished, and the patient passes into a condition of complete apathy; he is incapable of recognising his relatives and becomes oblivious to everything going on around him. Physically he gradually becomes a wreck of humanity; a tottering, feeble gait is followed by inability to stand or to sit, until ultimately the patient lies motionless in bed, with expressionless face, emaciated body and cold, livid, contracted limbs.

Physical signs. Changes in the pupil, in the action of the facial muscles and the tongue, together with disturbances in speech are particularly characteristic.

Pupillary changes occur early; they comprise irregularity in outline, inequality in size and shape, and impaired reaction to light. In many cases the pupils whilst giving no response to light, react to accommoda-

tion (the Argyll-Robertson phenomenon); occasionally the reaction to accommodation is also lost. Paralysis of the external ocular muscles may occur, but probably only when there are co-existent syphilitic lesions of the brain; transient weakness, however, of these muscles is not uncommon in uncomplicated general paralysis. Primary optic atrophy also occurs.

Fig. 206. Photograph illustrating a condition of marked exaltation and grandiose delirium; at the time the photograph was taken the patient was saying that his brain was geared up to 900,000. (Mott).

The expression of the face may be natural, or one of self-complacency, of depression or of excitement: sometimes there is a sudden change in the physiognomy, thus an expression of wearied depression may be rapidly replaced by one of wild excitement. Even in repose the face, if carefully observed, often shows spasmodic movements, as a sudden elevation of the eyebrow, or

twitchings or tremor of the lower facial muscles. Such movements are brought out or accentuated when the patient speaks, shows his teeth, or protrudes his tongue; and apart from actual spasm or tremor there may be evidence of inco-ordination in the associated actions of the muscles around the month. On being asked to protrude his tongue the patient complies in an exaggerated manner; the organ is jerked out and in, and these movements may be continued until the patient is told to desist.

The speech, even at an early stage, may be altered; at first there is simple hesitancy without any fault in articulation. The hesitation increases and the patient begins to have a difficulty in finding the right words; the first words of a sentence may be distinct and correct, the last words indistinct and slurred. Consonants, especially the linguals and dentals, are blurred; words or syllables are repeated, dropped out, or are imperfectly uttered. The defects in articulation are most conspicuous in the attempted utterance of certain test-words, such as artillery, territorial, parallelogram, or electricity.

A fine tremor affects the hands, and is often associated with inco-ordination of movement. These irregularities

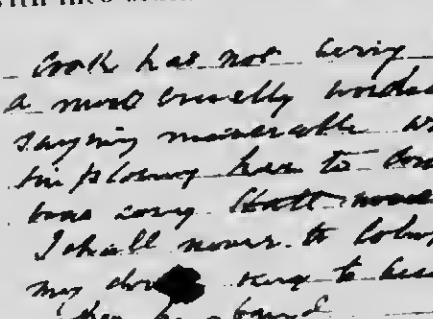


Fig. 207. Letter written by a General Paralytic.

in muscular contraction are shown in the patient's handwriting; letters and syllables are misplaced or omitted, while owing to defects in memory and ideation, mistakes may be made in the writing of the most ordinary words.

At first voluntary power is unaffected, but as the

disease progresses the muscles gradually become weak until eventually there is more or less complete paralysis. The condition of muscular tone varies; in many cases it is increased and is associated with exaggeration of the tendon reflexes; in tabetic cases there is hypotonus with loss of the knee-jerk.

Seizures, either apoplectic or epileptic in character, are apt to occur; they may precede the other symptoms of general paralysis by months or even years, but more frequently they occur during the developed disease. An apoplectic attack is characterised by a sudden disturbance of consciousness, varying in degree from simple stupor to profound coma. When consciousness returns it may be found that the patient is affected with monoplegia or with hemiplegia, and sometimes with aphasia. The paralysis, whether of speech or of limb, is usually transitory, lasting from a few days to a few weeks.

Epileptiform attacks may take the form either of local Jacksonian or of general epileptic seizures. In the Jacksonian seizures the fit often begins with conjugate deviation of the head and eyes, and is continued by clonic spasms of the face or the limbs on one side of the body. In the general seizures there may be isolated fits, or batches of fits leading to the status epilepticus; sometimes the attacks resemble those of minor epilepsy, and are followed by the post-epileptic automatism. During a severe seizure the face is flushed and the temperature is raised sometimes to hypopyrexia. It is probable, as suggested by Mott, that many epileptic seizures depend upon an associated toxæmia, such as that derived from tuberculosis, pneumonia, cystitis, a loaded rectum or from bed-sores.

Course and prognosis. General paralysis begins insidiously, progresses steadily and ends fatally, recovery in a genuine case probably never occurring. The average duration of the disease is about three years; life is rarely maintained for more than five years, and frequently comes to an end in a year or two. Some

cases run a course of only a few months, death occurring either from the intensity of the mental changes, or from some intercurrent complication. A rapid downward progress is often determined by the frequent occurrence of epileptiform seizures.

Sometimes the progress of the disease appears to be arrested: improvement may occur and occasionally to such a marked degree that the doctor is blamed for having put the patient under control. During the remission, which may last for a year, the only signs of the disease may be slight tremor of the face or the hands, sluggish pupils and an abnormal knee-jerk. Remissions and variations in individual symptoms are also observed: thus inequality of the pupils may disappear, or exaggerated knee-jerks may be replaced by responses which are normal or diminished.

Clinical types. (1) The *exalted type*. In this form every variation may be observed between exaltation with grandiose ideas and acute maniacal excitement; occasionally the condition resembles that of acute delirious mania.

(2) The *depressed type*, which often begins with symptoms of neurasthenia and is characterised by the presence of melancholic, or of hypochondriacal delusions.

(3) The *demented type*, in which there is progressive mental deterioration without any pronounced exaltation or depression. Sometimes the patient, recognising the impairment of his memory, and the lowering of his capacity for work, expresses a wish to enter an asylum as a voluntary boarder.

(4) The *tabo-paretic type*, in which the symptoms of general paralysis are preceded by those of tabes. Attacks of pain may be insanely interpreted, thus the patient may believe that he is being tortured by unseen agencies, by electricity, or by poison.

(5) The *convulsive type*. In this variety convulsions are the most prominent features of the disease. Each batch of fits is followed by an increase in the impair-

ment of both mind and body, the disease running a rapidly fatal course.

Much rarer types are the *stuporose*, in which mentation is at a minimum, and stupor gradually passes into complete dementia; and the *circular*, in which periods of excitement alternate with periods of depression.

Pathology. In a case of advanced general paralysis the following post mortem appearances may be observed. There is considerable emaciation of the body and limbs; the viscera and the whole nervous system are also wasted. Visceral gummata, or signs of tertiary syphilis on the surface of the body are rarely present. The lungs may show signs of pneumonia, gangrene, or of tuberculosis. The aorta is often atheromatous, and in a condition of nodular fibrosis. The skull-cap is thickened, and its density contrasts with the fragility of the ribs which in many cases can be easily broken. The dura mater is adherent to the skull; on opening it a considerable quantity of cerebro-spinal fluid escapes. Frequently, and especially when the patient has suffered from epileptiform seizures, hæmorrhagic pachymeningitis is present. The pia-arachnoid is opaque and thickened, and in stripping it from the brain, portions of the cortex are torn away with it. The whole brain is wasted and shrunk, and its weight is much diminished. The frontal and central convolutions are particularly wasted; their sulci are wide and deep and their grey matter is much reduced in thickness; it is over these regions that the pia-arachnoid shows the greatest thickening. There is usually a great excess of the cerebro-spinal fluid in the ventricles as well as in the subdural space; the ependyma of the ventricles often presents a granular appearance.

A microscopical examination of the brain reveals changes in its membranes, vessels and nervous elements. The pia-mater is infiltrated with plasma cells and lymphocytes, and many of its vessels are degenerated. The vascularity of the cortical grey matter is increased, and there may be evidence of a new formation of capil-

laries. The cortical arterioles show proliferation of the endothelium, degeneration of the muscular coat, pro-

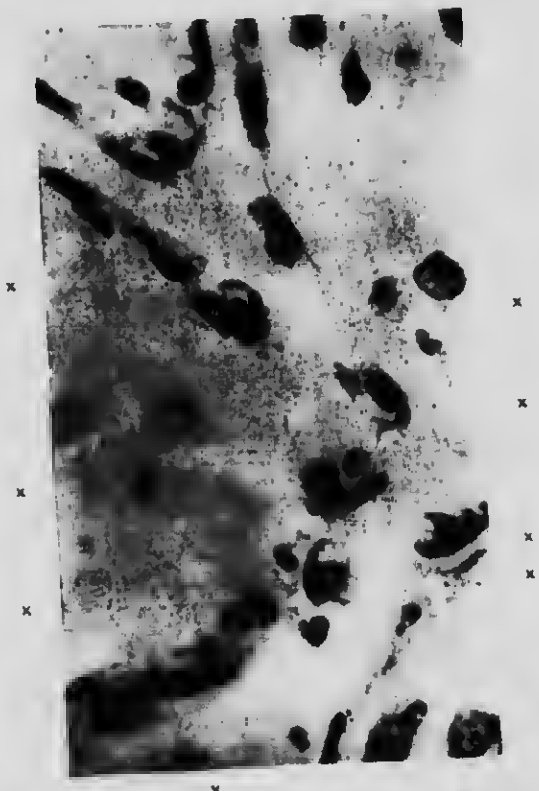


Fig. 208. A small Cortical Vessel in the Occipital Lobe of a General Paralytic, showing Typical Plasma Cells (×) upon it. Note the oblong, angular shape with a clear space in the cytoplasm and the laterally-situated nucleus with its darkly stained chromatin bodies (× 800). (Stoddart and John Turner).

liferation of the perivascular nuclei, and cell-infiltration of the adventitial lymph spaces. Changes in the neuroglia are also conspicuous; its cells and fibres are increased in number, and many of the cells are large and have a characteristic spider-like appearance. Very noticeable too are the changes in the pyramidal cells of the cortex; many of them are totally destroyed, whilst others are altered in shape, with their branches broken or atrophied, and present the various stages of chromatolysis. The association neurons, especially those of the

frontal lobe, are severely and extensively affected. Similar degenerative changes in the vessels and neurons are to be found in the basal ganglia, the cerebellum, the

Fig. 209. A Glia or Spider Cell from the Cortex of a General Paralytic's Brain. The stout, vascular fibres ending in fan-like expansions by which they are attached to the walls of the blood-vessels are well shown. A film preparation (× 600). (Stoddart and John Turner).

nuclei of the oculo-motor nerves, and in the pons, medulla and spinal cord. In the cord, the pyramidal tracts and the posterior columns are the systems which most frequently show degeneration.

Treatment. In all cases of general paralysis the patient should live a quiet life in the country, being placed under suitable care and strict supervision. If the mental changes are marked, the sooner the patient is certified and sent to an asylum the better it will be both for himself and his friends. In the early stages of the disease, especially in cases which have not been previously treated for syphilis, an energetic course of mercury and iodide of potassium is to be recommended.

Townsend claims to have obtained good results in general paralysis by the daily administration of urotropine, Ford Robertson by the use of an antiparalytic serum.

The treatment of the convulsive seizures is that of ordinary epilepsy (see p. 498). Mott believes that a

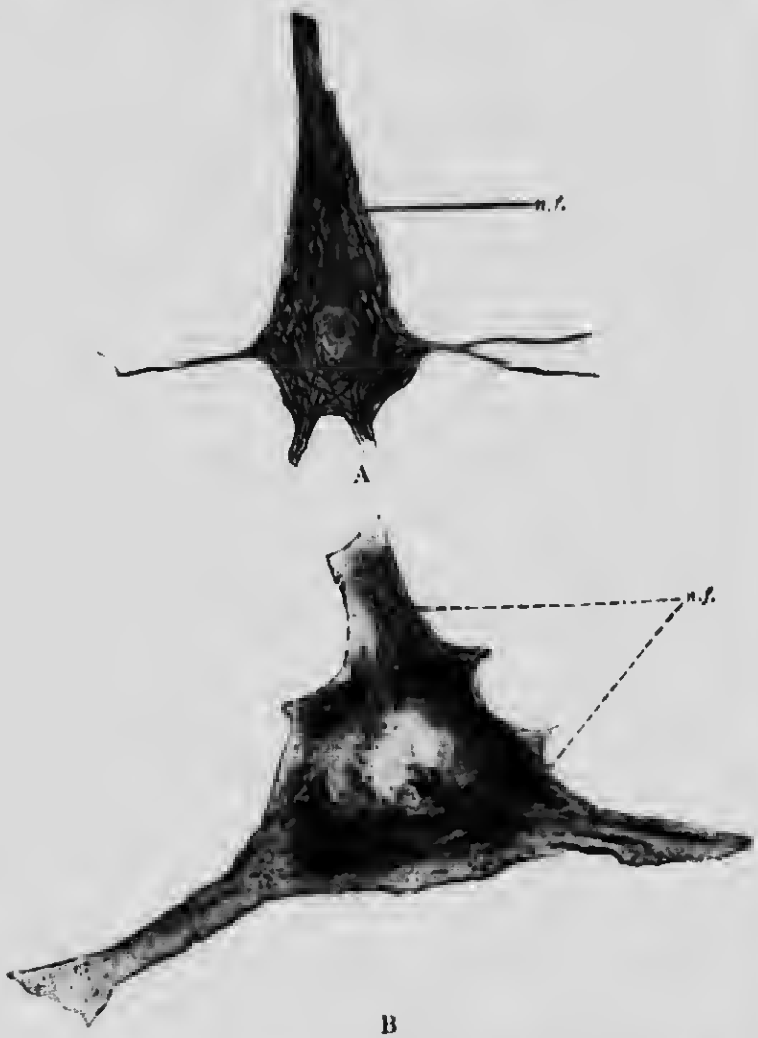


Fig. 210. Two cells stained by the Cajal method: (A) a normal Betz cell showing the nerve-fibrils (n.f.) which form the conductile structures that pass from the dendrites through the cells; (B) a Betz cell in general paralysis which has undergone degeneration and decay, and the nerve fibrils (n.f.) are broken up and destroyed. Magnification 500 diameters. (Mott).

loaded bowel, which causes portal congestion and fæcal intoxication, is one of the most potent causes of congestive and epileptiform seizures, and therefore advises that all paralytics should take some kind of purgative twice a week.

