CIHM
Microfiche
Series
(Monographs)

ICMH
Collection de
microfiches
(monographies)

Technical and Bibliographic Notes / Notes techniques et bibliographiques

Tha Institute has attampted to obtain the best original copy available for filming. Faatures of this copy which may be bibliographically unique, which may alter any of the images in tha raproduction, or which may significantly change the usual method of filming, ara checkad below.

L'Institut a microfilmé le maillaur axamplaira qu'il lui a été possible de se procurar. Las détails de cet exemplaire qui sont peut-êtra uniques du point de vua bibliographiqua, qui peuvent modifier une image raproduita, ou qui peuvent exiger une modification dans la méthode normale de filmage sont indiqués ci-dassous.

- ☐ Coloured covers/ Couvertura de coulaur
- ☐ Covars damaged/ Couvertura andommagée
- ☐ Covars restored and/or laminated/ Couvartura restaurée at/ou pelliculée
- ☐ Cover title missing/ La titra de couvertura manqua
- ☐ Coloured maps/ Cartes géographiques en coulaur
- ☐ Coloured ink (i.e. othar than blua or black)/ Encre de coulaur (i.e. autra que bleue ou noira)
- ☐ Coloured plates and/or illustrations/ Planches et/ou illustrations an couleur
- ☐ Bound with other material/ Reliá avec d'autres documents
- ☑ Tight binding may cause shadows or distortion along interior margin/ La raliure serrée peut causer de l'ombre ou da la distorsion la long de la marge intérieure
- ☐ Blank laaves added during restoration may appear within tha taxt. Whenever possible, these have been omitted from filming/ Il se peut que cartaines pages blanches ajoutées lors d'una restauration apparaissent dans le texta, mais, lorsque cela était possible, ces pages n'ont pas été filmées.

- ☐ Coloured pages/ Pages de coulaur
- ☐ Pages damaged/ Pages endommagées
- ☐ Pages rastored and/or laminated/ Pages restaurées at/ou pelliculées
- ☑ Pages discoloured, stained or foxed/ Pages décolorées, tachatées ou piquées
- ☐ Pages detached/ Pages détachées
- ☑ Showthrough/ Transparence
- ☐ Quality of print varies/ Qualitá inégala da l'imprassion
- ☐ Continuous pagination/ Pagination continue
- ☐ Includes index(es)/ Comprand un (des) index

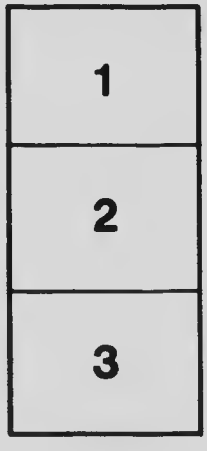
  Title on header taken from:/ Le titra da l'en-têta provient:

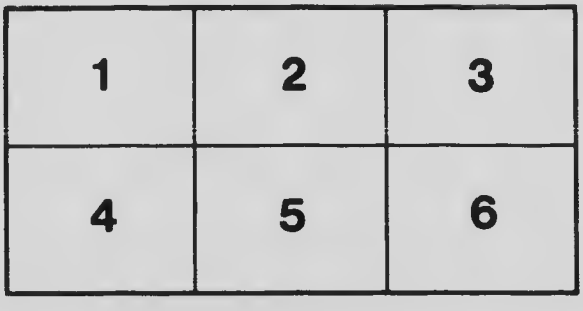
- ☐ Titla page of issua/ Page de titra de la livraison
- ☐ Caption of issua/ Titre de départ de la livraison
- ☐ Masthead/ Générique (périodiques) da la livraison

☐ Additional comments:/ Commantaires supplémentaires:

This itam is filmed at the raduction ratio checkad below/
Ce document est filmé au taux de réduction indiqué ci-dessous.

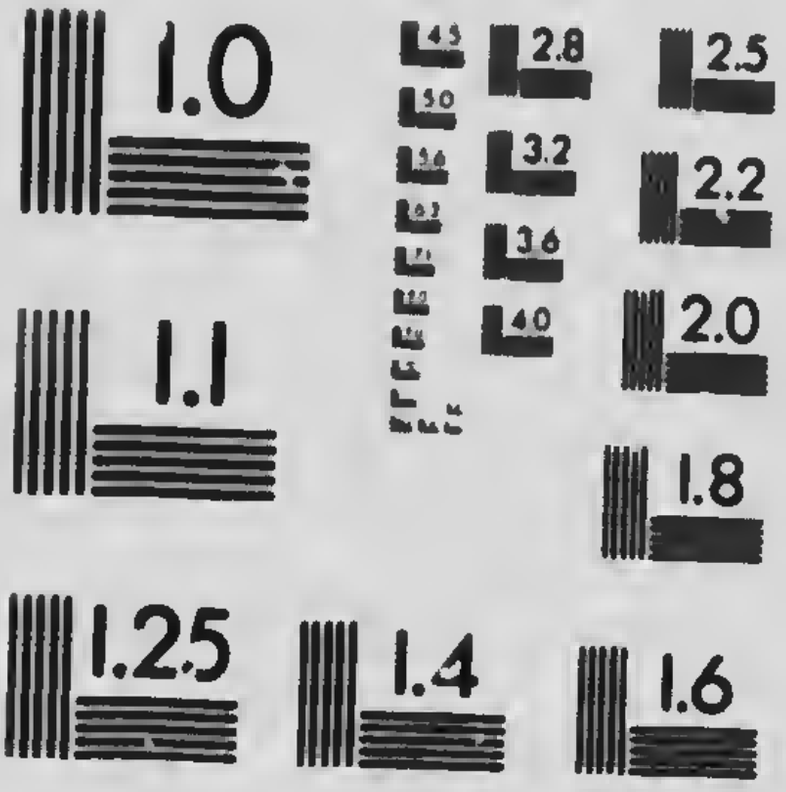
10X	12X	14X	16X	18X	20X	22X	24X	26X	28X	30X	32X
☐	☐	☐	☐	☐	☐	☐	☑	☐	☐	☐	☐





MICROCOPY RESOLUTION TEST CHART

(ANSI and ISO TEST CHART No. 2)



APPLIED IMAGE Inc

1653 East Main Street
Rochester, New York 14609 USA
(716) 482 - 0300 - Phone
(716) 288 - 5989 - Fax

A CLINICAL MANUAL
OF THE MALFORMATIONS
AND CONGENITAL DISEASES OF THE FŒTUS

A CLINICAL MANUAL
OF
THE MALFORMATIONS AND CONGENITAL DISEASES OF THE FŒTUS

BY

PROFESSOR Dr. R. BIRNBAUM

CHIEF PHYSICIAN TO THE UNIVERSITY CLINIC FOR WOMEN AT GOTTINGEN

TRANSLATED AND ANNOTATED

BY

G. BLACKER, M.D., B.S., F.R.C.P., F.R.C.S.

OBSTETRIC PHYSICIAN TO UNIVERSITY COLLEGE HOSPITAL
AND THE GREAT NORTHERN CENTRAL HOSPITAL
TEACHER OF PRACTICAL MIDWIFERY AT UNIVERSITY COLLEGE HOSPITAL MEDICAL SCHOOL ETC.

WITH 58 ILLUSTRATIONS IN THE TEXT, AND 8 PLATES

TORONTO
THE MACMILLAN COMPANY OF CANADA LTD.
1912

Printed in Great Britain

AUTHOR'S PREFACE

Malformations and Congenital Diseases are of interest from several points of view: thus they have an anatomical, pathological, physiological, clinical—more especially in relation to Surgery and Obstetrics—and medico-legal importance.

While it is true that all the facts relating to these various branches can be found scattered throughout different text-books and manuals, as well as in monographs, yet it is difficult for the student and for the busy practitioner—or, indeed, for anyone interested in the subject—readily to acquire a knowledge of them. A work dealing completely with the whole subject is still a desideratum. In this book I have attempted to supply that deficiency: although I am well aware that I have not exhausted the subject: such a complete treatment of the whole matter could only be accomplished by the ready co-operation of workers in all the branches of medicine concerned. Neither I nor my publisher have had any intention of supplying such a treatise.

Nevertheless, I have endeavoured, as far as I deemed it necessary for this work, to collect and make use of all the available material in the general literature. As a gynecologist, I have treated the obstetrical side of the question as of most importance and in greatest detail—next to the anatomical and pathological. A large number of malformations have only, or at least primarily, a purely obstetrical importance.

While, on the one hand, the carrying out of the work has been greatly facilitated by the numerous exhaustive treatises which exist—I need only mention the works of Schwalbe, Ahlfeld, Marchand, Kleinhans, Hohl, Ziegler, Strassmann, Kaufmann, Taruffi, St. Hilaire, Dareste, Förster, Ballantyne, Tillmanns, and Küstner—yet, on the other hand, it has been rendered the more difficult, since in this branch of medicine such an enormous amount of literature exists, which for the purpose of this book could only be utilised to a small extent. With

regard to the surgical treatment of malformations, I have contented myself with referring the reader to text-books on surgery.

The monograph of Hohl has been of the greatest use to me in compiling this work, as has also been the newer and more comprehensive work of Schwalbe—which, however, at the time of going to press, has not been completed.

I have not included in this book a consideration of all the congenital diseases, as, had I done so, the size of the work would have been considerably increased. Thus, for example, congenital syphilis (except for a few short allusions), the congenital infectious diseases, and asphyxia neonatorum have not been considered; nor have the congenital diseases and anomalies of the fœtal membranes and placenta. All these conditions are fully described in text-books on obstetric medicine.

The illustrations are for the most part original photographs taken from specimens in the collection of the Göttingen Women's Clinic. To my chief, Herr Geheimrath Runge, my heartiest thanks are due for the willingness with which he has placed at my disposal the material of his clinic for the preparation of the photographs: specimens from his collection, and medical journals. A few are from the Anatomical Institute, and in this connection I have also to thank Herr Geheimrath Merkel for so kindly giving me permission to make use of his specimens. I desire also to thank Dr. Finger, who has taken the majority of the photographs.

R. BIRNBAUM.

GÖTTINGEN.

TRANSLATOR'S PREFACE

In undertaking a translation of this book, I was influenced mainly by the fact that no work of precisely similar scope had been published in the English language. Ballantyne's classical work—a mine of information, to which I cannot sufficiently acknowledge my indebtedness—is concerned mainly with the subject of Teratology from the pathological point of view, and hardly considers the important question of treatment; nor does it discuss the subject to any great extent from the clinical aspect. It seemed to me, therefore, that Professor Birnbaum's book would supply a want, and that a translation of it might prove useful both to the practitioner and to the student.

I have ventured to add a considerable number of notes to the original work, partly explanatory of the text and partly containing additional information. These, I hope, will increase the usefulness of the book, and at the same time will help the reader to understand the close relationship which exists between the facts of normal embryology and the mode of development of many of the malformations and monsters described.

The addition of this new matter has increased the size of the book by some 100 pages. Four of the original illustrations now appear as plates, and four new plates and three new illustrations in the text have been added.

I wish to express my thanks to Mr. E. G. Heumann for his valuable assistance in the translation of the first half of the book; I, however, am responsible for the whole of the work and for any faults which may be found in the English version. To my friend Dr. Clifford White, I am greatly indebted not only for his assistance in looking over the proofs, but also for many most valuable suggestions and corrections. I have also to thank my colleagues, Mr. Morriston Davies and Mr. Cooper, for taking some photographs for me.

LONDON. G. BLACKER.

CONTENTS

ILLUSTRATIONS

FULL PAGE PLATES

ILLUSTRATIONS IN TEXT

MALFORMATIONS

CHAPTER I

The Causation of Malformations

A malformation is generally defined as a condition which may be attributed to more or less important deviations from the normal course of intra-uterine development. (On the history of malformations the reader should consult the works of Hohl and Schwalbe.)

If the case is one of very marked abnormality of form, with pronounced deformity of the individual, it is termed a 'Monster'; while less conspicuous deviations, which affect only circumscribed portions of the body, are termed 'Anomalies' (Ziegler and Schmans).

Schwalbe's definition is perhaps a better one. He defines a malformation as an alteration occurring during fœtal development—e.g., a congenital alteration of one or more organs or systems of organs, or of the whole body, which does not come within the range of variation of the species.[1]

The difference between malformations and congenital diseases, which are also considered in his work, is defined by Schwalbe in this way: namely, a malformation is an alteration of a part already existing, or a permanent abnormal condition; while a congenital disease, on the contrary, represents a condition which may continue to progress after birth. A fœtal disease, however, may also, although very rarely, produce a lasting change of form or a malformation.

In the great majority of cases malformations are found affecting only

[N.B.—All the footnotes throughout the book are by the translator.]

[1] An alteration, for example, which does not come within the possible variations in form of any organ or organs in normal conditions. The possible variations which may be considered as normal for any particular organ can naturally only be determined as a matter of observation and experience.

'For the monstrosity belongs to the class of things contrary to Nature: not any and every kind of Nature, but Nature in her usual operations' (Aristotle, *De Generatione Animalium*, book iv., 4 (1910); transl. by Prof. Arthur Platt).

a single individual, and on this account they are termed 'Single or Autosite Malformations.'

In a small number of cases, however, two individuals are affected by the malformation, and then it is termed a 'Double Malformation.'

The aetiology of malformations depends either upon internal causes or upon the action of external influences. I will not enter here on a consideration of the very interesting questions of the mechanism of their development, experimental teratology, or of their regeneration. On these subjects the reader should consult Schwalbe.

As nearly all malformations originate in the first three months of embryonic life[1]—that is, at a time when the development of the form of the embryo mainly takes place, whereas the so-called fœtal diseases (for example, fœtal rickets or fœtal endocarditis), attack more especially the already formed fœtus—these need not be considered in discussing the aetiology of malformations. Among the internal influences are included inheritance, atavism, and so-called variation of the germ. By a malformation the result of inheritance is meant a case of disturbance of development such as has occurred in the ancestors of the individual: for example, supernumerary fingers and toes, supernumerary nipples, abnormal hairiness of the skin, hare-lip, spina bifida, and multiple fibromata in the course of the nerves. Latent inheritance is defined as the condition in which an individual transmits the malformations of his parents to his descendants without having been affected by them himself. Thus, for instance, malformations of the male sexual organs may be transmitted through the female members of the family to their descendants. In the case of crossed inheritance, characteristics of the father are to be found in the daughter, and those of the mother in the son. So-called collateral inheritance is also met with, for example, in hæmophilia, when the father and mother of the hæmophiliac are healthy whereas an uncle is a bleeder. If there are no malformations in the parents of the individual in question, but they occurred in the case of previous generations, and yet were absent in the intermediate members of the family, the condition is termed atavism. Schwalbe suggests that the term atavistic inheritance should be replaced by avitary inheritance.[2]

In the so-called primary variations of the germ, both the influence of inheritance and external influences are wanting, and the malformation

[1] It would probably be more correct to say during the first six weeks of intrauterine life—that is to say the embryonic period or formative stage during which the parts and organs of the embryo are formed. Teratology is in reality the pathology of the embryo (Ballantyne), or embryology in disorder, which has for its result the development of monstrosities and malformations.

[2] These facts require for their explanation some assumption such as that they are already determined in the germ, and this introduces the so-called 'germinal factor' into the consideration of the causation of malformations and deformities.

occurs for the first time in the family. In this case, it is possible that either one of the two cell nuclei—the female pro-nucleus or the male pro-nucleus, or even both of them—are abnormal, or that through the union of two normal nuclei an abnormal variety may result.

Among external causes the following play an important part: mechanical influences—such as concussions, blows, or long continued pressure as a result of want of space; for example, through anomalies of the amnion, or of the uterus, such as a uterus bicornis[1] or pregnancy in a rudimentary horn. Further, from an analogy with experimental investigations, failure in the supply of oxygen and disturbances of nutrition, chemical influences,[2] the effects of poisons, variations in the temperature; and psychical influences, such as maternal impressions, must also be taken into consideration.

Concussions of the abdomen—especially those affecting the uterus—may have a directly dangerous influence upon the embryo; for, from this cause, haemorrhages may occur between the ovum and the uterine wall, giving rise to loosening or actual separation of the ovum from its attachments, and so leading to disturbances of nutrition. Haemorrhages of this kind, with resulting damage to the ovum, may be set up by various causes: thus, in nephritis of the mother, through toxic influences on the vessels by disease of the decidua, by myomata of the uterus; by certain infectious diseases affecting the mother, such as typhoid fever; and by certain poisons. In the majority of cases, death of the foetus occurs; and its premature expulsion soon follows. With regard to other mechanical causes it is difficult to prove that changes in the temperature, an insufficient supply of oxygen, or chemical influences, are conditions dangerous for the life of the intra-uterine ovum.

Psychical causes, such as the above-mentioned maternal impressions, cannot come into play in the manner in which they are usually understood by the public. Schwalbe, however (quite rightly) lays stress upon the fact, and it cannot be denied that mental emotion, more especially in sensitive natures, may lead to the premature expulsion of the foetus. In the same way the possibility also exists of some resulting interference with the process of normal development.

[1] See a case recorded by F. v. Winckel, *Munchener Medicin. Wochenschr.* (1896), vol. xliii., p. 390.

[2] For a full account of the influence of various chemical substances upon the development of malformations, the reader should refer to the writings of Féré, who has done much to elucidate this point. His results clearly prove that certain toxic substances can, by their influence upon the embryo, lead to malformations. Whether these are brought about by the action of a diseased amnion, or are due to disturbances of nutrition, remains for further investigations to show: but that such an influence may exist is certain.

As regards any further effect, maternal impressions may be regarded as fables and rejected as unscientific.[1]

Finally, as regards the effect of poisons on the developing ovum, the possibility of such an influence cannot be denied, for example, through over-indulgence in alcohol or the continued administration of powerful drugs (Schwalbe). I have already mentioned that fœtal diseases probably do not play an important part in the production of malformations. Of all the known causes, the pressure exercised by the uterus and the fœtal membranes, as a result of a deficiency in the amount of liquor amnii, is undoubtedly the most important (oligohydramnios). The damage caused by this condition manifests itself mainly in the extremities (club feet, curvatures of the limbs, congenital dislocations, wry neck, and anomalies of the vertebral column).

According to the views of some authors—e.g., v. Winckel—anomalies of the amnion play the most important part in the production of malformations. The so-called amniotic or amniogenic malformations, which are thought to owe their origin to such anomalies, are considered at the present day to be of great importance. Thus many authors believe that, more or less, all—or, at any rate, all external—malformations have an amniotic origin. Such anomalies of the amnion are pathological narrowness of the amniotic cavity, abnormal adhesions and band formations, defects of the amnion and hydramnios. Want of development of the amnion undoubtedly may lead to serious malformations through pressure[2] on the embryonic germ.

Schwalbe mentions several instructive examples of this kind. It is supposed that the defective formation of the amniotic cavity is due to imperfect secretion of the liquor amnii. As is well known, the amnion

[1] After a very complete and careful summary of all the evidence, both in favour of and against the hypothesis of maternal impressions, to the two questions—(1) Does a definite impression upon a pregnant woman's mind often or ever cause a defect in the fœtus closely resembling the thing producing the impression? and (2) Has the state of the mother's mind during gestation any effect upon her unborn infant's development?—Ballantyne answers the first in the negative and the second in the affirmative, and with these conclusions the great majority of English writers are in complete agreement.

[2] The view that pressure played an important part in the production of monsters is to be found in the writings of Hippocrates, and still more certainly in Aristotle (see *De Generatione Animalium*, book iv., 4 (1910); transl. by Professor Arthur Platt). It must be borne in mind, in reading the passage in which Aristotle describes the production of monsters in the fowl, that he thought the chick was formed from the white of the egg. As Ballantyne remarks, more than two thousand years elapsed before the views of Hippocrates and Aristotle found expression and acceptance in the works of Paré.

It must be remembered that any effect pressure may have will depend very largely upon the amount of liquor amnii present, and that the same malformations may occur in cases of hydramnios and oligo-hydramnios. As Ballantyne points out, the obvious result of pressure, as seen in a fœtus papyraceus, is not necessarily teratological.

at first, lies in close relationship with the embryo, and its separation from it takes place later through the agency of the secretion of the liquor amnii.

To such a deficiency of the amnion the following malformations may be attributed: anencephalus, cyclopia, synecha, and other marked defects of the cephalic and pelvic extremities; curvature of the spine, and malformations of the limbs (Dareste and Schwalbe). Further than this, the amnion may be more or less absent (Dareste); such a defect may exist primarily, or it may be the secondary result of a rupture of the amnion and of the chorion, or of the amnion alone. With the first condition rupture of both membranes of the ovum is associated so-called 'Hydrorrhœa uteri gravidi'[1] which develops most commonly from the third to the fifth month of pregnancy. In the majority of cases this results in an abortion. In a very few cases the pregnancy continues until the sixth or the eighth month, and the fœtus may even be born alive (Stoeckel). One such case has been described by Fleck from the Göttingen Clinic.

Almost always after the rupture of the membranes of the ovum, the fœtus escapes from the amniotic cavity forming the so-called 'Grossesse extra-membraneuse.' The membranes shrink and retract to the surface of the placenta. Owing to the deficiency of the liquor amnii the possibility of movements of the fœtus taking place is much curtailed, and thus it often happens that it is born with malformed extremities or ankylosis.[2]

In a few cases, rupture of the amnion alone has been noted in the first half of the pregnancy. In this case, the amnion retracts to the fœtal surface of the placenta, and the fœtus is then surrounded only by the chorion 'Fœtus extra-amniotique.' The cause of this isolated rupture of the amnion is not clearly known. Possibly the band-like adhesions which are found in most cases between the amnion and the fœtus play an ætiological rôle (Stöckel). According to some authors, however, these bands only form after the rupture of the amnion. In such a case the pregnancy usually proceeds to term. As a result of the extensive amniotic adhesions severe malformations may be met with in the child.

An early hydramnios may also be a cause of malformations through pressure on the embryonic rudiment. The view still widely held at the present day that hydramnios only occurs in the later months

[1] This should be termed 'Hydrorrhœa uteri gravidi amnialis,' to distinguish it from the variety of hydrorrhœa due to excessive secretion from the glands of the decidua vera and capsularis, so-called 'Endometritis decidualis.' A further point of distinction is that in this variety of hydrorrhœa, uterine hæmorrhage nearly always occurs, either at the same time as the escape of fluid or alternately with it. Stöckel has collected eighteen cases of this kind.

[2] Meyer-Ruegg has collected twelve cases.

of pregnancy is entirely erroneous. In pregnancies which are far advanced, hydramnios can no longer be considered as a cause of malformations. Yet hydramnios, as we shall see, is a very frequent accompaniment of certain malformations. This is especially so in cases of hemicephalus, hydrocephalus, and in the majority of fissure formations—such as hare-lip, spina bifida, ectopia vesicæ, etc.[1]

I will take this opportunity of considering briefly the causes of hydramnios when met with in association with malformations (see also Küstner, Seitz, and others).

According to Küstner, hydramnios is only met with in cases of spina bifida and hemicephalus when the defective portions of the brain and spinal cord are not covered with epithelium, or when the floor of the ventricle, or the floor of the spinal canal, lies exposed. We may also suppose that the increased amount of fluid results from the rupture of an hydrocephalus, or of a spina bifida, while some authors also attribute the origin of an hemicephalus to such a rupture.

While this possibility must be admitted, yet Küstner points out correctly that the formation of an anencephalus from an hydrocephalus takes place at a very early period of fœtal life when the cerebro-spinal fluid is present in only a very small amount (a few tablespoonfuls).

It is easier to understand the formation of the hydramnios, if we accept the explanation of Lebedeff of the development of an anencephalus. He attributes the formation of an anencephalus to an abnormally marked curvature of the embryonic rudiment, so that the head end grows excessively in length. As a result of this the transformation of the medullary plates into a medullary tube is hindered, or the already-formed medullary tube opens out again (see Ziegler). The development of an hydramnios is then capable of a very simple explanation. The fluid is secreted by the portions of the central nervous system not covered by epithelium. Küstner considers the following explanation a very probable one. In an anencephalus, the floor of the fourth ventricle either lies freely open, or is exposed to the action of the liquor amnii. In this way a stimulus is set up analogous to that produced by the experimental puncture of the medulla, as a result of which, as is well known, an increased secretion of urine and excretion of sugar is produced. According to this view, one portion

[1] We may sum up, then, the supposed causes of malformations by saying that they are to be explained by one or other of the following three factors—namely: (1) the germinal factor, in which is found the explanation of twins and double monsters, and the transmission of inherited malformations; (2) the action of the amnion; (3) changes in the nutrition whether the causes of this are to be found in the action of the amnion, or in the influence of certain chemical bodies, toxic or otherwise.

of the hydramniotic fluid must consist of the fœtal urine secreted in excess. In other malformations, too, frequently accompanied by hydramnios, various body-cavities may be exposed: namely—the abdominal cavity, the thoracic cavity, and the bladder.[1]

Seitz further points out that a large number of malformations may be attributed to the amniotic bands (described in detail further on), and that these again are associated with anomalies, and, probably also (in the early stages of intra-uterine existence), with inflammatory conditions of the amnion; so that structural alterations of the amnion may also play a part in the production of hydramnios.

Hydramnios, moreover, is found relatively frequently in some fœtal diseases and abnormalities which result in disturbances of the circulation: stenosis of the umbilical vein, phlebitis and thrombosis of the umbilical vein, multiple torsions of the umbilical cord, cirrhosis of the fœtal liver associated often with ascites, anomalies and diseases of the heart, inflammation of the lungs, and other affections. In association with anomalies of the umbilical cord the occurrence of hydramnios is sufficiently clear. In fœtal diseases, too, the mechanism of its origin, (which is the same) is easy to understand; thus, through the occurrence of disease of any particular organ, an obstacle may be opposed to the flow of the blood returning through the umbilical vein to the fœtal body. In this way congestion in the umbilical vein is brought about and a transudation of fluid occurs into the amniotic cavity from the ramifications of the veins on the fœtal surface of the placenta (Küstner).[2] Into the question of the pathology and ætiology of hydramnios in the case of uniovular twins, I do not propose to enter. For further details the reader should consult text-books on Obstetrics.

We must now consider the most important and frequent anomalies of the amnion: namely—amniotic bands and constrictions. They undoubtedly play a most important part in the production of malformations. Schwalbe gives some very instructive illustrations of malformations of amniotic origin.[3]

[1] It must be remembered that such anomalies often occur without any hydramnios.

[2] For further information the reader should refer to Ballantyne's discussion of this question. He points out that hydramnios is in reality the persistence of a state which is normal in the early months of pregnancy; for at the fourth month the liquor amnii weighs more than either the fœtus or the placenta and membranes. He regards the condition as sometimes due to a chemical irritant coming from the mother or formed in the fœtus, or it may be caused by changes in the umbilical vein, or it may be the result of changes in the maternal blood, or it may be in part fœtal urine or cerebro-spinal fluid.

[3] Schwalbe lays down two criteria for the recognition of malformations as of amniotic origin—namely: (1) that amniotic bands or adhesions are present in the neighbourhood of the malformation; and (2) that it is of such a nature as could be produced by their action.

Adhesions between the amnion and the surface of the embryo are frequently present at the time of birth in the form of membranes, bands, and cords. It is indeed often possible to determine the ætiological relationship of such bands to the existing malformations. These bands are commonly known as Simonart's ligaments. If the adhesions between the amnion and the embryo are very extensive then serious malformations of the cranial or facial portions of the head may ensue (see Fig. 1). Simonart's ligaments have, further, an obstetric significance (see chapter on the congenital anomalies of the skin). Thus difficulties in delivery may arise when the amniotic bands pass from the fœtus to the placenta, as in this way a close attachment of the fœtus to the uterus may occur. Hein describes a case of a band-like attachment between the placenta and the dura mater. The child was born as a footling presentation, and on attempting to hand the screaming child to the nurse Hein found to his astonishment that the head, although born, was still closely attached to the mother. On making an internal examination he found a firm band attached to the head which led to the after-birth. The placenta was easily expressed by Credé's method. According to Hohl, the head is the most frequent site of attachment, when there are abnormal adhesions of the fœtus to the placenta or to the membranes.

In the Göttingen Clinic Museum there is a very interesting preparation of this kind of which complete details are, however, wanting. On referring to Fig. 1, it will be seen that there is a very close attachment between the placenta and the dura mater in the shape of a broad adhesion of the amnion with the surface of the head. The vault of the skull is completely wanting, and the short umbilical cord is inserted into the broad amniotic membrane which stretches from the placenta to the skull of the child, and in this the vessels of the cord spread out on their way to the placenta (velamentous insertion). There is also a condition of exencephalus, the eyes are present but the left one is displaced outwards and forwards, and the nose is rudimentary and malformed, otherwise the child is normally developed and at full term. A very similar specimen is figured by Stöckel in 'v. Winckels Handbuch der Geburtshülfe,'[1] vol. ii., pt. 3, plate 3.

[1] In the University College Hospital Medical School Museum there is a good example of a malformation—no doubt the result of an amniotic adhesion. The specimen (No. 802) is that of a female fœtus, with anencephalus, absence of the nose, maldevelopment of the upper lip, and prolongation of the mouth so that it forms a large triangular opening with the apex upwards. Adherent to the upper angle of the mouth and to the membrane covering the base of the skull is an amniotic band. The liver, spleen, stomach, and intestines protrude through an opening in the abdominal wall. Both feet are in a condition of talipes equinovarus, the right radius is wanting, and there are only two digits and a rudimentary thumb on the right hand.

If the amniotic bands are cord-like, then whole extremities or portions of them may be constricted and separated and even become absorbed—

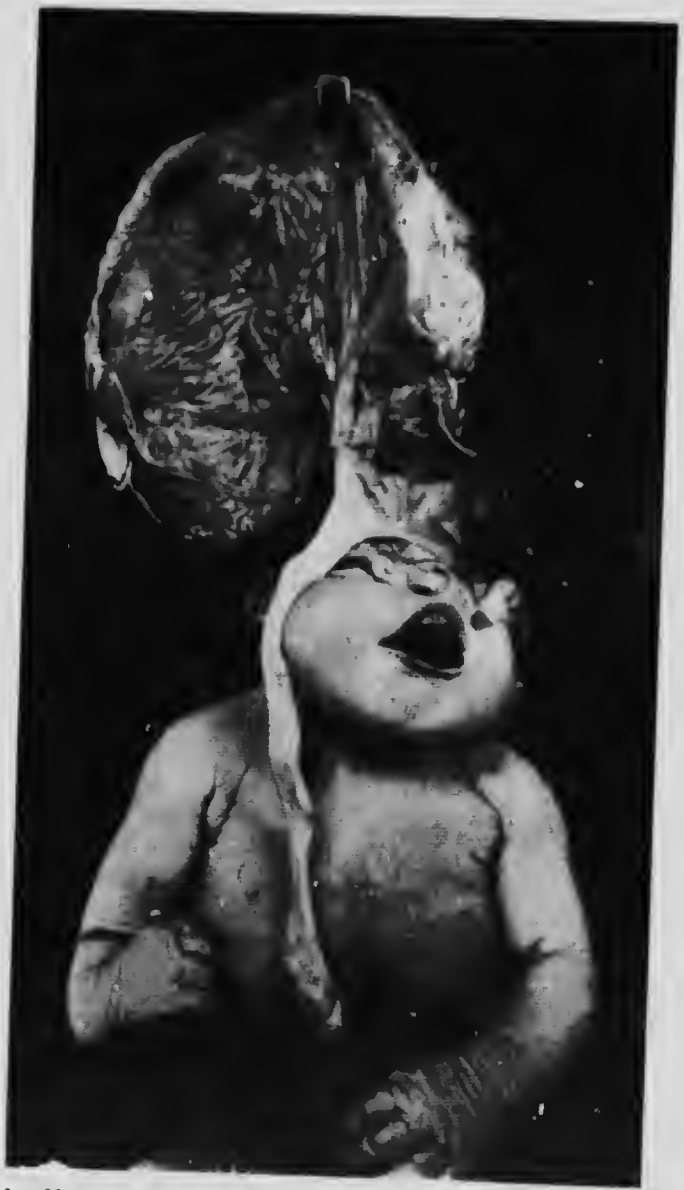


Fig. 1.—Union of the Amnion with the Head, Abnormally Short Cord. (Specimen from the collection of the Göttingen Women's Clinic.)

a condition known as spontaneous amputation; or the cords may lead to mutilations—more especially of the fingers and toes (Fig. 2).