Mental excitement may be lessened by the use of warm baths, and by the occasional employment of paraldehyde, or of sulphonal; sometimes the hypodermic injection of one-hundredth of a grain of hyoscine is beneficial.

When the patient becomes bedridden every care must be taken to prevent the formation of bedsores, and distension of the bladder; retention of urine requires the regular use of a sterilized catheter.

SECTION XIX.

Neurasthenia.

NEURASTHENIA is a disorder of the nervous system, unassociated with any evidence of organic changes, which gives rise to disabilities both as regards mental and physical work, to disturbance of sleep, and to a train of subjective sensory symptoms, accompanied by much morbid introspection.

Etiology. The disease occurs more frequently in males than in females, and between the ages of twenty-five and fifty than at other periods of life.

The most important antecedent is prolonged mental strain, especially when this is associated with worry, disappointment or anxiety. Sometimes the symptoms have developed after a sudden shock, as that caused by the hearing of bad news, or the witnessing of a catastrophe. Railway and other accidents in which a physical jar, with or without injury to the body, is accompanied by emotional shock, are often followed by a definite group of symptoms known as "traumatic neurasthenia."

Acute febrile illnesses are responsible for many cases of neurasthenia; influenza, even a mild attack, is especially prone to set up the disease. Sometimes the symptoms of neurasthenia are the only manifestations of the early stages of serious disease, such as cancer, tubercle, general paralysis, kidney disease or myasthenia gravis. Digestive troubles are at the root of many neurasthenic conditions; anæmia, excessive child-bearing, prolonged lactation or any other condition which lowers the general vitality is liable to bring on the disease. The excessive use of alcohol or tobacco leads to nervous exhaustion, but of all toxic agents morphia and cocaine are the most potent in the production of neurasthenia.

In many cases no adequate cause can be discovered,

the symptoms beginning in a person whose health and surroundings appear to be quite satisfactory: thus I have known the disease to develop in a robust farm labourer who had not been subjected to any form of mental disturbance. In such cases we have to fall back for an explanation on some inherent predisposition to neurosis, and there can be no doubt that heredity plays a part in the causation of the disease. For although it is unusual to obtain a family history of serious nervous disease, such as epilepsy or insanity, it is common to find that the parents had a weak constitution or had suffered from headache, nervousness, or other evidence of nervous instability. Probably in the majority of cases the patient is born with less than normal resistance to emotional, intellectual, or physical stress. Sometimes there is reason to believe that the higher centres of the brain which are affected in this disorder have been attacked by a toxic agent either introduced from without, or developed within, the body as the result of some faulty metabolism.

Symptoms. The symptoms of neurasthenia vary much both as regards character and degree of severity. In many cases they are chiefly psychical, in others they indicate disturbance of motor, sensory or visceral functions. But whatever their character they are mainly subjective, and they closely resemble the temporary sensations experienced by healthy persons who have undergone great fatigue either of body or of mind.

Sometimes the physical condition appears to be good, the patient looks robust and may be stout and ruddy; more frequently he is sparely built and of pale or sallow complexion, and very often there is an appearance of abated vigour and of mental depression. Occasionally there is obvious emaciation.

Signs of *mental disturbances* are usually prominent. In manner the patient may be quiet and morose, or restless and fidgety. Restlessness is a common feature and is frequently accompanied by an uncontrollable tendency to discuss symptoms in detail. Such a patient

will often bring a long written account of his state of health, and of the various ailments from which he has suffered since infancy, and then after a protracted interview with the doctor, he will write to say that owing to the hurried nature of the conference he would like to call again in order to explain matters more fully.

The chief manifestations of the exhausted, weary brain are incapacity for work requiring mental effort, morbid fears and various cephalic sensations. An explanation of the incapacity is to be found in weakened powers of volition and attention and to some extent in deficient memory. The stimulus of unusual excitement may enable the patient to get through a spell of hard work, but the effort is of short duration and is followed by profound exhaustion.

Apart from unusual stimulus work is ill-done and even its commencement may bring on a sense of fatigue. The patient is unable to concentrate his thoughts, he reads without understanding, he writes with difficulty, words not coming to him so readily as formerly, business becomes distressing and he feels unable to attempt the slightest task. Very often his memory is impaired, he finds it difficult to recall the names of persons well known to him, he forgets the names of familiar objects and in conversation may appear unable to complete a simple sentence. Such a condition, however, may be compatible with a power to grasp as well as formerly any abstruse problem presented to him.

Certain morbid fears are prone to occur. Some are indefinite in character, as when a patient is haunted by a vague sense of impending calamity. Others assume a more definite form; there may be a dread of becoming insane, or of becoming poor even when such a fear is quite groundless. Some patients suffer from a dread of not falling asleep at night, which may be so overpowering that sitting in a chair the whole night is preferred to going to bed. Some fear to be alone, others to be in company. Some dread being in open places

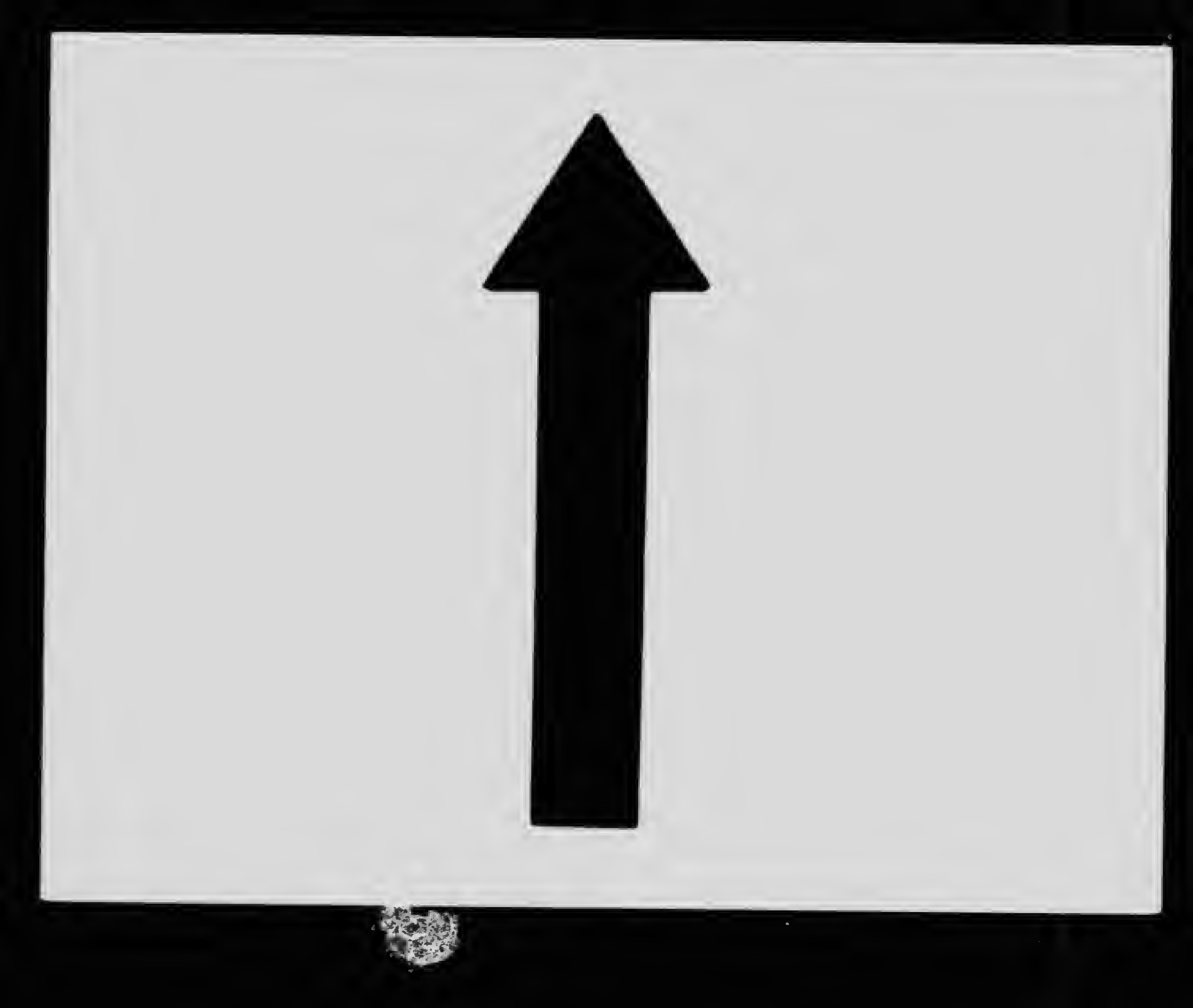
agoraphobia; others being in enclosed places such as a narrow street between tall houses—*claustrophobia*. Associated with these mental perversions there is often a sense of unreality, the patient feels as if in a dream. Sudden dazed feelings, sinking sensations, or actual vertigo may be experienced.

Headache is common and usually is most severe on the vertex or the occiput, the pain often extending down the back of the neck. But more prominent than pain are curious cephalic sensations. The head feels heavy, or too full or empty; it feels as if there were a tight band round it, or as if it were too light; sometimes a patient refers curious sensations to the centre of the brain, he may say that his head feels as it were about to open.

Insomnia is an almost constant symptom. A patient may fall asleep soon after going to bed and awake with a start in the early morning, or he may lie awake for hours and then drop into a troubled sleep. Sometimes the amount of sleep is small, though as a rule it is greater than the patient believes to be the case. It is apt to be disturbed by dreams of a disagreeable character, and the patient often rises unrefreshed and suffering from a dazed aching head.

The *special senses* are frequently affected. The patient may complain of buzzing or of other abnormal sensations in the ears; or of spots before the eyes and of spectral illusions. Sometimes there is undue sensitiveness to bright lights or to noises, which make the patient irritable, and tend to increase e.g. headache. Vision may be dimmed owing either to feebleness of accommodation, or to speedy exhaustion of the visual nervous mechanism: the visual field is of normal extent but as the retina is easily fatigued, the perimetrical examination should be made quickly, otherwise perception fails and the fields appear to be contracted.

The *motor system*. The muscles are flabby and wanting in tone and soon tire; there is an incapacity for much muscular exertion, an inability to stand long



or to walk far. The patient often complains of weakness in the legs, but no real paralysis can be detected. A fine tremor of the hands is common, and occasionally slight twitchings are observed in some of the facial muscles. Jerkings of the limbs, as the patient is about to fall asleep, are often particularly annoying.

The superficial *reflexes* are present though often difficult to elicit, sometimes they are exaggerated; the plantar reflexes are always flexor in type. The knee- and wrist-jerks are usually increased; true ankle clonus does not occur.

Sensory system. In addition to headache, pains in the back and limbs are frequently experienced. The backache is aggravated by exertion, and is often associated with considerable spinal tenderness, or with deep-seated burning or sore sensations. The patient may complain of numbness, tingling and formication in the limbs, and sometimes of intense itching; there may be considerable cutaneous hyperæsthesia.

Gastro-intestinal disturbances. The appetite is often capricious; it may be excessive or diminished. The patient may say that he is never really hungry or that he never feels as if his stomach were quite empty. Digestion is often slow and associated with flatulence, or with a feeling of distension, or of sinking in the epigastrium. In some cases the secretion of hydrochloric acid is increased. As a rule the bowels are constipated; sometimes the colon is distended with gases. Membranous colitis is occasionally present.

Circulatory disturbances. Loss of vaso-motor tone is indicated by cold hands and feet. Blushing is easily produced by slight physical or mental disturbance. Profuse sweating on the least exertion is often a distressing symptom. Cardiac palpitation is common, and is rapidly started by muscular or mental efforts. The patient is often conscious of the beating of his heart and arteries; he may complain of pumping sensations through the blood. Pseudo-anginal attacks are apt to occur. The pulse, usually low in tension, tends to be

increased in frequency and during attacks of palpitation may reach 120 or 140.

Sexual disorders. It is rare for the male neurasthenic not to allude to the condition or functions of his genital organs; the varieties of sensations and perversions are innumerable and are apt to be described in detail without shame or reticence. Common complaints are feelings of weight or of dragging in the pelvis, of throbbing, aching, coldness or numbness about the penis and testicles. Sometimes the testicles are unusually tender and are stated to swell at times.

Lessened sexual power is a frequent symptom; incompetence in the act of coition due either to imperfect erection, to premature ejaculation of semen, or to other abnormality may be associated with frequent nocturnal emissions. In both sexes the sexual appetite may be much diminished. Even when coitus is normally performed and not too frequently repeated, it is often followed by great prostration, disturbed sleep and fidgety sensations in the legs. In this way the mental equilibrium, already unstable as regards the expenditure and repair of nervous force, tends to become still further disturbed.

Micturition is often unusually frequent, although the quantity of urine is not increased. In some cases the urine is pale, of low specific gravity and of neutral or alkaline reaction. In others it is scanty and concentrated and loaded with lithates; uric acid, indican, earthy phosphates and oxalates may be present in considerable quantities.

Pathology. The exact changes underlying neurasthenia are unknown. In all probability the disorder depends on molecular changes in certain groups of cortical cells, changes which are closely allied to those that have been shown to follow great fatigue or prolonged experimental stimulation. In many cases these changes, owing to inherited weakness of nerve tissue, are easily initiated even by a slight strain or by other adverse influence. The molecular changes following healthy

fatigue are transitory, and so too they may be in some cases of neurasthenia: but if frequently produced or of long duration they may lead to an increase, with imperfect removal, of the poisonous products of tissue metabolism. Such products may even set up sclerosis of the surrounding tissue: this is a possible explanation of the severity and persistence of symptoms in some cases of neurasthenia.

Diagnosis. As a rule simple neurasthenia is easily recognised, but it must be remembered that neurasthenic symptoms may be set up by serious disease in any part of the body; hence in all cases it is of paramount importance to make a thorough examination of the patient, and to repeat such examination at intervals, before an absolute diagnosis is made.