The freely floating appendages which are found at times attached to the amnion or the fœtus, are thought to originate from Simonart's ligaments, and no doubt they are derived from the tearing of these ligaments.

A case of this kind, associated with spontaneous amputation, occurred a few years ago in the Göttingen Women's Clinic. The mother was twenty-three years of age (Jr. No. 15996), had had five children and her previous pregnancies had all run a normal course. She was admitted into the clinic in labour, with hæmorrhage as a result of the separation of a normally situated placenta. At the birth the child was in the position of a complete footling presentation with the right leg, which was amputated below the knee joint, prolapsed. The completely healed amputation stump, with the joint of the knee lying above it, was mistaken at first for a shoulder presentation. The further course of the labour was quite normal. The fœtus (see Fig. 2) was 35 cm. long and weighed 1100 grm. Owing to other amniotic constrictions, besides the spontaneous amputation below the right knee-joint, the index and middle fingers of the right hand were missing, while the index and middle fingers of the left hand were also slightly mutilated. From both hands some Simonart's bands about 3 cm. in length were hanging.

Fig. 2. — Spontaneous Amputation. Malformation of the fingers as the result of amniotic bands. (Specimen from the collection of the Göttingen Women's Clinic.)

With regard to the origin of these cord-like or flat adhesions of the amnion to the fœtus, there is a good deal of dispute as to whether they occur as the result of simple coalescence or as the result of inflammatory reaction.

Küstner calls attention to the fact—and in my opinion he is correct—that it is not necessary to take into account the conditions which might cause inflammation. As the epithelial cells in early embryonic life show no keratinisation, it is quite possible for adhesions between the amnion and the epidermis to occur, if their epithelial surfaces are in contact with one another for a sufficient length of time and sufficiently intimately—a condition which is fulfilled when the development of the amnion is imperfect, when the liquor amnii is scanty, or when some displacement of the amnion (such as a folding of it) occurs. As time goes on these primarily simple epithelial adhesions become firmer, organised, vascularised, and thus persistent. If later on an increased accumulation of the amniotic fluid takes place, the adherent surfaces become separated again and the remains persist as shreds of amnion adherent to the skin, or the adherent portions are drawn out by the separation of the amnion, and form the so-called bands or cords. If the external portions of the embryo at the time of this occurrence are undergoing development then the adhesions cause an arrest of this process, and the embryo remains at the stage of development already reached. In this way arise cranioschisis, fissures of the face, and hare-lip. In another way these adhesions favour the formation of fissures with ectopia of the internal organs, since they hinder the union of the edges and so bring about ectopia of the bowels and ectopia cordis. For the clear explanation of this condition, we have to thank Küstner. Schwalbe believes that possibly such adhesions can only occur after shedding of the epithelium, but he believes also that they can occur in the manner described. For further details I must refer the reader to the very interesting remarks of Schwalbe upon this subject.

In rare cases it has been found that the amputated extremities acquired attachment to other parts of the body—e.g. a foot to the breech (Ahlfeld, 'Monatsschr. f. Geburtsh. u. Gynäk.' (1905), p. 191). A very interesting and unique case, probably of this kind, has been published by Th. Landau in the 'Berliner klin. Wochenschr.' (1909), p. 55. The case in question is that of a malformed fœtus of about five months, the sex apparently female, with an indication of labia but no vaginal opening, atresia ani, the trunk cylindrical in shape, double club-foot, and the left upper extremity almost wanting (Micromelus). The left upper arm and forearm measured 5 cm. in length, while the hand was 1·5 mm. long. The upper end of the trunk is rounded and ball-like in the form of a cupola, while the head is entirely wanting, and apparently the condition is one of acardius acephalus. The head indeed is present, but it is attached at the insertion of the umbilical cord into the placenta, and forms a very striking contrast to the rest of the body as it is only the size of a hazel-nut, and slightly shrivelled. The eyes, the nose, and the opening of the mouth can be distinguished readily

(see illustration by Landau). Attached to the head is a short and slightly rotated neck which is firmly implanted on the amnion so that it appears as if it grew out of this structure. The development of the fœtus corresponds to that of the fifth month; that of the head at most to the second month. Landau regards this case as one of intra-uterine amputation of the head—either the result of the coiling of the cord round the neck at an early date or the result of the action of an amniotic band. The umbilical cord was, however, quite normal, and there was no Simonart's ligament present. The head, after amputation, evidently healed with its raw surface attached to the amnion. A further explanation is possible if we accept Graf v. Spee's view of the mode of origin of the amniotic cavity.[1] According to this view, the cavity is formed in a primarily solid mass of cells placed at the dorsal aspect of the embryonic cell mass, as a result of a process of gradual absorption. During this process it is possible for bridges of tissue, cords or bands, to remain, which might bring about such an amputation as occurred in this case.

A head severed in this way may become absorbed, and then the condition would be one of pure acephalus.

Before I conclude my remarks on the causation of malformations I will just refer to those malformations—or, better, anomalies—in which certain conditions of the embryonic period persist into extra-uterine life: for example, the persistence of the ductus Botalli, or of the foramen ovale.

The sex of malformations in a curiously large number of the cases is feminine, as the present work will show. As Schwalbe points out, this preponderance is due to the disproportionately greater frequency of female double monsters. Among the double monsters the proportion of female to male is as 3 to 1; and in single monsters, according to Marchand's statistics, of 158 there were 55 female and 103 male; not 93, as is erroneously stated in Marchand's work.

It is not an easy matter to give correct statistics as to the frequency of malformations. Some figures only include the more marked forms of malformations, while others include even the slightest anomalies, such as colour of the hair, etc. Many malformations are indeed only discovered for the first time at an autopsy. For more accurate statistics it would be necessary to determine first how far the various

[1] The examination of other early human ova has shown that Graf v. Spee's view as to the mode of origin of the amnion is probably correct; and the case, therefore, with which bands and bridges of tissue can remain is evident. It must be remembered, however, that we have no exact knowledge of the mode of formation of the amnion in the human embryo. In the Teacher-Bryce ovum, the youngest yet described, estimated to be about thirteen to fourteen days old, the amnion was already formed. It may, therefore, as Ballantyne suggests, be possible to speak of some disease of the amnion—'amniotitis,' for example—as producing malformations of the embryo, the formation of which is preceded by that of the amnio

conditions mentioned should be included. The following figures are given in Schwalbe's work: Chaussier (1812), 132 in 22,293 births; Isidore Geoffroy St. Hilaire, 1 in 3000 births; Puech, 7 in 778 births (he also found, among 100,000 births, 434 simple anomalies, 61 single malformations, and 2 double malformations); Schworer, 1 in 455 births; v. Winckel, in Dresden, among 10,056 new-born children, 156 anomalies; in Munich 232 among 8149 (see also the table given by Schwalbe).

Very frequently, malformations are found combined in the most various ways. Generally, it is difficult to determine whether this is a matter of purely accidental occurrence or of some definite relationship. French writers maintain that they have discovered definite laws with regard to such occurrences (Schwalbe).

Classification of Malformations

the classification of malformations, I have followed the scheme which is mostly in use at the present day; but I would say here that in the course of the work, I have not followed this classification closely or entirely, for other practical reasons. An additional ground for doing so is that I have included Congenital fœtal diseases within the scope of this book. In my descriptions, I have followed mainly a classification concerned primarily with the various regions of the body. A classification on a natural system is not possible at present, since we are not yet sufficiently acquainted with the mode of origin of the various malformations. The most commonly employed system of classification of malformations is as follows. (For the classifications of Marchand, St. Hilaire, and others, see Schwalbe.)

A. Single Malformations

1. Malformations due to arrest of development (Monsters by defect).

To this class belong cases of hypoplasia of the whole body or of parts of the body; agenesia or aplasia of single organs or of parts of the body; malformations due to arrest of development in the form of fissures or duplicatures—namely: fissures in the median line of the thorax or of the abdomen, fissures in the face, doubling of the vagina and uterus, fusion or union of organs lying in relation to one another—for example, the kidneys or the eyes.

2. Malformations due to excess of development (Monsters by excess).

This group exhibits the opposite condition to the foregoing. Under this heading belong partial and general excess of growth, increase in the number of the mammary glands (polymastia), spleen, supra-renal capsules, fingers and toes, teeth, ribs, and vertebræ.

3. Malformations due to errors of development (Monsters by altered relation).

To this group belong cases of transposition of the viscera. In cases of complete transposition of the viscera, all the organs of the thoracic and abdominal cavities, which normally are placed on the left side, are transposed and placed on the right side of the body. Situs transversus may also affect single organs only (situs irregularis). The displacement of the heart to the opposite side is called dextrocardia. The cause of situs inversus completus lies probably in some mechanical cause—such as abnormal rotation of the fœtus.[1] Abnormal displacement of organs affects more especially the abdominal cavity: thus, for example, dystopia renis, ectopia testis, abnormal position of the intestine—especially of the colon (Ziegler). In this group many authors include the various malformations of the heart and of the large vessels; but, as a rule, these are better included among malformations due to arrest of development.

4. Malformations due to displacement of tissues and persistence of fœtal structures: Teratomata, dermoids, and cysts.

5. Malformations due to the fusion of sexual characters: Hermaphroditism.

3. Double Malformations

For the classification of these, see p. 392.[2]

[1] While experiments on animals have shown that situs inversus can be produced by external causes acting upon the embryo in the very earliest stages of its development, it by no means follows that cases of situs inversus, as seen in the human subject, have such an origin. There are two possible explanations for these cases: some condition inherent in the fertilised ovum and producing its result when the ovum is undergoing division into its two primitive blastomeres, or some cause acting upon the ovum from without—in the great majority of the cases still at a very early stage of its development, but possibly in some cases at a somewhat later stage. As to the exact nature of the causes which bring about situs inversus we have no accurate knowledge.

[2] For the sake of comparison, and because on the whole it is the simplest and the best of all the classifications, that adopted by Ballantyne is given here in outline. It is in large part founded upon that employed by Taruffi in his *Storia della Teratologia*.

Terata (Monstrosities and Anomalies)

1. Monosomatous: a single individual involved.
   - (*a*) Pantosomatous: the anomaly affects the whole or nearly the whole body. This includes dwarfism, giantism, hemi-atrophy, and hemi-hypertrophy.
   - (*b*) Merosomatous: the anomaly affects only a part of the body. This includes the anomalies of the various regions of the body, and of the placenta, cord, and amnion.
   - (*c*) Heterotaxic: the anomaly affects only the arrangement of the parts of the body—transposition of the viscera, for example.
2. Polysomatous: two or more individuals involved.
   - (*a*) Twins entirely separate, but in a single chorion.
   - (*b*) Twins united only by the vessels of their umbilical cords.
   - (*c*) Twins united more or less completely, symmetrically and asymmetrically, Double Monsters.
   - (*d*) Triplets, Quadruplets, etc.

LITERATURE

Ziegler, *Lehrb. der allg. u. path. Anat.* (Jena, 1905–06); Schwalbe, *Grundriss der path. Anat.* (Wiesbaden, 1910); Schwalbe, *Die Morphologie der Missbildungen des Menschen u. der Tiere*, pt. i., *Allg. Missbildungslehre* (Jena, 1906); v. Winckel, *Volkmanns Samml. klin. Vorträge*, Nos. 251, 252; Küstner, *Mullers Handbuch der Geburtshülfe* (1888), vol. ii.; Seitz, *v. Winckels Handb. der Geburtshülfe*, vol. ii., pt. 2., p. 1028; Lebedeff, *Virchow Arch.* (1881), vol. lxxxvi p. 263; Simonart, *Journ. des Connais. méd. prat.*, June 1846; Hein, *Zeitschr. f. Geb. u. Gyn.* (1881), vol. vi., p. 352; Hohl, *Die Geburten der missgestalteter kranker u. todter Kinder* (Halle, 1850); Spiegelberg, *Lehrb. der Geburtshülfe* (1891), p. 545; Dareste, *Rech. sur la production artif. des monstruosités* (Paris, 1894), 2nd ed.; Stöckel, *v. Winckels Handb. der Geburtsh.*, vol. ii., pt. 3., p. 1453; Fleck, *Archiv. f. Gynäk.* (1902), vol. lxvi., p. 672; Ballantyne, *The Fœtus: Antenatal Pathology and Hygiene* (1902), p. 405.

CHAPTER II

Arrest of Development of the whole Embryonic Rudiment

As the result of marked disturbances of development, the fœtus generally dies sooner or later, or develops normal in form, but with imperfect development of all its parts (Microsomia, Dwarfism).

In the first case a miscarriage usually soon occurs. The dead fœtus, however, may remain in the uterus for a considerable period of time, while the membranes—and especially the placenta—undergo further growth and increase in size,[1] so that a disproportion exists between the ovum and the embryo (Missed labour, Travail manqué).

The membranes frequently are completely permeated with blood (Blood mole), with the result that such an altered ovum when discharged may very closely resemble blood-clot. The true nature of this blood-clot can usually, however, be recognised by the presence of the cavity of the ovum lined by the smooth amniotic membrane. If the blood pigment becomes absorbed from the blood mole, it has a greyish-white flesh colour (Fleshy mole). In some instances the formation of a subchorionic hæmatoma occurs in dead ova which have been long retained. These hæmatomata form nodular projections under the membranes and have been described by Breus as hæmatomatous moles, or hæmatoma subchoriale tuberosum.[2] The embryo becomes partially or totally absorbed after infiltration with leucocytes, so that it is either entirely absent or its remains can be recognised floating in the amniotic cavity.

According to Berlet and Engel (quoted by Ziegler), the leucocytes which are present in the tissues of the fœtus are derived from the fœtal blood. The chief cause of the complete absorption of the fœtus is, however, to be found in autolytic processes, leading after

[1] While it is quite likely that the placenta may undergo some increase in size as a result of degenerative changes—such as fibroid changes or vesicular changes in the chorionic villi—yet it is probable that no true growth of the placenta—that is to say, of a normal character—ever occurs after the death of the fœtus.

[2] According to Berry Hart, the formation of the hæmatoma is due to the clotting of the blood in the secotinal sinuses, followed by the death of the fœtus: which is therefore a secondary condition, and not the cause of the formation of the mole.

the death of the fœtus to the formation of ferments derived from the fœtal body and from the placenta, which bring about the conversion of the albuminous constituents of the tissues into a less complex body soluble in water. Such ferments capable of splitting up albumen and starch, have recently been found in the normal placenta. In other cases where no fœtus is found it is possible that it has not been absorbed, but that it has been discharged unobserved in the urine or in the stools. If an already fully formed fœtus dies and is not discharged soon after its death, various processes can occur in it. The most frequent is maceration (Fœtus sanguinolentus). This is, however, only possible in the absence of organisms, and necessitates, therefore, unruptured membranes.

FIG. 3.—MACERATED FŒTUS. (Specimen from the collection of the Göttingen Women's Clinic.)

The expression, 'macerated fœtus,' which is frequently employed for this condition, is not strictly accurate. During maceration, separation of the epidermis occurs in the form of larger or smaller blebs. These later burst, so that the reddish-brown subepidermal tissue becomes exposed in large patches (see Fig. 3). The blood pigment becomes diffused and permeates all the fluids and tissues of the body; while in the skull the sutures become loosened, so that the skull-bones lie loose in their covering of skin. In the same way loosening of the articulations of other bones occurs. As a result of this the fœtus becomes lengthened—a fact which must be taken into account in estimating its length for the purpose of determining its age. All the cavities of the body—especially the thorax and the abdomen—together with the subcutaneous tissues, become filled with

a dirty reddish-brown fluid.[1] The liquor amnii also acquires the same colour, which frequently becomes changed to a dirty green, from the discharge of meconium into it shortly before the death of the fœtus. The internal organs—especially the liver and the brain—are markedly diffluent. The lungs are preserved best, so that they can nearly always be inflated. On microscopic examination, however, well-formed and differentiated cells are but seldom seen.

The umbilical cord has a characteristic sheen, is considerably thickened, and of a reddish-brown colour. The fœtal membranes and the placenta are generally well preserved up to the time of birth. The stale smell of the macerated fœtus and of its organs is characteristic. According to Sentex and Runge, the degree of absorption of the blood pigment by the lens and the vitreous can be used as a means of determining the date of the death of the fœtus. It may be assumed, if the media are clear and refracting, that the fœtus has been born soon after its death; while if the vitreous is of a dark red colour, some eight to ten days have elapsed, depending upon the degree of the discoloration; while if the lens be already discoloured it is, at the earliest, some fourteen days since the death of the fœtus. According to Ahlfeld these assumptions are incorrect. The exact nature of maceration, and of the chemical changes occurring in connection therewith, is still entirely unknown.

In macerated fœtuses the cause of death can sometimes be ascertained: for example—such as small-pox, malformations, and especially syphilis; some 80 per cent. of all macerated fœtuses showing signs of syphilis—osteochondritis syphilitica, pemphigus syphiliticus, pneumonia alba, microscopic gummatous formations in the liver and other organs, and changes in the placenta.

Maceration of the child produces in the mother definite symptoms, which must be referred primarily to the absorption of toxic bodies originating in the macerated fœtus; these are shivering fits, elevation of temperature, debility, a bad taste in the mouth, and definite cachexia.

Further symptoms on the part of the mother after the death of the child are diminution of the girth of the abdomen—or at least no further increase in its size—as the result of the absorption of the liquor amnii, flabbiness of the breasts, cessation of fœtal movements, and a sensation of a foreign body in the abdomen, so that if the mother lies on the right side she feels as if something in the abdomen fell over to the right, and vice versa. On physical examination, the heart-sounds are absent in spite of repeated auscultation; the abdomen is softer, the position of the child is more difficult to define, and the fœtal parts can only

[1] It is to the appearance of the fœtus at this stage of maceration that the term 'sanguinolentus' is most applicable.

be felt with difficulty or not at all. If the abdomen is examined and measured on several occasions its diminution in size can be determined. If a thermometer is placed in the uterus, and afterwards in the vagina, the temperature in the case of a dead foetus is the same in both; but if the foetus is alive the temperature in the uterus is usually one-tenth of a degree higher, since the living foetus, as a result of its own metabolism, always has a temperature somewhat higher than that of the mother.

During labour, the loosened bones of the skull can be felt, the escaping liquor amnii is discoloured, and, on internal examination, portions of the separated epidermis may come away. Parturition occurs as a rule very easily, and, in the case of a multipara, almost unnoticed. As a result of the laxity of the articulations, prolapse of the foetal parts—for instance, prolapse of the foot by the side of the head, as a rule, an uncommon condition—is frequently observed. During the third stage of labour, manual removal of the placenta is often required because of adhesions. There is frequently, according to some statistics (Martin and v. Winckel), slight fever during the puerperium after the birth of a macerated foetus.[1]

FIG. 4.—MUMMIFIED FŒTUS PAPYRACEUS. (Specimen from the collection of the Göttingen Women's Clinic.)

Mummification of the dead foetus occurs much more rarely than

[1] This is in all probability to be explained by the entrance of saprophytic organisms into the uterine cavity after the rupture of the membranes, and a resulting slight degree of septic intoxication.

maceration. In such cases all the liquor amnii becomes absorbed and the fœtus desiccated. This process is observed most frequently in twins of which one has died *in utero* (Fœtus papyraceus).[1] Mummified fœtuses have a greyish-yellow colour, the skin is wrinkled, and through it the skeleton can be clearly seen. The placenta is small and hard—in rare cases also compressed (Placenta papyracea); the umbilical cord is thin and at the same time shrivelled, and the liquor amnii is absent (see Figs. 4 and 5).[2]

FIG. 5.—MUMMIFIED FŒTUS FROM A TUBAL PREGNANCY. (Specimen from the collection of the Göttingen Women's Clinic.)

Petrification of the fœtus, leading to the formation of the so-called lithopædion or stone child, is very rare. This process is most frequently seen in extra-uterine gestation. In cases of this condition the membranes and the surface of the fœtus itself become encrusted with lime salts. The interior of the fœtus does not undergo calcification, but becomes mummified. The process commences with the absorption of the liquor amnii, so that the membranes are situated close to the fœtus. According to Küchenmeister, three varieties of this condition may be recognised:—

1. The mummified fœtus is contained in the calcified membranes (Lithokelyphos).

2. The fœtus is adherent at some places to the membranes, and the calcification spreads from the membranes to the fœtus in these situations (Lithokelyphopædion).

3. The fœtus after rupture of the membranes escapes into the abdominal cavity, and a layer of lime salts of increasing thickness forms round the shrunken fœtus (True lithopædion, stone child).

Küchenmeister's conclusions as to the frequency of the different forms have been contradicted lately by Werth. If the case is one of the very rare intra-uterine lithopædion formation, the maternal membranes can, through incrustation, acquire a very firm attachment to the inner surface of the uterus. In many cases a lithopædion is found as a secondary condition accidentally at an autopsy. The process is observed more frequently among cows and sheep (see also

[1] In the majority of the cases the twins are derived from a single ovum. Two papyraceous fœtuses have been met with in triplets, the third fœtus developing normally to full term.

[2] Mummification is especially met with from the third to the sixth months of fœtal life, while after that period maceration is more common.

Seitz). A lithopædion in cases of extra-uterine gestation can be borne by a woman for as long as ten years without any symptoms, but in some cases it may give rise to long continued suppuration with even a fatal termination.

In the Göttingen Women's Clinic, we have been able to observe very carefully an interesting case of lithopædion formation. A patient, thirty-nine years old (Jr. No. 20401), gave the following history: At the age of thirty-six years she had been pregnant, and labour-pains had occurred at what was thought to be full term. A doctor, called in by the midwife on account of the slow progress of the labour, diagnosed a transverse presentation. As the cervix was still closed he went away, with the warning to call him when the dilatation of the cervix had made good progress. The pains ceased completely on the fourth day, as did also the movements of the child, so that there was no further need for the services of the doctor or the midwife. Subsequently the patient complained of slight pains, which spread from the right to the left side, with vomiting, and marked hæmorrhage at the periods. Some time later, at an examination undertaken on other grounds, the doctor found a tumour above the symphysis pubis, and ordered the patient into the Women's Clinic at Göttingen. The provisional diagnosis of a lithopædion, made from the history the patient gave, was confirmed by an X-ray photograph. The patient refused to undergo a suggested operation.

In the museum of the Göttingen Clinic there is a very beautiful preparation (No. 110) of a lithopædion (see Fig. 6), which was discovered at an autopsy on a woman aged seventy-six years who died of marasmus. It was proved that the woman had carried this lithopædion for over forty years. Before her death she stated that her husband, who had been in the royal stables of King Jerome, was killed by the kick of a horse. On the receipt of this news she had, while pregnant, fallen downstairs on to her back. For the first few years after this event she experienced, whenever she changed her position, a sensation of a lump falling from one side to the other. On several occasions she had undergone medical treatment for attacks of peritonitis. During the last ten years of her life she had no special pain. At the autopsy, undertaken on the fifteenth of March 1849, at Kassel, the body was found to be very thin, the legs œdematous, the abdomen very retracted; and situated beneath the umbilicus there was a hard tumour, which could be felt with ease through the thin abdominal walls. On opening the abdomen an irregular oval tumour, about the size of a head, was found with its narrow end pointing towards the true pelvis, and placed between the uterus and the rectum. It was closely united by strong adhesions to the right ovary and the broad ligament. The uterus, the left ovary, and left tube, were normal. The tumour was removed, opened

with a saw, and a lithopædion found corresponding, when measured, to a seven-months' fœtus.[1]

These processes of maceration, mummification, and lithopædion formation only occur within the uterus when it contains no organisms —that is, when the bag of membranes is intact. When the membranes are no longer intact, organisms can readily enter the uterus either through the use of instruments, or by the finger, or by the gradual upward spread of the germs themselves. The contents of the uterus then very quickly undergo decomposition, a stinking discharge occurs, and, if the decomposing masses are not speedily removed, death may ensue as a

Fig. 6.—Stone Child. (Specimen from the collection of the Göttingen Women's Clinic.)

result of acute sepsis. In the majority of cases, the fever present is of the sapræmic variety (putrefaction fever), and when the stinking masses are removed it soon subsides.

In very rare cases intra-uterine skeleton formation occurs. In this case the process of disintegration is confined to the uterus, the soft parts are gradually removed, with a free discharge of the secretion set up by their destruction, and the bones alone remain behind. A longer

[1] Specimen No. 705, University College Hospital Medical School Museum (Obstet. Sect.), is that of a lithopædion, removed from the right broad ligament of a woman forty-three years of age who died of bronchitis. The condition was not suspected during life. The macerated fœtus is enclosed in a thick membrane, on the inner surface of which there is some calcareous material (see *Trans. Path. Soc. London*, vol. xxxviii., p. 445).

or shorter time afterwards the bones are eliminated, either spontaneously or with artificial aid.[1]

Nanism Dwarfs

Under this heading—the arrest of development of the whole embryonic rudiment—may be considered the formation of dwarfs (Nanism). In this case a fœtus develops normal in form, but with imperfect growth of all its parts.[2] In such cases the development of the bones more especially is arrested at a very early stage. In this class should be included, really, only the cases of true dwarf formation. The dwarfism produced by curvature of the spinal column and of the extremities, or as the result of bone diseases, is something quite different.

Kaufmann distinguishes between dwarfs developed in proportion, and dwarfs developed out of proportion: or genuine and false dwarfs.[3] In the true dwarfs the whole build of the body, and, above all, the surrounding soft parts, corresponds to the neat and symmetrical bones. The development of the whole body has progressed very slowly from the earliest period (*ab ovo*), or has come to a standstill prematurely in early childhood. The developing zone of cartilage is quantitively deficient; the epiphyseal nuclei, however, develop normally, and the union of the epiphysis and diaphysis occurs at the proper time. To this group belong, for example, the Lilliputian forms of the annual fairs, and the type described by Paltauf. In this case there remains attached to the bones, in part unused, the cartilaginous material serving for their increase in length. Certain bone sutures and cartilaginous joints are preserved, and certain centres of ossification are still present, while others are missing. To this class belong, further, the myxœmatous dwarfs, or those without a thyroid, as well as the dwarfs met with in cases of endemic cretinism; and further, with the above-mentioned restrictions, the dwarfs of imperfect proportions produced by chondrodystrophia (see p. 244), osteogenesis imperfecta (see p. 246), microcephalus, hydrocephalus, and rickets.

The dwarfs have a great interest for the obstetrician, inasmuch as they supply the type of the dwarf pelvis (Pelvis nana). This is an example of the highest degree of the generally contracted pelvis. It

[1] Skeleton formation can occur both with and without the occurrence of suppuration. In the latter case the soft parts are probably destroyed both by the action of phagocytes and of certain cellular enzymes.

[2] Ballantyne uses the term 'Microsomia,' which he defines as 'monstrous smallness' of all the parts of the individual—whether that individual be an embryo, a fœtus, a child, or an adult.

[3] With regard to their probable origin, dwarfs may be divided into three main classes: those due to primary defect of the unfertilised ovum; those due to secondary defect of the fertilised ovum; and those due to some arrest of development occurring during the ante-natal or post-natal period.

precisely resembles the pelvis of early childhood. The individual bones are, as in the child, united by masses of cartilage. The conjugata vera, the measurement between the upper border of the symphysis and the sacral promontory, may measure as little as 5 cm. or less, so that in such cases Cæsarean section must be considered as an absolute or relative indication.[1] Occasionally, one can detect, even in the living, at the lines of synostosis in the acetabula, or at the ischio-pubic junctions, irregularities or nodular projections, evidences of an abnormal process of ossification (Sonntag, Breus, and Kolisko).

Literature

Breus, *Das tuberöse subchoriale Hæmatom der Decidua* (1892); L. Seitz, '*Die Veränderungen von Fötus und Plazenta nach dem Tode der Frucht, usw.*' *v. Winckel, loc. cit.*, p. 15; Sentex, *Des altérations, que subit le fœtus après sa mort, etc.* (Paris, 1868); M. Runge, *Lehrb. d. Geburtsh* (1909); Martin, *v. Winckel*, quoted by Seitz, p. 1273; Kücheumeister, *Arch. f. Gynäk.* (1881), vol. xvii., p. 153; Werth, *Die Extrauterinschwangerschaft, v. Winckels Handbk. d. Geburtsh.*, vol. ii., pt. 2, p. 655 (with bibliography); Kaufmann, *Lehrb. d. spez. path. Anat.* (1907); A. Paltauf, *Über den Zwergwuchs.* (1891); E. Sonntag, *Die Pathologie des knöchernen Beckens, v. Winckels Handbk. d. Geburtsh.*, vol. ii., pt. 3, p. 1789; C. Breus and Kolisko, *Die path. Beckenformen* (1903–04); Ahlfeld, *Lehrb. d. Geburtsh.*, p. 551 (1903).

---

[1] At the present day most obstetricians would consider Cæsarean section as absolutely indicated in a dwarf pelvis with a conjugata vera measuring as much as 8 cm. to 8·5 cm., or 3⅛ inches to 3⅜ inches.

CHAPTER III

Malformations the result of defective closure of the Cerebro-Spinal Canal

If the bony spinal canal is only partially closed it will lead to Rhachischisis, Spina bifida. In this condition there is no skin in the region of the cleft (Adermia), and the spinal cord may be entirely or partially wanting (total or partial Amyelia). In total rhachischisis the whole, or nearly the whole, of the arches of the vertebræ are absent, and the spinal cord is also more or less wanting. Such marked cases of rhachischisis are nearly always combined with hemicephalus (see the next chapter).

FIG. 7.—SIMPLE SPINA BIFIDA. (Specimen from the collection of the Göttingen Women's Clinic.)