The early stages of cerebral tumour and of general paralysis are occasionally represented only by neurasthenic symptoms, and even when the possibility of these diseases is before one's mind the diagnosis may be very difficult. This is particularly the case with *general paralysis*. Defects in articulation and much impairment of memory are not common in neurasthenia, but when present and associated with slight twitching of the facial muscles and with exaggerated knee-jerks it may be impossible for a time to exclude general paralysis. It is to be noted that the neurasthenic dwells on his symptoms and describes them minutely, whereas the general paralytic is often unaware that he is ill, and talks rather of his successes and projects than about his state of health. In a doubtful case it is desirable to make a lumbar puncture and examine the centrifuged deposit of the cerebro-spinal fluid, when if there is a marked lymphocytosis general paralysis may be diagnosed with assurance.

On the other hand it is possible to diagnose serious disease when neurasthenia is alone present. Thus exaggerated knee-jerks and a shuffling gait might be taken as evidence of lateral sclerosis, but this ought to be excluded if ankle clonus and Babinski's reflex are

not obtained. Again in some cases of neurasthenia the knee-jerk is not obtained until the reinforcement method is employed; this might raise a suspicion of tabes.

Between hysteria, neurasthenia and hypochondriasis no sharp boundary lines can be drawn: the characteristic features of typical cases must be looked for.

In *hysteria* we meet with contractures, definite areas of anæsthesia, concentric contraction of the visual fields, globus and convulsive attacks. These symptoms do not occur in neurasthenia.

Hypochondriasis implies a fixed conviction that certain visceral sensations indicate disease of certain organs; it tends to pass into melancholia in which the patient suffers from actual delusions regarding his physical condition.

The muscular weakness and the feelings of fatigue which occur in neurasthenia are present also in *myasthenia gravis*, but in the latter disorder muscular fatigue is much more rapidly produced, and when the disease is established we expect to find partial ptosis, paresis of the palate, the myasthenic reaction and other characteristic symptoms.

With regard to *traumatic neurasthenia* it is important to distinguish cases in which the condition is simply the result of mental shock and is therefore similar to that of non-traumatic neurasthenia, from cases in which an injury to the head has produced definite cortical lesions, such as minute lacerations, punctiform hæmorrhages and other changes which collectively constitute contusion of brain tissue. Now although the symptoms of a bruised brain may closely resemble those of a severe and advanced case of neurasthenia certain differences may be usually observed. Instead of an uncontrollable tendency to discuss their symptoms in detail, sufferers from a bruised brain are dull and apathetic, and show a tendency to drowsiness and stupor rather than to insomnia. The neurasthenic is able to commence work and to do it accurately for a time, but he soon tires, he lacks concentration and exhibits indecision in all his

actions. But the patient who has a bruised brain is unfit to follow any employment, he is too apathetic as a rule to show signs of worry or anxiety, and when his cortical lesion is severe and extensive, apathy may be associated with profound loss of memory and other signs of mental deterioration.

Prognosis. Neurasthenia is a chronic malady, the duration of which is to be measured by months or years. Its severity and persistency vary much in different cases and a full consideration of every circumstance connected with the patient is necessary before even a probable prognosis can be given.

The outlook is favourable when no hereditary tendencies to neurosis can be traced, when the previous health of the patient has been good, and when the onset of the attack is acute and can be assigned to a definite and removable cause. The outlook is unfavourable, when the disease is of long standing, when it begins in childhood or in advanced life, and when the patient's circumstances do not allow of prolonged treatment, involving a long absence from home and business.

Neurasthenia does not shorten life, but it does much to add to its miseries, and to lessen its achievements. On the other hand an absolute cure is sometimes obtained, and in every case one may promise periods of partial or of complete recovery.

Treatment. The first essential in the treatment of neurasthenia is to ascertain by means of a thorough examination of the patient whether there is any source of peripheral irritation or of toxic infection, which may have a causal relation to the condition. For although it is rare for neurasthenia to depend solely on such a cause, its symptoms may be aggravated thereby. The condition of the eyes, the teeth, the mouth, and the naso-pharynx must be investigated and any defect or morbid condition removed or corrected. Some form of dyspepsia may require treatment, or there may be a movable kidney which it is desirable to support or fix,

or disease of the uterus or ovaries which requires some operative procedure.

The chief cause of neurasthenia, however, is usually a psychical shock or strain as a result of grief, anxiety or worry. To remove such a cause may be impossible, also the patient may find it difficult to spare the time necessary for treatment. Nevertheless in all well-marked cases, complete rest from work mental and physical—must be insisted on. The duration of this rest and the plan of treatment adopted will vary with each case. The mildest forms of the disease are often cured by a short restful holiday with change of scene, or even by giving up two or three afternoons a week to golf or other moderate exercise. But in severe cases of long standing, a prolonged rest must be secured, which it is well to combine with the other factors which collectively constitute the "rest cure," or the treatment originally advocated by Weir-Mitchell. This consists in complete isolation of the patient from all relatives and friends, rest in bed, massage, electricity and abundant nourishment. The treatment is best carried out in a nursing home and should be continued for a month or six weeks or longer according to the needs of the case. The psychological influence of the nurse is great; it is therefore important to select one who is known to be observant, tactful, and kind as well as firm. Massage alone or combined with faradism is of great service; it takes the place of exercise and enables the patient to digest and assimilate a large quantity of food. In many cases massage should be performed for an hour twice daily, but its duration and intensity must be regulated for each patient. In addition to ordinary meals, half a pint of milk every three hours is often well taken.

The benefit derived from a course of the Weir-Mitchell treatment, carried out with care and suitably modified for different cases, is generally very great. Under its influence sleep returns, nerve tone is restored, and the patient loses the symptoms which previously

had so greatly distressed him; frequently a complete cure is obtained.

It is however always desirable for the treatment to be followed by a few weeks' residence near the sea or in a dry hilly country. The sea-side is more suitable for the winter months, a mountainous region for the summer. As a rule sea air is less beneficial than mountain air, and may be harmful to dyspeptic and bilious subjects. Sea voyages and much travelling, involving long railway journeys, are not to be recommended.

Many cases of neurasthenia are benefited by a course of hydro-therapeutics in the form of wet sheets, packs, sitz baths and spinal douches; the various applications being selected and graduated according to the condition of the patient. As an adjunct to such methods the faradic bath used daily for about twenty minutes, with the current of sufficient strength to produce tingling of the skin will often lessen the nervousness, improve the appetite and lead to sleep. The high frequency current is also deserving of trial, when other methods have failed to give relief; it is said to be particularly effective in the gastro-intestinal type of the disease.

Treatment by drugs is quite subsidiary to the above methods. As a general rule tonics are the most useful, arsenic and iron being especially valuable in cases associated with anæmia. Arsenic also is often of great benefit when there is gastralgia, and in combination with strychnine when there is disturbance of the cardio-vascular system. In some cases the glycerophosphates of sodium and lime are of service. The formates also have been recommended; forty to fifty grains of sodium formate may be taken daily in divided doses. For constipation, abdominal massage usually suffices. Purgatives may do harm, but a laxative—senna, cascara, aloes and belladonna—is frequently beneficial.

Valerian, in the form of the ammoniated tincture or in some of the concentrated forms known as bornyval, gynoval and valyl, is said to give relief in cases presenting symptoms of cerebral disturbance, such as

vertigo, impaired memory and mental confusion; and also in cases with various visceral discomforts. Nitroglycerine too is sometimes effectual in clearing away the cloud which appears to envelope the neurasthenic's brain.

Before prescribing hypnotics all other means of producing sleep must be tried, and the doctor must satisfy himself that the sleep is as defective as is stated. A warm bath or half an hour's "pack" at bedtime often promotes sleep; when the patient awakes in the early morning hours a little food will frequently induce sleep. Should these remedies fail and insomnia become obstinate, small doses of veronal, trional or sulphonal may be administered. In some cases nothing acts better than the bromides, either given alone, or in combination with chloral.

SECTION XX.

Hysteria.

Hysteria is a psychical disorder, which often leads to disturbance of the lower centres of the brain and spinal cord and of the sympathetic system. The field of consciousness appears to be limited so that certain impressions are excluded, are not admitted to the patient's own personality; thus a patient with a paralysed limb may have no perception of its existence; another patient may show a failure of memory limited to some particular event or person. There may be a complete dissociation of the mental processes, as in the condition known as dual personality, in which the patient lives in two different states; in the one state she is natural, in the other abnormal, and in neither state has she any recollection of what occurred in the other. Hysteria is expressed clinically by the most curious and varied symptoms; these often closely resemble the manifestations produced by actual lesions in the nervous centres and in many cases the diagnosis is very difficult. The exact nature of the cortical disturbance is unknown, but from the readiness with which it appears to be brought about in certain individuals an underlying neuronic instability may be assumed, an instability which is more frequently inherited than acquired.

Etiology. Heredity is undoubtedly a most important factor; it may be shown either directly in the transmission of emotional instability, or indirectly by the existence of epilepsy, insanity or of other neuroses in the family history of the patient. This being the case it is not surprising to find that hysteria occurs more frequently and in severer forms amongst the Latin, Slavonic and Jewish than amongst the more phlegmatic Anglo-Saxon races.

Hysteria occurs most frequently between the ages of

puberty and thirty and especially between fifteen and twenty. Occasionally it is met with in young children, and in women at the menopause. Females are much more commonly affected than males; in males boys are more subject to the disorder than adult men.

The cortical instability which predisposes to hysteria being for the most part inherited, its degree necessarily varies much in different cases so that every transition may be assumed between a neuronic system that is nearly normal and one that is disturbed by the slightest cause. When the congenital instability of the cerebral cortex is slight, the most adverse circumstances may be required to produce hysteria, but when it is considerable the slightest change in the environment may induce an attack. Bearing this in mind the influence of the following exciting causes must be considered in relation to the previous psychical condition of the patient. The most frequent antecedent is some mental shock: this may be a sudden fright, the witnessing of a bad accident, the reception of bad news, disappointment in love or the prolonged anxiety and strain associated with the nursing of a sick relative especially if the illness ends fatally and leads to much emotional shock.

Injury sometimes initiates an attack, especially if the accident, as in a railway collision, is one to create great alarm. The injury itself may be quite insignificant—it is the associated fright that is the potent factor. Traumatic hysteria, commoner in men than in women, is often severe in type.

In a predisposed person emotional mental shocks are prone to set up hysteria when the health has been lowered from any cause, as by unfavourable hygienic conditions, an acute or a protracted illness such as influenza, tuberculosis or syphilis; or poisoning by alcohol, lead or other toxic agent. In many of these cases a psychic impression can be traced, thus a patient who is conscious that he has become the victim of a poison is naturally alarmed and apprehensive.

Diseases of the generative organs have long been

associated with hysterical symptoms; their influence has doubtless been exaggerated, yet it is true that they often cause more anxiety than disease of other organs.

Lastly it is of the greatest importance to remember that the development of lesions in the brain or the spinal cord are apt to set up hysterical manifestations or to exaggerate those already present; for example, such manifestations may completely mask the early symptoms of disseminated sclerosis, of cerebral meningitis, or of intracranial tumour.

Symptoms. The manifestations of hysteria are numerous and their combinations infinite in variety, hence no single clinical picture can adequately depict the symptomatology of this disease.

The underlying cortical instability may be conspicuously expressed by morbid self-consciousness, by emotional and other psychical disorders; or it may be latent and inconspicuous, the manifestations of its presence being indirect, and apparently related to disturbances of lower centres, as shown by paralysis, anæsthesia and other local phenomena. Thus on the one hand many typical hysterical subjects pass through life without ever exhibiting any definite local hysterical signs, or "stigmata"; and on the other hand it is not uncommon to find a limited anæsthesia, or a contracted limb, unassociated with any obvious mental disturbance, when the diagnosis of hysteria has to be based on the peculiarities presented by the sensory or the motor symptoms. But in most cases there is ample evidence as to the psychical nature of the malady.

The symptoms of hysteria may be broadly grouped into those which characterise the convulsive or paroxysmal attacks, and those which occur between such attacks or in their absence. Of the latter the psychical, including the general mental state of the patient, are the most important and therefore the first to be mentioned.

Psychical Symptoms. The mental condition of the hysterical patient varies with the peculiarities of the individual character, but common characteristics are a

lack of self-control, a deficient power to resist the calls of inclination and to inhibit the emotions. The will power appears to be lessened, yet sometimes the greatest strength of will is displayed as when the patient carries out some fixed purpose, the accomplishment of which is difficult. Such an effort, however, is not frequently repeated as the will soon loses its resisting power and becomes dominated by emotional impulses, and by a desire for sympathy and attention; these whilst gratifying to the patient steadily increase her self-consciousness and her dependence on the moral strength of her friends. The mind is not necessarily feeble, indeed the intelligence is often clear and sharp; in conversation the patient tends to be very loquacious especially as regards the description of her abnormal sensations and ailments. Nor is the memory usually affected, though in some cases gaps may be observed; thus it may be limited to past events, or to certain periods of time, the occurrences of other periods being forgotten owing to the greatness of introspection at the time. The tendency to dwell on her sufferings leads to their exaggeration and sometimes to actual shamming; certainly the line between malingering and true hysteria is often difficult to draw.

The conception of a symptom either by hearing of it, or by seeing it in another person may lead to its occurrence, thus the idea of a helpless limb enters the mind and the patient becomes unable to move one of her own limbs, or the thought of a definite pain leads to its presence. Again the influence of undue solicitude on the part of friends is often pernicious, thus if they speak of a poor appetite the patient may try to live without food, if the doctor remarks that the urine is scanty, the patient may affect that the secretion has stopped. In some cases profound depression is a marked feature, in others there is a tendency to outbursts of great excitement which may culminate in hysterical mania; sometimes the patient falls into a trance and exhibits tendencies to somnambulism.

Sensory Symptoms. In hysteria every variety of sensory disturbance—subjective and objective—is met with. Pains are readily produced; they may occur in any part of the body, and often correspond in position with tenderness on pressure either superficial or deep. In the head pain may occupy any position; most frequently it is vertical and of a fixed boring character as if a nail were being driven in, hence the term "clavus hystericus." Spontaneous pains in the spine, the joints, the inframammary region or in other parts are also common. Pain down the spine may be associated with tenderness to both light and deep pressure; sometimes the condition simulates that of caries or of malignant disease of the vertebrae. Pain about the hip, intensified by movement, together with tenderness on palpation and contraction of the surrounding muscles, may suggest disease of the joint; this, however, is excluded by measurement of the limb, which shows that there is no real shortening, and by an x-ray examination.

Sometimes the patient complains of numbness and tingling and creeping sensations in the limbs or of a feeling down the spine as if cold water were trickling along it. One of the commonest subjective sensations is the "globus hystericus," that is a feeling of something rising in, or constricting the throat; it is often attended by a difficulty in breathing or by a sudden sense of suffocation. Hyperæsthesia affecting the whole body is rarely met with, but hyperæsthesia limited to certain areas on the thorax and abdomen is common, and occasionally it affects one half of the body. The pain may be produced by the lightest touch or only by deep pressure. Deep seated tenderness is often present especially in certain localities, as over the dorsal spines or in the inframammary region and is particularly common and characteristic in the ovarian region. Pressure there or indeed over hyperæsthetic spots in any part of the body may cause great distress and give rise to fainting, to globus or even to convulsive attacks, hence these hyperæsthetic areas have been called "hystero-

genic." Sometimes pressure, or the faradic current, applied to these areas will arrest an attack. In traumatic hysteria the seat of a slight injury may become excessively tender.

The *special senses* may be unduly sensitive; there is intolerance of light whilst smell and hearing may be preternaturally acute: a patient for example may hear conversation at a distance inaudible to healthy persons, or perceive sounds so keenly as to have much mental suffering.

Anæsthesia, a common symptom, may be easily overlooked, for the patient seldom complains of it, often because she does not know of its existence. As a rule it is complete, all forms of sensation being affected; sometimes it is partial, sensation being lost to either pain or touch, or whilst lost to pain is only diminished to touch. In rare instances the whole body is anæs-

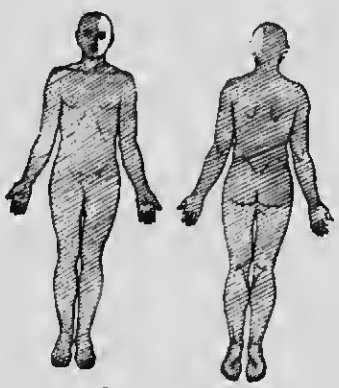


Fig. 211.

thetic (see fig. 211), but usually only one half of the body, or some segment of a limb is affected. A peculiarity of the anæsthesia is that it does not correspond in distribution to that of nerve roots or of spinal segments (see fig. 212); thus when the hand or foot is affected and the anæsthesia extends for a variable distance up the limb, a sharp line of demarcation can be drawn between the sensitive and insensitive skin, a condition named "glove or stocking anæsthesia."

In hysterical hemianæsthesia, which occurs most frequently on the left side, every form of sensation in the skin, the accessible mucous membranes, the muscles, bones and other deep structures is more or less com-

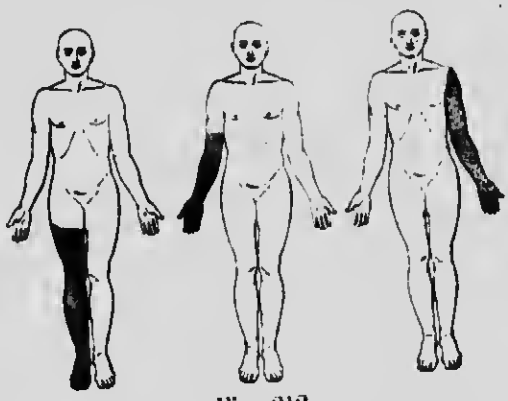


Fig. 212.

pletely lost, the loss being sharply bounded by the middle line of the body (see fig. 213). The special senses are likewise implicated; the sense of taste is lost on the affected side, that of smell on both sides; hearing

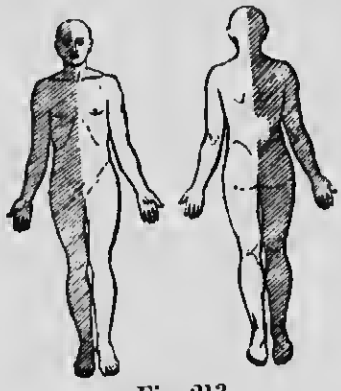


Fig. 213.

is impaired, whilst that on the other side is often abnormally acute. Sight is also impaired; the most common disorder is restriction of the field of vision, especially for blue, the eye on the anæsthetic side being most markedly affected—"crossed amblyopia." Occasionally taste is lost on both sides, a condition

practically unknown except in hysteria. A striking peculiarity of hysterical anæsthesia is that in some cases when the patients are blindfolded and told to say "Yes" when they feel, and "No" when they do not feel, they promptly say "Yes" when normal parts, and "No" when anæsthetic parts are touched.

The muscular sense is often preserved, so that anæsthetic fingers can be used for needlework, even when the eyes are closed. Other peculiarities are:—That the degree of anæsthesia may vary from day to day, that it tends to increase after an examination of the sensibility, that it may disappear during sleep and that ovarian tenderness persists on the affected side. Sometimes the anæsthesia lasts for years, and it may disappear quite suddenly.

The anæsthesia may pass from one side of the body to the other; this transfer may occur without obvious cause or it may be induced by the application of gold and other metals, or a blister, a magnet or the faradic current. The transfer is temporary; in a few hours or a day or two the loss of sensation reverts to the original side. In some cases sensation is perverted, thus a touch on the radial side of the arm may be felt on the ulnar side, or a touch on one side of the body is referred by the patient to a corresponding spot on the other side—"alloeheiria."

Motor Symptoms. *Tremor* is a common symptom; it varies much in character, being fine or coarse, rhythmical or irregular. One peculiarity is that it shows abrupt transitions from one type to another, a fine quality for instance being suddenly replaced by jerky spasms. It is also apt to increase in intensity under the influence of any emotional change, whilst it may cease entirely when the mind is diverted from the affected part. Perhaps the commonest form is a rapid fine movement of the hands which in some cases simulates that of paralysis agitans but differs from it in being aggravated by voluntary movements of the arms, in its more variable intensity and character, in the

absence of the cigarette rolling movements of the fingers and in its occurrence at an earlier period of life. In another variety, tremor is wider in range and shows every transition between coarse rhythmical movements and actual clonic spasms. Occasionally such tremor occurs only on voluntary movement and may then resemble the tremor of disseminated sclerosis, when it will be necessary to consider the other symptoms of the two diseases before a diagnosis can be made.

Hysterical tremors are often associated with paralysis and contractures, and are sometimes intensified by attempts to move the affected limbs. They may appear or disappear after convulsive seizures, or they may intermit without apparent cause.

Rhythmical or irregular spasmodic movements are also observed; sometimes they resemble those of chorea, sometimes those of "habit spasm." As a rule they are more sudden and quick than those of true chorea, and are always increased by attention being directed to them. The closest simularity is observed when a case of chorea has been watched and imitated by a hysterical patient: the movements, however, are more transient and tend to disappear when the example is removed. Sometimes a spurious form of torticollis occurs; the neck muscles are usually affected on both sides, the head being jerked sideways, forwards or backwards. In some cases involuntary movements are more elaborate and purposive in character. Thus a patient in the erect posture may begin to progress by a series of jumping or hopping movements—"saltatory spasm"; or there may be bowing movements, or alternate flexion and extension of a limb. Occasionally there are constant movements of the face or limbs which may resemble those of athetosis, though as a rule they are more rapid.

Persistent *spasmodic contracture* of a limb is a common symptom, sometimes spasm affects other parts of the body. The contracture may follow and complicate paralysis or may occur independently of it. Usually started by emotional excitement, by a fit or by some

local injury or by pain, it occasionally develops spontaneously. The spasm is often very strong and may be difficult or impossible to overcome; sometimes indeed the attempt to relax it aggravates its intensity. Variable in degree at different times the muscular contractions may suddenly disappear, to reappear in the same or in another limb. Sometimes they persist during sleep, but relax when the patient is deeply under the influence of chloroform. Contracture of the upper limb is of flexor type, that of the lower limb is of extensor type, but in exceptional cases flexor contracture at hip and knee occurs which may become permanent. Occasionally only one segment of a limb is affected, thus the hands and fingers may be rigidly contracted, the attitude in some cases resembling that of tetany. The spasm may be limited to certain portions of a muscle, giving rise to local swellings. A "phantom tumour" of the abdomen appears to depend in some cases on spasmodic contraction of parts of the recti and obliqui muscles, and in others on spasm of the diaphragm with relaxation of the recti or of the whole abdominal wall, when if the intestines are distended with flatus the abdomen becomes greatly enlarged. Another variety of spasm is hysterical trismus due to contracture of the muscles of mastication, which may prevent the teeth being separated for more than a quarter of an inch. Such trismus may develop spontaneously or follow a fit; it is usually transient but tends to recur. In some cases of hysteria an appearance of ptosis is produced by contracture of the orbicularis palpebrarum, which may be unilateral or bilateral; occasionally a spasmodic convergent squint is observed.

Convulsive or *paroxysmal attacks* may occur; they are of several varieties, and generally are the result of some unusual emotional excitement. It is convenient to describe the mildest and the severest type of paroxysm, it being clearly understood that every intermediate variety may be observed.

Hysteria minor is the term applied to the mildest form of paroxysm. As a rule it begins with some

disagreeable sensation, as palpitation, globus, or dizziness and fainting. Such sensations are frequently followed by a feeling of choking and difficulty in breathing, when the patient may have a convulsive seizure, evinced by violent movements of the limbs, arching of the spine and throwing of the head from side to side. She does not fall suddenly to the ground but sinks on to a sofa or chair and appears to be unconscious. After a few minutes recovery slowly ensues, with much emotional display, with eructations of wind and a copious evacuation of pale limpid urine. There is never complete loss of consciousness and after the fit the patient usually remembers—it may be imperfectly—all that has happened.

Hysteria major. Hystero-epilepsy. Hysteroid convulsions are terms given to the severest forms of convulsive manifestations. These seizures are much rarer in England than in France; they are vividly described by French writers, who divide a typical attack into four stages.

The first or epileptic stage, preceded or not by globus, palpitation or other visceral or mental disturbance is frequently ushered in by a sensory aura, such as some curious sensation in the hypogastrium in the ovarian region or in the soles of both feet, which ascends to the throat or head, after which the patient falls down. All the muscles become rigid, the features are distorted, respiration is embarrassed and the attack closely resembles the first stage of epilepsy. The muscular tonicity is followed by irregular contortional movements which are quite unlike the clonic spasms seen in epilepsy; these in their turn are replaced by general relaxation of the muscles and apparent coma. It is to be noticed that the patient is rarely injured by falling; that the tongue is not bitten, although occasionally both the tongue and the lips may be accidentally injured by the teeth during the act of falling; that the pupils are normal and that the paroxysm is sometimes arrested by faradisation or by pressure over the ovarian regions.

After a short interval the *second stage* or the "phase de grandes mouvements" begins. This is characterised by the most varied and grotesque attitudes, the most striking being the attitude of crucifixation and that of the "arc de cercle." In the attitude of crucifixation the body and legs are straight, whilst the arms and hands are held at right angles to the trunk. In the "arc de cercle" there is marked opisthotonus which may be so exaggerated that the trunk is arched forwards and the patient rests on the top of her head and the soles of her feet. In this stage screaming, biting, tearing and all kinds of struggling movements may be witnessed, their severity being proportional to the efforts made to restrain them.

Fig. 214. Attitude during an attack of hysteroid convulsions in a boy aged twelve.

In the third stage, or the "phase des attitudes passionelles," expression may be given by delirious talk and by attitude to every possible emotion or idea, as anger, fear, grief, ecstasy, beatitude, or the most intense voluptuousness.

During the fourth stage the paroxysm gradually subsides. The patient begins to recognise her surroundings, but still rambles and occasionally is subject to various hallucinations; she sees rats or other animals, or hears voices and conversations. As the fit passes off she is often troubled with painful muscular spasms of the limbs, with hiccough or eructations of flatus, which may be followed by the passing of much pale urine. An average duration of the whole attack is from a quarter to half an hour.

In this country it is extremely rare to see a complete fit of this type in all its stages; abortive forms are much commoner. Sometimes hysteroid fits occur in series like epileptic fits; this status hystericus, as it is called, differs from the status epilepticus in the absence of pyrexia and of danger to life.

Hysteroid convulsions may follow true epileptic seizures but in such cases there is usually a history of epileptic fits which were not followed by these convulsions. They may be set up by organic brain disease, hence in all seizures apparently hysterical, a careful search should be made for any evidence of more serious disease.

Paralysis. This may be paraplegic, monoplegic or hemiplegic in distribution; with the exception of the larynx, paralysis rarely affects parts supplied by cranial nerves.

Paraplegia is the most common form; as a rule it is sudden in onset and can be traced to emotional disturbance. The paralysis is rarely absolute and is rarely limited to a particular group of muscles. It is rather a general inability of the limbs to perform a complex co-ordinated movement such as walking: thus a patient while able to move the legs in bed with normal power and co-ordination is unable to stand, or he can hop or jump although unable to walk. Inability to walk or stand, associated with freedom of leg movement when lying down is called "astasia abasia." Sometimes the power to move the legs in bed appears to be diminished.

but the degree of weakness is not constant, the limbs are drawn up slowly or jerkily and it may be difficult to estimate their strength.

An attempt to perform a particular movement may be opposed by the contraction of the opposing muscles, thus if the patient tries to extend the knee, the hamstrings become strongly contracted; or if he tries to flex the knee, the quadriceps is felt to be strongly in action. Similarly if the patient attempts to strongly flex the elbow a feeble contraction of the biceps is preceded by a well-marked contraction of the triceps. As pointed out by Beevor, this contraction of the antagonists before the prime movers, is characteristic of hysterical paralysis and does not occur in organic disease.

When a group of muscles such as the dorsi-flexors of the ankle is alone paralysed, serious disease should be suspected; the condition may be due to hysteria but there are usually indications either as regards variations in the power of ankle flexion at different times, or in the position of the foot, that the nature of the weakness is different to that caused by serious disease.

Paraplegia of varying distribution and character may be transient or may last for years; the muscles are either flaccid or rigid, they may waste from disuse, but degenerative atrophy does not occur, and the electrical reactions remain normal. In prolonged cases, disuse of the limbs may lead to changes in the joints and tendon sheaths, when movement will be still further hampered. Anæsthesia, varying in extent, often accompanies the paralysis, sometimes its upper margin is a transverse line across the hips. When the patient is able to walk, certain peculiarities in the gait are observed. Frequently the limbs appear rigid and difficult to move; there is an exaggerated show of effort and the patient shuffles cautiously forwards with the soles close to and parallel to the ground; in other cases the heel is unduly raised and the foot doubles over, its dorsum trailing along the ground (see fig. 215).