A partial spina bifida is usually situated in the lumbo-sacral region,[1] or in the upper part of the vertebral column; while the intervening portion remains unaffected. When a cyst-like tumour protrudes from the spinal canal through such a partial cleft of the vertebral arches it is termed a spina bifida (better cystica, since the term spina bifida alone indicates merely the cleft in the vertebræ) or hydrorrhachis. An external and internal hydrorrhachis may be distinguished.[2]

[1] In this region the medullary plates close last.

[2] Three main varieties of spina bifida may be distinguished: namely—a meningocele in which the membranes alone form the sac; a meningo-myelocele in which the sac contains the spinal cord—usually flattened out, as well as the membranes with fluid between the two; and a syringo-myelocele in which the cord has a dilated central canal, which contains fluid and forms part of the wall of the sac.

In a hydrorrhachis interna or myelocele[1] the central canal of the spinal cord is markedly dilated, and, as a result of this, the substance of the spinal cord and its coverings protrude through the cleft in the bones. Such a tumour is described as a hydrorrhachis externa or spinal meningocele[2] when it is composed of the outer skin, the membranes of the spinal cord, and a collection of fluid between the substance of the cord and its membranes.

A spina bifida cystica is distinguished according to its site as cervical, dorsal, lumbar, lumbo-sacral, or sacral. They occur most often, as has been already mentioned, in the lower part of the spinal column: namely—in the lumbar and sacral regions. The protrusion forms usually a more or less rounded tumour the size of a plum, an apple, or a child's head, which has either a broad sessile attachment in the middle line or a narrow pedicle. The skin covering it is either normal or ulcerated, or presents an altered, scarred, and puckered appearance. At times the skin is almost completely absent over the whole surface of the tumour—a fact of considerable importance in the treatment of the condition, since it renders the closure of the skin after operation more difficult.

The tumour feels soft, often elastic, and in many instances fluctuates. If a communication exists with the spinal canal it becomes smaller on pressure. Through the increased tension of the cerebro-spinal fluid produced in this way, extending as it does ultimately into the ventricles of the brain, mild symptoms of irritation of the brain and spinal cord are produced—such as general muscular cramps, marked bulging of the fontanelle, irregular breathing, slowing of the pulse, and vomiting. Other malformations are very frequently found with a spina bifida: namely—club feet, gastroschisis, eversion of the urinary bladder, and encephalocele. Walterhöfer describes a case of spina bifida with prolapse of the rectum and uterus. I think it is not improbable that these conditions stand in an ætiological relation to one another, and that the insufficiency of the ligamentary apparatus of the uterus, and of the musculature of the pelvic floor may be produced in this way: namely—the nerve fibres innervating these muscles are damaged or paralysed by the compression produced by the spina bifida. This explanation seems all the more probable since we observed the same condition, a short time ago, in a case of spina bifida anterior (see p. 28). Bürger, too, accepts this explanation for a similar case.

The variety usually described is the spina bifida posterior. In other rare cases, the sac may come forward through a defect in the bodies of the vertebræ (Spina bifida anterior). A very

[1] That is, really a syringo-myelocele.

[2] It is not clear here whether the author includes under this term both a meningocele and a meningo-myelocele.

interesting case of this kind has been described by Kroner and Marchand. In a twenty-year-old multipara, a tumour was found springing from the posterior wall of the small pelvis, and extending up to two fingers' breadth above the umbilicus. After puncturing and emptying the tumour, episthotonus, meningitis, and death occurred. The autopsy revealed a spina bifida anterior. The large sac described communicated by a fine opening on the anterior surface of the sacrum with the spinal canal.

Fig. 8.—Spina Bifida Cystica.

A further case is recorded by Neugebauer. In a virgin, twenty-two years old, marked constipation was present, the bowels being moved only every three to four weeks. The small pelvis was filled by a fluctuating tumour the size of two fists situated between the rectum and the anterior wall of the sacrum, and appearing to take origin from the latter by a broad base. All the pelvic organs were displaced against the symphysis pubis, and the uterus was displaced upwards—the uterus and vagina being double. There was a defect of the right half of the fourth and fifth sacral vertebræ and of the coccyx. An exploratory puncture of the tumour gave exit to cerebro-spinal fluid. The patient did not return, and died suddenly, four weeks afterwards—probably as the result of rupture of the cyst.

Another very interesting case, although not confirmed by an autopsy, has been published by Nieberding. The case was that of a girl, eighteen years of age, who, on account of abdominal discomfort, was admitted into the Gynæcological Polyclinic. On examination, a tumour was found which almost filled the small pelvis, pushed up the uterus, and sprang from the anterior wall of the sacrum. The rectum passed in front of the tumour towards the right (see the very similar case observed in the Göttingen Women's Clinic). An exact diagnosis could not be made; but the pain was so severe that Nieberding determined to perform laparotomy. After opening the abdominal cavity, however, the operation was soon abandoned, as the tumour, which was situated deep in the small pelvis, could only be reached with great difficulty. The wound was therefore closed, and a further vaginal operation decided upon. Before doing this, however, Nieberding punctured the tumour from the vagina. The fluid obtained was clear, like water. The whole contents of the cyst, two litres in all, were then evacuated by Dieulafoy's apparatus. Shortly afterwards, the patient developed very serious symptoms—vomiting and severe pains in the head and neck—and

from these a diagnosis of anterior meningocele was made. The cyst, after three days, had completely filled again. The patient refused any further operative treatment. (For the literature on the subject, see Nieberding.)

I may now report a very similar case observed and operated upon in the Göttingen Women's Clinic. So far as I can ascertain from the literature (see the cases collected by Nieberding), this is the ninth recorded case of anterior meningocele. A. R. (Jr. No. 23725), a girl twenty-two years of age, said that formerly she had been in good health. Menstruation commenced when she was seventeen years old, and had always occurred regularly. The last period occurred six months ago. She further said that for some time she had been under medical treatment for kidney and bladder disease. She complained, on admission, of marked emaciation, frequent pains in the abdomen, scalding on passing water, strangury, stinking urine, and a feeling of weight in the abdomen. The bowels were generally confined

Examination showed a very debilitated and emaciated patient. The urine was very foul, markedly purulent, and microscopically contained numerous leucocytes, colon bacilli, and bladder epithelium. An examination of the genital organs, under an anæsthetic, revealed prolapse of the anterior vaginal wall, descent of the uterus, with elongation and hypertrophy of the cervix—the body of the uterus being retroverted and somewhat enlarged. Behind the uterus a cystic tumour could be felt which almost filled the small pelvis, and was completely fixed. The rectum passed transversely to the right; and, on the left, one could feel the tumour described bulging into it, the bowel being placed almost entirely in front of the tumour. The ovaries were not definitely recognised. A certain diagnosis could not be made as a result of the examination. The possibility of a retro-peritoneal or a retro-rectal ovarian tumour was discussed. An operation carried out under lumbar anæsthesia revealed the following condition.

On opening the abdomen the markedly hypertrophied bladder was first encountered, reaching up midway between the symphysis pubis and the umbilicus. Behind this was found a tense elastic tumour of a dark blue colour, about the size of a man's head, which passed down deep into the true pelvis and was attached to the anterior wall of the sacrum. The parietal layer of peritoneum, which covered the tumour in front and above, was split and peeled off. The rectum, displaced forwards by the tumour, was pushed aside without any difficulty. On attempting to enucleate the tumour, however, it burst and gave exit to a clear thin fluid. The cyst was gradually detached further, and

a structure which appeared to be a small pedicle, tied and divided. After arrest of the hæmorrhage and removal of the fluid as far as possible from the abdominal cavity, ventrofixation of the uterus was performed after Olshausen's method, and the abdomen closed. The patient died on the seventh day after the operation, with increasing weakness of the heart's action. After the operation, the provisional diagnosis was made of spina bifida anterior. This supposition was confirmed by the autopsy. The condition was that of a spina bifida anterior lumbosacralis. I will not enter here into a detailed account of the autopsy as the case will be published elsewhere.[1]

As has been mentioned above, the contents of a spina bifida are very different, according as to whether the case is one of hydrorrhachis interna or externa. This distinction must be very clearly made, more especially in considering the treatment. As it is never certain which of the two forms is present, simple removal of the sac after ligature is contra-indicated. Between the cavity of the sac and the spinal canal either a more or less narrow aperture is present, or the communication between the two is entirely obliterated—naturally a very favourable condition for treatment.

A spina bifida is a not very rare malformation. According to Chaussier, one case occurs in every 1,000 children; while according to Demme, one case is met with among 630 (Biedert and Fischl). The ætiology of such malformations is explained by interference at an early date, with the development of the embryo and agenesia[2] (Ziegler), or hypoplasia of the medullary plates[3] which should form the vertebral arches. Whether this defect is primary, or, as Ahlfeld thinks, is secondary to an increase in the amount of the fluid contents of the vertebral canal, is still a matter of opinion. According to Leser, both

[1] A case of this kind has been described by J. S. Fairbairn, *Journ. Obst. and Gyn., Brit. Emp.*, vol. xx., No. 1, p. 1 (1911). An attempt was made to remove the cyst through the perineum, but this proved impossible, and the patient, a girl of eighteen years of age, died about seven weeks after the operation from meningitis. Such a condition forming a pelvic tumour is very likely to give rise to errors in diagnosis. In a case recorded by Grossmann (*Jahrbuch f. Kinderheilkunde*, vol. lxiii., p. 224 (1906)), a Röntgen-ray photograph showed the defect in the sacrum—and such a means of investigation might enable a correct diagnosis to be made in a doubtful case. It is interesting to note, as Fairbairn points out, that these tumours have been met with almost exclusively in children, or young women.

[2] The term 'agenesia' should be limited to the mal-development of structures or organs of the body which have never been formed, or, at any rate, of which no vestige remains; while the terms 'aplasia' and 'hypoplasia' are applied to structures or organs of the body which have been developed, or of which vestiges still remain.

[3] As the closure of the medullary plates precedes in point of time the upgrowth over them of the mesoderm which forms the structures superficial to the cord, the primary condition must be the want of union of these plates.

origins are possible.[1] The prognosis of this condition is not good, as children born with a spina bifida are generally not capable of living. In numerous cases children thus affected are born dead or die soon after birth. If they survive, a gradual increase of the tumour is generally observed, and as a result signs of pressure occur—more especially on the spinal cord and the cauda equina. Paralysis, anæsthesia, disturbances of the bladder and of the rectum, and bedsores, may be present. Still more frequently, rupture of the sac takes place with resulting inflammation of its walls, and death from suppurative meningitis. The most favourable result after rupture of the sac is the formation of a persistent fistula. In very rare cases a spontaneous cure has been observed from inflammation with obliteration and shrinking of the sac by scarring. According to Biedert, of thirty-two cases of spina bifida twenty-five died in the first week after birth.[2] As a result of the very bad prognosis of this condition, attempts have been made, especially recently, to obtain a cure by various means—more especially by operation. As a matter of fact, in a good many cases, a cure has been obtained by operative measures. But it must be pointed out that even after the most successful operations a number of the children die from increasing hydrocephalus. In most of these cases, however, the hydrocephalus is the primary condition. A cure occurs most certainly when the communication between the cavity of the sac and the vertebral canal is obliterated, or at least very narrow. In myeloceles[3] where the substance of the spinal cord takes part in the formation of the wall of the sac, the prognosis is very bad.

Treatment must have in view the removal of the contents of the

[1] It is, of course, quite possible that the non-development of the medullary plates may be due to an excessive accumulation of fluid in the spinal canal in the early weeks of pregnancy. It would have to be within the first three weeks, since after that period the medullary plates have met and closed. It is, however, more probable, and this is the view generally taken at the present time, that the increase in the amount of fluid is a secondary condition, probably the result of congestion or inflammation of the membranes of the cord. While the defective development of the medullary plates forms an adequate explanation of many cases of spina bifida, other factors must, however, be taken into consideration to explain such conditions—as syringo-myelocele, and the changes in the skin met with in cases of spina bifida occulta. These factors would appear to be a tendency to over-growth, and at the same time a faulty direction of this tendency. In this way would be explained the increased length of the spinal cord so common in these cases, the hypertrophic changes present in the skin over a spina bifida occulta, the increase in some cases in the amount of cerebro-spinal fluid, and the varying relations of the dilated central canal to the rest of the spinal cord in cases of syringo-myelocele. The occasional duplication of the cord, and the presence of cartilaginous or bony projections in the spinal canal, are also evidence in favour of a tendency to irregular overgrowth.

[2] Of 649 children that died of spina bifida in England in 1882, 612 died within the first year. Of 99 cases not operated upon, the majority died within the first few weeks; only 20 living to be over five years of age.

[3] That is, a syringo-myelocele.

sac and then the obliteration of the sac itself (Leser). The result which can thus be attained is, in the best possible conditions, the hindering of any increase in size of the tumour. In closed meningoceles, and base with a narrow communication, puncture may be employed, followed by the injection of an irritating fluid: for example, ½ to 1 c.c. of tincture of iodine. During this procedure, any existing opening there may be in the vertebral column must be kept closed, whenever it is possible, by means of pressure with the finger, and at the end of the operation a tight binder applied. The injection must be repeated if a definite obliteration and shrinking of the sac is to be brought about.[1]

If such interference, however—as is often the case—does not lead to any result, an incision must be made into the meningocele, with the strictest aseptic precautions; the superfluous coverings excised, and an attempt made to close it by suture. I saw recently, in a patient twenty-five years old, to whom I was called in her first confinement, a scar about 10 cm. long in the lumbar region of the spinal cord, which had resulted from an operation for spina bifida. The scar was quite firm, and there were no resulting disturbances, such as paralysis or anæsthesia, present. The pelvis was narrowed, by some 1 to 2 cm., in the antero-posterior diameter of the inlet. It would be difficult to say whether the primary uterine inertia, present at the time of the labour, bore any relation to the existence of the spina bifida.

Schirmer describes two cases from the surgical clinic at Greifswald in which, after opening and extirpation of the sac, a permanent cure was obtained by the implantation of a portion of bone, with its periosteum, in the defect in the spinal canal.

Cragin reports three cases of spina bifida treated by operation. The first was cured by the operation; the second died of marasmus after the operation; in the third there was an enormously large spina bifida which ruptured during delivery. As a result of this, such severe bleeding occurred that the medical man was compelled to plug it with iodoform gauze. Healing occurred after an operation undertaken five days after birth.

Herman reports the cure of a spina bifida as large as a man's head (Meningo-myelocele). The child in question was operated upon the day after birth. The tumour was removed through an incision in the healthy skin, after the separation of numerous nerve fibres which had to be replaced in the spinal canal. The arachnoid was sutured, and the defect in the bones closed with two thin plates of bone loosened from the lumbar vertebræ surrounding the opening. These were united in the middle line with silkworm-gut sutures. Two flaps of fibrous tissue

[1] Although it is true that very good results have been obtained by the injection of iodine in a solution of iodide of potassium and glycerine, yet at the present day most authorities would be in favour of the much more surgical procedure of excision of the sac.

and muscle were then formed and used to cover the line of suture of the bones, and finally the skin was united. A fistula, which formed spontaneously on the third day after the operation, healed without treatment in a month. (I need not refer to any more similar cases; they may be found in the surgical literature on the subject.)

In my opinion, the method employed by C. Keller of removing a spina bifida with a narrow pedicle by ligature is, in view of the facts related above, entirely irrational and indeed wrong, since, as has already been pointed out, important nerve trunks—such as the cauda equina—are very likely to be removed at the same time.

According to Biedert, by operative treatment the mortality of this affliction has been reduced to 30 per cent.[1]

In the Göttingen Women's Clinic, we operated a short time ago on a child with spina bifida sacralis which was about the size of a small apple. The child presented by the vertex, and the cyst which had remained intact during labour, burst soon afterwards. At the operation, undertaken some hours after birth, the excess of skin in the sac was removed with careful preservation of the nerves of the cauda equina lying beneath and in part attached to the wall. The sound skin on both sides was then drawn together and united with silver wire and catgut sutures. The temperature during the first fourteen days after the operation was quite normal and the child fed well at the breast; but during the first few days, however, complete anæsthesia and almost complete loss of movement of the lower limbs set in. The child lay during this period with its legs constantly extended, and with almost complete incontinence of fæces. Further than this, during the same period, an acute hydrocephalus commenced. The occipital suture gaped for ½ cm., and through the frontal suture a bulging swelling occurred. The child was treated daily for ten minutes with the faradaic current. The symptoms of hydrocephalus soon retrogressed, and the loss of power of the lower limbs improved; while the slight reaction to the faradaic current remained very definite. On the eleventh day after the operation the skin and scar showed some reddening. The condition of the child changed for the worse during the next few days, so that its appearance impressed one unfavourably, and it did not take its nourishment. On the eighteenth day after the operation the temperature rose to 102·8°, and, with a further rise of temperature on the twentieth day, death occurred. The autopsy showed, besides suppurative meningitis which had spread to the base of the brain, a right-sided ascending suppurative pyelonephritis, the

[1] Sachtleben reports eighteen cases from Mikulicz's Clinic with satisfactory final results in 38·9 per cent. Moore collected 385 cases treated by excision, among which the mortality of cases operated upon since 1885 was 24 per cent.; but he expresses the opinion that if all the cases had been followed up, the total mortality a few months after the operation would have been fully 50 per cent.

duration of which was estimated to be at least fourteen days, besides marked hypertrophy of the bladder wall.

When labour occurs, the children are usually born spontaneously, since the sacs, even when of large size, are often yielding and flaccid, or burst spontaneously during the passage of the child. In a good many cases, however, a spina bifida causes some—although at times only temporary—obstruction to labour. It is remarkable that these children, in spite of the displacement of the centre of the body-weight towards the breech, are usually born in a cephalic presentation. The cause is indeed a simple one: namely—that the breech and the spina bifida together cannot find room in the lower uterine segment. Errors in diagnosis often occur during delivery. Thus the sac may be mistaken for the distended bag of membranes. In this case the parts of the child will not be felt through the membranes, and, further, the tumour moves with the foetus on external manipulation while the membranes do not. The outer covering of the tumour in question can also be felt becoming continuous with the skin of the foetus. The tumour does not lie so centrally in the genital canal as the bag of membranes, and its consistence remains the same during and between the pains. The mistake of taking a spina bifida for an hydrocephalus, or for the skull of a macerated foetus, can be avoided by recognising the absence of hair. It must be remembered, however, that in an hydrocephalus, the development of hair is often very deficient as a result of trophic disturbances.

Lastly, confusion with double monsters, and tumours of various other kinds—especially such as take origin from the body of the child—is not unlikely: for example, tumours of the breech and tumours of the spinal column. In order to avoid as far as possible such serious mistakes it is advisable, when the delivery of the child is delayed, to examine with the whole or the half-hand under an anæsthetic. Since, as has already been pointed out, many children with spina bifida cystica are operated upon at the present day with good results, it is important not to proceed too hurriedly with the removal of the hindrances to birth produced by such tumours—that is, by performing mutilating operations. In some cases, however, puncture of the sac is essential. In many cases the obstruction to birth can be overcome if the spina bifida is, by rotation of the child, made to occupy the roomy hollow of the sacrum. In footling presentations, in making use of this suggestion, the thigh should be brought down to carry out or to render this rotation easier.

Literature

Walterhöfer, *Diss. München*, 1905; Kroner and Marchand, *Arch. f. Gynäk.* (1884), vol. xvii., p. 444; v. Neugebauer, *Monatsschr. f. Gynäk. u. Geburtsh.*, i., Jahrg., Nos. 4 and 6; and *Beitr. z. Geburtsh. u. Gynäk.* (1903), vol. ix., p. 198; Nieberding,

'*Meningocele sacralis ant.*,' *Munchner med. Wochenschr.* (1904), vol. li., p. 1384; Bickert and Fischl, *Lehrb. d. Kinderkrankheiten* (1902); Ziegler, *loc. cit.*, p. 15. Ahlfeld, *Die Missbildungen des Menschen* (1880); Lomer, *Die spez. Chirurgie* (1908); Spiegelberg, *loc. cit.*, p. 15; Schirmer, *Diss. Greifswald* (1902); Cragin, *Gynecol. Soc. of New York* (1903), *ref. Zentralbl. f. Gynak.* (1904), p. 1147; Herman, *Journ. de chir. et ann. de la soc. belge de chir.* (1905), No. 8, *ref. Zentralbl. f. Gynak.* (1906), p. 1389; C. Keller, '*Die Krankheiten der ersten Lebenstage*,' *Deutsche Klinik*, vol viii; Kaufmann, *loc. cit.*, p. 24; Schmaus, *loc. cit.*, p. 15; Tillmanns, *Lehrb. d. allge. u. spez. Chirurgie* (1907); Küstner, *Muller, loc. cit.* p. 15; Kleinhans, *v. Winckels Handb. d. Geburtsh.* vol. ii., pt. 3, p. 1080; Baginsky, *Lehrb. d. Kinderkrankh.* (1899); Strümpell, *Spez. Pathologie u. Therapie innerer Krankheiten* (1897); Hohl, *loc. cit.*, p. 15; Schanta, '*Die Beckenanomalien*,' *Muller, loc. cit.*, p. 15; Bürger, *Geb. u. Gynak. Gesellsch., Wien* (December 15, 1903), *ref. Zentralbl. f. Gynak.* (1904), p. 621; Sachtleben, *Zentralbl. f. Chir.* (1904), vol. xl., p. 341; Moore, *Surg. Gynecol. and Obstet.* (Aug. 1905), vol. i., p. 137.

Loc. cit. p. 15, p. 24, etc., refers to the lists of the literature on the corresponding pages of this book.

CHAPTER IV

Defective Formation of the Skull-cap, and the Associated Disturbances in the Development of the Brain

ACRANIA, HEMICRANIA, CRANIOSCHISIS, MICROCEPHALUS, ANENCEPHALUS, EXENCEPHALUS, AND CEPHALOCELE

These malformations occur less frequently alone than combined with one another.

In acrania the bony and membranous cranial vault is more or less wanting, and the base of the cranium is covered only with a reddish membranous covering. Hemicrania is a somewhat less marked degree of acrania, as is also cranioschisis, in which the defect frequently involves also the arches of the vertebræ (Craniorhachisis). In the latter the vertebral column is usually also shortened and bent so that the head is drawn backwards, and the face turned upwards (see under hemicephalus).[1]

In microcephalus, the skull-bones which are undeveloped are normally united with one another.

Acrania (sometimes called), hemicrania, cranioschisis or craniorhachischisis is generally associated with total anencephalus or absence of the whole brain. The base of the cranium is then covered with a red spongy mass resembling granulation tissue. In other cases, part of the brain, or even the whole brain, protrudes through the fissure in the vault of the skull and is placed external to this (exencephalus); in this case the portion of brain is covered only by the pia mater, or, less commonly, by the external skin.

Micrencephalus occurs with microcephalus. In this condition the brain is very small and often imperfectly developed. Microcephalus[2]

[1] The term 'anencephalus' is commonly used by English writers for the condition described by the author under the name 'hemicephalus.'

[2] The view that microcephalus may be due to the previous existence of an hydrocephalus has now been abandoned by most writers on the subject, and the condition is rather regarded as the result of a partial atavism, or arrested development of the brain itself with resulting changes in the bones and sutures. If this view is correct, it is obvious that not much benefit can be derived from such operations as craniectomy.

can, however, also develop from an hydrocephalus; and Ahlfeld points out that in this condition of hydromicrocephaly clear signs of an abnormal collection of fluid are present in the cranial cavity. This collection at an early embryonic period has formed an obstacle to the development of the brain, which, later—as a result of the absorption or escape of the fluid—has been partially overcome, and has led to the formation, not of an hydrocephalus but of an hydromicrocephalus. If the cranial vault is for the most part closed, but partial defects are still present, the membranes of the brain, or even portions of the brain itself, can protrude in the form of a hernial sac (Hernia cerebri, Cephalocele).

Fig. 9.—Hemicephalus. (Specimen from the collection of the Göttingen Women's Clinic.)

That children with acrania, cranioschisis, and accompanying anencephalus are not capable of living, does not require any further proof. In exceptional cases, however, they have lived for a few days (see p. 39).

In cases of microcephalus, attempts have been made, on the recommendation of Lannelongue (Leser), to obtain good results by the excision of strips of bone (Craniectomy). According to Tillmanns, too much must not be expected from this operation, since in the majority of cases of microcephalus congenital malformations of the brain are present. According to this author, craniectomy should be reserved for those rare cases in which the microcephalus is the result of premature closure of the sutures and fontanelles.

Hemicrania and hernia cerebri have a special interest for the accoucheur. Hemicrania, or—as it is still called by obstetricians—anencephalus, hemicephalus, cranioschisis, craniorhachischisis, acrania, Frog's head, Toad's head, or Cat's head,[1] is a not uncommon

[1] These terms are generally used synonymously, although strictly they do not always mean the same thing.

malformation. It has, however, only been observed three times in twelve years, by Kleinhans, in the clinic at Prague. In the Göttingen Women's Clinic, it has been met with three times[1] among 3,700 confinements, and nine times in the Midwifery Polyclinic. Of these cases, I observed two, in the last two years, among 200 births in the polyclinic. Küstner saw a woman who was delivered three times of a hemicephalus.[2]

In this malformation the whole of the cranial vault is wanting, and at the same time, as a rule, the whole of the brain. In many cases the abnormal cleft extends to the arches of the vertebræ, sometimes as far down as the lumbar region. In such cases total or partial amyelia (absence of the spinal cord), or adermia (absence of the skin), is present over the whole course of the malformation. Less frequently the spinal cord is present but is flattened against the anterior wall of the spinal canal. In the place of the cord there may be found in the region of the vertebral cleft a thin whitish membrane; in the place of the brain, the dark bluish-red congested mass resembling granulation tissue,[3] already described. The cranial nerves are usually present, but the nerve fibres are absent in the optic nerves and in the retina. For a very complete account of the condition of the central nervous system

FIG. 10.—RADIOGRAM OF AN ANENCEPHALUS.

[1] Among 325 cases of monstrosity and fœtal disease examined by Ballantyne, there were forty-six cases of anencephalus, or 14 per cent.

[2] Ballantyne mentions the extraordinary case placed on record by Martin of six anencephalic fœtuses born to one mother, and points out that these children are most commonly of the female sex. Cases of the repeated occurrence of such a deformity in the same woman lend some support to the view that the tendency to the defect may be present primarily in the ovum.

[3] Area cerebro-vasculosa.

in these deformities, the reader should consult Ernest, 'Schwalbe, Die Morph. der Missb.,' vol. iii.,' pt. 2, pp. 92–100 (1909). The facial portion of the skull is almost always well preserved. The defect in the skull is usually surrounded with a fringe of hair. The eyes markedly protrude, as the result of the want of development of the forehead or of the upper wall of the orbit (Goggle eyes); the neck is abnormally short, so that the head appears to be attached directly to the shoulders. Similarly, the ears are attached just above the shoulders. The head is, as already mentioned, more or less bent backwards and is set firmly in the nape of the neck; as the tongue frequently protrudes between the margins of the gums, the repulsive appearance of these monsters is still further increased. Besides the anencephalus, other malformations are often also present: for instance, umbilical hernia, hare-lip, and cleft palate.[1]

Fig. 11. — Hemicephalus with Fissure of the Vertebral Column. (Specimen from the collection of the Göttingen Women's Clinic.)

As to the origin of an anencephalus there are widely divergent views. V. Winckel, Ahlfeld, and others, believe that it can originate from the early rupture of a well-marked hydrocephalus (about the fourth week).[2]

[1] The suprarenal capsules are often found to be maldeveloped in cases of anencephalus. Thus in nineteen fœtuses examined by Karl Biesing they were more or less defective in all. Elliott and Armour have shown that the large size of the suprarenal gland during fœtal life is due to a peculiar hypertrophy of the cortex, which commences very early and continues until birth. Immediately after birth, this special mass of cells commences to degenerate, and at the end of the first year all trace of it has disappeared. In a case of anencephalus, they found that the small size of the suprarenals was due to the absence of this layer of fœtal cortex. The gland therefore, in this condition, develops in the same manner as does that of animals, in which the special fœtal cortex is wanting.

[2] Against this view may be urged the facts that hydrocephalic embryos are usually expelled prematurely, that the eyes are often well developed, that the base of the skull in an anencephalus is usually convex and not concave—as it should be if a condition of hydrocephalus had been present; that the remains of the brain—the so-called area cerebro-vasculosa—is usually covered with a layer of ectoderm, or even of skin; and that there may be present, associated with anencephalus, defects of the spinal cord and other organs which can have no causal relationship to a hydrocephalus.

Dareste and Perls (see p. 5) attribute the cause to pressure acting on the skull from without, produced by the head-cap of the amnion, which lies close to the cephalic flexure and so hinders the formation of the skull-cap.[1] Besides the production of a hemicephalus by cerebral dropsy—which, according to Ahlfeld, as already mentioned, is the most frequent cause of the condition—it is, according to the same author, also possible for a typical acrania to originate from time to time as a result of amniotic adhesions. Ahlfeld thinks the process is as follows: 'As a result of the adhesion of the membranous skull to the inner wall of the ovum, a stretching and enlargement of the cranial cavity occurs, which leads to a marked increase in the amount of cerebral fluid (secondary cerebral dropsy), and finally also leads to rupture of the cranial vault' (Ahlfeld). Lebedeff[2] regards a defect of development of the medullary canal in early embryonic life as the cause.

The hemicephali, as may be seen from the above observations, are not capable of long surviving their birth. They are born either macerated, or recently dead, or die soon after birth. Küstner, Wichura, Arnold, Sternberg, and Latzko, however, have seen cases of hemicephalus live for several days after birth. This is only possible when the defect of the skull—and especially that of the brain—is not too great.

During pregnancy hydramnios is not infrequently observed with this malformation (see p. 6). If twins can be excluded for certain, it is important to think of the possibility of a malformation, and especially of a hemicephalus, when the woman in question has already on a previous occasion given birth to such a fœtus. On the whole, external examination does not reveal very much in these cases. While Ahlfeld has once been fortunate enough, in a case of hemicephalus with a breech presentation, to recognise the deficiency of the vault of the skull through the abdominal walls, this is only very exceptionally possible under the best conditions: namely—with little liquor amnii, and lax abdominal and uterine walls.

[1] It is not difficult to reconcile this hypothesis with our present views as to the mode of origin of the amnion in the human subject, but it is certainly curious if this is the true explanation of the development of an anencephalus that it is so uncommon a malformation among birds.

[2] He has observed folds of the medullary plates, both transverse and longitudinal, which he thinks may produce a bending of the cerebro-spinal axis in such a way as to prevent the closure of the medullary canal, or to cause it to re-open after it has once been closed (Schwalbe). He further suggests that the differences found in different cases may be due to the varying angle which the cephalic pole forms with the trunk at the time that the pressure is exerted. No doubt the true explanation of this malformation, as of a spina bifida, is to be found in an arrest of development, either due to some primary defect in the ovum, or the result of causes acting upon it from without—such as amniotic deformities.