The knee-jerks are often greatly exaggerated; true

ankle-clonus is absent, but a spurious form is common, consisting of a few irregular imperfect movements, which occur only when the ankle is extended. The plantar reflex is difficult to elicit and is often absent; when present it is always of the flexor type.

Hysterical *hemiplegia* may follow a violent emotion;

Fig. 215. Gait in a case of left-sided hysterical hemiplegia. The marks on the left leg are scars of self-inflicted burns. (Purvis Stewart).

it is common after railway accidents, but no relation can be traced between its frequency, and the severity or the site of any injury received. The onset is usually gradual, and the left side is more often affected than the right. In a typical case the left limbs are weaker than their fellows, but by no means powerless; the leg is often weaker than the arm; no weakness can be detected in the movements of the face or of the thorax during inspiration. The weakened limbs often show contractures, the arm usually being rigidly flexed and the leg extended. In rare cases spasm may affect the face and the tongue, causing the latter to deviate either towards or away from the paralysed side. Hemianæsthesia, profound in degree, usually accompanies the hemiparesis, the face and special senses being involved as well as the trunk and limbs. A patient suffering

from these unilateral symptoms may be excitable, apathetic or otherwise abnormal in manner, and may complain of various head symptoms or of queer sensations down the affected side, but very often he is quite unaware of the presence or of the profoundness of the anæsthesia.

Monoplegia is much more frequently due to hysteria than to an organic lesion of the cortex. The hysterical variety is distinguished by the variable nature of the paralysis and by its usual association with profound anæsthesia, which in the arm may cease abruptly at the shoulder joint, and in the leg at the knee or hip. In crural monoplegia the plantar reflex is either absent or flexor, but never extensor, in type.

One of the commonest forms of paralysis is that of the *adductors of the vocal cords*; there is aphonia and the patient talks in a whisper. The glottis may be closed in coughing: sometimes singing is well performed, but in attempts to speak the cords remain wide apart and do not approximate. The condition is often cured by the application of a strong faradic current to the larynx, or even by the introduction of the laryngeal mirror.

Occasionally there is mutism—whispering speech being lost as well as phonation; attempts to speak may be accompanied by grotesque facial contortions. Very rarely a form of sensory aphasia is met with, which is difficult to distinguish from that due to coarse lesions in the cortex.

Visceral, vaso-motor and trophic symptoms. In addition to the globus hystericus, the patient may be troubled with spasm of the pharynx leading to difficulties in swallowing or with spasm of the œsophagus accompanied by regurgitation of food. Frequently vomiting is a troublesome symptom; usually it is unattended by pain or nausea. Sometimes it is set up by real gastric disturbance, as from an ulcer, when it may persist as a morbid habit and may be easily started by any emotion of disgust. In severe cases, especially if anorexia also

is present, the patient falls into a condition of extreme marasmus and may die from exhaustion. In some cases loss of appetite and abstinence from food are genuine symptoms, but more frequently they are fraudulent, the patient fasting at meals and eating in secret as in the well-known cases of "fasting girls."

Every variety of dyspepsia is met with in hysteria; flatulence with noisy eructation is very common, and is often associated with obstinate constipation. In severe cases the inactivity of the bowel seems to depend on serious derangement of its muscular walls.

Incontinence of urine or of fæces does not occur, but retention of urine is not uncommon. In many cases a large quantity of pale urine, of low specific gravity, is passed; and sometimes micturition is extremely frequent. Very rarely the secretion is lessened and even completely arrested,—hysterical ischuria and anuria. The suppression may persist for several days, without any toxic symptoms; it is often attended by vomiting, the amount of liquid vomited varying inversely with that of the urine passed.

Attacks of extreme rapidity of breathing, fifty to eighty respirations per minute, are sometimes observed. When laryngeal spasm is also present, there may be stridor, cyanosis and other signs of dyspnœa.

Hysterical patients are also liable to suffer from palpitation, with pain, giddiness and actual fainting. Sometimes an anginal attack is simulated: the exciting cause of the seizure and the absence of vascular degeneration distinguish the condition from true angina pectoris.

As a rule the temperature is normal: occasionally very high temperatures have been observed, but it is probable that these are due to the patient's tampering with the thermometer, by means of a hot water bottle, a poultice or in some other way.

The skin of a paralysed limb is often blue and cold: it may appear to be œdematous but does not pit on pressure. When pricked or cut no bleeding may occur.

a curious feature of hysteria which has long been recognised.

Course and Prognosis. It is almost impossible to define the course or to foretell the duration of hysterical symptoms. An anæsthesia, a paralysis or a contracture may suddenly disappear, often in consequence of some profound emotion, or it may persist for months or even years. The chance of recovery from any particular symptom bears no proportional relation to its duration: the symptom may suddenly pass away whether it has lasted a few days, several months, or longer. Nor, when all the symptoms have vanished, can we speak with certainty regarding the possibility of their return. Undoubtedly in hysteria there is a tendency to remission and relapse of symptoms, a feature, it must be remembered, that is perhaps still more strikingly manifest in disseminated sclerosis. Speedy recovery may generally be expected in cases of fits and paralysis occurring in young persons as a result of emotional shock, and also in cases of traumatic hysteria as soon as the question of compensation is settled; moreover there is no hysterical symptom that may not be cured. Life is rarely imperilled, except in severe cases of inanition due to anorexia or to persistent vomiting.

Diagnosis. There is little or no difficulty in saying that certain symptoms are hysterical in character but there is often a great difficulty in deciding whether they are set up by some definite lesion in the nervous system, or whether they represent hysteria only. The early stages of disseminated sclerosis are particularly apt to be associated with hysterical manifestations; the early symptoms of cerebral tumour also may be similarly masked. Hence it is of the utmost importance in all cases of hysteria to search for signs of the presence of more serious disease, such as optic neuritis or atrophy, nystagmus, a fixed pupil, paralysis of cranial nerves, absence of the knee-jerk, sustained ankle clonus, Babinski's reflex, incontinence of urine, and paralysis limited to a definite group of muscles. But even when

signs of organic disease are absent and we have decided that the patient is suffering from hysteria only, we must not close our minds to the possibility of future developments. The hysterical symptoms may have a background of structural changes the signs of which will become manifest at a later period. Subsequent examinations of the patient must therefore be as thorough as the first examination, and the observer must eliminate from his mind the bias produced by his first diagnosis of hysteria; only in this way can mistakes be avoided.

Treatment. In the treatment of hysteria the personality of the physician counts for much, as well as his manner in dealing with the patient. His attitude must be one of quiet sympathy, combined with tact and firmness. He should explain clearly to the patient the nature of her complaint, neither magnifying nor making light of her symptoms and should encourage her to believe in the certainty of recovery. Having obtained both the confidence and the co-operation of the patient, the principle of the treatment to be adopted should be explained to her and the necessity for every instruction to be faithfully followed.

The exact details of treatment will vary in almost every case, but some degree of separation from relatives and friends is usually advisable. In mild forms of the disease a change of scene and companionship may suffice, but when the symptoms are severe and persistent more complete isolation is required. This is best attained by residence in a nursing home or hospital, where the other methods which constitute the Weir-Mitchell treatment can be satisfactorily carried out. These, as already explained in the section on neurasthenia, comprise rest in bed, abundant nourishment and massage.

The faradic current, especially as conveyed through a wire brush, is frequently useful in aiding to restore motility and sensibility, and in cutting short hysterical convulsions. Sometimes too a seizure is modified, or arrested by cold water in the form of a bath or a spinal douche.

But whatever methods are adopted the importance of helping the patient to regain her lost will power must be kept constantly in mind. For example, when paralysis affects the lower limbs, the patient should be set on her feet and firmly encouraged to try to walk. At first she will have to be well supported. If this exercise is systematically gone through two or three times a day, an increased security in gait will gradually ensue, and as the patient regains confidence in her powers, progress towards complete recovery may rapidly occur.

Medicinal remedies, although of little value in the treatment of the malady itself, are frequently useful for the relief of symptoms. The bromides and preparations of valerian or assafoetida have a beneficial influence on many hysterical manifestations, and especially when there is much excitement. A mixture containing bromide and digitalis will often reduce the number of hysterical attacks. Arsenic and iron may be required for anæmia, whilst quinine and cod liver oil are helpful during convalescence. Narcotics such as morphia, sulphonal and chloral, must be avoided as much as possible; sleep is sometimes induced by the administration of some indifferent substance.

As to the value of hypnotism in the treatment of hysteria there is much difference of opinion. There is always the danger that the mental instability may be increased; nevertheless many authorities speak favourably of the method, and it is at least deserving of a trial when symptoms are very urgent, severe and obstinate.

SECTION XXI.

Syphilis of the Nervous System.

In the preceding pages many of the effects of syphilis on the nervous system have been described; others, however, remain for consideration: and, further, the importance of syphilis in relation to nervous diseases makes it desirable to devote a separate section to a general survey of the subject.

Much light has been thrown on the syphilitic origin of many diseases of the nervous system by the discovery of the spirochæte pallida, by the examination of the cerebro-spinal fluid obtained by lumbar puncture, by the sero-diagnostic investigations of Wassermann and by the numerous clinical and pathological records of many eminent observers.

The more carefully the effects of syphilis on the nervous system are studied, the greater appears their tendency to generalisation; cranial and spinal syphilis may occur separately, but in a large number of cases there is evidence of their close association. Sometimes their development is simultaneous, and yet the indications of cranial and spinal syphilis may vary greatly; thus the manifestations of cranial syphilis may be marked, while those of spinal syphilis are slight, or *vice versa*. In some cases the symptoms of the one variety, which alone perhaps are present, may subside; and then, after an interval of varying duration, the symptoms of the other variety become manifest. For example, a syphilitic myelitis may be ushered in by transitory symptoms of brain disturbance; the myelitis may pass away, to be followed at a later period by an attack of hemiplegia.

Another striking fact is that while syphilitic affections of the nervous system may occur at any period after infection, a large number of them, and especially the severe and generalised forms, occur within the first year

or two. As suggested by Mott, the membranes of the brain and spinal cord are probably infected at the time when the cutaneous rash appears. It is true that the outbreak of the exanthem is not commonly attended by obvious symptoms of nerve-disturbance, but it is also true that occasionally at this time such symptoms do occur, as for example headache, giddiness and irregularity of the pupils, although in many cases they may be so slight as to be easily overlooked.

The widespread distribution of the nervous disorders is also evinced by the later manifestations of syphilis, namely, tabes and general paralysis. These diseases, which depend on degeneration of various nerve tracts and cells, are sometimes found in combination. They are not due directly to the action of the syphilitic virus, but indirectly to its effects in lowering the vitality of the fibres and cells concerned: hence they are called para-, or meta-syphilitic diseases.

The various effects of syphilis on the nervous system may be classified as follows:—

Effects of Acquired Syphilis.

1. Intracranial and Spinal lesions characterised by the presence either of specific inflammatory affections of the membranes and vessels; or of new formations or gummata
2. Progressive degenerations of neuronic tissue, which usually develop some years after the initial lesion. Tabes; general paralysis (see Sections VIII. and XVIII).
3. Peripheral neuritis (see p. 186).

Effects of Hereditary Syphilis.

INTRACRANIAL DISEASES FROM ACQUIRED SYPHILIS.

Pathology. The chief lesions are arteritis, meningitis and gummata.

Arteritis. The most characteristic change is proliferation of the intima: frequently the middle and adventitial coats of the artery are also thickened. The

resulting narrowing or obliteration of the lumen causes anæmia in the distribution of the affected vessel, but if thrombosis does not take place there may be only transitory symptoms of brain disturbance. The arteries most commonly affected are the internal carotid, the middle cerebral and its branches, the basilar and the vertebrals. A spreading thrombosis in one of these vessels leads to softening in the area cut off from its blood supply. When the cortical arteries are involved the softening may be slight or even absent, the collateral circulation maintaining the nutrition of the affected area. The nucleus of each of the cranial nerves from the third to the twelfth is supplied by a separate artery, and in the case of the third nerve the individual parts of its nucleus have their own blood supply, hence thrombosis of a particular vessel will lead to degeneration only of the group of cells supplied by it.

In association with the arteritis there is frequently much perivascular infiltration, which is sometimes attended by the formation of small gummatous nodules in the walls of the vessels. Occasionally the weakened wall bulges and an aneurysm is formed; in relation to the giving way of the vascular coats the influence of atheroma must not be overlooked, as this change is liable to occur at a much earlier age in syphilitic than in non-syphilitic persons. The veins also may suffer, their calibre undergoing a progressive diminution; this is another factor in the production of malnutrition of the brain.

Meningitis occurs in two forms, namely as pachymeningitis or as leptomeningitis; frequently the two conditions are combined. Probably the commonest variety of meningeal syphilis is a diffuse gummatous leptomeningitis at the base of the brain. As a rule the meningitis is associated with disease of the vessels, and sometimes with gummata in various parts of the brain. In some cases the neoplastic formation extends down the spinal canal, involving the spinal roots as well as the cranial nerves; there is thus a generalised cerebro-spinal

meningitis; in severe cases the whole of the base of the brain and the spinal cord are covered with a gelatinous substance which encloses the vessels, the roots and the nerves.

Gummata may originate in an osteitis or a periostitis of the skull, or may develop in the membranes: sometimes they begin in the dura mater and invade the bone. When a gumma begins in the pia-arachnoid, it tends to extend along the sheaths of the vessels into the brain. Like any other local swelling, a gumma leads to an increase of intra-cranial pressure: it irritates and then destroys nerve-elements, and by pressure on vessels or by involvement of their walls it leads to localised areæ of softening. Multiple gummata are commoner than a single gumma. The growth may be circumscribed or it may occur in a diffuse form in association with meningitis. It may be found in any part of the nervous system, but more frequently it involves the cerebral cortex and the base of the brain: at the base it implicates the optic chiasma, the interpeduncular space and the cranial nerves, especially the second and the third.