The diagnosis of the condition can be made during labour almost with certainty on an internal examination, when the hemicephalus is in a cephalic presentation. A characteristic feature is the sharp bony border just above the protruding eyes. In the majority of cases it is possible to feel the sella turcica and the commencement of the clivus.[1] Ahlfeld was able to make a diagnosis, in one case, by feeling the foramen magnum. Negri and Viana point out a further very interesting and almost pathognomonic symptom in cases of anencephalus: namely—that pressure exerted upon the base of the cranium produces well-marked movements on the part of the fœtus.

A hemicephalus presents most frequently, during parturition, by the cephalic pole. According to Hohl's statistics, of twenty-nine cases of hemicephalus in fifteen there was a cephalic presentation; in seven, a footling presentation; in six, 'abnormal presentations'; and on one occasion a placenta prævia was present. In the majority of the cases, the position of the head varies with the nature of the malformation. If this is only present in the head, and not in the cervical and dorsal portions of the vertebral column, then the head is usually situated in the normal position on the cervical vertebræ and, owing to the short neck, is almost fixed midway between flexion and extension. During labour, then, the finger may feel the characteristic base of the skull presenting (see Küstner). If the defect is also present in the upper part of the vertebral column, then, as a result of this, there is shortening of the posterior part of the neck in its long diameter (Inieencephalus), and during delivery the head is born with the face presenting.[2]

Ahlfeld distinguishes, as a 'nose presentation,' a position of the face which occurs when only a slight degree of hemicephalus is present, when the vertebræ are not defective and the base of the skull forms a movable articulation with the vertebral column, so that the necessary over-extension of the head is rendered possible. This is a position which occurs normally among the young of animals. Küstner does not

[1] The clivus is the grooved area of bone extending from the anterior margin of the foramen magnum to the summit of the sella turcica. Pinzani has recorded a case in which the diagnosis was made on internal examination with the membranes unruptured four days before birth.

[2] Ballantyne gives a very good account of the curious deformity, iniencephalus, which he thinks is not so uncommon as is usually supposed, and which is characterised by imperfect formation of the occiput in the neighbourhood of the foramen magnum, spina bifida of considerable extent, and retroflexion of or backward bending of the spine.

Specimen No. 815, Univ. Coll. Hosp. Med. School Museum, is a good example of this condition. The fœtus presents retroflexion of the spine with slight hydrocephalus, and a posterior meningo-encephalocele. The lower part of the sac of the encephalocele corresponds to the lumbar region. The arches of the vertebræ are absent throughout.

consider such a borrowing of words from veterinary obstetrics justifiable. If the hemicephalic fœtus is not too large, presentations of the shoulder and base of the skull may be observed (see p. 43 for a case observed by myself).

Not uncommonly, hemicephalus forms an obstacle to delivery.[1]

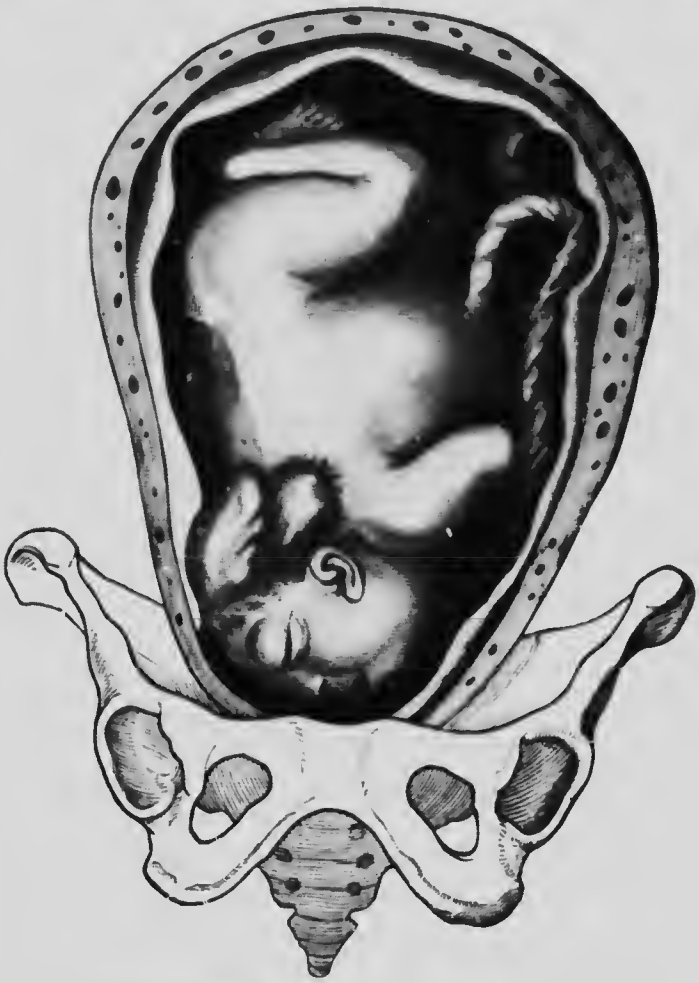


Fig. 12.—Hemicephalus Presenting by the Face. (After Küstner.)

Such an obstruction can, for instance, occur when the head is attached to the neck in a position of very marked extension, as in the inencephalus already mentioned. Ahlfeld pictures, on p. 418 of his 'Text-book of Midwifery,' such a variety of hemicephalus. Usually, however, the obstacle to delivery is the result of difficulty in the birth of the

[1] Hohl records twenty-four operations in forty cases.

monster from the abnormal breadth of the shoulders—a condition frequently observed in cases of hemicephalus in relation with an excessive body-weight (8 to 10 lb.).[1] This marked development of the body-weight is due, according to some authors (see Stumpf, 'v. Winckels Handb. der Geb.,' vol. iii., pt. 3, p. 353), in part to an increase in the duration of pregnancy, which is said to result from the fact that the imperfectly formed skull in the anencephalus does not exert so marked a stimulus as usual on the uterus. Either the shoulders do not enter the pelvis at all, or they become wedged in the pelvic cavity.

As the skull, from its imperfect development and form, generally cannot be utilised as a point of application of the force used—namely, forceps or manual traction—the attempts at extraction must be made on the shoulders. For this purpose one or more fingers are placed in the axilla, or a blunt hook is employed and applied in the axilla. If this method does not succeed, then the posterior arm should be brought down and traction made on this, which has the further advantage that by this means the circumference of the shoulders is diminished. As a last resource the division of one or both clavicles may be practised—cleidotomy (v. Herff). To avoid these difficulties, Spiegelberg recommends version, in cephalic presentations, when there is delay in the advance of the child, and extraction by the feet—a method which naturally can only be employed when the shoulders are not already wedged in the pelvis. In these cases, Spiegelberg employs also the methods described when the craniоclast fails, as will always be the case if the solid blade cannot be placed in the mouth and the fenestrated blade over the vault of the skull (Fritsch). Olshausen and Veit also recommend version.

Marie Ode Henri de Fleurian has made detailed observations, on the vital reactions of cases of hemicephalus. According to this author, the anencephalus is a monster in which all the various biological manifestations can be observed that the normal new-born fœtus shows.

These vital reactions are, however, always accompanied by an almost total absence of the brain. As a result of this, the centres concerned with movements are absent, and the cerebellum is also often deficient, only the pons Varolii and the medulla are practically normal. In the last, however, the pyramidal tracts are often wanting. It would seem, therefore, that all the manifestations observed, whatever they may consist of, are nothing more than simple bulbo-spinal reflexes. Arnold has carried out some very interesting observations on a hemicephalus which survived for three days after birth. It weighed six pounds, the pulse and respiration were normal, and on introduction of the fingers into the mouth attempts at sucking were made. Fluids were drunk,

[1] It is not uncommon for an anencephalic fœtus to weigh from 4000 to over 5000 grammes (8·75 to 11 lb.).

and micturition and defæcation were normal. The child cried seldom, but moaned a good deal. A neurological investigation, specially undertaken, showed in the first place pronounced reflex excitability. Mechanical and electrical stimulation of the superficial surface of the brain gave the following results: When the part lying above the lamina cribrosa was stimulated with a blunt pin, convulsive quivers occurred over the whole body, while stimulation of the parts lying posterior to this gave less marked results (for the results of the stimulation of other motor centres, see the conclusions and figures given by Arnold). From these observations of Arnold the very interesting fact is shown that portions of the brain are capable of conducting and functionating, in which histological examination shows no sign of any normal brain substance. Arnold himself says: 'I must admit, I had formed quite another opinion as to the organisation of the cerebrum from the results of the electrical stimulation.'

In another case of hemicephalus, Sternberg and Latzko examined the physiological functions. The child cried loudly, and the sucking reflex was present. In spite of the absence of the pyramidal tracts, co-ordinated movements of the extremities were present. The reflexes —especially the grip reflexes—were present, but warding-off movements were absent. Closing of the lids on the eyes were noticed, but there were no movements of the ocular bulbs, nor did the pupils react. The temperature was always subnormal.

In the Göttingen Women's Clinic, during the last twenty years, twelve cases of anencephalus have been observed in the Clinic and the Midwifery Polyclinic—three in the former and nine in the latter. In five cases, birth took place in a vertex presentation, in three cases in a face presentation, in two cases in a footling presentation, in one case in a transverse presentation; while in one case, the head and the shoulders passed into the pelvis at the same time and were born together. Hydramnios was noticed six times; in one case there were six litres present, and in one as much as ten litres of fluid. In nine cases, the hemicephalus lived a shorter or longer time; in one instance, twenty-four hours. In two cases the fœtus was recently dead, and one was macerated. Half the women were primiparæ, half multiparæ.

It is noteworthy that on several occasions the midwife had diagnosed a foot presentation when an anencephalus was present. In one case it was noted that there was an unusual stiffness of the whole body present, although the heart was beating strongly and well marked movements of the fœtus were present.

Of the clinical importance of the other malformations mentioned at the beginning of the chapter, there is nothing specially to be said. In exencephalus the misplaced brain in a cephalic presentation may be mistaken in certain circumstances for placental tissue.

Literature

Ahlfeld, *loc. cit.*, pp. 24 and 31; Lannelongue, quoted by Leser, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 31; Küstner, *Müller, loc. cit.*, p. 15; Kleinhans, *loc. cit.*, p. 34; v. Winckel, *Lehrbuch. d. Geburtsh.* (1893); Dareste, *loc. cit.*, p. 15; Perls, *Lehrbuch. d. allg. Pathologie* (1879); Lubarsch, *loc. cit.*, p. 15; Wichura, *Jahrb. f. Kinderheilk* (1902), p. 131; Negri and Viana, *Zentralbl. f. Gynäk.* (1907), p. 974; Hohl, *loc. cit.*, p. 15; Spiegelberg, *loc. cit.*, p. 15; Fritsch, *Lehrbuch. d. Geburtsh.*; Olshausen and Veit, *Lehrbuch. d. Geburtsh.*; E. Schwalbe, *loc. cit.*, p. 15; Marie Odile Henri de Fleurian, *Diss. Bordeaux*, 1903; Arnold, *Ziegler's Beitr.*, vol. xi.; Sternberg and Latzko, *Deutsch. Zeitschr. f. Nervenheilk*, vol. xxiv.; Stumpf, *loc. cit.*, p. 44, vol. iii., pt. 3; Karl Biesing, *Diss. Inaug., Bonn*, 1880; Elliott, T. R., and Armour, R. G., *Journ. Path. and Bact.* (1911), vol. xv., p. 481; Taruffi, *Storia della Teratologie*, vol. vi., pp. 97–160 (1881); Ballantyne, *The Embryo: Antenatal Pathology and Hygiene*, pp. 332–351 (1904).

Hernia Cerebri, Cephalocele, Myelocele, Encephalocele, and Meningocele

A hernia cerebri is formed when a portion of the contents of the skull protrudes, in the form of a hernia-like sac, through a partial defect in the otherwise closed vault of the skull. The cause lies, in the majority of cases, in defects of ossification, or in local areas of diminished resistance in the membranous skull-capsule (Ziegler). In other cases, the contents of the skull are caused to protrude by the action of amniotic adhesions, by which some portion of the membranous skull is drawn out. In other cases, again, a defective formation of the primary cerebral vesicle is present, or neoplastic conditions which are to some extent responsible for the formation of an encephalocele (Tillmanns).[1] Other cases are the result of a circumscribed hydrocephalus (see the case described below from the Göttingen Clinic). The combination of a general hydrocephalus and meningocele has also been observed. Thus Schultze reports the case of a merchant nineteen years of age in whom, besides a hydrocephalus, an occipital meningocele was present. The patient showed a satisfactory degree of intelligence.[2]

[1] While all these conditions can be explained as due to defects of development affecting some parts of the primitive brain or skull, yet, as has been mentioned in considering the origin of a spina bifida, some other condition—such as overgrowth—is probably also present, since in certain cases of encephalocele—for example, the parts of the brain lying outside the skull—may be represented by similar normally developed parts within the skull.

[2] In view of the fact that in the adult, with defects of the skull, the intra-cranial contents do not protrude unless there is some increase in the intra-cranial pressure, it is probable that in these congenital protrusions either there is primarily an excess of the cerebro-spinal fluid, or there is some obstruction to its outflow by the normal channels.

The size of the hernia varies considerably, from that of a pea to that of the head of a child. The condition is called a meningocele when the arachnoid and pia mater only protrude as a result of the collection of fluid in the subarachnoid space. In a meningo-encephalocele more frequently met with portions of the brain are found at the same time in the hernia. An encephalocele[1] is a hernia cerebri which contains only pia mater and true brain substance without any collection of fluid. In the hydrencephalocele[2] the brain matter contained in the hernia is thinned out by the presence of an abnormal quantity of fluid. A hernia cerebri is found most frequently at the occiput close above the foramen magnum (Hernia occipitalis),[3] and at the root of the nose in the middle line (Hernia anterior). Frequently also, it occurs in the region of the lateral[4] fontanelles (Hernia lateralis); at the base of the brain, (Hernia basalis inferior); and lastly, in the region of the sagittal suture and the large fontanelle (Hernia sagittalis superior). In this position they may be mistaken for dermoids, which are frequently situated in the region of the sagittal suture, or of the large fontanelle. If they are situated at the base of the skull in the sphenoid, they may, after displacing the palate, protrude downwards and forwards, even out of

FIG. 13.—HERNIA CEREBRI OCCIPITALIS. (Specimen from the collection of the Göttingen Clinic.)

[1] No distinction is usually made between a meningo-encephalocele and an encephalocele.

[2] Better called an encephalocystocele, the collection of fluid usually communicating with the ventricle. In these cases the layer of cerebral tissue may be so thinned out that it can only be recognised by histological examination, and for this reason these cases are often thought to be simple meningoceles.

[3] With this condition cervical spina bifida is very constantly associated.

[4] That is, the postero-lateral fontanelle.

the mouth, and lead to great diagnostic difficulties (Hernia cerebri palatina).

Hernia cerebri are always congenital malformations; they form fluctuating tumours, as already mentioned, of varying size. The more

FIG. 14.—HERNIA CEREBRI OCCIPITALIS. (Specimen from the collection of the Göttingen Women's Clinic.)

fluid they contain the more transparent are they to transmitted light. The skin covering them is either normal, or altered by scarring, or abnormally thinned. As a result of irritation it may become adherent to the underlying tissues.[1] Very frequently at the point of origin of the hernia a pedicle is present. The respiratory movements and pulsa-

[1] Besides skin, the coverings generally consist also of the cerebral membranes, modified or unmodified—that is, arachnoid and pia mater.

tions of the brain can often be observed in these tumours. They become particularly prominent if the child cries. By pressure on the hernia, and resulting compression of its contents, it is usually possible to produce the recognised symptoms of cerebral compression: namely slowing of the pulse, vomiting, irregular breathing, cyanosis, and convulsions. The completely shut off meningoceles, present in rare cases, may be mistaken for congenital cysts.

Children with large herniæ cerebri, in the majority of cases are not capable of living, since a large part of the brain may be displaced: as

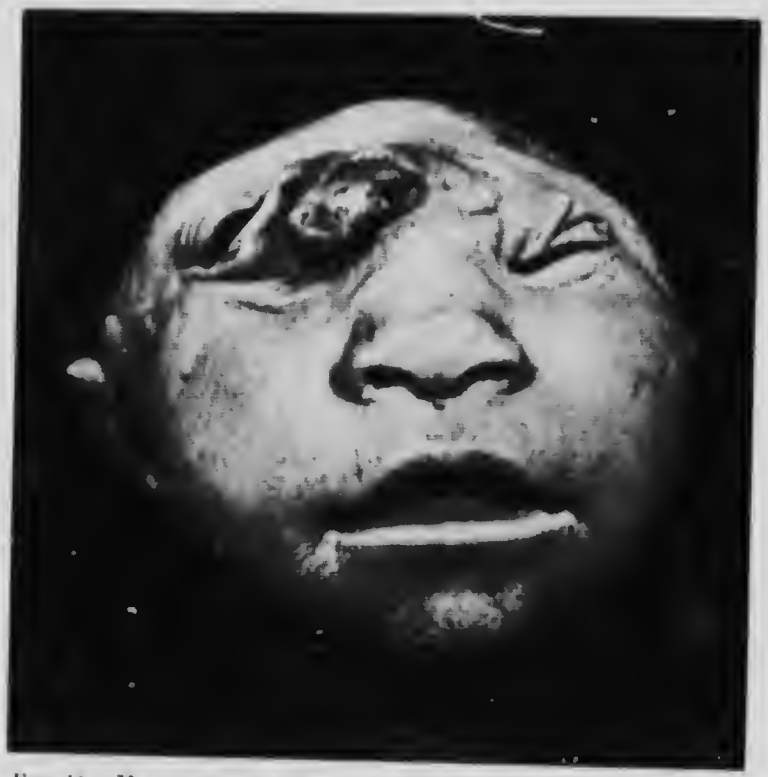


FIG. 15.—HERNIA CEREBRI ANTERIOR. (Specimen from the collection of the Göttingen Women's Clinic.)

a result of which marked cerebral symptoms may be present—such as paralysis, contractions, idiocy, and convulsions. Only children with small herniæ can continue to live without such disturbances. Often the tumour bursts during or soon after birth. In other cases it may continue to grow and burst later: as a result of which a purulent meningitis may occur. In very exceptional cases they may also remain stationary. Thus Heineke records meningoceles in patients 12 to 17 years old (Tillmanns). The frequency of meningoceles is about 0·3 per 1000

(Moscow Findelanstalt).[1] Small meningoceles covered with thick skin or membranes can remain without affecting the individual concerned; but the further course of such cases left to themselves is more unfavourable (according to Biedert and Fischl) than is that of those operated upon. Thus of 80 encephaloceles, 32 died within the first seventy-seven days, 13 at an unknown age, and only 6 survived the second year of life; while of 77 cases operated upon, 14 were cured.

As regards treatment, attempts may be made in the case of quite small meningoceles with a small aperture to replace the hernia and to keep it replaced with a compressory bandage. The following method is a better one: After the narrow neck of the sac has been closed by a subcutaneous suture (Leser), the contents are sucked out with a syringe, and the sac is then injected with tincture of iodine, Lugol's solution,[2] or alcohol. In this way an endeavour is made to obtain a permanent obliteration of the inner opening of the sac through the inflammatory reaction set up. Of late, both meningoceles and encephaloceles have been treated by operation. When there is a portion of the brain in the hernia, then radical operations can only be carried out when the removal of the prolapsed portion of brain does not produce serious after-results. The small frontal and occipital encephaloceles are best suited for such treatment. All those cases of hernia cerebri, where large portions of the brain lie in the sac, or where other severe malformations are present (for example, microcephaly) should be excluded from any operative treatment. Taking these factors into consideration, a large number of meningoceles and encephaloceles have been operated upon with good results. For instance, Cameron has reported a successful operation on a meningo-myelocele on the day of birth.

During delivery, these malformations seldom if ever give rise to any trouble, since the sacs are as a rule so flaccid that they can pass, without difficulty even when of large size, through the pelvis and the soft parts (see, for example, the cases recorded by Zorn, Leopold, Austerlitz, Thieme, Façce, and Schäffer, and a case occurring in the Göttingen Women's Clinic). At times, they burst during delivery. Nevertheless, in some cases, obstruction to delivery occurs necessitating puncture of the sac, so that the prognosis which, to start with, is not very good, becomes very bad (see the case from Zweifel's Clinic).

Whenever possible, puncture should be avoided for the reason that children with meningoceles are in certain circumstances capable of living. In cephalic presentations with failure of the expulsive forces

[1] Bathantyne quotes Trélat as having seen three cases among 12,000 labours; while in his own series of 325 specimens of fœtal disease and deformity, there were eight instances of cephalocele.

[2] Iodine, 2 parts; iodide of potassium, 3; water, 40. (Liquor Iodi, B.P. 1885.)

In suitable conditions the application of the forceps should be considered, and in pelvic presentations the careful extraction of the child. If necessary, in the interests of the mother—for example, with thinning of the lower uterine segment—without considering the child, either the hernia must be punctured or the sac, if necessary, removed with scissors or a curved bistoury. The exact diagnosis often presents great difficulties during delivery. Confusion may arise with double monsters, other kinds of tumours, teratomata, and even with the membranes and the placenta. In many cases the hernial sac has been mistaken for the breech (case in the Göttingen Polyclinic). If the expulsion of the child is delayed and the diagnosis is uncertain, it is best to introduce the whole or half-hand, under anæsthesia, as in this way a certain diagnosis will most quickly be arrived at.

Fig. 18.—Hydrencephalocele Palatina (After Leser.)

In a certain number of cases, obstruction to delivery may occur through the hernial sac causing faulty positions and presentations of the child: for example, pelvic presentations, face presentations, or brow presentations. A hernia cerebri in the region of the occiput can prolong the posterior arm of the head lever, so that the same conditions obtain as in a case of a dolichocephalic skull. The result is a face or brow presentation.

A very remarkable case of this nature was observed in the Göttingen Polyclinic (Lina, R., 'Polyclinical Journ.,' 1898, No. 48). Help from the Polyclinic was sought in the case of a primipara twenty-four years of age on account of a brow presentation. On the arrival of the assistant, the symptoms of a threatened rupture of the uterus were present, the retraction ring was felt two fingers' breadth below the umbilicus, the abdomen was very tender, and the patient very excited. Internal examination showed that the cervix was fully dilated, the membranes ruptured, the child alive and presenting in the first brow presentation. Attempts to convert the brow into a vertex or face presentation were unsuccessful. As the lower uterine segment was very distended, perforation of the living child was

carried out, and the child then extracted with the cranioclast. After the birth of the child, a very large hernia cerebri occipitalis was found. The head presented the typical moulding of a brow. In this case, then, the hernia cerebri had led to a brow presentation as a result of the elongation of the posterior extremity of the head. In the second place, the combination of the brow presentation and the hernia caused very marked obstruction to delivery, with over-distension of the lower uterine segment. Davies has also reported a case of dystocia as a result of the presence of an encephalocele. It was the case of an octipara in whom, with a face presentation, labour did not progress. On performing version, a large fluctuating tumour was found at the occiput. On attempting to extract the after-coming head, the tumour burst and gave exit to a stream of fluid resembling pus. Extraction was then easily accomplished. The sac turned out to be an encephalocele occipitalis containing almost one litre of fluid. In it lay the occipital condyles. The woman had already given birth to a child with a frontal meningocele and spina bifida.

Thies reports a similar case from Zweifel's Clinic. In this case a hydrencephalocele occipitalis containing 1800 c.c. of fluid led to a face presentation. The forceps applied, on account of threatening asphyxia of the child, failed to deliver, as the head could only be drawn down as far as the region of the large fontanelle. The tumour was then recognised, and extracted with great stretching of the lower uterine segment. After this, the labour proceeded without difficulty. The child breathed only a few times.

A further case is reported by Goldberger. The patient, a tertipara, was twenty-six years of age, and the membranes had been ruptured for twenty-four hours. The external os admitted four fingers, and in it there was a smooth flat body not containing any portions of bone. On an internal examination with the whole hand a tumour was found springing from the skull. Podalic version was performed, and a full-term well-developed dead child extracted with difficulty. The tumour growing from the squamous portion of the occipital bone by a broad base was smooth and elastic, about the size of a child's head, and had a circumference of 36 cm. The base was covered with a hairy scalp.

In the Göttingen Obstetrical Polyclinic, a case of encephalocele occipitalis occurred a short time ago which has been described in detail by Rödelius. In this case of hydrencephalocele occipitalis, the hernia cerebri (about the size of a small child's head) was born first, followed by the head in an occipital presentation. The child lived three days, took the breast, passed urine and meconium in considerable quantities, and did not show any signs of paralysis. At the autopsy, the small fontanelle was found to be round in shape and about the size of a sixpenny-piece.

In the hernia, between the brain substance and the membranes, about 150 grm. of blood-stained fluid were found. The enlarged cerebellar hemispheres were contained in the sac, while the vermis had remained within the skull. In the centre of the cerebellar hemispheres there was a considerable quantity of blood-stained fluid.

Fig. 17.—Cyclopia. (Specimen from the collection of the Göttingen Women's Clinic.)

The cause of the formation of the hernia in the above cases is not altogether easy to explain. It may be that the cerebellar substance had from the very beginning shown a tendency to excessive growth, and that the fluid in the hemispheres had collected secondarily (as a result of disturbances of nutrition): but it is also possible that the pathological collection of fluid in the cerebellar hemispheres was the primary factor (see the observations of Rödelius).[1]

Fig. 14 is a hernia cerebri anterior, noteworthy, as it is taken from a preparation in the Göttingen collection which v. Osiander showed at a meeting of the Royal Society of Science in the year 1812. Osiander depicts in epic breadth, in ten printed pages, the course of the birth, the malformation, and its causes. The child died on the fourteenth day after birth of disturbances of nutrition, caused, as Osiander thought, by the continued anger of its mother. 'The mother was angered because almost all day she was visited by people—often in a most

[1] Encephalocystoceles of the frontal region usually communicate with the anterior horn of the lateral ventricle, those of the superior occipital region with the posterior horn, and those of the inferior occipital region with the third ventricle.

impetuous fashion—who came singly or in troops, not only to satisfy their entirely useless curiosity but also to tell her the most fantastic tales about the child: for instance, that it would have an ox's horn, whatever else it might develop.'

An autopsy showed that anteriorly, in the glabella, there was an opening, through which the right hemisphere of the cerebrum had passed together with the membranes and formed the small horn-like process. The brain itself was very poorly developed, and dropsy of the cerebral ventricles was also present. As in the month of June several fœtal monsters were born, Osiander suggested as a cause of these malformations, 'the favouring effect of the warm summer and autumn weather of the previous year on the breeding of men and beasts, by which more pregnancies ensued. For the same reason, too, more births had occurred in the first half of the year in the Göttingen lying-in Hospital than usually occurred in three-quarters of the year. The warm winter was also so favourable to the development of the fruits of the womb, that those which usually came to an end as early and unrecognised abortions, and those in other years occurring as monsters, in this year reached a degree of development which they would not have attained in climatic conditions less favourable for breeding and growth. No wonder, then, that among so large a number of births there should be many exhibiting fœtal monstrosities.'

Cases of cyclopia, synopsia, synophthalmia, cyclencephalus, arhinencephalia,[1] occur as a result of faulty development of the anterior part of the brain.[2] This consists chiefly in an arrest of development of the anterior cerebral vesicle. As a result, the large brain and its ventricle remain single and a faulty separation of the optic vesicles also occurs. The two eyes may be completely fused (Cyclopia), or the two eyes united together may lie in a single cavity (Synophthalmia): the rudimentary eye thus formed lies in the region of the root of the nose (for more detailed changes, see v. Hippel). Usually the nose is at the same time stunted, and forms a snout-like skin appendage, without any bony framework, which is placed above the eyes (Ethmocephalus). The bones of the nose and the nasal cavities

[1] Congenital absence of one or both olfactory lobes.

[2] The frequent association of malformations of the visual and olfactory organs is explained by the fact, that anything interfering with the development of the primitive fore-brain, at the time of the outgrowth of the two optic vesicles, will also interfere later with the development of the olfactory organ, which appears towards the end of the third week as an area of thickened ectoderm on either side of the fore-brain. These deformities are to be explained, on embryological grounds, as the result of some failure of development. If the actual cause, as some authors believe, lies in pressure exerted on the head end of the embryo by the amnion, then we have the interesting fact that this pressure on the one hand brings about the more or less complete fusion of the eyes in cyclopia, and on the other hand the failure of separation of the two cerebral hemispheres in arhinencephaly.

are nearly always absent, the cavity of the mouth also often remains undeveloped, and occupies but a very small space.[1]

In spite of the maldevelopment of the brain, the frontal region of the skull has usually its normal circumference, owing to the fact that the amount of fluid in the ventricle is increased (see Ahlfeld). The cranial nerves which supply the eyes and the nose are either undeveloped, or rudimentary, or single. Cyclopia is one of the rarer malformations. Fœtuses presenting this deformity are incapable of living long, as a result of the maldevelopment of the brain, although some have lived for weeks and months, and one even for ten years (quoted by v. Hippel). This malformation does not possess any obstetric importance. Only in face presentations can diagnostic errors occur, as a result of imperfect examinations, owing to the altered anatomy of the face.[2]

Literature

Ahlfeld, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Ziegler, *loc. cit.*, p. 15; F. Schultze, *Nothnagels Handb. d. spez. Pathologie und Therapie*, vol. ix., No. 3, p. 208; Kaufmann, *loc. cit.*, p. 24; Kleinhans, *loc. cit.*, p. 34; Küstner, *Müller, loc. cit.*, p. 15; Biedert and Fischl, *loc. cit.*, p. 34; Keller, *loc. cit.*, p. 34; Hohl, *loc. cit.*, p. 15; Cameron, *Glasgow Obstet. and Gynæc. Soc.* (Feb. 1904), *ref. Zentralbl. Gynäk.* (1904), p. 1194; Zorn, *Diss.*, 1903; Leopold, *Geb.-gynäk. Gesellsch. Dresden* (April 16, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 560; Amsterlitz, *Diss. München* (1903); Thies, *Geb. Gesellsch. Leipzig*, (Aug. 17, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 1601; Thieme, *Diss. München* (1904); Façce and Schäffer, *Ref. Zentralbl. f. Gynäk.* (1905), p. 1096; Johansen, *Münchner med. Wochenschr.* (1896), No. 1; Davies, *Brit. Med. Journ.* (April 22, 1907); Rödelius, *Diss. Göttingen* (1909); Osiander, *Göttinger geb Anzeigen*, vol. ii., p. 1377; Unger, '*Zyklopie*,' *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxvi., No. 3; E. v. Hippel, '*Missbildungen des Auges*,' *Graefe-Saemisch*, vol. ii., p. 114; Driessen, *Operation einer Hydrokephalozele, Niederländ. gynäk. Gesellsch.* (Dec. 13, 1908), *ref. Zentralbl. f. Gynäk.* (1909), p. 344 (interesting discussion); Cushing, H., *Keen's Surgery* (1911), vol. iii., p. 111; Ballantyne, *loc. cit.*, p. 44; pp. 393–403; Taruffi, *loc. cit.*, p. 44; (1891) vol. vi., p. 565; (1894) vol. viii., p. 372; Hippel, E. von, *Schwalbe, loc. cit.*, p. 15; (1909) vol. iii., pt. 1., p. 29.

[1] The essential features of a cyclops are the absence of a well-formed nose in its normal position, the presence of a single eye (often with traces of duplicity in it) in the middle line of the face, and the defective development of the skeleton of the anterior and upper part of the face and of the anterior part of the brain (Ballantyne).

[2] Specimen No. 814, Univ. Coll. Hosp. Med. School Museum, is a good example of a cyclops fœtus. The trunk and limbs are well developed, and so is the head, except in the orbital region. In the situation of the nose is a large eye, resulting from the fusion of the two organs, and showing a transversely elongated cornea, and below the globe a body—probably the fused caruncular. Above the eye is a proboscis—the fronto-nasal process—with a transverse aperture, the skin at its end showing well-marked sebaceous glands.

CHAPTER V

Hydrocephalus

By hydrocephalus is understood an abnormal collection of fluid, amounting to five litres or more, in the ventricles of the brain, or in the subarachnoid space, as a result of which an extraordinary enlargement of the fœtal skull occurs, so that it may measure 60 to 70 cm. or more in its circumference.[1]

The more frequent variety, where the collection of fluid is within the ventricles, is distinguished as hydrocephalus internus; the other more uncommon variety, as hydrocephalus externus.[2]

Congenital hydrocephalus is nearly always of the true primary idiopathic form in contrast to the secondary form, usually extra-uterine and resulting from meningitis or tumours. Hydrocephalus has been observed also in patients with intra-cranial teratomata and congenital dermoid tumours of the brain. Hydrocephalus arising as a result of faulty development of the brain need not be considered here. In the great majority of cases a pronounced hydrocephalus, giving rise to difficulty during delivery, is already present during the period of embryonic life.[3]

Less commonly, it originates as a consequence of birth. But, even in these cases, it is generally held that the cause[4] of the condition is to be sought during the period of intra-uterine development. Nevertheless, the view cannot be entirely given up that dropsical disease of the ventricles of the brain may be set up by the trauma of birth.

[1] Anton has recorded a case in which the circumference of the skull was 85 cm. and its cubic capacity 8300 c.c.

[2] External hydrocephalus may be associated with aplasia of the brain; it may be an accompaniment of the internal variety, the result of inflammation of the ependyma, or of venous congestion from tumours of the neck, or of the base of the brain.

[3] V. Winckel figures a three-months' fœtus with hydrocephalus, anasarca, and ascites.

[4] Ballantyne believes that ante-natal hydrocephalus is generally due to an embryological arrest of development, to which may be superadded in the fœtal period a disease affecting the malformed parts; but he admits the possibility of hydrocephalus arising, without a preceding malformation in the fœtal epoch, from a disease, just as it sometimes does in post-natal life.

Of the real cause of hydrocephalus, as yet we know nothing definite. According to some authors, it is to be regarded as a malformation of the brain. As is well known, the brain develops from membranous vesicles, the walls of which gradually become developed into the brain substance; from these vesicles, filled with fluid,[1] the ventricles are formed, while the brain matter undergoes further development in a marked degree. During this process, either the development of the brain may be defective, or the accumulation of fluid may be excessive and so hinder the growth of the brain substance. Frequently, a widening of the spinal canal occurs, with the formation of an hydromyelia or an hydrorrhachis. As factors favouring the conditions described, habitual drunkenness of one of the parents, or syphilis,[2] are given; while some authors consider trauma, obliteration of the foramen of Magendie,[3] and fœtal rickets, as causes. According to Schultze, no other explanation than that of a fœtal meningitis is to be thought of in connection with the development of an hydrocephalus. The frequent presence of changes in the delicate membranes of the brain, and in the plexuses, seems to uphold his theory.[4]

Gaboil regards some infection on the part of the mother as the most frequent cause: namely, inflammation of the lungs, small pox, influenza, or chronic infections such as syphilis and tubercle. The inflammation of the lining membrane of the ventricles, present in some cases—which, according to Schultze, appears as if strewn over with fine grains of sand, or impressed with net-like ridges—may be wanting in other cases. At the present day, one is inclined to regard these changes in the ependyma as secondary rather than primary.

Hydrocephalus has been frequently found to occur in several children of one family (see a case from the Göttingen Clinic). Thus Frank reports a case of a family with six, and another family with seven children,

[1] The cerebro-spinal fluid, in all probability, is a secretion from the ependymal cells of the choroid plexuses, and in normal conditions escapes from the cranial cavity by passing into the sinuses—especially into the longitudinal.

[2] Of 362 cases of hereditary syphilis studied by Hochsinger, thirty-four were hydrocephalic. Syphilis may produce hydrocephalus, either by leading to some malformation of the brain, or by its direct effect in producing inflammation of the membranes, and so interfering with the outflow of the cerebro-spinal fluid from the lateral ventricles.

It must be remembered, however, that some of the lower animals, not susceptible to syphilis, may have hydrocephalic offspring.

[3] Not infrequently, it is possible to tap the lateral ventricles through the lumbar subarachnoid space; in such cases the block is not at the foramen of Magendie but probably at the points of exit of the fluid from the cranium.

[4] The hypertrophic condition of the plexuses present, in some cases, may be regarded as the result of imperfect involution, since at the third month of fœtal life they almost fill the ventricles, and even in the later months are relatively much larger than they are after birth.

all of whom were born with hydrocephalus. Göhlis, too, describes another case in which a woman gave birth six times running to hydrocephalic fœtuses at the sixth month, then she was delivered of three full-term children, of whom two at the age of three months, and one at the age of one and a half years, died of congenital hydrocephalus (Schultze). Very frequently other malformations — such as encephalocele, porencephalus, microcephalus, spina bifida,[1] club foot, ascites, hydramnios,[2] defect of one kidney, disease of the suprarenal bodies,[3] or diaphragmatic hernia—are found to exist with an hydrocephalus. Another very frequent complication is anophthalmia. In association with the faulty development of the fore-brain the optic vesicles are often also maldeveloped. As a result of this, only rudimentary eyes are found in the orbits (Ahlfeld). It may be taken for granted that the same cause must be assumed for these frequently associated malformations as for the hydrocephalus. Weinberg has recently described a case of congenital internal hydrocephalus associated with phocomelia, which

FIG. 18.—HYDROCEPHALUS. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] Of sixty cases collected by v. Winckel, in eighteen there was a spina bifida; and of 330 cases of spina bifida, forty-one were accompanied by hydrocephalus (Schwalbe).

[2] The frequent association of hydramnios gave rise to the view of the old authors that 'the dropsy of the ovum' could be transmitted to the fœtus.

[3] According to Czerny, the medullary substance may be absent in the suprarenal glands in some cases of hydrocephalus.

be regarded as a result of the hydrocephalus. Liese has reported a similar case.[1]

The hydrocephalic brain bears a proportion relative to the quantity of the fluid present. The latter is found in largest part in the lateral ventricles, and may be present in this position to such an amount as to convert the cerebral hemispheres into large membranous sacs. In such a case, the substance of the brain is either absent altogether, or is present only in a very thin layer (up to 2 mm. in thickness), in which the distinction between the grey and the white matter is entirely lost.[2] The cerebellum and the cranial nerves are usually unaltered. The nerve nuclei, however, may be markedly altered; and, as a result, disturbances of the organs innervated by the affected nerves, originating from the base of the brain may occur. The vault of the skull is markedly widened, in correspondence with the amount of fluid present, the bones are more or less separate from one another, and, as a result, the sutures and fontanelles are enormously increased in width. In some cases of hydrocephalus, intercalary or Wormian bones are found in the greatly enlarged sutures or fontanelles; or the sutures are filled with needle-like spicules of bone. These small bony plates, which occur as the normally placed centres of ossification of the skull-bones, as a result of the marked distension of the skull are not able to unite with one another; and so single islets of bone are left (strewn, as it were) on the periphery of the larger bone-plates (Ahlfeld).

The enlarged skull is usually globular in form, and the temporal fossæ are no longer concave, but are obliterated. The frontal, parietal, and occipital bones are displaced outwards to a considerable degree, and extend beyond the base of the skull. The bones themselves are thin and in large part translucent. The small face always forms a marked contrast to the enormously distended skull. As Schultze points out, the lower jaw appears to hang on to the skull as a narrow triangle with its apex pointing towards the chin. The eyes are usually displaced downwards and forwards, as the roof of the orbit is pushed down. On this account, too, more than usual is seen of the white sclerotic of the eyes. The upper eyelid often appears too short, so that during sleep the eyes cannot be closed. The eyebrows are displaced upwards. Under the skin of the scalp, and especially of the temporal region, there is a network of tortuous and enlarged blue veins, due in part to the

[1] Even in extreme cases of hydrocephalus, however, the fœtus may be otherwise well developed and attain a considerable size; thus Anton met with a fœtus weighing 3900 grm. Such a case forms an interesting illustration of the fact that the body may develop normally, even when there is marked malformation of the brain.

[2] In the highest degrees of development the skull becomes converted into a skin-like sac filled with fluid, with the atrophied remains of the brain substance lying at its base.

thinness of the skin, and in part to the disturbance of the circulation within the skull (Tillmanns).

The fluid contained in the ventricles is serous, colourless, or at times of a light yellowish-red colour, and shows only slight traces of albumen. Its specific gravity lies between 1001 and 1009.

Hydrocephalus results very frequently in intra-uterine death as a

Fig. 19.—Radiogram of an Hydrocephalus.

result of the cerebral disturbances present. A large number of the children die at or soon after birth, from the interference rendered necessary (perforation or tapping). But if the hydrocephalus—for instance, after having been tapped—is not too large to pass through the pelvis, such children can be born alive.[1] In these cases the further prognosis depends upon whether the process comes to a standstill or progresses. In some very rare cases, where the disease is completely arrested, the children, provided no other results of the hydrocephalus are present, may continue to develop normally; and it is a well-recognised fact that many of our best-known men in science and art have been

[1] Of sixty cases collected by v. Winckel, seventeen of the children were born alive, and of these seven died soon after birth.

hydrocephalic to a marked degree (Helmholtz, Cuvier, and Menzel). Thus Taylor, Christian, Göhlis, and Monroe have reported instances of intellectual capacity, in cases of hydrocephalus, normal or even above the average (Schultze).

Further, in very rare cases, an hydrocephalus may become gradually cured by the absorption of the fluid after the condition has ceased to progress; or the soft skull-cap may give way spontaneously, or as a result of trauma, and the watery contents having escaped, closure and healing of the wound may occur. Spontaneous rupture has also been observed through the nasal, optic, and aural cavities. In such cases there will, however, always be a suspicion that in reality cysts of the brain, or similar circumscribed collections of fluid, were present (hygroma of the dura mater, for example).

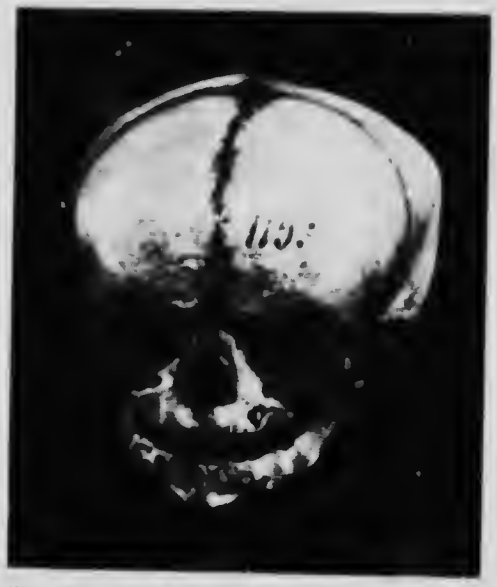


FIG. 20.—NORMAL SKULL. (Specimen from the collection of the Göttingen Women's Clinic.)

Unfortunately, an unfavourable result is more usual. The circumference of the head gradually and continuously increases, so that even in the first year of life it may measure 60 to 80 cm. and over; while in normal children, during the first year of life, it measures only about 45 cm. In the rapidly progressing cases, an increase in the circumference of the head of 2 cm. or more can be detected every two to three weeks. The children then fall ill, either of some intercurrent diseases, stomach or intestinal disturbances, or pneumonia; or they die, as the result of increasing general wasting, or epileptiform convulsions; or, finally, with the recognised symptoms of pressure on the brain: somnolence, vomiting, and slowing of the pulse. At the beginning of the illness, or when the process continues to progress after birth, faulty development of the intelligence is the first thing noticed. They hardly learn to speak at all, or only imperfectly; are sluggish and dirty, and cannot concentrate their attention on anything. This condition of impaired intelligence may finally develop into complete idiocy.

Besides these purely psychical phenomena, disturbances in the motor centres soon occur—spasms, contractures, and increased reflexes. The children do not learn to walk or stand, or they have a staggering gait. Nearly always choked disks are present, with or without optic atrophy; and resulting disturbances of vision, imperfect sight or even complete blindness, nystagmus and strabismus often exist at the same time. Death ensues usually in the first year of life.

The treatment of such cases of hydrocephalus as survive after birth is, according to the published results, a thankless task. The application to the skull of grey ointment, tincture of iodine, tartrated antimony, and silver salts (Credé) has been employed without good results. Just as little benefit has followed the internal administration of iodide of potassium and similar preparations. In a few isolated cases, antisyphilitic treatment has resulted in a cure. Recently an Italian physician, Somma, has recommended the treatment of hydrocephalus with sunlight. In this method, the children are exposed to the rays of the sun for fifteen to twenty or thirty minutes daily.

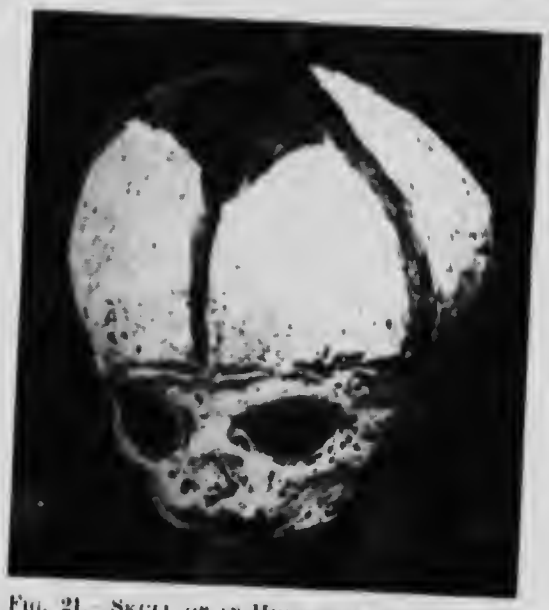


FIG. 21.—SKULL OF AN HYDROCEPHALUS. (Specimen from the collection of the Göttingen Women's Clinic.)

Better results have been obtained by Quincke's method of lumbar puncture, in which the hydrocephalic fluid is withdrawn in part, by puncture through the space between the second and third lumbar vertebræ. This mode of treatment requires to be repeated at stated intervals. I have not succeeded in checking the progress of the disease in several cases treated by this method. The immediate effect is most surprising, but it does not last long. Similar observations may be found in many places in the literature. Lastly, attempts have been made to influence the process, by tapping followed by compression of the skull, but in most cases without any result. The procedure can only be undertaken when severe damage to the brain

is not present and when the collection of fluid in the ventricles is very marked.

A further condition, necessary for a permanent result from the procedure, is such a degree of softness of the skull as to render compression

Fig. 22. Hydrocephalus in Cephalic Presentation.
(After Bumm.)

possible after the puncture has been made. This operation must only be undertaken with the strictest aseptic precautions. To avoid any damage to the longitudinal sinus, a fine trocar is introduced into the ventricle, at a distance of 2 to 5 cm. from the sagittal suture. According to the degree of the hydrocephalus, 50 to 100 or 200 c.c. of the fluid are removed. The puncture, as a rule, must be repeated many

times; and, after it has been carried out, a compression bandage is applied, and compression continued for several weeks. Some authors have recommended drainage after puncture[1] (v. Bergmann and Kocher).

In obstetrics, the importance of hydrocephalus is very considerable, although the frequency of this malformation or disease is not very great. Schuchard observed one case of hydrocephalus in 753 births; v. Winckel, eight cases in 15,000 births, or one in 1875; Kleinhans, one in 1600; and Merriman, quoted by Kleinhans, one in 900. In the Göttingen Women's Clinic, during the last twenty years eight cases of hydrocephalus have been observed among 4,200 births, or one in 525, which is a very high percentage. The percentage rose considerably during the last year, as in that year four cases were met with. A further case was admitted into the clinic after the doctor had severed the neck of the child, which was born in a breech presentation as far as the neck, and had left the head *in utero*.

If we now pass on to consider the course of labour in cases of hydrocephalus, it is obvious that the primary difficulty arises from the excessive size of the head. The failure to recognise the existence of an hydrocephalus, like the failure to recognise a transverse presentation, is a very serious error in obstetric practice. The result of this error is nearly always rupture of the uterus, from over-distension of the lower uterine segment, and resulting haemorrhage into the peritoneal cavity, or septic infection. The spontaneous birth of an hydrocephalic child is only possible when it is a case of a very slight, or, at the most, of a medium degree of severity. In the higher degrees this can only occur if the foetus is already dead or macerated, or if the hydrocephalus bursts spontaneously and so brings about the diminution in the size of the skull which is necessary before spontaneous delivery can occur.

Such a rupture of an hydrocephalus has been observed most frequently in breech presentations. The spontaneous delivery of an hydrocephalus of not too great a degree can occur in the case of a breech presentation, provided that the intracranial fluid does not fill the skull but tensely.

[1] The condition has been treated by drainage of the ventricle into the subarachnoid space. A suitable metal tube of gold or platinum is bent to a right angle, and one arm passed into the descending cornu of the right lateral ventricle, while the other arm is fixed by sutures to the inner surface of the dura mater.

This method is a sound one if the foramen of Magendie is blocked; but in the majority of the congenital cases this is not so. In such cases, the method recommended by Cushing should be employed. As a first step, a lumbar puncture is performed; then a simultaneous lumbar and ventricular puncture, to determine that the child can stand the withdrawal of the fluid—if it is clear that the ventricle can be emptied by the lumbar route; and then a laparotomy and laminectomy are performed, and the abdominal cavity placed in communication with the subarachnoid space by a trephine opening through the body of the fifth lumbar vertebra into which a silver tube is placed. Cushing has carried out this method in twelve cases with a considerable measure of success.

The hydrocephalic skull is then able to undergo the change of form necessary for the passage of the head more readily than in the case of a cephalic presentation. In a few rare cases, where a spina bifida has existed as well as an hydrocephalus, rupture of the former has occurred with escape of the whole of the intracranial fluid contents and, finally, spontaneous delivery.

The variety of the presentation of the head is of importance in regard to the progress of the labour. If the head engages in the pelvis with its greatest occipito-frontal diameter, it is much more unfavourable than when it is markedly flexed and occupying an oblique [illegible]; in the latter case, moulding can occur more easily than [illegible] variety. Hydrocephalus of moderate degree[1] is more [illegible]; in these cases the head does not engage in the pelvis [illegible] the pelvic brim. In the case of a very large hydrocephalus with its enormously thinned bones and brain substance [illegible] the thickness of paper—can be pressed into the pelvis [illegible] one form, namely, very much elongated. Or it may even [illegible] case it sometimes happens that an hydrocephalus internus [illegible] into an hydrocephalus externus as the fluid collects under [illegible] of the head.

How often interference during birth is necessary, in cases of hydrocephalus, the statistics of Hohl and Schuchard demonstrate. Of seventy-seven cases collected by Hohl, artificial aid was necessary for the termination of labour in sixty-three; and, in seventy-three cases collected by Schuchard, in sixty-two; while of seven cases in the Prague Clinic it was necessary in all of them. In nine cases recorded by H. Veit, artificial delivery had to be practised on each occasion.

The importance of this complication is also very evident from these statistics. Of Hohl's seventy-seven mothers, twenty-one died; of Schuchard's seventy-three, thirteen died; and of Veit's nine, four died. These figures also show that rupture of the uterus, as has been already mentioned, is the primary cause of these unfavourable results. Of Hohl's seventy-seven cases, four women died of rupture of the uterus; of Schuchard's seventy-three cases, twelve; and of Veit's nine cases, four. Besides rupture of the uterus, other severe lacerations and urinary fistulæ have been met with, produced most frequently through the slipping of the widely separated blades of the forceps. Moreover, women who have escaped the dangers of rupture of the uterus may die from womb infection, which occurs very readily on account of the repeated examinations necessary, the long duration of the labour, and the many attempts at delivery. Fatal post-partum hæmorrhage

[1] The clinical fact is well established that it is not the most marked forms of hydrocephalus which give rise to the greatest difficulties during delivery.

from atony of the uterus has also been seen in cases of hydrocephalus. Of late the prognosis has improved—thanks to the better practice of asepsis, and to the better obstetric teaching of practitioners and midwives (see Kleinhans).[1] Of the seven cases in the Prague Obstetrical Clinic only one mother died of tetanus puerperalis, and among the eight mothers in the Breslau Clinic there were no deaths. Of the eight mothers in the Göttingen Clinic, there were no deaths; but it must be admitted that only four of these cases were at all severe. The one case of hydrocephalus, in which the mother died of sepsis, was admitted to the clinic with the decapitated hydrocephalic head *in utero*, and the bad result cannot be laid to the charge of the clinic, more especially as in this case the uterus had already ruptured before the patient's admission.

Cases of hydrocephalus present, in the majority of cases, by the head. Pelvic presentations are also very frequently observed, according to some statistics, in from 2 to 27 per cent. of the cases.[2] In the eight cases under observation in the Women's Clinic at Göttingen, cephalic presentations occurred six times; in one case the head was macerated and hydramnios was also present, and in two cases there was a breech presentation. Very rarely is there a transverse *lie* with an hydrocephalus. In the twenty-two cases collected by Hammerschlag from the Königsberg Clinic, a cephalic presentation occurred sixteen times—once a face presentation, and on five occasions a breech presentation. According to Küstner, a shoulder presentation is prevented by the abnormal size of the head; and the shoulder is concealed, as it were, behind the head, as it cannot project beyond it. As a result, the part of the body lying below the shoulder must present, and 'it only requires then the correcting influence of the first uterine pain to change the transverse presentation, with the flank presenting, into a presentation of the lower pole of the body, or the rupture of the membranes to bring about the prolapse of the foot.' This occurs the more readily as the hydrocephalic head is best accommodated in the fundus of the uterus. According to v. Bardeleben, a further factor in the development of a pelvic presentation is this: that the specific gravity of the hydrocephalic head is less than that of the normal head, and therefore from the very beginning there is no marked inclination for it to sink downwards. The different presentations in hydrocephalus are of immense importance in relation to the course of the labour. Almost all the cases running an unfavourable course for the mother are those in which the hydrocephalus presents by the head. This will be made clear by the following consideration. In a cephalic presentation, dangerous thinning of the lower uterine segment occurs very readily, since

[1] Of thirty cases, recorded by Hoffmann and Bertino, only two mothers died.

[2] Of 273 cases collected by Kleinhans a pelvic presentation occurred in seventy-eight, or 29 per cent.

FIG. 23.—HYDROCEPHALUS IN BREECH PRESENTATION.
(After Bumm.)

the whole of the hydrocephalic child and a large part of the liquo amnii is placed above the pelvic inlet, and lies in the cavity of th uterus. The body of the uterus then gradually retracts, as the resul of the presence of the obstacle to delivery, and rupture of the uterus occurs, especially when attempts have been made to deliver by forcep or by version. In pelvic presentations, on the other hand, a large part o the original contents of the uterus (namely, the body of the child as fa as the head and part of the liquor amnii) have escaped from the uterus and dangerous thinning of the lower segment is therefore scarcely possible. Further, it usually happens that the after-coming head o the hydrocephalic child in a pelvic presentation assumes a favourable triangular shape—broad above, narrow below (Spiegelberg).

The diagnosis of an hydrocephalus during labour is not made in any very large number of cases. In some, indeed, as will be shown later, the diagnosis may be exceedingly difficult. If the case is one of hydrocephalus in a multipara, then the fact that the head does not enter the pelvis, although good pains are present and previous labours have taken place spontaneously, should lead to a suspicion of the presence of an hydrocephalus. Apart from this, it can only be a question of an osteomalacic pelvis having developed comparatively recently, or of a tumour of the soft or hard parts of the pelvic canal. Besides the position of the head, on external examination, the increased circumference of the abdomen should be taken into account. In my opinion, however, too much stress must not be laid upon this sign, as in cases of hydrocephalus of medium degree which, as has already been pointed out, are especially to be feared, the abdominal circumference may be very slightly increased, especially if there is but little liquor amnii.

The feeling of parchment crepitation of the bones of the skull, said to be pathognomonic of hydrocephalus, can only be recognised on external examination in the most favourable conditions—that is, with thin, flaccid, abdominal and uterine walls, and little liquor amnii. This crepitation of the bones of the skull, as Fritsch points out, can also be recognised sometimes in macerated fœtuses. I have been able—and so have other gynæcologists—to recognise this sign in many normal living children born prematurely. On the other hand, cases of hydrocephalus also occur where the skull-bones are not thinned but thickened.

The symptom given by Stratz—that in cases of hydrocephalus, when the head presents, the fœtal heart-sounds can be heard distinctly above the level of the umbilicus—does not appear to me to be a reliable one. The increased frequency of the fœtal heart-sounds described by Keilmann was not present in a number of the cases of hydrocephalus which I have seen.

Fabre (see Jammes) asserts that in cases of hydrocephalus, when the head presents, there is nearly always a circular furrow between the head

and the body (*coup de hache circulaire*). This symptom is, however, often absent (Hammerschlag, and in several cases observed by us). On the one hand, in cases of hydrocephalus, this sign may be obscured by very thick abdominal walls; and on the other hand, in cases where the head is not hydrocephalic, the retraction ring may be well marked.

More distinctive signs are obtained in most cases on internal examination. In the slighter and medium degrees of hydrocephalus it is usually possible to recognise the more or less markedly enlarged sutures and fontanelles, as well as the soft and smooth feel of the movable skull-bones and the irregularity of their margins. It must be remembered, however, that in some cases the enlarged sutures and fontanelles are ossified. If the case is one of a well-marked degree of hydrocephalus – especially when the coverings are very thin – the region of the posterior fontanelle may protrude like a bladder and even project into the cavity of the vagina, so that it may be mistaken for the bag of membranes. This mistake can be made the more easily since the skin covering an hydrocephalus is often very sparsely covered with hair – a condition to be explained by trophic disturbances. The supposed amniotic sac has not infrequently been punctured by mistake.

On the other hand, in many cases, an hydrocephalus has been diagnosed when tumours of other kinds were present—such as exencephalus, hernia cerebri, tumours of the breech—particularly when the membranes were especially thick. A mistake is also possible with a macerated fœtus presenting by the head when the skin of the scalp, as the lowest pole of the ovum, has become sac-like and filled with watery blood-stained fluid. This occurred to me in one case—which, however, was soon cleared up on bimanual examination. The further sign usually given as an important diagnostic feature of hydrocephalus—namely, that *ballottement* of the head cannot be obtained when the membranes are intact—is not of much importance, as it may be present or absent in cases of moderate degree, especially when hydramnios is also present. Lastly, a diagnostic puncture may be employed; but this method is not to be recommended on account of the danger of injuring the blood-vessels or the brain. A correct diagnosis can always be made if a bimanual examination is carried out under anæsthesia with an empty bladder. In this way the excessive size of the head when between the two hands can at once be detected. The suggestion to introduce the whole hand into the uterus, so as to explore the head directly, appears to me (as well as to Hammerschlag) too risky, in view of the fact that dangerous thinning of the lower uterine segment is often present.

In pelvic presentations, the diagnosis is usually first made when the child is born as far as the shoulders and its further delivery, either spontaneously or artificially, meets with difficulty.

In this case, with normal pelvic measurements, an hydrocephalus should at once be thought of, especially when a spina bifida and club feet—so frequently associated with this condition—are seen to be present. A bimanual examination in this case too will speedily render the diagnosis certain. Since rupture of the uterus in cases of hydrocephalus in pelvic presentations—as has already been pointed out—is not to be feared, the introduction of the whole hand into the uterus for the purpose of making a certain diagnosis is permissible.

When considerable force has been used in an attempt to extract the child, in a case of hydrocephalus presenting by the breech, it has happened that the head has been torn off; and when rupture of the uterus has occurred as a result of attempts at version, the head has even slipped into the abdominal cavity.

If we return to a consideration of the diagnosis of hydrocephalus, we must agree with Hammerschlag when he says that neither the abdominal circumference, nor the absence of *ballottement* of the fœtal head, the increased frequency of the fœtal heart, the recognition of the gaping sutures and fontanelles, the parchment-like crepitation of the bones, the failure of the head to enter the normal pelvis, nor the circular furrow between the head and the body, form, when taken singly, certain diagnostic signs. Only by the recognition of several of these signs together is one in a position to make a certain diagnosis. I must once more lay stress upon a bimanual examination as of the greatest importance.

With regard to the treatment of cases of hydrocephalus during labour, there is no definite agreement. According to Fritsch the hydrocephalus, whether alive or dead, should be perforated with Smellie's scissors, as soon as the cervical canal is sufficiently dilated. Then the child should be seized by the scalp, or a finger introduced into the opening, and the head extracted, which can usually be accomplished without difficulty. According to the same author, instruments for extraction are not necessary; while the use of midwifery forceps is contra-indicated because of the danger of slipping. After perforation of the after-coming head, pressure should be made from the abdomen to drive out the fluid through the long and narrow canal. In such cases Fritsch recommends perforation through the palate, because in this method the edges of the opening do not tend to come together, and so to convert it into a valve-like opening or to close it up. From these recommendations, it is evident that Fritsch pays no attention to the life of the child, and in the treatment of hydrocephalus lays the greatest stress upon everything being done during the delivery to preserve the life of the mother. Other authors pay more attention to the life of the child in view of the fact that it is quite possible for a hydrocephalic child to remain alive after birth, and even, as a

result of the arrest of the condition, to become completely cured. From this point of view, some authors recommend the use of forceps when the head is engaged in the pelvis and the bones are hard and there is therefore no possibility of the forceps slipping. Even under these conditions the use of the forceps in hydrocephalus does not appear to me to be advisable; for the practitioner can easily be deceived as to the condition of the head in such a case, and so it may readily happen that he does serious harm with the forceps. The cases of hydrocephalus where delivery has fortunately been ended by the forceps have, in the majority of instances, been recognised later on as cases of slight degree. From these observations it may be concluded that the use of the forceps in cases of hydrocephalus should be entirely rejected. According to Runge the application of the forceps, either before or after perforation of the head, contravenes the most important indications for the forceps operation, and must be regarded as bad practice.

As a diminution in the size of the skull in hydrocephalus sufficient for the treatment of the case can be obtained—according to various authors—by the employment of puncture, these authorities employ this procedure rather than perforation (Küstner, v. Winckel, Olshausen, and others).