Symptoms. In studying the morbid anatomy of intracranial syphilis we are struck by the tendency for the changes to be generalised rather than localised. Membranes and vessels may be separately affected, and only in one part, as on the convexity or at the base of the brain, but this is not the rule: usually both these tissues are affected and often in many places.

The wide distribution and the numerous combinations of the morbid changes throw light on the variety of the individual symptoms, on their grouping and on their tendency to remissions and relapses. When we consider the circulatory disturbances going on in a syphilitic brain, we can readily understand the transitory duration of many symptoms, their reappearance and the development of new ones. There is no essential difference between the character of the brain disturbance produced by syphilis and that produced by other causes. The same patient at different periods may present symptoms

of epilepsy, of meningitis and of tumour. The diagnosis is based rather on the association and succession of symptoms, on their multiplicity and their tendency to asymmetry in distribution.

The recognition of intra-cranial syphilis in its earliest stages is of the greatest importance, for it is at this period that treatment is of most avail, frequently abating or arresting the morbid changes and thus postponing or preventing the onset of paralysis or of other serious manifestations.

The most common prodromal symptom is headache, which is often severe, is worse at night, is increased by mental activity and may last for some time. The pain may be referred to the frontal or the parietal region, and in cases of meningitis of the convexity it is associated with tenderness on pressure; sometimes a slight thickening of bone from periostitis can be detected. Insomnia, often the result of headache, may occur independently. Attacks of somnolence lasting for many hours are also common: they tend to be very prolonged when endarteritis is widely distributed. In some cases there are attacks similar to those of alcoholic intoxication; the patient talks and moves about in an excited fashion, and afterwards may have no recollection of what has occurred. Vertigo and epileptiform seizures may also occur at an early period of the disease. A tendency to irritability of temper and to other signs showing a lack of self-control is often observed. Much less energy than formerly is evinced by the patient for both mental and physical work; frequently also his memory and power of ideation are much diminished.

The above-mentioned symptoms, which indicate early changes in the vessels or membranes, may subside under treatment, or they may become more marked, and may be accompanied by other more definite signs of arteritis, meningitis or of a gummatous tumour.

Arterial thrombosis is not always preceded by symptoms of vascular disturbance. The onset may be quite sudden, the patient in apparently good health becoming

aphasic, or paralysed on one side; the aphasia or the hemiplegia may be either temporary or permanent. Thrombosis commonly occurs in the lenticulo-striate branch of the middle cerebral artery, hemiplegia of the ordinary type being produced. Thrombosis, however, may occur in other arteries, the symptoms varying with its situation and degree (see p. 620).

Syphilitic meningitis occurs in two forms—an acute and a chronic form. The acute variety is rare, and usually is really an acute exacerbation of a chronic meningitis, which may not have given rise to obvious symptoms of its presence. The clinical manifestations of the chronic form vary greatly, according to the situation of the disease and to the degree and extent of disease of the vessels and the cranial nerves, with which it may be associated. There are two main types, namely, meningitis of the convexity and meningitis of the base of the brain.

In *cortical meningitis*, which is usually accompanied by some degree of encephalitis, headache is apt to be violent, and obstinate, and localised to a particular part of the head, pressure on which is often very tender. In addition to pain and the other symptoms of meningitis there may be signs that certain portions of the cortex are specially involved. The motor region being frequently affected, unilateral convulsions are common; beginning in one portion of a limb they may spread to other parts and eventually both sides of the body may be affected, when consciousness is often completely lost. Attacks of general convulsions are only distinguished from true epilepsy by the occurrence in the intervals, of nocturnal headache, loss of memory, transient aphasia and other signs of irritated or functionless cortical neurons.

In many cases convulsions are followed by a monoplegia or a hemiplegia; sometimes an old hemiplegia, due to softening of the internal capsule from thrombosis of a syphilitic vessel, is associated with attacks of Jacksonian epilepsy from a cortical meningitis. In

other cases the parietal or the occipital portions of the cortex are specially implicated, giving rise to disturbances either of sensation or of vision. In cortical meningitis mental symptoms are common: sometimes they are predominant, the memory fails, the speech becomes indistinct and dementia gradually ensues.

Basal meningitis. Of the general symptoms, headache is one of the earliest and most constant; it may precede other symptoms by many weeks or months. It tends to occur in acute paroxysms, is deep-seated, and cannot be localised, nor is it attended by local tenderness as is the headache of cortical meningitis. Vomiting is also a frequent symptom: sometimes it is absent throughout the whole course of the disease. Vertigo, staggering and attacks resembling *petit mal*, or migraine are not uncommon. Some form of psychical disturbance is a prominent feature in basal meningitis. A somnolent condition in which the mental faculties are at a low ebb is particularly characteristic: the patient appears to be in a state of stupor, or as if semi-intoxicated from alcohol: he may be roused to answer questions in a drawling, sleepy manner, or to perform certain actions such as sitting up in bed in response to requests. Owing to his mental condition he appears to have no control over his sphincters and passes his excreta in bed. There is thus a partial dementia, which may clear up for a time to again relapse. The course of the disease is characterised by marked oscillations and remissions in the mental disturbance. Periods of excitement may alternate with periods of depression in which a tendency to suicide may develop; delusions of persecution or of being poisoned may lead to acts of violence. Such a condition may be preceded or followed by an attack of unconsciousness; sometimes the mental condition remains normal for a long time and then suddenly the patient becomes comatose and dies.

General, unilateral or partial epileptiform seizures often occur: when the meningitis involves the posterior fossa and extends into the spinal canal, tetaniform

spasms and opisthotonos may be present. The local symptoms, those due to implication of the cranial nerves, are of greater diagnostic importance. Visual disturbance and oculo-motor paralysis are the most frequent. Choked disc, optic neuritis leading to optic atrophy, or disorders of vision apart from changes in the fundus may occur. Choked disc affects both eyes and indicates a general increase of intracranial pressure from the presence of a gummatous tumour. Optic neuritis due to extension of the inflammation to the nerve is often more intense on, or is limited to, one side; blindness is partial or complete according to the number of fibres in the optic nerve that are affected. If the chiasma or the optic tracts are invaded by the gummatous process there may be much visual disturbance without any changes in the fundus; bitemporal or binasal hemianopsia pointing to disease of the chiasma, homonymous hemianopsia to disease of the optic tract. A noticeable feature of the blindness in syphilitic basal meningitis is the great variation in its degree, all possible forms of limitations and irregularities in the visual fields being observed.

The third nerve is more commonly affected than any other cranial nerve, partial paralysis of the muscles supplied by it being more common than complete. There may be ptosis of one or both eyes, paralysis of one pupil, or paralysis of all the muscles except the levator palpebræ. In explanation of the fact that the paralysis is often limited in distribution it is to be noted (1) that each group of ganglion cells in the nucleus of the third nerve has its own arterial branch, so that if this alone is diseased the nerve supply to one or two muscles only may be impaired; and (2) that the fibres of the third nerve run separately through the crus and therefore may be separately damaged. Sometimes the paralysis is associated with hemiplegia on the opposite side of the body owing to implication of the crus by gummatous exudation. Paralysis of the external rectus, usually only on one side, from disease of the sixth nerve, is also

common. The fourth nerve is affected more rarely, and always in association with other cranial nerves. In some cases there is more or less complete ophthalmoplegia, which may be caused by occlusion of the arteries supplying the nuclei of the oculo-motor nerves.

The fifth nerve is rarely affected alone; its sensory portion is more commonly involved than its motor. The seventh and the eighth nerves are usually affected together, as a result of pressure from a gumma, of meningitis or of syphilitic disease of the petrous bone. Sometimes they are implicated along with other structures in the pons, either by a gumma or by softening from endarteritis; there may then be a crossed paralysis, the face being paralysed on the side of the lesion and the limbs on the opposite side. When the meningitis affects the side of the medulla the tenth, eleventh and twelfth nerves may be involved, giving rise to paralysis of the tongue, palate and vocal cord on one side. Disturbance of the heart's action, of respiration, deglutition and articulation may also be present.

In some cases basal meningitis runs an acute and fatal course, death being usually due to disease of the vagus; in others the disease may last for some years, when its course is characterised by remission and recrudescence of many of its symptoms.

Gummata. The symptoms of a single large gumma do not differ from those of other intracranial tumours, except perhaps as regards their greater variability from time to time. There is a tendency for optic neuritis to appear early, to develop rapidly and to become intense.

The symptoms of multiple gummata are numerous and varied; in the early stages there may be local or general convulsions, in the later, motor and sensory paralysis. Considerable mental disturbance is often a noticeable feature.

SPINAL DISEASES FROM ACQUIRED SYPHILIS.

The majority of cases of spinal syphilis occur during the first four years after infection, and it is common to

meet with symptoms of myelitis or of meningo-myelitis during the first year. It is significant that a lymphocytosis of the cerebro-spinal fluid is often found during the eruptive period; indeed it seems probable that the membranes and vessels of the nervous system are frequently, if not always, affected during the secondary period, the disease in many cases being unattended by any clinical manifestations, while in others it is sufficiently developed to give rise to symptoms of slight sensory or motor disturbance, which are overlooked or are attributed to other causes. Such slight degrees of meningeal or of vascular disease may subside or they may lie dormant for months or years, and then under the influence of cold, injury or alcoholism may be lit up afresh and may develop into a pronounced spinal cord affection.

The syphilitic virus may attack the vertebræ, the membranes, arteries, veins or the lymphatics. In spinal, as in cranial syphilis, it is disease of these tissues which irritates and destroys the nerve elements in the spinal cord or in its roots; they suffer secondarily in consequence of morbid changes in the surrounding tissues, and even in Erb's variety of spastic paralysis (see p. 117) it seems probable at least as regards many of the cases, that the tract degenerations are secondary to previous inflammatory lesions of the spinal vessels.

Spinal syphilis is less common than tabes, and as a rule occurs much sooner after infection; thus it frequently comes on during the first three or four years, whilst tabes develops usually after the eighth year. Occasionally a case of spinal syphilis is met with as early as the fifth month after infection, or as late as the fifteenth year. The age of the patient is usually between twenty and forty: males are more frequently affected than females.

Syphilitic disease of the vertebræ is very rare: it occurs in the form of periostitis, osteitis, exostosis, gummata, necrosis and caries. The disease may originate in the vertebræ, or, in the case of the cervical spine, it

may occur as a result of extension from syphilitic disease of the skull, or from a deep syphilitic ulcer of the throat.

In some cases the symptoms are limited to tenderness over the affected bones, pains and stiffness on movement of the spine, and radiating pains along the spinal roots coming from the diseased parts. In other cases there are symptoms of compression myelitis which, when dependent on syphilitic caries, may be accompanied by a deformity of the spinal column similar to that produced by tuberculous caries. The diagnosis is based on the absence of signs of tuberculous or of malignant disease in any part of the body, on the evidence regarding previous syphilitic infection, and on the results of anti-syphilitic treatment. In malignant disease, the pain is usually more severe, the course of the illness is more rapid, and there is a greater degree of emaciation. It must be remembered that tuberculous caries may occur in a syphilitic subject.

Meningitis. Very rarely, cases are met with in which there is evidence, both clinical and pathological, of a syphilitic chronic meningitis without any signs of implication of the cord. As a rule all the membranes are affected, and, at an advanced stage of the disease, are fused together forming a thick sheath of fibrous or of gummatous material, which surrounds and is often adherent to the cord. Exceptionally the inflammatory changes are limited to the dura mater or to the pia-arachnoid. In pachymeningitis the inflammation either begins on the inner surface of the dura mater or spreads to its outer surface from syphilitic disease of the vertebræ.

It is rare to meet with meningitis along the whole length of the cord: usually it is limited to, or is most intense in, one region. In the cervical region it may be an extension from meningitis at the base of the brain: thus sometimes symptoms of cervical hypertrophic pachymeningitis (see p. 315) are combined with signs of paralysis of some of the lower cranial nerves. Meningitis in the lumbar, or in the sacral region, causes pains in the legs, followed by anæsthesia, and sometimes by

disturbance of the functions of the bladder, rectum and genital organs.

There is no essential difference between the symptoms of syphilitic and those of non-syphilitic meningitis. Pain in the back and stiffness in its movements; radiating pains, patches of hyperæsthesia and anæsthesia from implication of the posterior roots, together sometimes with atrophy of the muscles supplied by the affected anterior roots, all these symptoms are common to both forms of spinal meningitis.

Points in favour of the syphilitic variety are an increase of the pain at night, absence of pyrexia, and the presence in some cases of cerebral symptoms, such as headache, hemianopsia, diplopia and changes in the pupils. A confirmation of the diagnosis would be given by finding a decided lymphocytosis of the cerebro-spinal fluid, and a positive Wassermann's reaction.

Meningo-myelitic. This is the commonest variety of spinal syphilis. Sometimes its manifestations are preceded by signs of cerebral syphilis.

The symptoms due to changes in the spinal membranes precede by several days, or sometimes by several weeks, those due to implication of the spinal cord. The meningeal symptoms comprise pain in the back, often worse at night, spinal tenderness and signs of implication of the spinal roots. Extension of the morbid process to the spinal cord is indicated by weakness of the legs, and by sensory and visceral disturbances. Paraplegia may develop suddenly, rapidly or very gradually: it may be either partial or complete. Sometimes there are several attacks of temporary paresis of the legs, before the development of severe paraplegia. Anæsthesia may or may not be present; it may be either partial or complete, both as regards its distribution and the kind of sensation affected. There may be loss of sensation to all forms of stimulation, to pain and temperature only, or to temperature only. In some cases, sensation to temperature is diminished or lost before that to touch or to pain: occasionally the loss is limited to cold stimuli. The

vibrating sensation may be lost when all other forms of sensation are preserved.

An early affection of the bladder is somewhat characteristic of syphilitic myelitis, retention of urine often preceding weakness of the legs. Sometimes, however, the development of paraplegia is followed by paralysis of the bladder and rectum; there may be retention with dribbling of urine, or paralytic incontinence of both urine and fæces. The bladder affection leads to cystitis which may be followed by pyelo-nephritis and septicæmia. Bed-sores are apt to form on the buttocks or the sacrum. The sexual functions are often impaired.

The distribution and character of the symptoms vary with the site of the lesion, which most frequently is the dorsal region of the cord. They are similar to those which occur in cases of non-syphilitic myelitis and have been already described (see p. 272).

The course of meningo-myelitis presents many variations. In some cases there is recovery, either partial or complete; in others the symptoms fluctuate in intensity and in distribution from time to time, or they remain stationary for many months or even years. Death may occur at an early or at a late period of the disease; the chief causes are septic poisoning from a bed-sore or from pyelo-nephritis, respiratory paralysis from an upward extension of the lesion, or the development of some lung complication, such as pneumonia.

Acute myelitis. In some cases of spinal syphilis the development, character and distribution of the symptoms correspond to those of acute myelitis or even of spinal hæmorrhage. There is a rapid or sudden onset of paraplegia which in the course of a few hours or a day or two may become complete. Paralysis may also invade the lower intercostal muscles and very rarely the arms. Various forms and degrees of anæsthesia may be present: sometimes the temperature sense is alone affected. The knee-jerks are exaggerated, diminished or lost; their condition may vary at different times in

the same case. There is often paralysis of the bladder and rectum.

Frequently premonitory symptoms usher in the paraplegia; they are apt to occur for a longer time before the onset of paralysis, and to be more variable in intensity, than in non-syphilitic cases of myelitis. They comprise headache, vertigo, diplopia and other signs of cerebral disturbance, as well as pains in the back and legs, numbness and tingling in the legs, and retention of urine. The occurrence of retention of the urine for some days or weeks before the development of paraplegia is much commoner in syphilitic than in other forms of myelitis.

In one of my cases the onset of paralysis was almost as sudden as in spinal hæmorrhage. The patient, with a secondary eruption on his body, suffered one morning from severe pain in the lumbar region; the pain lasted for one hour. A few hours later the patient on attempting to rise from his chair, found that both of his legs were paralysed; retention of urine occurred about the same time.

Acute syphilitic myelitis may develop either a few months or several years after infection: as a rule it occurs during the first three years. The prognosis is very unfavourable; many cases end fatally in a short time, others show improvement if only for a time, while in a few cases there is partial or even complete recovery.

The symptoms depend on syphilitic disease of the spinal blood vessels which in some cases leads to softening, in others to degeneration of the nerve-elements. In a case examined by Williamson many of the vessels were dilated and obstructed by thrombi, while around the thrombosed vessels there was softening with hæmorrhagic infiltration of the tissue.

Gumma of the spinal cord, or of its meninges. Very rarely cases occur in which there is clinical evidence of a localised meningitis or of an intra-medullary tumour, and still more rarely the diagnosis has been confirmed by the finding of a localised gummatous

meningitis or of a gumma within the cord. Gummata occur more frequently in the meninges than in the cord; the chief symptoms of the former condition are pain in the back and signs of implication of the spinal roots. When a gumma begins within the cord, the development of paraplegia is slow and gradual, and root symptoms are generally absent. In some cases the symptoms are those of a unilateral lesion (Brown-Séquard's symptom-complex) but they rarely conform strictly to this type.

Miscellaneous forms. Almost every variety of disease affecting the nervous system may be simulated by the effects of syphilis; this is not surprising when we consider the tendency of the virus to attack vascular tissues, and the frequent extensive distribution of the vascular lesions. The following rare varieties may be briefly mentioned: (1) *Triplegia* or paralysis of one arm and both legs; this is a combination of hemiplegia and paraplegia, due to the simultaneous or the successive occurrence of a cerebral and spinal lesion. (2) Cases resembling *disseminated sclerosis*, the symptoms indicating the presence of multiple lesions in the nervous system. True nystagmus, intention tremor and scanning speech would be in favour of disseminated sclerosis, while immobility of the pupils, or marked ocular paralysis, especially if it preceded the other symptoms, would suggest the presence of syphilitic lesions. (3) Cases resembling *primary lateral sclerosis*: spastic paraplegia being present without anæsthesia and without any disturbance of the functions of the bladder or rectum. As a rule, however, the condition is preceded or followed by other symptoms which indicate, as does also the pathological examination, that the changes are not strictly limited to the pyramidal tracts. (4) Cases resembling various forms of *atrophic paralysis*. Syphilitic disease of branches of the anterior spinal arteries may produce symptoms similar to those of anterior poliomyelitis. A meningo-myelitis in the cervical region may lead to a condition resembling

either progressive muscular atrophy or amyotrophic lateral sclerosis. As a rule the resemblance is not a close one; usually there are symptoms other than those caused by disease of the anterior horns or of the lateral columns, which show that the lesions are not limited to these parts of the spinal cord. (5) Cases resembling *syringomyelia*, in which the symptoms depend on the effects of meningo-myelitis; sometimes there are cavities in the cord, the result of softening from syphilitic disease of the vessels. (6) Cases resembling *tabes*, syphilitic pseudo-tabes. The symptoms ataxia, absent knee-jerks, pains and anæsthesia depend on a meningo-myelitis affecting the posterior roots and the posterior columns. These symptoms appear at an earlier period after infection than in true tabes, and their development is more rapid. They are also more variable in character and are more subject to regressions and remissions; thus at one time the knee-jerk may be absent, at another present. Optic neuritis and post-neuritic atrophy are not uncommon, whereas primary optic atrophy occurs in tabes. Lymphocytosis is more marked than in tabes; it may disappear under the influence of mercury. The other symptoms of pseudo-tabes may also improve or entirely pass away under anti-syphilitic treatment.

Treatment. The general treatment of diseases of the nervous system which are non-syphilitic in origin is equally applicable to those which occur as a result of syphilis and need not be further considered. Attention may here be directed to some of the specific remedies and to the method of their administration. The most efficacious are mercury and the iodides. Recently a number of new preparations, having arsenic as their chief ingredient, have been recommended.

Mercury may be given by the mouth, by inunction or by intra-muscular injection. *By the mouth* the best preparations to use are Hyd. c Creta to which one or two grains of Pulv. Ipecac. Co. may be added to prevent diarrhœa; corrosive sublimate, usually in the form of

liquor hydrarg. perchlor.; and the green iodide of mercury. The chief disadvantages of the ingestion method are the uncertainty in the action of the drug, and the tendency to gastro-intestinal disturbances.

The method of inunction is the simplest and the least likely to give rise to complications. The best ointment to use is the unguentum cinereum composed of equal parts of mercury and lanoline, with a sufficiency of olive oil. A drachm of this ointment should be rubbed daily for fifteen to twenty minutes into the skin, preferably after the patient has taken a warm bath; a fresh region of skin should be selected each day. At Aachen a course of treatment consists usually of sixty immersions in the hot sulphur water, followed by sixty inunctions.

Intra-muscular injections of mercurial salts are strongly advocated by some authorities. The chief advantages of this method are that a known quantity of mercury is administered and absorbed, that less frequent applications are necessary and that a more rapid action on the syphilitic lesion is obtained. The method is therefore to be recommended, in conjunction with the mercurial inunction and the administration of the iodides, when the symptoms of cerebro-spinal syphilis are severe and urgent. The soluble preparations of mercury, which require to be injected more frequently than the insoluble, are the perchloride, one-eighth of a grain; the succinimide, one-sixth to one-quarter of a grain; and the cyanide, one-fourth to one-half of a grain. The insoluble preparations are metallic mercury in doses of one grain; salicylate of mercury, half a grain; and calomel, half to two-thirds of a grain. The grey oil or mercurial cream prescribed by Lambkin contains one grain of metallic mercury in every ten minims; this quantity may be injected once a week for a period of about two months and then repeated after an interval of a few weeks.

The *iodides* prescribed either as the potassium or the sodium salt, in daily doses of from ten to ninety grains, are of great value in the treatment of cerebro-spinal

syphilis, especially in the later stages of the disease. The most beneficial results are usually obtained by combining the administration of the iodides with mercurial inunction.

It is too soon to express a definite opinion with regard to the value and safety of salvarsan in the treatment of syphilis. The administration of some of the other preparations of arsenic, such as atoxyl, arsacetin and soamin, has been followed by serious toxic effects, as, for example, blindness from optic atrophy. Lambkin, however, has successfully treated a large number of cases of syphilis with a preparation of atoxyl and mercury, and without the slightest toxic effect.

EFFECTS OF HEREDITARY SYPHILIS.

The following account of hereditary syphilitic disease of the nervous system is based on a paper written by the author in *Brain* 1883, on an article by Sir Thomas Barlow and the author published in Tuke's *Dictionary of Psychological Medicine* 1892, and on the more recent investigations by Mott and other observers. At the outset it may be noticed: (1) That diseases of the nervous system produced by congenital syphilis are met with much less frequently than those produced by acquired syphilis. (2) That although a few broad differences can be made out as to the character and distribution of the lesions in the two groups, there is the same tendency to generalisation of the disease and to combination of the various lesions—pachymeningitis, leptomeningitis, gummatous deposits and endarteritis. (3) That a less abrupt separation is to be observed between the early and the late effects of hereditary than of acquired syphilis. In the adult, the parasyphilitic affections—tabes and general paralysis—are very rarely found in association with specific inflammations. In the child these affections appear to be the outcome, at least in some cases, of definite specific lesions, which sometimes are associated with gummata in the viscera

and syphilitic periostitis of one or more of the limb bones.

Intracranial Congenital Syphilis.

Pathology. Diseases of the nervous system as a direct result of syphilitic disease of the *bones* are very rare, the rarity being particularly marked in the case of the vertebræ. The brain is more liable to be affected by disease of the cranial bones from congenital than from acquired syphilis. This is due to the massive thickening, often accompanied by great exfoliation of bone, which is a characteristic feature of congenital syphilitic osteitis. In one case under the care of Henry Humphreys so marked was the thickening at the base of the skull that many of the foramina were distinctly narrowed. Such dense sclerosis of the cranial bones occurring in early life must tend to hinder the growth of the brain and is doubtless a factor in the production of juvenile dementia.

The *dura mater* may be greatly thickened, assuming in some places an almost cartilaginous consistence: it may be the seat of inflammatory deposits and sometimes there are adhesions gluing together the brain to the membranes and the membranes to the bone. The inflammatory process is generally an extension from periostitis of one or other cranial bone; accompanying the pachymeningitis there may be hæmorrhage, giving rise to laminæ of fibrin. In the *pia-arachnoid* every variety of inflammatory deposit may be found; rarely there is an acute meningitis represented by patches of greenish lymph on the vertex or the base of the brain: more commonly a chronic meningitis occurs, which may lead to great fibroid thickening and even to calcareous changes. Occasionally gummata and endarteritis are found in the inflammatory deposit. The *endarteritis* so characteristic of acquired syphilis is also common in hereditary syphilis. The small cortical arteries may be diseased when the large basal ones are healthy, or *vice versâ*, or both may be affected together.

The most striking changes are found in the *cerebral cortex;* occasionally softening is present but much more frequently hardening. The sclerosis either involves one or more convolutions or it occurs in the form of small nodular masses; but as a rule it is widespread, affecting large portions of the hemispheres, and is frequently associated with atrophic changes. It may extend for a short distance into the white matter, and very rarely it has affected the basal ganglia and other portions of the brain, but in the majority of cases it is limited to the cortex. "The narrowing of separate convolutions, the alteration of consistency to that of cartilage and the very slight alteration of colour towards a brownish-pink are very characteristic features" (Barlow and Bury).

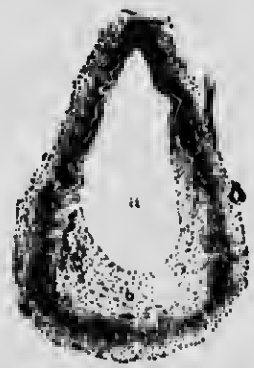


Fig. 216. Middle cerebral artery; a, lumen; b, thickening of inner coat.

In some cases the atrophy and fibrosis of the convolutions are probably the result of chronic meningitis, in others of deficient supply of blood in consequence of periarteritis and endarteritis, but sometimes the sclerotic atrophy is not accompanied by obvious changes in either the membranes or the vessels. Its explanation is then difficult: it may be the result of acute encephalitis; thus a case is reported by Ashby, in which complete idiocy resulted from meningo-encephalitis in a syphilitic infant, the disease ending fatally in six months. But it is remarkable how seldom signs of acute encephalitis have been found, nor is there satis-

factory evidence that encephalitis from syphilis is more than an occasional cause of the spastic diplegias and hemiplegias of infancy (see p. 99). This is probably not owing to the rarity of syphilitic encephalitis, but because the disease proves fatal either in utero or shortly after birth, in consequence either of septicæmia set up by the spirochætes, or of an associated meningitis or hydrocephalus. It is of interest to note that the sclerosis and chronic meningitis may be present after a very short illness. Barlow found extreme fibroid thickening of the pia mater in an infant two months old; Still found thickening of the pia mater and cortical sclerosis in one case after seven weeks' illness, and in another after an illness of only thirteen days. In such cases it is tempting to infer that there was a fœtal encephalitis which gradually led to the development of the chronic changes.

A moderate degree of *hydrocephalus* is not uncommon; frequently it is associated with meningitis at the posterior base or with chronic changes in the brain. Sometimes it appears to have been produced by inflammation of the ependyma of the ventricles and of the choroid plexus, but usually it is the result of syphilitic lesions interfering with the outflow of the cerebro-spinal fluid. Mott bearing in mind the importance of early treatment in congenital syphilis, and the liability of a syphilitic infant to infect a wet nurse, advises that the Wassermann reaction of the blood-serum should be tried in all cases of hydrocephalus in which syphilis cannot with certainty be excluded, or the hydrocephalus accounted for by other causes.

Gummata, both large and small, are occasionally found, and usually in combination with syphilitic disease of the membranes and vessels. In the case of a boy, aged fifteen months, reported by Barlow, there were symmetrical gummata on the third, fourth, fifth, sixth, seventh and eighth cranial nerves at their point of exit from the brain-stem. Cnopf mentions the case of an infant, aged six months, who had convulsions and

hydrocephalus; at the meropsy gummata were found in the corpus striatum on both sides, as well as sclerosis of the cortex.