This procedure is based primarily upon the fact that children have remained alive after puncture, without any danger resulting to the mother from this method of assisting delivery. The objection that a tapped hydrocephalus is not suited to bring about the necessary dilatation of the cervical canal and of the remaining soft parts, is contradicted by clinical observation and experience. Küstner recommends—when the diagnosis is certain—that puncture should be carried out as soon as possible: that is, when it is possible to introduce a slightly curved trocar, some 6 mm. in diameter, through the cervical canal. He gives the somewhat obvious advice to carry out the puncture as aseptically or antiseptically as possible, so that the wound in the skull may not be infected from the vagina. As much of the fluid as will, is then allowed to flow out, and the further progress of the delivery left to the natural forces. If the child is dead, perforation should be carried out, or the puncture-wound enlarged and the cranioclast employed, when the method—usually quite successful—of introducing the finger into the perforation opening does not succeed. In the same way Ahlfeld suggests the use of a fine trocar for carrying out the puncture, so as to avoid injury to the cerebral sinuses. According to Spiegelberg, a knife may be employed in place of a trocar.

Puncture is of very little value in the treatment of an hydrocephalus presenting by the breech, as the child may die in the meantime. In this case perforation need only be considered, and is best carried out through the tabular portion of the occipital bone, a lateral fontanelle, or the foramen magnum. In cases where the mouth can be reached more

easily over the perineum, perforation through the hard palate might perhaps be tried. When forcible attempts at delivery have been carried out, the head may at last become diminished in size either as the result of rupture of the cranium and escape of the fluid beneath the pericranial aponeurosis, or by tearing of the cervical vertebræ and its escape into the subcutaneous cellular tissue of the neck (see Kustner). The perforation of the after-coming head may present great difficulties when, in a contracted pelvis, it is situated high above the pelvic brim, so that it can only be reached with the perforator with great difficulty or not at all. For these cases, the spinal method of emptying the fluid, according to Van Huevel, may be employed. In this method, the lower cervical or dorsal portion of the vertebral column is divided transversely with a knife and the spinal canal opened, a female catheter is then introduced into the cranial cavity and the fluid evacuated. This method has been employed many times with success and may be strongly recommended (Ahlfeld and others; see also Kleinhans). It is a method similar to that recommended by Colmstein for the diminution in size of the after-coming head when placed high up, removal of the brain in this manner being practised, followed by washing out of the cranial cavity under considerable pressure. If, as so frequently happens, a spina bifida is present at the same time as an hydrocephalus, this may be opened and a catheter pushed from it into the spinal canal.

Roorda Smit delivered an hydrocephalic head, which had been left intact, by perforating the base of the skull through a fissure of the palate which was present. A midwife and a quack had cut off the head after fruitless attempts to deliver the child, which was presenting by the breech.

As to the methods to be employed after perforation, opinions differ widely. If the head is more or less engaged in the pelvis, or at least in the pelvic inlet, and the condition of the mother necessitates immediate delivery, then either expression by pressure on the fundus (after Kristeller's method) should be carried out, or the head should be extracted manually, after introduction of one finger into the perforation. If these measures fail, most authorities recommend the employment of the craniochast. Hammerschlag and others, however, do not consider the use of the craniochast a very suitable method, as this instrument is so likely to tear out. Hammerschlag recommends, therefore, if expression and manual delivery of the head fails, the introduction of a vulsellum into the scalp and continuous traction being made thereon with a weight, say, of three pounds. An operative procedure recommended by v. Winckel is deserving of notice. He suggests that the bones of the skull should be removed with bone-forceps, and that then the extraction of the remaining soft parts be undertaken, so as to avoid injuries to the

maternal soft parts, which can so readily occur from the projecting margins of the bones. In case this mode of extraction fails, the arm can be brought down and the body delivered by this.

G. Veit suggests, from a consideration of the circumstances, that one might be in a difficult position in the case of a well-engaged hydrocephalic head, when immediate delivery is necessary in order to save the life of the child, and he recommends, therefore, that podalic version should be performed, as the extraction of the after-coming head after puncture is not difficult. Numerous obstetricians support this view—namely, Schröder, v. Winckel, Olshausen, and others. Fritsch rejects version, as does Küstner, who raises objections—in my opinion well justified, and such as I have many times expressed—against this method: namely, that the overstretched lower uterine segment may rupture when the hand is introduced for the purpose of performing version. Runge has also grave objections against version. When we remember how seldom children with pronounced hydrocephalus survive after birth, and how bad the later prognosis is, the regard paid to the life of the child—if we may judge from the literature—appears to be exaggerated. A case of hydrocephalus tapped during delivery and continuing to live is a great rarity. Thus Spiegelberg has not succeeded in finding a single instance in the literature in which it is proved that a child continued to live after it had been tapped for hydrocephalus. It cannot be permissible, therefore, in cases of such rarity, to suggest a method of delivery for preserving the life of the child which may, in certain circumstances, endanger the life of the mother.

As further, in the extremely rare instances in which a case of hydrocephalus survives after tapping, the condition almost always tends to progress; the children, as has been pointed out, falling victims to a progressive idiocy with ultimately a fatal result, it may indeed, be the best treatment, if such an operation, whenever possible, is undertaken as offers the best chances for the mother—that is to say, perforation of the head of the child and the necessary subsequent measures already described. No regard whatever should be paid to the life of the child in a case of well-marked hydrocephalus. Anyone who has seen at an autopsy the marked changes present in the tissues of the child's brain produced by the abnormal collection of fluid in the ventricles—and more especially the atrophy of the cerebral hemispheres and of their important centres, until the tissues are only of the thickness of paper—cannot help smiling at the methods of delivery still suggested by many in cases of hydrocephalus, designed with a view to preserving the child's life. Similar views to these are held, too, by Runge and Kleinhans.[1]

In the last twenty years in the Clinic at Göttingen, eight cases of hydrocephalus have been observed during delivery. In a further ninth

[1] Practically all English authorities would concur in these views.

case the woman was admitted in labour with an hydrocephalic head, after the doctor who had attended her had decapitated the child, which was presenting by the breech and was born as far as the neck. In another case, an acute hydrocephalus occurred in connection with delivery. Of the eight cases observed, seven were multiparæ and one a primipara. There was nothing to be noted in the history of any of the women which could be considered to account for the onset of the hydrocephalus. One of them had been treated for several weeks, in the early months of her pregnancy, in the Göttingen Women's Hospital, for excessive vomiting of pregnancy. One patient, with nine children, had already given birth to an hydrocephalus at her eighth confinement. The pelves were all normal, with the exception of one markedly flat pelvis. In six cases there were cephalic, and in two breech presentations. In two cases, labour occurred spontaneously. In one of these the child was macerated, and its head was very much elongated after birth (Jr. No. 20204). The other child, a female (Jr. No. 14992), was born prematurely at the eighth month, its head measuring 31 cm. in circumference. The third stage of labour and the puerperium were normal in both cases. Post mortem examination of the two cases showed hydrocephalus internus, chronic granular ependymitis with hæmorrhages under the pia mater, the pleura, and the pericardium. There were no signs of syphilis present.

The third case of hydrocephalus, a male child (Jr. No. 16793), was delivered by expression. The third stage and puerperium were normal. The measurements of the head immediately after birth were as follows: fronto-occipital diameter, 12·5 cm.; bi-parietal, 11 cm.; bi-temporal, 8·5 cm.; circumference, 40 cm. The weight of the child at birth was 3,100 grm., and on leaving the hospital, 2,770 grm. On the seventh day after delivery, marked œdema of the hands and feet of the child was observed. At the same time a curious contraction in a position of flexion of the upper and lower extremities became evident; the muscles were as hard as a board; the overlying skin, however, was unaffected, as it was quite soft, and, on the extensor surface, freely movable. The child cried with a feeble hoarse cry. An attempt to bring the limbs from the flexed into the extended position failed. There were no changes at the umbiliens. Gradually the œdema and contracture disappeared, so that the condition lasted altogether about eight days. Such appearances are often noted in cases of chronic hydrocephalus and are characteristic. They are produced by the degeneration present in the pyramidal tracts, but can also occur without this—simply as a result of the pressure of the increased fluid.

In the fourth case (Jr. No. 18956) there was present a marked degree of hydrocephalus. The mother, twenty-six years of age, was a tertipara, and there was nothing of note in the history. She was admitted

in labour, twenty-four hours after the rupture of the membranes. On examination, she was found to be a strong woman, her face showing signs of severe pain. The labour pains were strong and very painful; the pulse-rate, 110; and temperature, 38·5° C. Attempts at delivery had not been made. The urine drawn off by a catheter was blood-stained, but contained neither shreds nor casts. The abdomen was very markedly distended, and extremely tender on examination. Somewhat above the mid-point, between the umbilicus and the symphysis pubis, there was present on either side a well-marked transverse groove. Reaching up to about the same level, and completely filling the false pelvis, there was a hard, smooth, tumour—the abnormally large head. The breech was at the fundus uteri. The fœtal heart-sounds could be heard, normal in frequency, on the right side above the umbilicus. The whole uterus was markedly elevated. On internal examination the cervical canal was dilated to the size of the palm of the hand; and occupying it was a smooth elastic tumour, in which no irregularity could be detected, and which closely resembled a distended bag of membranes. Above, the tense membranous sac could be felt to pass over into an irregular bony margin. A diagnosis was made of hydrocephalus presenting in the second vertex position and impending rupture of the uterus. Under anæsthesia, perforation and craniotomy was performed without any difficulty. On perforation of the head, two litres of fluid were evacuated. The child, a female, weighed (without the two litres) six pounds; and, besides the hydrocephalus, there was present a spina bifida and a club foot. There was slight post-partum bleeding due to atony of the uterus, but the puerperium ran an uninterrupted course.

In the fifth case (Jr. No. 23544), the patient, a secundipara, had been treated, during the first months of pregnancy, on account of excessive vomiting of pregnancy. The child, a male, presented in the first vertex position. On account of thinning of the lower uterine segment, perforation and craniotomy were performed without any difficulty. Besides the hydrocephalus there was present a cleft palate, club foot, amniotic ring-like constrictions of the fingers, syndactyly of the toes, and a double facial fissure. The third stage of labour and the puerperium were normal.

The sixth case (Jr. No. 23834) was a very interesting one from other points of view. The mother was a quadripara, and the previous labours had taken place normally. In the last months of pregnancy slowly increasing hydramnios and œdema were observed. The soft skull presented and the fœtus was very movable—twelve to fifteen litres of liquor amnii being present. Version was performed, and the child was extracted deeply asphyxiated, and could not be revived. Plugging of the uterus was required on account of atony. The puerperium was normal. The fœtus, about five weeks premature, was

of the male sex and 42 cm. long. Hydrocephalus of medium degree was present, the circumference of the head being 35 cm. There was very marked swelling of the anterior and lateral portions of the neck (see Plate I.). As a result of this, a position of the head was present which is observed in cases of congenital struma and in many cases of henicephalus (see p. 40)—that is, the head is depressed downwards and backwards between the shoulders. Post-mortem examination revealed an hydrocephalus internus, no struma, no abnormalities of the cervical vertebræ, only a very marked development of the subcutaneous fat, and very extensive œdematous infiltration of the subcutaneous and fatty tissues. The circumference of the neck measured 30 cm. This curious attitude of the child, in addition to the hydrocephalus, rendered its delivery very difficult.

In the seventh case (Jr. No. 24292) the mother was a nonipara, her eighth child had also been hydrocephalic; the others were normal. She was seen by the doctor eighteen hours after the rupture of the membranes, because the labour had made no progress. The child presented in the first breech presentation, and perforation of the after-coming head was performed. The child, a female, had also a flat foot and a spina bifida. The third stage of labour and the puerperium were normal.

In the eighth case (Jr. No. 24674) the mother, a secundipara, was admitted from the Midwifery Polyclinic. The child, a female, presented by the breech, and perforation of the after-coming head was carried out. Besides the hydrocephalus, it presented agenesia of the vermis of the cerebellum, a posterior lumbo-sacral meningocele and lumbo-sacral lordosis, agenesia of the sixth and seventh ribs on the left side, and status lymphaticus. The third stage of labour and puerperium were normal.

In the ninth case (Jr. No. 24419), a quadripara was admitted into the clinic with an hydrocephalic head retained *in utero*, the rest of the body of the fœtus having been removed. A doctor was called on account of hæmorrhage which, on his arrival, was only moderate. An internal examination revealed a placenta prævia. The child was in a breech presentation and apparently had been dead for some days. A foot was brought down and the bleeding immediately ceased. The body could only be extracted as far as the head. The child, which had a spina bifida, was then decapitated, as the medical attendant suspected the presence of locked twins. The patient, who had a temperature of 100° and a feeble pulse of 140 to 160 per minute, presented a very collapsed appearance on her admission into the clinic. The rupture of the uterus, which was certainly present, had not been recognised by the obstetric assistant, who had omitted to examine the uterus. The diagnosis of an hydrocephalus was at once made, and the head easily and rapidly delivered by perforation and

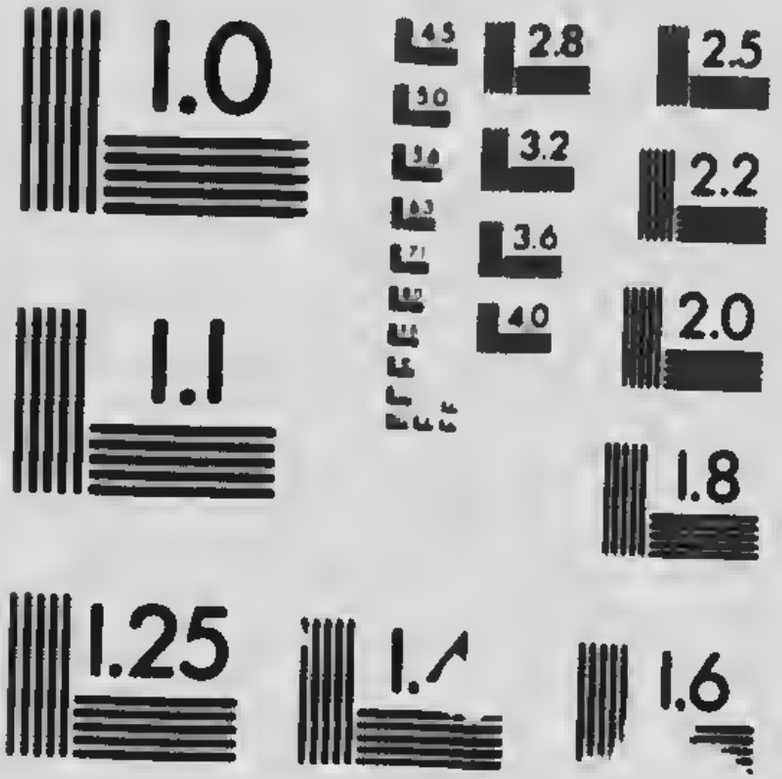
PLATE I.

MARKED HYDROCEPHALUS, WITH ŒDEMA AND EXCESSIVE FORMATION OF FAT ON THE ANTERIOR SURFACE OF THE NECK

(Specimen from the collection of the Göttingen Women's Clinic.)

MICROCOPY RESOLUTION TEST CHART

(ANSI and ISO TEST CHART No. 2)



APPLIED IMAGE Inc

1653 East Main Street
Rochester, New York 14609 USA
(716) 482 - 0300 - Phone
(716) 288 - 5989 - Fax

manual extraction. The woman at first improved; but the pulse-rate and temperature very soon rose again, and she died, on the eighth day after delivery, with gradually increasing coma and heart failure. Haemolytic streptococci were found in the blood, and an autopsy showed a complete rupture of the uterus, peritonitis, endocarditis, and an infarct in the spleen.

In the tenth case (Jr. No. 20484), the mother, a primipara, had suffered many years previously from typhoid fever, but all the organs were sound. Parturition occurred very rapidly, especially after the rupture of the membranes. The child, a female, healthy at the time of delivery, weighed 3,160 grm., measured 50 cm. in length, and the circumference of the head measured 35 cm. Very soon after birth an acute hydrocephalus developed. The sutures gaped to more than one finger's breadth, and the circumference increased daily by 1 cm. or more. No improvement occurred, in spite of lumbar puncture on two occasions. On the fifteenth day after delivery, the mother left the hospital at her own wish. The child's weight was then 2,800 grm. The aetiology of the hydrocephalus remained unknown. It could not be determined whether parturition acted as a trauma on an especially vulnerable brain, or whether the cause was present before birth and the process had lain latent until the time of delivery. Possibly it was a case of acute widespread thrombosis of the veins of the brain and of the scalp – especially of the sinus longitudinalis – or of the vena magna Galeni, as has been described in cases of acute hydrocephalus (see Schultze).

In the pathological collection of the Göttingen Women's Hospital there are two specimens of hydrocephalus from the same patient – a complete child (Specimen No. 113), and an hydrocephalic skull (Specimen No. 47a). In the specimen of the complete child with hydrocephalus, Osiander performed perforation through the occipital bone on May 4, 1800, and evacuated about four litres of fluid. A year later, the mother was again delivered of an hydrocephalic foetus.

Obstetrical Injuries to the Skull

As an appendix to the preceding section we may now proceed to consider injuries to the skull occurring during birth. A detailed consideration of this question may be found in Keller, 'Deutsche Klinik'; Küstner, 'Müller's Handbuch'; Birnbaum, 'Volkmann's Vorträge'; and Stumpf, 'v. Winckels Handbuch.'

A caput succedaneum is, if we may so term it, the most frequent injury during delivery. It occurs in the majority of cases after rupture[1]

[1] In two frozen sections, Barbour has observed the commencement of the formation of a caput succedaneum before the rupture of the membranes.

of the membranes, in living children, on that portion of the body which is situated for a long time during delivery either in the cervical canal, the vagina, or the vulva. It is found therefore, in a first-vertex presentation, on the right parietal bone;[1] in a face presentation, on the face; and in a breech presentation, on the breech. It is formed by an œdematous blood-stained infiltration of the skin, and of the cellular tissue between the scalp and the periosteum (in cephalic presentations). Small extravasations of blood are also found in the same situation. These changes occur at the periphery of the girdle of contact of the child's body with the cervix, vagina, or vulva, and are the result of the venous hyperæmia present in this position.

A caput succedaneum is a perfectly harmless condition, which disappears a short time after birth—at the latest, on the second day. The small (mainly punctiform) hæmorrhages[2] remain usually for a few more days and allow, even after this lapse of time, of certain definite inferences being drawn as to the mechanism of delivery. In very rare cases it has been found that exciting causes of infection, especially the organism of erysipelas, have penetrated through small abrasions of the skin, and have led to severe and even fatal illness. Ehrendorfer (see Stumpf) has observed necrosis of the compressed portion of the scalp in two children of the same mother, owing to fibroid degeneration of the lower uterine segment.

In a cephalhæmatoma—the next most frequent head injury—there is a collection of blood between the bones of the skull and the periosteum, occurring after birth and leading to the formation of a circumscribed fluctuating tumour. Besides this variety, which is distinguished as a cephalhæmatoma externa, there may be also, in rare cases, an effusion of blood between the bones of the skull and the dura mater, which is distinguished as a cephalhæmatoma interna. Small extravasations of blood may also occur before birth.[3]

In a series of twenty cases collected by Kee, in nine instances a cephalhæmatoma externa was present at the same time as a cephalhæmatoma interna. This ratio appears to be very high in comparison with the experience of other authors. Both these conditions occur either as the result of some injury to the skull, or from the presence of

[1] With the head normally flexed, at the posterior end close to the sagittal suture. When internal rotation has occurred, and the head is delayed for some time on the pelvic floor, another, or secondary, caput may form upon the part which presents at the vulva—namely, in a first vertex presentation near the posterior fontanelle.

[2] These small hæmorrhages may occur into the skin, pericranium, periosteum, and the bone, and show the original position of the caput, even in cases in which the serous exudation has to some extent changed its position, as may happen when the child has been lying for a long time on one side only.

[3] A sub-aponeurotic cephalhæmatoma may also occur. In this case there is a diffused extravasation of blood under the pericranial aponeurosis.

congenital clefts, the result of defects of ossification, which are found more especially at the seat of election of cephalhæmatomata—namely, on the parietal and occipital bones. An external cephalhæmatoma may at times spread into the interior of the skull.

It is remarkable that cephalhæmatomata are found nearly always in vertex presentations where labour has been rapid and easy.[1] Only in a small number of cases are they met with in association with severe or slight injuries to the head, especially after forceps operations. A short time ago we met with a cephalhæmatoma on the occiput, after a very difficult delivery in a breech presentation, without any recognisable injuries to the bones. In the majority of cases they occur in primiparæ.

Their position is most frequently on the right parietal bone, corresponding to the greater frequency of a first-vertex presentation.[2]

Fritsch explains the occurrence of this condition in the following manner: 'During a pain a definite segment of the head enters the cervix; in the intervals between the pains, while the skull, freed from the pressure, retreats, the scalp is held fast by the closely applied circular ring of muscle fibre of the external os. In this way there occurs a recurring movement of the sub-periosteal tissues against the bones and, associated with this, a stretching and tearing of the vessels. The compression, which the maternal soft parts exercise before birth, is naturally wanting after delivery, so that an effusion of blood occurs between the skull and the periosteum. This displacement of the soft parts occurs more especially during the rotation of the occiput to the front.' Thus it is evident why in a vertex presentation the anterior parietal bone (in a breech presentation, the occipital bone) is the seat of the sub-periosteal bleeding. According to Küstner, it is small fractures or fissures of the bones which lead to these hæmorrhages.[3]

[1] This statement is, on the whole, somewhat too absolute. Thus there is no doubt that the condition is predisposed to by primiparity, the size of the child, a contracted pelvis, and malpresentation—all conditions which rather favour a prolongation of labour, or require some artificial interference. The older writers laid stress upon the supposed fact that these effusions are met with so frequently in easy labours, but that the labour is often—to some extent, at any rate—abnormal. Of sixty-eight cases observed by v. Winckel, in thirty-three the membranes ruptured prematurely; in ten, probably prematurely; and in only twenty-five at the right time; and the lives of 41 per cent. of the children were endangered during the labour. The fact remains, however, that a cephalhæmatoma may occur in an apparently perfectly normal delivery.

[2] Of ninety cases of cephalhæmatoma collected by Beck, in thirty-three the tumour occurred in the same position as the caput succedaneum.

[3] Féré has found, in two-thirds of all the cases he examined, fissures due to defects of ossification, chiefly at the upper posterior angle of the right parietal bone. It is obvious that such defects render the bones readily liable to injuries, and that in such injuries any blood-vessels running through these clefts will be easily torn. In every one of six cases of cephalhæmatoma, which Féré had an opportunity of examining, such a fissure or cleft existed.

Försterling holds that the rubbing of the skin and the periosteum against the bones can only occur when a parietal bone slips slowly past the promontory of the sacrum under considerable pressure. In this way the skin is held fast against the walls of the mother's pelvis while the bones continue to move on. By this mechanism Försterling explains a case from the Halle Clinic where two cephalhæmatomata were present at the same time. That besides this movement of the aponeurosis on the bones in certain circumstances, a condition of asphyxia, occurring during birth, forms an important ætiological factor in the development of the blood effusion, goes without saying, when we remember how markedly the veins are filled with blood in such cases. This venosity of the blood may also be the reason why it remains fluid in the cephalhæmatoma. Further, it cannot be denied that possibly an abnormal vulnerability of the vessels may favour the occurrence of the bleeding, while the occurrence of movement between the periosteum and the bones is favoured, according to some authors, by excessive hairiness of the scalp.[1]

In contracted pelves, cephalhæmatoma occurs infrequently, because the disproportion between the head and the pelvis renders the slipping backwards and forwards described extremely difficult. If tearing of the vessels does however occur, then the continued compression of the head leads to the formation of thrombi in the injured vessels, which prevent the occurrence of any bleeding after the birth of the child.

A cephalhæmatoma forms, clinically, a tumefacing tumour, varying in size from that of a nut to a fist, and, in contrast to a caput succedaneum, never oversteps the sutures and fontanelles as in this position the periosteum is firmly attached. It is met with about once in 250 deliveries. The effusion of blood is situated usually only on one bone, and, as has been pointed out, usually on the right parietal bone; but it may be present at the same time on several bones. In a series of 127 cases (collected by Henning), in fifty-seven the right parietal bone, in thirty-seven the left, in twenty-one both parietal bones, in seven the occipital bone, in three the frontal bone, and in two the temporal bones were the seat of the cephalhæmatoma. Usually the swelling appears some days after birth, when the caput succedaneum has in large part disappeared. In the majority of cases the tumour increases in size during the first days after birth, so that it appears more tense. Then there follows usually a period of quiescence, which passes over into the stage of retrogression. The effusion of blood occurs immediately after birth only in cephalhæmatomata, the result of injuries to the bones.

Healing requires a considerable time, and lasts from three to ten or

[1] In 25 per cent. of the cases observed in Pinard's Clinic, the hair of the child was 3 to 5 cm. long.

even fifteen weeks. After the tumour has been present for several days, in nearly all cases a well-marked and characteristic change can be found; namely, it becomes surrounded by a more or less definite bony wall, while at the same time the blood gradually undergoes absorption and the feeling of fluctuation gives way to a doughy sensation. After complete absorption, even after a long time, a slightly uneven periosteal formation of bone can be recognised (Keller). In large cephalhæmatomata with long delayed absorption, a thin, bony lamella forms over the whole extent of the separated periosteum, and this gives the so-called parchment-like crepitation.

The child's general health is not influenced by this affection when it runs its normal course, but in very extensive effusions of blood a marked degree of anæmia may be present. In a child with large bilateral effusions of blood on both parietal bones, we observed marked anæmia with resulting disturbance of the general health.

In a few cases, suppuration of the hæmatoma has been observed as a result of organisms finding their way in through superficial lesions of the skin, and leading to infection of the collection of blood. In such cases, eczema of the head is especially to be feared. The suppuration is generally confined to the collection of blood; but, in some instances, necrosis of the bones, meningitis, phlegmonous inflammation, and general septicæmia has occurred.

If cerebral symptoms are present with a cephalhæmatoma—such as stupor, irregularity of the pulse, slight cyanosis, shallow respirations, and inability to suck—the presence of a cephalhæmatoma interna or of a cerebral apoplexy should be suspected.

The diagnosis will present no difficulties if it is remembered that the tumour never passes beyond the sutures,[1] and if the peripheral bony wall is felt. Herniæ cerebri exhibit respiratory movements and frequently show pulsation, while they are partly reducible, with symptoms of cerebral pressure—namely, slowing of the pulse, vomiting, and interference with respiration. Simple abscesses are accompanied by fever, heat, redness of the skin, and great tenderness.

The prognosis is good when internal bleeding is not present at the same time, or suppuration with its sequelæ does not occur.

Treatment must, in the great majority of cases, be purely expectant. The skin over the cephalhæmatoma should be preserved from injury by covering it with wadding or some similar substance. Attempts may be made to hasten the absorption by the application of alum acetate solutions. In the majority of cases, puncture or incision of the sac is to be avoided, as, besides the danger of infection, there is the risk of

[1] In the few cases in which such a limitation is not present, either the hæmorrhage is really subaponeurotic, and not subperiosteal; or, as a result of some injury to the bone, the attachment of the periosteum to its edge has been torn through.

secondary hæmorrhage, which has been met with on several occasions (Runge). Some authors (Olshausen, Monti, and others) recommend that puncture, followed by aspiration of the blood, should be carried out in those cases where the cephalhæmatoma is very large and shows no tendency to absorption. They base this treatment on the supposition that the bone, separated from its periosteum, is cut off from its supply of nutriment and may in these circumstances become necrotic. After puncture,[1] they apply gentle compression with a bandage. In the case of a suppurating cephalhæmatoma, the sac should be widely opened and plugged with iodoform gauze. If a cephalhæmatoma interna is present at the same time with the symptoms already described, relief to the brain may be obtained by emptying the external tumour, as very frequently there is a communication between the two collections of blood.

On the skin of the skull or face, so-called pressure-marks or pressure-grooves are not infrequently found; as a rule, however, they are found only with contracted pelves, or with very large children, or, more rarely, after forceps operations. They consist of red spots and streaks, which are usually caused by the sacral promontory, and indicate clearly the part of the skull which has been in contact with this bone. These marks extend most frequently from the large fontanelle, or from the sagittal suture, along the coronal suture and then bend forwards towards the temple.[2] From these changes in the skin it is often possible, even several days after birth, to recognise the manner in which the head passed through the pelvis and the shape of the pelvis itself. In some cases two parallel marks, produced by a double promontory, can be seen passing from the sagittal suture towards the ear.

In rare cases, pressure-marks are also found due to the symphysis, especially when the pubic articulation is unduly prominent. These run usually in the long axis of the bones, and are situated above the squamous portions of the temporal bones. Finally, these skin-marks can also be caused by exostoses on the sacrum and by the horizontal ramus of the pubis.[3] All these pressure-marks occur most commonly in flat pelves, rarely in generally contracted or funnel-shaped pelves.

[1] The best method of doing this is to introduce a long narrow knife, with the edge uppermost, from the margin of the swelling into its centre; and through the incision thus made, the semi-fluid contents can usually be squeezed out.

[2] The change in direction is due to the head having engaged in the brim in the extended position, and then, before completely passing through, becoming more flexed. In some cases the mark runs at first towards the parietal eminence and then turns towards the temple, indicating that the head has been extended and squeezed in the direction of the forehead before the occiput descended.

[3] In the latter case the marks are usually on the anterior parietal or the frontal bone and are not opposite the mark due to the promontory.

In funnel-shaped pelves, the pressure marks are produced by the ischial spines. They take the form of light-red linear streaks, which extend on both sides from the parietal eminences in a straight line to the anterior or the lateral angle of the large fontanelle, or directly towards the outer canthus of the eye (Schauta). In the neighbourhood of the pressure-marks some observers have found coincident œdema of the eyelids (Litzmann, Küstner, and others). These pressure-marks and grooves are present, as a rule, only on the fore-coming head, as the after-coming head passes the contracted portions of the pelvis too quickly.

If a part of the scalp is subjected to long-continued pressure, the skin in this area may become gangrenous. A round slough with reddened edges is then formed, which separates after suppuration, and the resulting wound ultimately cicatrises. None of these injuries result in any damage to the child provided that the wounds do not become infected. In this case the formation of an abscess or of a phlegmon may be observed. The treatment consists, therefore, in the avoidance of any such infection, dressing with iodoform, the application of alum acetate fomentations, and the use of suitable ointments.

The changes in the skin which occur after the separation of amniotic bands (Simonart's ligaments) are of a different nature. They consist of localised defects of the skin or hair, without any necrosis or inflammation, which are either cicatrised, or are granulating, when they have occurred shortly before the birth of the child (see Congenital Diseases and Malformations of the Skin, Chap. XVII. p. 296).

In connection with this, I may mention a case described by Küstner, in which a bungler mistook the caput succedaneum for the bag of membranes and, as it did not rupture in the usual manner, incised it intra-genitally with a sharp knife. The epicranial aponeurosis was split sagittally over the whole skull and turned down over the ears, so that the child was born scalped. In spite of this severe injury, complete healing occurred. In other cases, death from sepsis or meningitis has been observed. Slight injuries of the scalp have also been noticed as the result of rough examinations.

Injuries to the bones of the skull in the new-born child are of special importance, and under this heading we have to consider fractures, fissures, and depressions.

The most frequent of these are gutter- or spoon-shaped depressions of the skull-bones. They are found, in cases of contracted pelves, almost always on the posterior parietal bone, which is pressed against the promontory of the sacrum. Most commonly they occur spontaneously; but they may also occur when the fore-coming or after-coming head, lying movable above the brim or wedged into the pelvic inlet, is delivered with forceps or by pressure from above. They may also be observed, in delivery by the breech, in contracted pelves. In the gutter-like

depressions, most commonly a simple bending-in of the bone is present—seldom a fracture. They are situated most frequently on the edge of the parietal bone adjoining the coronal suture. The spoon-like or funnel-shaped depressions of the skull-bones are much more dangerous to the life of the child. They are found on the frontal or parietal bones between the parietal eminences and the large fontanelle or the coronal suture, and form deep depressions of the bones. At their deepest portion, a true cephalhæmatoma is not uncommonly found; while, at the periphery of such depressions, fissures of the bones often occur.[1] Such fissures, like the very rare cases of tearing of the sutures, have given rise in some cases to traumatic meningoceles or encephaloceles. Depressions on the frontal bones are uncommon, and occur most frequently when the forceps, improperly applied, have dragged the frontal bone forcibly past the sacral promontory. On the parietal bones, they are usually the result of the uterine contractions alone. In a very few cases (Lomer, Hoffmann, and Rembold), such indentations have been met with on the temporal portion of the orbits, associated with a flat pelvis and marked descent of the sinciput. As a result of the pressure on the bone, in several cases exophthalmus has occurred. Among the cases collected by Lomer is that of a woman who, on two occasions, had given birth to a child with such an injury. Lomer explains the injury in the following manner: namely, that in marked transverse compression of the skull the bones are displaced over one another at the sutures, and the very thin bones of the orbit, not being able to follow this movement, become fractured. Rembold also met with this lesion twice in the same woman, who had a funnel-shaped pelvis.[2]

All these injuries may constitute a grave source of danger to the child's life when the bones are fractured, and especially when a comminuted fracture is present. In such cases, tearing of the middle meningeal artery, of the longitudinal sinus, and bursting of the sutures can occur—more particularly of the squamous suture with tearing of the transverse sinus. As a general rule, however, one may say that even marked deformities of the cranial vault can be recovered from quite readily. Further than this, the later mental development does not appear to be affected (Keller). In later years, however, the flattening of the skull-bones can often be definitely recognised.

Of sixty-five cases collected from the literature (Olshausen and Veit), with spoon-shaped depressions, twenty-two children, or 34 per cent., were born dead or dying; ten, or 15·4 per cent., died soon after birth

[1] The presence or absence of fractures no doubt depends upon the degree of ossification present.

[2] Franke has recorded a case in which a woman gave birth to five children, all with the same injury to the skull.

as a result of the injury; and thirty-three, or 30·8 per cent., remained alive without—with few exceptions—their later state of health being in any way affected.

It has been recommended to treat the deeper depressions by operative measures—a recommendation which could only be justifiable if signs of damage to, or of pressure on, the brain were present. The prognosis of such operations in new-born children is bad, since surgical measures are badly borne by them. The proposal to draw out deep depressions with an air-pump would probably encounter great difficulties, especially of a technical character. According to Munro Kerr, depressions can sometimes be got rid of, with a very audible click, by pressure applied to the skull in a diameter at right angles to that compressed. Bumm suggests drawing out the depressed portion by traction exerted with a fine corkscrew which has been bored through the bone.[1] Boissard introduces a sound from the suture, between the bone and the dura mater, so as to lever up the depressed portion and so avoid a trephining.[2] Bursting of the squamous suture, already shortly alluded to, and separation of the temporal and parietal bones, may occur in the fore-coming head as the result of traction with the forceps; but it occurs most frequently when the after-coming head in pelvic presentations is dragged with considerable force through a contracted pelvic inlet. This injury is fatal if the transverse sinus[3] placed beneath the bones is injured, and haemorrhage is set up at the base of the skull. The mechanism of this dangerous injury is readily understood when we remember that the broad upper part of the skull is firmly attached to the narrower lower portion, and that the greatest force is exerted upon the squamous suture, which is the least able to stand it.[4]

Even more dangerous for the child is the separation of the epiphysis of the occipital bone (first described by Schröder) and the separation of the tabular portions of the occipital bone from the condylar portions. This occurs, in the majority of cases, during the extraction of the after-coming head; and, as a result of the lateral compression, the condylar portions are loosened from the tabular portion of the bone and also displaced on them.

Fritsch attributes the injury to a forcible pull on the pelvis of the child, either alone, or occurring in conjunction with marked displacement

[1] Hastings Tweedy uses the point of a vulsellum for the same purpose.

[2] In a considerable number of instances such cases have been treated successfully by operation: an incision being made along the margin of the depressed area of bone, the latter cut through, and then elevated with a flat elevator.

[3] Or basilar sinus.

[4] When the sutures are very lax, and the bones in consequence very movable, haemorrhage may readily occur from the pressure exerted by the lower anterior angle of the parietal bone on the great anastomatic vein, and for this reason Herbert Spencer regards this region as the most vulnerable part of the child's head.

of the tabular portion of the occipital bone under the parietal bones, which may occur in marked narrowing of the pelvis in its transverse diameter. As a result of this injury, tearing of the transverse sinus with fatal hæmorrhage into the interior of the skull may result, or direct compression of the medulla from the displacement forwards of the tabular portion of the occipital bone, so that the cervical cord may be regularly guillotined, as Olshausen points out.

Besides these typical injuries of the skull, a large number of atypical injuries have also been observed. For example, in difficult forceps extractions, fractures of the skull may occur generally in the form of a so-called star fracture[1] as a result of too marked compression of the head by the forceps, when it has been applied to the head in an antero-posterior diameter; and, in the same way, marked injuries may occur to the occipital and frontal bones. In difficult extractions in pelvic presentations, multiple transverse fractures of the tabular portions of the occipital bone are more frequently met with.

The great danger of these injuries to the skull lies in the coincident injury to the substance of the brain, or injury to a vessel with resulting bleeding into the cavity of the skull.[2] As a general rule, however, hæmorrhages on the convexity of the brain — as I shall explain later on — provided that they are not very extensive, are usually readily recovered from. They are, however, always fatal when a large hæmorrhage occurs at the base of the brain. Rupture of the sutures leads almost always to injuries of the sinuses. Besides the already-mentioned rupture of the squamous suture with damage to the transverse sinus, tearing of the frontal and sagittal sutures with damage to the longitudinal sinus has been observed on many occasions.

Hæmorrhages into the interior of the skull are not only observed after injuries of the bones of the skull, but also after spontaneous, easy, and rapid deliveries.[3]

This is especially the case in injuries of the sinuses (Olshausen). The cause of these injuries is to be found in the fact that, in long-continued difficult deliveries, the skull-bones undergo a very marked displacement upon one another, which occurs especially in contracted pelves, but

[1] Such star-shaped fractures usually start from the parietal eminence or from the frontal bone, and may be very extensive in cases where the forceps has been applied and has repeatedly slipped off.

[2] Spencer found hæmorrhage into the pia mater or arachnoid in fifty-three, or 40·7 per cent., of 130 still-born children examined. Of twelve children delivered by forceps and still-born, cerebral hæmorrhage was present in every one. In 143 post-mortem examinations of new-born children, Wallich found meningeal and cerebral, or cerebral, hæmorrhage in fifty-eight.

[3] Küstner has described a case of cerebral hæmorrhage in a child delivered by Cæsarean section.

which may also occur even in normal conditions. As this displacement primarily affects the frontal and parietal bones, so injury to the vena longitudinalis, and to the veins passing from the upper surface of the brain to the sinus, occurs most frequently. Intra-cranial hæmorrhage may further occur at the base of the brain, into the internal meninges, into the cerebral ventricles, into the brain substance itself, or into the space between the skull and the dura mater. Keller rightly remarks that a coincident passive hyperæmia in the interior of the skull may form a contributory factor in the production of intra-cranial hæmorrhage. Such intra-cranial congestion occurs most frequently as the result of asphyxia, or when the veins of the neck are subjected to mechanical compression: for example, in congenital goitre or in cases where the cord is wound round the neck. Kustner and others have observed intra-cranial hæmorrhages in many such cases. That premature children are especially predisposed to this condition goes without saying, as their soft skull-bones are readily compressible and their vessels less resistant; and this applies also to syphilitic children.

If the hæmorrhage is slight, and situated on the convexity of the brain, it may occur without any symptoms. If marked hæmorrhages are present, collapse, slowing of the pulse, somnolence, shallow respiration, tremors of the extremities, sudden crying out, numbness of the limbs, and inability to suck may be noticed. With such symptoms, the child may die soon after birth. If the child is born asphyxiated, it will be noticed that the asphyxia cannot be treated successfully.

If an intra-cranial hæmorrhage is suspected, Schultze's swinging method should not be made use of; but an endeavour must be made to resuscitate the child by the use of other methods. The diagnosis, of intra-cranial hæmorrhage from pure asphyxia is made by the absence of any injuries to the skull in the latter condition, the presence of marked bulging of the fontanelles in cases of hæmorrhage; and it can, in certain conditions, be made by lumbar puncture. In slight hæmorrhages, especially on the surface of the brain, the symptoms may entirely disappear and the child completely recover.

In other cases (from pressure on certain cerebral centres), hemiplegia, spasms, and contractures, may occur. If a basal hæmorrhage has not ended fatally—as most of them do—paralysis of the oculo-motor nerves, of the trochlear, the abducter, or the facial nerve, may remain.

In this place, mention must also be made of the spinal hæmorrhages met with in rare cases.[1] They occur, as a rule, as the result of severe

[1] Litzmann met with spinal meningeal hæmorrhage in thirty-three of eighty-one necropsies on new-born children; and among Spencer's cases, in forty-four the spinal cord was examined, and in thirty of these there were hæmorrhages present; in twenty-one, outside the theca. His cases show that spinal hæmorrhage is greatly favoured by presentation of the lower extremity.

asphyxia: but they are very frequently of traumatic origin, especially in cases of breech presentation which have been delivered artificially. In such cases, the hæmorrhages may take place not only into the spinal cavity, but also, in rare cases, into the substance of the cord. Stumpf points out that spinal hæmorrhages may occur under the same conditions as cerebral; as in compression of the head the cerebro-spinal fluid may be driven into the spinal canal, and so lead to injuries in this situation. If at the same time other severe injuries—for instance, to the medulla—are not present, the prognosis is not unfavourable. Opisthotonus, contracture of the extremities, hemiparesis and paresis, and spasms, are given as symptoms of spinal bleeding. Latterly, neurologists have made such hæmorrhages responsible for the later development of certain diseases of the cord—such as syringomyelia.

Traumatic injuries to the skull and brain are frequently stated by neurologists to be ætiological factors in the development of diseases of the brain and nerves in after years. In this case, the injury either acts directly through a lesion of the brain or indirectly: for example, as the result of effusions of blood with their sequelæ. This holds good especially for the spastic forms of cerebral paralysis (congenital spastic paraplegia, Little),[1] and also for epilepsy, chorea, idiocy, athetosis, and deafness.

That such a relationship may exist in certain conditions, appears to be established from a number of separate observations. At the same time it may be affirmed that the importance of hæmorrhage as a cause of various cerebral palsies of children has been overestimated (Finkelstein).

The solution of these problems fails, as a rule, on the one hand, because of the failure in continuity of the observations (obstetrician and neurologist), and, on the other hand, from the uncertainty of the diagnosis: for example, between hæmorrhage and congenital disease of the brain. Such cerebral and nerve affections are seen also when intra-cranial hæmorrhages and direct injuries to the brain can be excluded. As possible ætiological factors, long and difficult deliveries—especially with forceps—asphyxia, premature, and precipitate delivery may all play a part. Schultze observed, in two children, late idiocy where severe asphyxia was present at birth. It must be admitted, indeed, that asphyxia alone cannot produce such conditions, but rather that the sequelæ of this condition—and especially intra-cranial hæmorrhage—play an important rôle.

[1] The greater frequency of paralysis of the lower extremities is explained by the fact that the largest of the cerebral veins open into the superior longitudinal sinus in the region of the central convolutions, and thus are especially liable to damage when they are passing over the motor centres for the lower limbs.

It cannot be denied that all the conditions already mentioned may act as factors in the causation of cerebral and nerve disease; but, for the reasons already put forward, it is not possible to make any certain statements.[1] To explain the cases where the mere trauma of a normal delivery is thought to exercise a harmful influence, one must either assume that pre-natal diseases of the brain are not infrequently present; or that there exists a predisposition of the affected individual, in the form of a ready vulnerability of the brain. Finally, an hereditary taint is undoubtedly of considerable importance—such as neurosis, alcoholism, tubercle, syphilis, and venereal disease (Finkelstein; see also the explanations of Stumpf).

Unilateral facial paralysis occurs not uncommonly as the result of forceps extraction. The paralysis is caused usually in this way: namely—the forceps seizes the head in a transverse diameter, so that the end of the blade exerts a pressure on the trunk of the facial nerve, at its exit from the stylo-mastoid foramen. At the same time a pressure-mark on the skin is often present in this region—that is, on the skin over the margin of the lower jaw. In rare cases the pressure of the forceps on the pes anserinus major, lying over the masseter muscle, leads to incomplete facial paralysis. Facial paralysis occurs the more readily, the more the handles of the forceps are pressed together during extraction—and especially when the hand is removed from the lock and applied to the handles, during the later stages of the operation. It is, therefore, a well-known fact that such paralyses occur more often, the less dexterity the accoucheur in question possesses. Küstner remarks that facial paralysis occurs more easily with forceps in which the distance between the ends of the blades is very slight, and in which the blades are well curved and look directly towards one another at their extremities, than with forceps in which there is a considerable distance between the ends and which have only a slightly marked cephalic curve.

Peripheral paralysis of the facial nerve occurs also in cases of necrosis of the scalp resulting from labour with a contracted pelvis, when this is situated in the neighbourhood of the stylo-mastoid foramen, the peripheral portion of the nerve becoming involved in the inflammatory swelling (Knappe). Vogel has recorded two cases of facial paralysis where, with an exostosis of the symphysis and an anterior parietal presentation, a direct pressure was exerted on the trunk of the nerve at its exit from the skull, in consequence of which paralysis occurred.

[1] According to Shuttleworth, protracted pressure during parturition is a frequent cause of mental impairment. In no less than 27·25 per cent. of the cases collected by Beach, there was a history of protracted parturition, forceps delivery having been effected in 3·98 per cent. only.

Franke has observed facial paralysis in an anterior parietal presentation with a markedly pendulous abdomen, and explains the paralysis by the pressure of the shoulders against the region of the ear (Keller). In the Göttingen Clinic, we observed, a short time ago, paralysis of the facial nerve in a case of a generally contracted flattened pelvis, which occurred in this way: namely, the pressure was exerted upon the nerve by a projecting symphyseal cartilage; the head, in a position of marked anterior parietal obliquity, lying with the small fontanelle deep in the pelvis. Delivery occurred spontaneously, with strong labour pains.

B. Schultze has recorded a case of facial paralysis from a hæmatoma of the sterno-cleido-mastoid muscle at its insertion into the skull. Olshausen explains some cases in this way: namely, that amniotic bands, adherent to the face, have been the cause of the paralysis.[1] It is very remarkable how seldom this paralysis occurs after spontaneous and easy delivery. In one such case, diphtheria of the nose and throat (acquired intra-partum) was found. In another case (observed by Olshausen), no definite cause could be discovered. Central paralysis of the facial nerve has also been observed: for example, when depressions of the bones, or fractures with intra-cranial hæmorrhages have produced pressure on the facial nerve nuclei. As the centre of movements of the arm and leg, and also that for the hypoglossal, lie in the neighbourhood, these may, in certain circumstances, also be affected.

When a child with facial paralysis is observed, it will be seen that the eye on the paralysed side cannot be completely closed (Lagophthalmus). Moreover, the paralysis of the facial muscles of expression will at once be noticed. The paralysed half of the face is lax and without expression: the lines on the forehead and the naso-labial folds are obliterated; the angle of the mouth hangs down, and saliva frequently dribbles from it. All these signs are especially well seen when the child cries. The nutrition of the child also suffers; sucking is either impossible or incomplete—even if the child seizes the nipple with the angle of the mouth on the sound side. In drinking, milk passes very readily into the nose as the result of the simultaneous paralysis of the soft palate. The differential diagnosis as to whether the lesion is of peripheral or central origin must be made by the well-known scheme of Erb (see Strümpell).[2]

[1] As Ballantyne points out, cases of ante-natal origin of this kind are generally more severe than those occurring during birth, and less readily recovered from.

[2] The central form may be distinguished from the peripheral by the persistence of the normal electrical excitability of both the nerves and the muscles, and the frequent absence of involvement of the upper branches of the nerve so that the orbicularis palpebrarum and frontalis muscles escape, while the emotional movements of the mouth are also as a rule retained.

I may point out, however, that the condition of the superficial petrosal nerve, and of the palate supplied by it, is of considerable importance.

The prognosis [1] of a simple case of peripheral paralysis is almost always good, the majority of the cases recovering in three to six days without any treatment. Olshausen has never seen a case of persistent facial paralysis. Henoch has, however, described two cases in which it did persist, and Seeligmüller has also seen this. In such a case, an irreparable and severe crushing of the nerve must have been present.

Treatment of this variety of paralysis is almost superfluous as cure takes place so quickly. Still, in cases in which it only slowly improves, the methodical application of the faradaic current to the nerve should be carried out.

Literature

Kaufmann, *loc. cit.*, p. 24; Schmaus, *loc. cit.*, p. 15; Ahlfeld, *loc. cit.*, pp. 24 and 34; Kleinhans, *loc. cit.*, p. 34; Küstner, *loc. cit.*, p. 15; *Text-books of Midwifery*, by Runge, Fritsch, Olshausen and Veit, Bumm, Spiegelberg, and v. Winckel; Schultze, *Nothnagels Handb. d. spez. Path. u. Ther.*; Tillmanns and Leser, *loc. cit.*, p. 34; Strümpell, *loc. cit.*, p. 34; Quincke, *Volkmanns klin. Vortrage* (new series), No. 67, and *Über Hydrokephalus*, *Verhandl. d. 10. Kongr. f. innere Medizin* (1891); Keller, *loc. cit.*, p. 34; Hohl, *loc. cit.*, p. 15; v. Bardeleben, *Geb.-gynäk. Gesellsch. Berlin* (November 27, 1903), *Zentralbl. f. Gynäk.* (1904), pp. 110 and 111; Schuchard, *Diss. Berlin* (1884); Cohnstein, *Arch. f. Gynäk.*, vol. vi., p. 503; Ahlfeld, *Berichte und Arbeiten.*, vol. i., p. 240; G. Veit, quoted by Küstner, p. 670; Hammerschlag, *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxvii., p. 415; Pacel, *Diss. Paris* (1907); Gabail, *Diss. Paris* (1902); Weinberg, *Diss. Bonn* (1902); Liese, *Geb. Gesellsch. München* (Oct. 21, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 879; Roorda-Smit, *Ref. Zentralbl. f. Gynäk.*, p. 1563; Jorns, *Diss. Berlin* (1903); Ballantyne, *Ref. Zentralbl. f. Gynäk.* (1905), p. 764; Flammand, *Diss. Paris* (1904); Jammes, *Diss. Lyon* (1905); Keilmann, *Deutsche med. Wochenschr.* (1895), No. 28, *Vereinsbeilage*, No. 46, p. 194; Stratz, *Geb. Gesellsch. Berlin* (March 1885); Anton, *Wien. Med. Jahrb.* (1888); Czerny, *Zentralbl. f. Path.*, p. 100 (1899); Cushing and Harvey, *Keen's Surgery*, vol. iii., p. 118 (1911); Hoffmann, *Inaug. Diss. Breslau* (1902); Barbour, *Anatomy of Labour*, p. 192 (1899); Beck, *Diss. Inaug., München* (1904); Munro Kerr, *Operative Midwifery*, p. 672 (1911); Spencer, *Trans. Obstet. Soc. London*, vol. xxxiii., p. 269 (1892); Seitz, *loc. cit.*, p. 15, vol. iii., pt. 3, pp. 73, 82, 85; Küstner, *loc. cit.*, p. 15, vol. iii., pt. 1, p. 497; Little, *Trans. Obstet. Soc. London*, vol. i., p. 293 (1862); Beach, *Clifford Allbutt's Syst. of Med.*, vol. viii., p. 876 (1910); Féré, *Revue Mens. de Méd. et de Chir.*, vol. iv., p. 112 (1880).

[1] According to Erb, if there is no change in the faradaic and galvanic reactions, the prognosis is good, and recovery usually takes place in fourteen to twenty days. If the faradaic and galvanic excitability of the nerve is lessened, while that of the muscle is increased to the galvanic current, recovery will take place in about four to six or eight weeks; while, if the reaction of degeneration is present, the prognosis is bad.

CHAPTER VI

Malformations and Congenital Diseases of the Face

Malformations of the face occur either alone or combined with other malformations—for example, hydrocephalus, fissures in the thoracic and abdominal regions, conjoined twins. They originate in the first few weeks of embryonic life as the result of faulty development of the face. In order that the reader may understand the various deformities more readily, I will give a brief account of the development of this region; and in doing so, have followed the very clear description of the subject given by Leser. In the embryo, the face and the neck are formed in the following manner: on either side symmetrical processes (the so-called visceral arches), which lie between the visceral or branchial clefts, grow forwards and unite with one another in the middle line. The neck develops from the three lowest visceral arches which unite with one another anteriorly. In the development of the upper jaw and the soft parts covering it—the cheeks and the tissues of the upper lips—the process takes place rather differently, as the horizontal processes growing towards one another—the so-called upper-jaw processes[1]—unite only, like the upper branchial processes of the neck, at the back in the soft palate and the palate bones,[2] while in front a vertical process growing down from the anterior end of the skull inserts itself between them.[3]

This frontal process is divided by two clefts (the nasal grooves) into three portions—a median and two lateral.

From the median portion is formed later the philtrum of the upper lip, the intermaxillary bones, and the vomer; from the lateral portions the lateral part of the upper lip and cheek on either side, with the corresponding underlying bones—the lateral segments of the upper jaw. From the first branchial arch[4] develops the lower jaw and the

[1] The maxillary processes.

[2] A horizontal plate grows inwards from the maxillary process and fuses with the plate of the opposite side to form the hard palate; while the soft palate is formed by a fold arising as a prolongation backwards of each horizontal plate into the pharynx.

[3] The fronto-nasal process.

[4] The mandibular process.

aperture of the mouth. By the union of these parts with one another, and in the median line, the anterior portion of the face is formed. If the union does not take place, abnormal fissures result. One of the nasal grooves most frequently remains open; or the fissure is situated in the region where, normally, the philtrum of the upper lip, the intermaxillary bone, and the vomer on the one hand, and the lateral portion of the upper lip and cheek, the upper jaw and gums, on the other hand, should unite with one another. In this way, lateral hare-lip and clefts of the palate occur. If the two upper-jaw processes do not unite posteriorly, then a medianly placed cleft of the soft palate occurs;[1] as, normally, these processes should unite in the middle line, as the lower branchial arches do. If the union between the process of the upper jaw and the lateral part of the frontal process does not occur, a cleft remains between the corresponding parts: namely, between the lateral part of the cheek and the outer part of the lip. This cleft reaches from the angle of the mouth to the inner angle of the eye, and may extend even further still—over the frontal and temporal region to the hair—and is known as an oblique fissure of the face.[2]

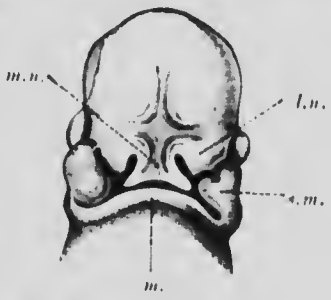


FIG. 24.—*m.n.*, Mesial nasal process; *l.n.*, lateral nasal process; *s.m.*, superior maxillary process; *m.*, manibular process.

The causes of these fissures are not well understood. The malformation is either primary and hereditary, or secondary and caused by adhesions of the amnion, or by the interposition of portions of tissue within the fœtal clefts. In other cases, the presence of an abnormal pressure within the fœtal skull may be recognised—for instance, in hydrocephalus and encephalocele. The effect of heredity[3] is said to be transmitted mainly through the mother. Of late years, there is more and more a tendency to attribute these fissure formations to anomalies of the amnion (see p. 11).

For the obstetrician and practitioner, the most interesting malformation is hare-lip. It is also the most frequent of these malformations. In the ordinary case there is a cleft in the upper lip; while

[1] And in this case, of the hard palate as well.

[2] The maxillary process fuses with the lateral nasal process, so as to form the outer wall of the nasal cavity, and with the globular portion of the mesial nasal process which forms the premaxilla and the median portion of the upper lip. If these processes fail to unite, a fronto-maxillary or oblique fissure of the face results.

[3] Ballantyne quotes Meckel's reference to Anna's observation of a man who had by his first wife eleven children, nine of whom were born dead, and two alive, with hare-lip; and by his second wife four children, two of whom had the same deformity, and one had a cleft palate.

in slighter cases there is only a shallow notch. A fissure of the alveolar border, and of the hard and soft palate, very frequently is present at the same time: malformations which are distinguished as clefts of the palate (cheilo-gnatho-palatoschisis). The fissures of the lips, and also of the bones, are almost always median; the fissures of the soft palate and of the uvula are always median.[1]

The hare-lip is usually unilateral, rarely bilateral, and most commonly it is situated on the left side. Ahlfeld gives as a reason for this

FIG. 25.—BILATERAL HARE-LIP WITH CLEFT PALATE. (Specimen from the collection of the Göttingen Women's Clinic).

that the position of the child on its left side during development causes a malformation of the left side of the face. According to E. Müller (quoted by Tillmanns), this malformation occurs more frequently in boys. The cleft in the alveolar borders and the hard palate may be bilateral,[2] while the cleft in the soft palate is always

[1] This statement is hardly correct: the fissure is usually placed extra-medianly, and is situated between the two parts of the premaxilla. It was formerly thought that it was placed between the premaxilla and the maxilla, but this is not so; and its position in the inter-incisor suture probably indicates that the premaxilla is formed from two ossific nuclei on each side. Thus the two incisor teeth are commonly placed one on either side of the fissure. A median fissure of both the upper lip and of the nose may occur, but this is a very rare abnormality, and not to be confused with the much commoner hare-lip.

[2] In this case there is a wide cleft, divided more or less into two parts by the lower border of the vomer.

single. The cleft in the palate may also occur by itself without any cleft in the lips or in the alveoli.

In bilateral complete clefts of the hard palate, a very striking deformity of the affected child may, in some cases, be present; since the vomer, as a result of excessive growth, may push forward the intermaxillary portions and the philtrum, so that they lie in a trunk-like form in front of the tip of the nose. In such a malformation the plastic operation necessary is rendered very difficult. In other cases the intermaxillary bone lies on the same level as the alveolar arches. In some cases, children have been born with a hare-lip already healed *in utero*; and in such cases a unilateral—generally, pigmented—scar can be seen[1] (Ahlfeld).

The malformations described, rarely ever claim an obstetric interest. As a rule, difficulties in diagnosis will only occur when children, with such malformations, present by the face during delivery. Well-marked clefts may then conceal the exact condition of things, or give rise to false diagnoses.

The result of these malformations is, primarily, disturbance of nutrition by rendering sucking difficult or even impossible, especially when a cleft of the hard palate is present at the same time. These difficulties are easily understood if we remember that sucking can only take place when the lips are hermetically sealed on the nipples. Only then, by the drawing down of the lower jaw, is the rarefaction of the air in the cavity of the mouth—necessary for the drawing in of the milk—rendered possible. Suckling, however, will often succeed, provided that only the upper lip is cleft, as the child seizes the nipple with the margin of the upper jaw. If a cleft of the palate is also present, sucking can only be possible when the cleft is very small, the nipple long and broad, the child healthy, and, lastly, the quantity of milk abundant in the mother or the wet nurse. The large majority of children with hare-lip, and especially with clefts of the palate, must be fed with a spoon. For this purpose beaked cups and so-called boats—that is, small porcelain vessels in the shape of a boat with a beak—are very useful for the administration of the fluid. In a few cases, especially if sufficient patience is exercised, a bottle will answer satisfactorily. In this case the hole in the rubber teat must not be too small, while the teat itself must go far enough into the mouth without exciting nausea (Keller). In spite of every care, the children die very frequently—almost always, indeed, in polyclinic practice—within the first few weeks of life, especially when the clefts are very extensive.

The causes of these bad results are evident. Even when the head is held high up during the spoon-feeding, the milk cannot be prevented from

[1] As Ballantyne points out, it is not clear that this is the real explanation of the anomaly, as no cicatricial tissue has been found.

passing into the nasal cavities and giving rise to a condition of chronic catarrh; and, as a result of this, aphthæ and thrush very frequently occur. The decomposed milk then passes with the catarrhal secretion into the stomach, and in this way severe disturbances of the stomach and intestines arise, from the effects of which children with hare-lip or cleft palate frequently succumb; while, by the aspiration of the secretions into the air passages and, further, into the lungs, an opportunity is given for the development of severe pulmonary affections. These diseases of the respiratory organs are also favoured by the fact that the air enters unwarmed and unfiltered. For the prevention of putrefactive processes in the buccal cavity, Keller recommends, as a prophylactic measure, the frequent use of a mouth-wash of weak boric acid solution. If the children survive these frequent complications, the later results (besides the external disfigurement) are indistinctness of speech, especially in cleft palate; and misplacement of the teeth, when the child has not been operated upon before the eruption of the first teeth.

The cure of this affection can only be obtained by operative procedures. The doctor is very soon asked by the parents when the operation should be undertaken. Almost all surgeons recommend that it should be done in the first few months of life—at any rate, before the eruption of the teeth. This advice applies especially to cases of simple hare-lip. In cases of cleft palate the operation does not so certainly succeed in the early months. In this case one should wait until the fifth, sixth, or seventh year of life, as the children only then possess sufficient intelligence to undergo the operation (Tillmanns, Leser, and others). Julius Wolff has, however, operated upon such cases, even in the first year of life, with good results.[1] The advantage of this early operation lies in the better development of the speech. Some authorities operate upon hare-lip soon after birth; others wait until the fifth or seventh week, or even until the fourth to the seventh month. The great advantage of an early successful operation lies in the fact that the affections of the stomach, intestine, and lungs—so dangerous to the child—are entirely prevented. A condition necessary for the success of the operation is the previous cure of any inflammatory affections of the mucous membrane of the mouth and nose (e.g. thrush and aphthæ). Severe catarrh of the intestine and any affection of the lungs must

[1] Arbuthnot Lane (among English surgeons) strongly advocates early operation. Of 399 cases operated upon by him, 144 were under one year of age; and of these, eighteen died—or a mortality of 12·5 per cent. It must, however, be remembered that the normal mortality of infants under one year is at least 10·9 per cent. The question of early or late operation is one upon which there is a considerable difference of opinion.