Symptomatology. In children who have inherited syphilis, *convulsions* are of frequent occurrence, and many infants born of syphilitic parents die of convulsions within the first two years of life; in some of these cases there is post-mortem evidence that the symptom was due to syphilitic changes in the brain. In others such changes were absent, and just as in adult epilepsy occurring in a syphilitic subject the question arises whether the convulsive seizures may depend on the presence of the spirochætes or their products in the blood, without any definite cortical lesions, so in juvenile epilepsy we may ask whether syphilis in the parents can interfere with the proper development of the highly differentiated plasma of the cortex, rendering it unstable and prone to discharge on the slightest excitation. But whether or not this may occur, there can be no doubt that in a large number of cases after a shorter or longer interval, fits in a syphilitic child are replaced or accompanied by other cerebro-spinal symptoms. A very early sign is exaggeration of the knee-jerk, and sooner or later unilateral spasm, paralysis of one or more limbs, ocular palsy and progressive mental defect are likely to develop.

The fits are usually bilateral, consisting of tonic and clonic spasms. In some cases laryngismus and carpopedal spasms occur, in others recurrent attacks of opisthotonos followed by persistent head retraction for varying periods, probably in consequence of inflammatory processes at the posterior base; in one infantile case such meningitis was found, with a softening gumma in its neighbourhood. It is important to note that syphilitic infants may have bilateral fits and laryngismus within the first year, associated with or shortly succeeding the snuffles and rash, and may then have a period of latency for months or years, and subsequently become affected with unilateral spasm or with paralysis.

Complaints of definite *headache*, worse at night, are

rarely made by children under ten years of age. There is, however, frequent evidence of great *irritability*. Syphilitic infants sleep very badly; they are subject to phases of continuous screaming, and in some of these cases diseased membranes have been subsequently found. There may also be cycles of unilateral convulsions, paresis and torpor ushered in by excessive irritability and stiffness of the neck with the head either retracted or held to one side.

Hemiplegia is not uncommon, being as a rule a result of endarteritis with sclerosis and meningeal thickening. It is usually preceded by unilateral convulsions, which may recur at varying intervals on the paretic side. Sometimes hemiplegia occurs without any initial spasm: the patient, without obvious warning, may fall down and lose consciousness for a time. In other cases there is some prodromal restlessness, irritability and vomiting, and then, without loss of consciousness, the patient suddenly loses power down one side of the body and shows impairment in speech. Attacks of hemiplegia are sometimes followed by marked torpor; in many of the initial attacks the paralysis, especially that of the face, clears up to a considerable extent. But there is a great proneness to subsequent seizures, after which a spastic condition may supervene; subsequently the other side of the body may become paralysed when one side may be spastic and the other limp, or more commonly both sides are more or less spastic. In one of the author's cases both pyramidal tracts were degenerated.

Defects of speech, of the nature either of dysarthria or of aphasia, occur; as a rule they are more temporary in the child than in the adult, though at a later period, when extensive degenerative changes, associated with mental failure, have supervened, speech may be considerably impaired or completely lost.

Any one or several of the *cranial nerves* may be affected, apart from disease of the brain or its membranes, by gummata or by interstitial neuritis. The nerve affection may be symmetrical, involving several

pairs of nerves, or it may be unilateral. Separate portions of both the third and fifth nerves may be paralysed, the other portions being intact. Two cases of ophthalmoplegia externa in congenital syphilis are recorded by Hutchinson; in one of them atrophy of the optic discs was also present. Very rarely the facial nerve is affected.

In addition to the early iritis and the interstitial keratitis the presence of *choroiditis disseminata* is a most important sign of congenital syphilis. The condition may be present when there are no nervous symptoms; it should therefore be looked for in all children in whom syphilis is suspected, for its detection would lead to the administration of mercury which might prevent the development of further lesions. Choroiditis is frequently associated with manifestations of intracranial syphilitic disease; it may be present in the youngest infant as well as in children of five or more years. Its degree varies from a few small spots of brownish exudation to marked atrophy with large aggregations of pigment. Vision is not necessarily impaired even when the choroidal disease is of great extent. Atrophy of the disc which is found in many cases of congenital syphilitic brain disease is frequently due to the participation of the disc in a general choroido-retinitis; sometimes it occurs independently as a primary affection.

The intractable *deafness* which may affect the subjects of congenital syphilis comes on usually between the periods of five years before and five years after puberty. It is bilateral, painless and unattended by otorrhœa, and is probably the result of damage either to the internal ear or to the auditory nerve. Many authorities believe that deaf-mutism is often due to inherited syphilis. It is possible, as the author suggested in the paper referred to, that deprivation of the sense of hearing by hindering the receptivity of the brain, may be an occasional factor in the production of mental failure.

Psychical changes. We have seen that the most common brain lesion in cases of hereditary syphilis is a diffuse affection of the cortex, in which one or more of the convolutions become hardened and shrunk and their cells atrophied, and that this condition is either secondary to a chronic meningitis or to an endarteritis, or gradually develops apart from obvious disease either of the membranes or the vessels. We have also pointed out that the symptomatology of brain syphilis in the child is largely made up of phenomena which might be expected to occur during the development and progress of such cortical changes; the instability of the large cells of the grey matter being expressed clinically by headache, screaming and convulsions, their destruction by paralysis, defects in speech and as many cases indicate, even at an early stage, as in Ashby's case, by failure of the mental faculties.

It is therefore not surprising to find that mental impairment is one of the most frequent and characteristic manifestations of hereditary syphilitic brain disease. The clinical type may be stated to comprise spastic paresis of the limbs, convulsions and a moderate degree of dementia. In these respects it presents a close resemblance to a type-case of "birth palsy," in which as a result of degenerative changes in the cerebral cortex the child is backward or demented, has spastic limbs and is subject to epileptiform seizures (see p. 103). According to the time of its development, its characters and associations the mental disturbance may be considered under the headings of idiocy and juvenile dementia, including general paralysis of the insane.

Idiocy. Congenital deficiency of mind from inherited syphilis is rarer than mental failure coming on in childhood, it may be owing to the number of infants who die before birth, at birth, or shortly after, in consequence of the effects of the invasion of the cranial cavity by the spirochæte pallida. Cases, however, of syphilitic children are met with whose mental functions have never perfectly developed, and such children may

subsequently be seized with convulsions and other symptoms of brain disease. The idiocy may be the result of a fœtal or an early infantile meningo-encephalitis, or of hydrocephalus. It is also probable that the syphilitic virus, without leading to demonstrable intracranial changes, may so devitalise the germ-cells as to arrest the development of the nerve-elements and thus lead to idiocy. In support of this view is the evidence afforded by examination of the blood-serum by the Wassermann reaction. An analysis of the various statistics collected by Mott shows that, in about eighteen per cent. of a large number of idiots, the blood-serum gave a positive reaction. Linser, who examined a series of children of syphilitic parents found that the blood-serum of two-thirds gave a positive reaction, while only one-third of the cases showed any other signs of syphilis. Mott points out that several investigators have found the specific organism in great numbers in the central nervous system of infants dying of congenital syphilis. Thus Ranke examined twelve cases of congenital syphilis, in which the infant was born dead or died soon after birth, and in nine he found numbers of spirochetes in the inflamed meninges and in the walls and lumens of the vessels.

There is therefore [illegible] reason to believe that syphilis plays a larger [illegible] production of idiocy than has hitherto been [illegible] by writers on insanity.

Juvenile dementia. In the majority of cases, however, mental failure comes on in childhood: the child when young is as bright and sharp as other children of the same age and cannot in anywise be called backward. Then at an age varying from five to fifteen his intellect begins to fail: the parents or teachers notice that he no longer learns his lessons as correctly as formerly, that his memory is failing, and that he is less vivacious, takes no interest in his work or his play: the condition then gradually develops into more or less complete dementia. In some cases there are maniacal attacks, or there are fits of excitement, or the patient is bad-tempered and

vicious; frequently there is evidence that he suffers from hallucinations or illusions. As a rule the cases fall into the class of simple intellectual failure; the patients are passive, deprived of memory, do not understand what is said to them and lapse into a purely vegetative existence. Such patients are rarely found in asylums; they are not sufficiently vicious or troublesome, they are apathetic and inoffensive, and are to be found dragging on an existence, aimless and devoid of interest and intelligence, at their own homes, or in union hospitals.

As already mentioned, typical syphilitic lesions are found in the cases of simple dementia, and it would appear from the researches of Mott that such cases are to be separated from cases of *juvenile general paralysis*. Mott points out that the morbid anatomy of general paralysis from congenital syphilis is identical with that of general paralysis from acquired syphilis, and that the clinical manifestations are almost identical. He believes that optic atrophy is commoner in the juvenile than in the adult form of the disease, and that in the child the disease usually runs a longer and slower course than in the adult, and consequently produces more complete dementia and paralysis. In the juvenile form "delusions of a sexual nature and grandiose delusions of wealth, strength and power may occur just as in the adult form, but only when the disease begins in adolescence: the reason being that ambition and sexual instincts do not become habitual passions dominating the will until after puberty, consequently if the mental decay has set in before that period these cannot become a content of consciousness."

In other respects there is a close correspondence between the symptoms of the two varieties. After a stage in which the child shows changes in its character and conduct together with signs of mental failure, the facial expression alters either to one of depressive apathy or to one of foolish contentment, and tremors of the face and tongue with characteristic affections of the speech and hand-writing become manifest. In many cases the

pupils are unequal and inactive to light. The knee-jerks may be either increased or lost; in addition to loss of the knee-jerks other symptoms of tabes may be present and the post-mortem examination may reveal degeneration of the posterior columns as well as of the cortical neurons. In a large number of cases the symptoms of general paralysis develop in children who are already feeble-minded. In such cases the mental symptoms are partly due to arrest of development of the brain in consequence of the action of the syphilitic virus, and partly to "the primary decay and death of the neurones associated with acute destructive changes, the result of congestive stasis and auto-intoxications."

The disease may begin at almost any age; in one of Mott's cases it began at the age of eight, in a case of Percy Smith at the age of thirty. It is therefore not unlikely that, as suggested by Mott, some cases of general paralysis developing in adult life, in which acquired syphilis can be excluded with certainty, may be the result of congenital syphilis.

It must be admitted that in the group of juvenile dementias produced by hereditary syphilis, it is not always easy to distinguish between cases of simple mental failure depending on specific inflammatory lesions, and cases of general paralysis depending on degenerative changes, known as para-syphilitic lesions. In the nervous diseases produced by acquired syphilis a trenchant distinction can be drawn between the early and the late effects of the virus, and it is noteworthy how rare it is to find syphilitic lesions in the bodies of persons who have suffered from either general paralysis or tabes; whereas in cases of juvenile general paralysis, the presence of syphilitic lesions is not uncommon.

A consideration of the evidence relating to cerebral syphilis suggests that the pathology of brain affections from congenital may be more complex than that from acquired syphilis. It seems reasonable to believe that the specific virus is apt to injure the developing brain in more manifold ways than the fully grown one, that

a greater variety of changes may be going on at the same time, an insidious degeneration of the neuronic tissue developing and progressing contemporaneously with, yet independently of, syphilitic inflammation of the extra-neuronic tissue, namely the vessels and membranes. Only in this way can we understand the infinite variety, and often the close association in time, of the cerebral manifestations produced by hereditary syphilis. But whatever degree of truth there may be in these considerations, we can have no hesitation in drawing a marked contrast between the most frequent features of brain disease due to congenital, and those produced by acquired syphilis. Dementia in association with convulsions and spastic limbs must be regarded as typical of hereditary syphilis; hemiplegia, and monoplegia with or without unilateral convulsions as typical of acquired syphilis in the adult. The morbid anatomy of the brain affections of hereditary syphilis consists mainly of chronic meningitis, endarteritis and cortical sclerosis and atrophy; whereas the common lesions in acquired syphilis are central softening from arterial disease and thrombosis, and cortical gummata.

Spinal Congenital Syphilis.

In addition to sclerosis of the pyramidal tracts, as a result of cerebral disease, independent spinal lesions are occasionally met with in cases of congenital syphilis. Periostitis of the vertebræ, pachymeningitis, meningitis, gummata and endarteritis have been found, but they are of rare occurrence. The clinical manifestations of such lesions may correspond to those of a meningo-myelitis, of a localised growth or of multiple lesions; frequently they are associated with symptoms of cerebral disturbance. Dixon Mann has recorded the case of a syphilitic boy who suffered from lumbar pain, paraplegia with exaggeration of the reflexes, paralysis of the bladder and sacral bed-sore. Under anti-syphilitic treatment the boy recovered; the condition was attributed to a local thrombosis of the vessels of the cord, which had

led to softening, but had not produced actual destruction of tissue. In some cases of congenital syphilis the clinical features of disseminated selerosis have been observed; in others those of Friedreich's disease.

Tabes is exceedingly rare under the age of twenty, but a number of eases have been recorded in which the disease began in childhood or in youtb. The reports of such cases usually sbow eitber that the parents bave suffered from syphilis, or that the tabetic children have presented signs of congenital syphilis. In a few of the cases the history pointed to acquired syphilis, the infection having been usually derived from a syphilitic nurse.

In juvenile tabes females are more frequently affected than males, the reverse being the case in adult tabes. Optic atrophy is a common symptom; disturbances of micturition are often present at an early period of the disease. There can be no doubt that very rarely tabes in the adult may owe its origin to congenital syphilis, or even to syphilis acquired in infancy. In the case of a man suffering from severe tabes under my care several years ago, it was quite certain that he had not acquired syphilis in the ordinary way; he had a protracted illness after vaccination in infaney, which the medical man who vaccinated bim regarded as due to inoculated syphilis.

Treatment. It is of paramount importance vigorously to treat the earliest exanthem stage of congenital syphilis, with the object not only of curing the affections of the skin and mucous membranes, but of limiting as far as possible the early damage to tissues which may lay the foundation of subsequent disease. In the early stages of congenital syphilis, mercury is best given by inunction; in the later stages grey powder is the best, but whenever active signs and still more when serious symptoms of the disease appear mercurial inunction should again be employed. To prevent any tendency to gastro-intestinal disturbance the grey powder may be combined with bicarbonate of soda and aromatic chalk powder, or

if diarrhœa is present, with suitable doses of Dover's powder. The iodides may be given as intermediate treatment, but as a rule they are not so well tolerated in children nor so effective as mercury.

In every case of congenital syphilis mercury should be continuously administered for at least a year, and for a longer period if there are any active signs of the disease. Only in this way can we hope to lessen the liability to the epilepsy, the paralyses and the psychical affections, the prognosis of which is of the gravest import, improvement seldom occurring even when the child is submitted to prolonged and energetic treatment.

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