PLATE II.

A FŒTUS WITH NON-DEVELOPMENT OF THE LOWER JAW (AGNATHIA); A VERY SMALL MOUTH (MICROGLOSSIA); AND DISPLACEMENT OF THE EARS DOWNWARDS ON TO THE NECK (PARTIAL SYNOTIA).

(Specimen No. 833a, University College Hospital Medical School Museum. Obstet. Sect. From a case under the care of Dr. L. O. Fuller.)

naturally be first improved by treatment. Before the operation, the child should be kept awake for several hours and well fed, so that it may sleep a long time after the operation, and not cry from hunger. Fissures of the face have also been operated upon with success on several occasions. With reference to the technique of the operations, the reader should consult text-books on surgery.

In very rare cases, a fissure of the lower lip has also been observed, resulting from faulty union of the processes of the inferior maxillæ (Ahlfeld).[1]

To this chapter belong also fistulæ and cysts of the lower lip. These are due to malformations in the region of the first branchial arch. The fistulæ are situated, generally symmetrically, on both sides of the middle line, and end in the red portions of the lower lip. The openings, which are surrounded by muscle fibres, secrete drop by drop a clear light fluid. Cysts of the lower lip occur when the openings of these fistulæ become closed.

If the development of the processes of the lower jaw, derived from the first visceral arch, is defective then incomplete formation or complete absence of the lower jaw occurs. This is very frequently the case in cyclopia and acrania. These malformations are termed brachygnathia and agnathia.[2] In such a case the ears may almost touch one another (synotia), and the cavity of the mouth is also very incompletely developed (see Plate II.). The oral opening of the trachea and œsophagus may be open or closed (Ahlfeld). Von Winckel attributes these malformations to anomalies of the amnion.[3] Von Klein has described briefly a case of synotia and agnathia. Induction of premature labour was performed on account of hydramnios, cystitis, and nephritis in a woman twenty-four years old who had had two children. Spontaneous delivery took

[1] In specimen No. 843, Univ. Coll. Hosp. School Museum—that of a kitten showing duplicitas posterior or dipygus—there is a double hare-lip, and the lower jaw and tongue have failed to unite in the middle line.

[2] There are three specimens of agnathia in the Univ. Coll. Hosp. Med. School Museum (Nos. 832, 833, and 833a). The first—a female fœtus—shows transposition of the viscera, a very small mouth and tongue, and the lobules of the ears meet in the neck anteriorly. The second fœtus shows also transposition of the viscera, with maldevelopment of the lower jaw and a small tongue. In this case there was difficulty in feeding, owing to the inability to suck, and choking from swallowing the tongue. In the third case (see Plate II.) there is well-marked agnathia and a small tongue, and the external ears are displaced downwards and forwards on to the neck (Synotia). It is interesting to note that there was transposition of the viscera in three of the sixteen cases collected by Taruffi.

It is a curious fact that agnathia is especially common in lambs.

The association of agnathia and cyclopia is so common that Taruffi places these cases in a separate class which he terms cyclops hypo-agnathus.

[3] He maintains that the mandible is usually present in a rudimentary form, and regards the condition as one of arrested development—the result probably of amniotic pressure.

place of a seven-months old male fœtus with the above-mentioned malformations. For epignathus, *see* p. 829.

Literature

Kaufmann, *loc. cit.* p. 24; Ziegler, *loc. cit.*, p. 15; Lasar, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, pp. 24 and 34; Biedert and Fischl, *loc. cit.*, p. 34; Biedert, *Kinderernährung* (Stuttgart 1900); Keller, *loc. cit.*, p. 34; Klein, *Gesellsch. der Aerzte Berlin* (May 22, 1908). *Zentralbl. f. Gynäk.* (1909), p. 27; Keith, A., *Human Embryology and Morphology*, p. 8; Arbuthnot Lane, *Proc. Roy. Soc. Med. London* (1911), vol. iv., No. 8, p. 105; Spencer and Lawrence, *Catalogue Univ. Coll. Hosp. Med. School Museum* (1911).

Macrostomia Congenital Buccal Fissure

Enlargement of the opening of the mouth is found most frequently with an oblique fissure of the face, and then takes the form of a cleft of the cheek.[1] It occurs as a result of the incomplete horizontal union of the processes of the upper jaw with the first visceral arch. In this condition, as in hare-lip, in certain circumstances several disturbances of nutrition may occur. The cure of such deformities must be carried out on surgical lines.

Microstomia

This condition is one of too small an opening of the mouth, as a result of the too extensive union of the processes of the upper jaw with the first visceral arch. At the same time, the lower jaw is usually remarkably small. This condition is extremely rare. In some instances the lips are completely united, and in such cases immediate operative treatment naturally is necessary. Congenital microstomia has nothing to do with similar conditions of the mouth developing at a later date —the result of syphilis and other ulcerative processes.

Milium Epithelial Pearls on the Hard Palate

In some 95 per cent. of new-born children, several small tubercles of a yellowish white colour, are found on the posterior part of the hard palate on either side of the middle line and are frequently mistaken by the inexperienced for thrush. These little tubercles were thought formerly to be similar to the comedones of the skin. Epstein was the first to show that they consisted of congenital depressions in the mucous membrane filled with epithelial masses. They are also found very frequently in the skin of the forehead and nose, and in these positions have the same origin. According

[1] Also known as the inter-maxillary or commissural fissure of the face.

to Bednar (quoted by Tillmanns), the appearance is, as if a bordeolum had been introduced under the epithelium. The thickenings are absorbed in the course of the first month. As a general rule the development of these epithelial pearls is harmless and of no importance. In a few cases especially when the cleansing of the mouth is inefficient small ulcers,[1] described by some authors as Bednar's aphthæ, develop on the floor of these epithelial pearls. They may render sucking very difficult, and are therefore especially prejudicial in a weakly and premature child, or in twins, as the thrush fungus is very prone to develop on their base.

Prophylaxis consists in very careful cleansing of the mouth, although this is best omitted, in our experience, in the case of new-born children when the mucous membrane is healthy. As a therapeutic measure, the touching of the ulcerated part with a stick of lapis mitigatus[2] may be employed. If sucking is rendered impossible by the condition, the children must be fed by hand until the ulcers are healed.

Precocious Dentition

In rare cases, children are born with teeth a condition very likely to be present in fissures of the lip and palate. In the great majority of cases the teeth fall out soon after birth, and at the same marked inflammatory changes are often present in the alveoli. they have to be extracted, especially when they interfere with breastfeeding. Such precocious dentition has been ascribed to many distinguished personages—Mohammed, Louis XIV., Mirabeau, Robespierre, Richard III. (Hiedert and Fischl, and Tillmanns). When the teeth are only loosely fixed, the condition is one of displacement of the enamel germ towards the surface; and, as the root is very short, the crown soon falls off. When, however, the teeth are firmly fixed, then the condition is one either of a premature formation of the germ-layer, or of increased rapidity of growth. Congenital teeth show a disposition to inflammation of the dental periosteum.

In the Göttingen Women's Clinic, I have only observed one case of precocious dentition among 4220 births (Jr. No. 13407). In this child, the left lower incisor tooth—it is usually the incisor teeth of the lower jaw—had erupted. Its growth after birth was very rapid; but it had already become loose at the time of the child's dismissal from the hospital. The nutrition of the child was apparently affected by the

[1] Henoch attributes these ulcers to friction with the finger in cleansing the mouth, or to the action of the teat, and calls attention to the fact that they are not syphilitic.

[2] Silver nitrate. This is best applied with a camel's-hair brush, and of the strength 1 in 15.

tooth, as its weight, which at birth was 3700 grm., had diminished on the eleventh day to 3440 grm., although no other cause for the loss of weight was present. In a few cases, giant growth has been seen in teeth of this character (Ahlfeld). Thus, in a child fourteen days old, an eye-tooth was observed, the point of which reached to the ala nasi of the same side. In another case, the tooth measured 2·9 cm. in thickness, 1·9 cm. in length, 1·8 cm. in breadth, and weighed 12·37 grm.

Defects of the Tongue

Complete congenital absence of the tongue is a very great rarity, and depends upon imperfect development of the first and second visceral arches.[1] Tillmanns has reported a case from the French literature. It is that of a girl, fifteen years of age, in whom in the place of the tongue, there was a small elevation in the floor of the mouth. Speech was but little affected; but nutrition became impaired, as chewing and swallowing were very difficult, and she was compelled, first of all, to push all her food to the back of the mouth with her fingers.

Bifid Tongue

A split tongue is a rather more frequent deformity. In such a case the tongue is divided by a more or less extensive longitudinal split into two halves.[2] In some cases the split extends as far as the root of the tongue. The treatment should consist in the freshening of the edges of the two halves and the subsequent suturing of the raw surfaces.

Adhesion of the Tongue—Ankyloglossia—Adherent Tongue

There are two varieties of this condition which must be differentiated carefully from one another. In some cases the tongue is firmly attached to the floor of the mouth by congenital bands passing from its margins, while in other cases the union of the tongue with the floor of the mouth is merely of a superficial epithelial character. Such a condition requires, in all circumstances, operative treatment. What is usually understood as

[1] The tongue really consists of two parts, the buccal and pharyngeal portions: the former is derived from the first mandibular arch, and the first interbranchial space by the upgrowth of a median tubercle (the tuberculum impar), while the floor of the primitive pharynx, growing up on either side of the tubercle, finally fuses with it and so forms the complete buccal portion. The pharyngeal portion of the tongue is developed from the united ventral ends of the second and third visceral arches. Congenital absence of the tongue mainly affects the buccal portion, and interferes with the speech to a less extent, as a rule, than in patients who have had the tongue removed.

[2] The occurrence of a fissure in the tongue is explained by its mode of origin as described above.

an adherent tongue—especially among laymen—is an altogether different matter. In this case the frænum linguæ is abnormally short and attached too far forwards, almost to the tip of the tongue. In the opinion of many medical men—especially of the older generation—the mobility of the tongue is so diminished thereby that sucking and, later on, speaking, are rendered very difficult. Although of late years it has been generally recognised that the employment of the term, 'adherent tongue,' has led to a misuse of the word—especially by anxious mothers and midwives—yet it is necessary in a small number of cases to loosen the tongue by cutting through the frænum with a pair of sterilised scissors.

In a recent work, Schleissner, on the ground of a large experience, criticises adversely the recommendation to loosen the tongue. According to this author, the assertion that the presence of a short frænum linguæ has any injurious influence upon sucking, speech, or the mental and bodily development, is a fable.[1] Schleissner, indeed, easily proves his contention. As, in the slight operation already alluded to, death has occurred from hæmorrhage and sepsis, any unnecessary interference should be avoided. It is of importance that these facts should be made clear to midwives, since they are the '*spiritus rector*.' Finkelstein is of the same opinion.

On the other hand, a frænum linguæ of excessive length has also been observed. According to some authors, in this condition attacks of choking may occur from swallowing of the tongue. These observations have, however, been contradicted by several authors.

Macroglossia—Prolapse of the Tongue—Glossocele

Macroglossia to a slight degree is not infrequent. The enlargement of the tongue is as a rule congenital, but the main increase in size takes place after birth. The affection frequently involves the whole tongue, less commonly one-half of it. The condition is primarily produced by well-marked cystic enlargement of the lymphatic vessels[2] (lymphangioma of the tongue).

At the same time an inflammatory infiltration of the underlying tissues, of the muscle, and of the connective tissue often occurs. The condition can further arise as the result of a cavernous enlargement of newly formed blood-vessels (Hæmangioma cavernosum) which may develop either in a circumscribed or a diffuse form (Telangiectasis). A mixed tumour can also form (Hæmato-lymphangioma: see Kaufmann), as the interstitial tissue between the ectatic lymph-vessels

[1] There can be no doubt as to the correctness of these views.

[2] A simple or muscular macroglossia may also be met with in which all of the muscles, or those of one-half of the tongue, enlarge—a condition which is especially common in idiots.

and the neighbouring veins may atrophy from pressure, and so blood may find its way into the lymph-spaces. As a result of compression of the umbilical cord, venous stasis may also occur in the tongue (Ahlfeld). Cystic tumours have been observed in this position originating from dilated mucous glands by obliteration of their ducts. The colour of the enlarged tongue in hæmangioma and hæmato-lymphangioma is dark blue.

A similar enlargement of the tongue is met with in chondrodystrophia fœtalis (see p. 244), in myxœdematous cretins, and in hemicephalus. As the mouth cannot be closed from the presence of the enlarged tongue, breathing and the taking of nourishment, owing to difficulty in swallowing, may be rendered very difficult; and, later on, speech will be interfered with. As a general rule, there is a continuous flow of saliva from the mouth. The teeth—especially the incisor teeth and their corresponding alveolar processes—at a later date are displaced forwards in a horizontal direction by the protruding tongue. From the pressure of the teeth, sore places occur from which infection may arise with resulting hyperplasia of the tissues. If the tongue remains permanently outside the mouth it becomes dry, friable, cracked, bleeds readily, and is covered with foul scabs.

Treatment consists in a wedge-shaped excision of the tissue and suture, or in puncturing with Paquelin's or the galvano cautery.

Rosenow has described a very interesting case of macroglossia of uncommon causation in a new-born child. The fœtus, which was 34 cm. in length, had growing from the side and upper aspect of the middle and posterior part of the left half of the tongue, a stalked tumour, which measured 8 cm. in length and breadth, and 6 cm. in thickness, and consisted of a proximal and distal portion separated from one another by a groove. Each of these cystic and soft masses of tissue contained a long bone with an epiphysis, and a piece of cartilage. Microscopical examination showed the presence of white brain matter and strands of cylindrical epithelium infiltrated with blood. Rosenow described the tumour as a parasitic double-formation, epignathus or epiglossus. The case is quite unique in the literature (see p. 330).

Ranula

Under this name are described a large number of congenital cystic formations situated beneath the tip of the tongue and on the lateral aspect of the frænum linguæ. They are either true cysts of the mucous glands, or arise from blocking of Wharton's duct of the sublingual gland [1]—for example, by salivary calculi. Most frequently

[1] Submaxillary gland. The sublingual gland discharges through the duct of Rivinus.

they occur through cystic changes in the glands of Blandin-Nuhn [1] which are placed in the tip of the tongue. Some arise also through errors of development in the fœtal branchial clefts. Dermoid cysts have also been observed in this situation. The contents of these cysts are usually viscid, of a stringy mucous character, and at times thin and watery; while, in the case of dermoid cysts, they are of a pultaceous consistence. They may be situated on one or both sides of the frænum, and their size varies between that of a pea and an almond. Their wall is generally thin and translucent; and they appear, according to Kaufmann, through the thinned mucous membrane of the mouth, like a bladder filled with water. These tumours may push the tongue out of its place and give rise to difficulty in mastication, swallowing, and speaking; and indeed may even render breathing difficult.

Suppuration of the cysts seldom occurs. Leser describes a case of a tumour of this kind which protruded in the floor of the mouth and formed a swelling in the neck behind the lower jaw. If the tumour causes pain it must be removed. Simple puncture is useless as the opening closes again; while extirpation frequently offers considerable difficulties.

Obstetric injuries of the mouth—especially of the angle of the mouth—of its floor, and of the tongue, are uncommon. They may, however, occur in face presentations, as the result of rough examinations, and in breech presentations from a clumsy use of the Veit-Smellie grip. In some cases the lower jaw has been fractured.[2]

Literature

Kaufmann, *loc. cit.*, p. 24; Biedert and Fischl, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34; Birnbaum, *Verletzungen des Kindes bei der Geburt.*; Rosenow, *Diss. Kiel* (1901); Schleissner, *Die angewachsene Zunge, Prager med. Wochenschr.* (1908), No. 16; H. Finkelstein, *Lehrbuch der Säuglingskrank.* (1908).

Malformations and Congenital Diseases of the Eyes

Complete absence of the eyes (anophthalmia), or the presence of only one eye (cyclopia), occurs almost always in non-viable 'monsters.'

Anophthalmia is, as we have already mentioned, an exceptionally frequent complication of hydrocephalus (see E. v. Hippel).

By microphthalmus is understood an abnormally small eye, it may be as small even as a pea or a millet-seed. In consequence of this maldevelopment—especially in the higher degrees—the power of

[1] Small acinous glands lying on either side of the middle line on the under surface of the tongue.

[2] Or dislocated.

distinguishing between light and darkness alone is retained. Coloboma of the iris and choroid, microcephalus and atrophy of one half of the face, are frequently associated with this deformity.[1]

Another condition often met with in association with microphthalmia and anophthalmia is the presence of cysts in the lower eyelids, which are filled with a clear fluid and stand in some relation to the rudimentary condition of the eyes.[2] Biedert met with two cases of microphthalmia, one soon after the other. Treatment is of no avail; cases, however, have been recorded in which the eyes subsequently increased in size.

Hydrophthalmus or buphthalmus[3] or exophthalmus is frequently congenital, and results from an increase in the intra-ocular tension, which leads to an enlargement of the eyes, blindness, and cupping of the optic nerves. The condition therefore corresponds to a congenital glaucoma, and can be improved by iridectomy. (For further details, the reader should consult text-books on ophthalmic surgery.)

Congenital defects of the eyelids are of considerable importance. The best known is a coloboma, affecting most commonly the upper lid. By this, is understood a defect of an irregularly triangular shape, the base of which is turned towards the free edge of the lid, and the apex towards the orbital margin. The defect should be remedied by a plastic operation (see v. Hippel).

Epicanthus is a congenital semilunar fold of skin, which arises from either side of the bridge of the nose, and covers horizontally the inner angle of the eye (see v. Hippel). In the Mongolian races, epicanthus is present in the adult, and produces in this case, the characteristic form of the opening of the eyelids. If the folds do

[1] A coloboma of the choroid is due to imperfect closure of the choroidal fissure. In normal conditions, the margins of the fissure, which is placed at the lower and outer part of the secondary ocular vesicle, unite and all traces of it disappear. The closure of the fissure is prevented by the persistence of the mesoderm, which occupies it in its early stages and conveys the blood-vessels into the interior of the eye. The persistent mesodermal tissue, which should develop into the choroid and the sclerotic, does not do so, but forms merely fibrous tissue. In many animals —as, for example, birds and fishes—patency of the ocular fissure is the rule.

[2] When the margins of the two layers of tissue, representing the pigment layer and the retinal epithelium forming the secondary vesicle, are prevented from uniting by the persistence of the mesodermal tissue, they grow in a faulty direction either inwards into the interior of the eye or more commonly outwards. In the latter case, a cyst-like structure is often formed, growing from the lower and outer aspect of the eye and forming a serous cyst in the lower lid, lined with retina and connected by a pedicle with the remains of the eye, which is commonly in a condition of microphthalmus.

[3] The so-called 'ox-eye' or glaucoma of childhood. The difference in appearance from the glaucoma of the adult is due to the fact, that the yielding nature of the sclerotic in the child renders possible the marked enlargement of the whole eye which takes place.

not diminish in size spontaneously they must be excised. An external epicanthus is also described. Other congenital abnormalities of the eyelids are paralysis of the levator palpebræ superioris with ptosis, resulting from imperfect development or complete absence of the muscle. The paralysis is, as a rule, bilateral [1] and can only be cured by operative treatment.

By distichiasis is understood a congenital formation of two rows of eyelashes in otherwise normally developed lids. The Meibomian glands are absent and in their place are found the normal eyelashes. Treatment by electrolysis is indicated.

Abnormal shortness of the lids has also been observed, and in rare cases their complete absence (ablepharia).

Symblepharon is the union of the palpebral conjunctiva with the bulbar conjunctiva. In marked degrees of this condition imperfect power of movement of the eyes occurs, with double vision and disturbances of nutrition, as the result of the imperfect opening of the lids. Treatment must be carried out on surgical lines.

Anchyloblepharon consists in the union of the two eyelids along their margins.

Congenital Anomalies of the Iris

Congenital closure of the pupil is a very rare anomaly. It occurs as a result of the persistence of the pupillary membrane after birth (Membrana pupillaris perseverans). On examination, a grey or brown tissue is found on the anterior capsule of the lens in the region of the pupil and attached to the iris by brownish filaments (see v. Hippel). Frequently, only isolated brown points are present on the lens capsule, or fine threads which pass from one margin of the iris to the other, or from the iris to the lens capsule. These are obliterated blood-vessels filled with blood pigment. The threads bear a great resemblance to the synechiæ [2] which persist after an attack of iritis.

The prognosis of this anomaly is good, as the free movements of the pupil are usually not interfered with; the threads readily stretching. By the instillation of atropine, the pupil can be dilated to a round shape and the threads torn by the pull of the iris muscle; the torn shreds ultimately becoming absorbed.

In polychoria, defects in the tissue of the iris are present; as a result of which accessory pupils [3] occur.

[1] In contradistinction to the acquired form which is usually unilateral.

[2] But do not (like the synechiæ) arise from the pupillary margin, but from outside it.

[3] These are not true accessory pupils, as they are not surrounded by the sphincter pupillæ.

Coloboma of the Iris

In these cases, a pear-shaped unilateral or bilateral cleft of the iris is present with its stalk directed downwards towards the margin of the cornea. The sphincter of the pupil, however, surrounds the whole of the pupil [1] as well as the coloboma (see Fuchs). In many cases, a coloboma of the eyelids, of the choroid, of the ciliary body, or even of the lens, exists at the same time. Colobomata are the result of the incomplete closure of the choroidal fissure of the fœtus.

At times, the iris is completely without pigment—such individuals being distinguished as albinos. In this case, the iris is transparent, and, on account of its numerous vessels, it has a fine delicate greyish-red colour.[2]

Irideremia—Aniridia

The iris may be partly or completely absent; in such a case there are usually also opacities in the cornea, the lens, and the vitreous. The pupil shines like a cat's eye, and at the same time marked nystagmus or tremors of the bulb occur, as in the albino. As the eyes receive too much light, treatment must be undertaken so as to shield them from the excess of light by the use of smoked glasses. Secondary glaucoma and separation of the retina is not uncommon.

Ectopia of the Pupil

The pupil may be placed eccentrically: most commonly, upwards and outwards from the centre of the eye; less commonly, upwards and inwards, or downwards and inwards. At the same time, the lens is often correspondingly displaced (Dislocation of the lens) and altered (Secondary cataract). All the described anomalies of the iris occur most frequently on both sides, and are not uncommonly hereditary.

As these conditions are often further associated with other anomalies of the eyes, the sight is generally affected to a corresponding degree.

Congenital Cataract

A congenital cataract is generally bilateral and often inherited. The cause is to be found either in some defect of development, or in an intra-uterine inflammation of the eye. After delivery, the anomaly may be either stationary or progressive. A partial intra-uterine cataract

[1] Congenital coloboma is thus distinguished from the artificial one made by iridectomy.

[2] In albinotic eyes, the pigment-bearing cells of the retina and uvea are present, but contain no pigment.

formation consists, most commonly, of a so-called anterior and posterior polar cataract (see Fuchs), in which there is a white spot in the middle of the pupil.

Less common is the lamellar cataract, in which a zone of opaque lens substance is placed between the clear nucleus and the clear cortical layer. Not uncommonly, a congenital cataract is recognised for the first time not immediately after birth, but some time later. This is to be explained, firstly, by the fact that new-born children have very small pupils, and secondly, that they often have their eyes closed. These cataracts, according to the opinion of most ophthalmologists, should be operated upon as soon as possible. Discission, which is the method of treating this anomaly, can be carried out with good results, even in the first week of life. If treatment is deferred, the retina fails to develop properly, and blindness or weakness of sight ensues. (For further details on these subjects see the text-books on Ophthalmology.)

Atresia of the Nasal Ducts in New-born Children [1]

This condition, according to Zentmayer, is frequently mistaken for, and described as, ophthalmia neonatorum purulenta. A swelling is present at the inner angle of the eye, and, on pressure, a whitish gelatinous secretion can be squeezed out of the affected nasal canals. In mild cases, the conjunctiva is not affected, and most frequently the affection is only on one side (see report by Ibrahim).

The following anomalies may be briefly mentioned: a persistent hyaloid artery [2] (canal of Cloquet),[3] congenital opacities of the cornea (leucoma and staphyloma), cryptophthalmus (the eyeballs are covered with a bridge of skin extending from the forehead to the cheeks), dermoids and teratomata of the bulbs and of the orbits, congenital pigmentary anomalies of the eyes (albinism and melanosis oculi), heterochromia (differences in colouring of the iris), and imperfect development of the optic nerves.

Inflammation of the eyes in the fœtus (the existence of which has been denied) has been established by the more recent investigations of Reis and Seefelder. According to these observations, every segment of the eye is liable to inflammation during the period of development.

[1] This is due to delayed perforation of the thin diaphragm of mucous membrane which in the fœtus closes the lower orifice of the nasal duct; as a consequence, decomposition of the retained gelatinous contents of the nasal passages occurs, leading to inflammation.

[2] In the new-born child, a remnant of the hyaloid artery (some 1 to 1½ mm. in length) can be recognised in the majority of cases.

[3] The canal in the vitreous, in which the hyaloid artery runs.

It is consequently very difficult to say, in many cases, whether any given anomaly of the eye is the result of such an inflammatory process or of a malformation (for further details, the work of Seefelder should be referred to).[1]

The injuries which the eyes may suffer during delivery, or as a result of the necessary interference in association therewith, are very numerous. By a rough examination—especially in face presentations—the eyelids and corneæ have been injured on many occasions. In one case, in which the eye was mistaken for the anus, as the result of such an examination, the bulb was completely destroyed (de Wecker). In another case, the bulb was dislocated, and, in spite of reposition, it suppurated (Bock; quoted by Stumpf). In perfectly normal births, subconjunctival or retinal hæmorrhages[2] may occur. Strabismus and exophthalmus have also been observed in normal deliveries, but more frequently in contracted pelves. Exophthalmus, further, occurs during delivery from retrobulbar hæmorrhages, and through indentations of the temporal portions of the orbit from improper application of the forceps.[3] The latter may also give rise to severe injuries to the eyes—such as large effusions of blood into the sclerotic and conjunctivæ, hæmorrhages into the anterior chamber, retrobulbar and intrabulbar effusions of blood, luxation of the lens and traumatic cataract, abscess of the orbit, microphthalmus, and phthisis bulbi (Stumpf). These hæmorrhages may lead to opacities of the cornea, and to paralysis of the nerves—especially of the abductor muscles. (See Stumpf, Schmidt-Rimpler, Birnbaum, and E. Runge.)

Literature

Fuchs, *Lehrb. d. Augenheilkunde* (1897); Biedert and Fischl, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34; Seefelder, *Über fötale Augenentzündungen, Deutsche med. Wochenschr.* (1908), No. 28; Schmidt-Rimpler, *Nothnagels, loc. cit.*, p. 53; Birnbaum, *loc. cit.*, p. 101; Stumpf, *loc. cit.*, p. 44, vol. iii., pt. 3, p. 482; Zentmayer, *Atresie des Tränennasenganges, ref. Ther. Monatsh.* (1909), p. 51; E. Runge, *Gynäkologie und Geburtshilfe in ihren Beziehungen zur Ophthalmologie* (Leipzig, 1908); E. v. Hippel, '*Die Missbildungen und angeborenen Fehler des Auges*,' *Graefe und Saemischs Handb. d. gesammten Augenheilkunde*, vol. ii., pt. 1, chap. 9 (with bibliography); Wolff, *Beiträge f. Augenheilk., Festschr, J. Hirschberg.*

[1] As a general rule, however, it may be asserted that malformations of the eye originate at a very early stage of development, whereas inflammatory processes only occur in the later stages of fœtal life.

[2] Paul estimates the frequency of retinal hæmorrhages in spontaneous deliveries at 20 per cent. and in contracted pelves at 50 per cent.

[3] In 112 cases of this kind collected by Wolff, in no less than nineteen the eyeballs were extruded from the orbits. These injuries naturally only occur in cephalic presentations; never when the child presents by the breech.

Malformations of the Ears

The pinna of the ear may be absent on one side, or, more rarely, on both sides; as a rule, however, small rudiments are present. At the same time, atresia of the external auditory meatus[1] and defects of the middle ear are often present, as well as malformations in the region of the first visceral arch, or of other more distant parts of the face, and neck: for example, hare-lip, clefts of the palate, congenital fistulæ of the neck (Moldenhauer). More frequently, partial defects alone are present: the auricle, for instance, may be abnormally small (Microtia), or abnormally large (Macrotia). The last is found especially in idiots. Ahlfeld describes one of these rare cases, and also reports one from the literature (Wreden) in which the right ear was 74 mm. long. Well-marked deformities of the auricle are generally found together with these defects. Thus it may be converted into a thick shapeless mass, or it may assume a rolled-up cylindrical or spindle-shaped form.

Doubling of the auricle has also been described (Polyotia): for example, in dipygus parasiticus. With reference to this, Ahlfeld points out that folds of the skin have been falsely described as double auricles. These folds are formed from the cheeks near the ears, and are to be considered as the result of a faulty closure of the visceral clefts leading to a process of hypertrophy commencing in the visceral arches. Such skin excrescences may be described as accessory auricles. They consist of skin, subcutaneous tissue, and cartilage, and are situated most commonly on the tragus, the lobule of the ear, and on the neck.[2] Very rarely dislocations of the auricle occur: namely, on the cheeks or the neck. (For synotia, see p. 95.)

A further congenital anomaly is a faulty position of the auricle, so that by standing out at right angles to the head it leads to an ugly deformity.

All these malformations, in selected cases, are suitable for treatment. In complete absence of the auricle a plastic operation with the formation of flaps should be considered, but unfortunately the results are nearly always very bad. In the majority of cases the only result is often an unshapely lump of flesh, although of late years better results appear to have been obtained. For these reasons many surgeons and otologists do not perform plastic operations, but allow

[1] The associated absence of these two structures is explained by the fact that the external auditory meatus is developed from the first cleft depression and a solid ingrowth of epithelium derived from it, while the auricle is developed from six tubercles, colliculi, which grow up round the first cleft depression from the hyoid and mandibular arches.

[2] For a full account of these 'pre-auricular appendages,' see Ballantyne, *Teratologia*, vol. ii., pp. 24 and 65 (1895). He regards them as due to budding from the margins of one or more of the branchial fissures or processes; or, in some cases, from the colliculi or helices themselves, and thinks that they have an amniotic origin.

the defects to be covered by the hair, or employ artificial ears of papier mâché or of metal. Slight defects can usually be corrected by appropriate operative measures.

In cases of macrotia, wedge-shaped pieces must be excised; while the disfiguring auricular appendages can readily be removed. An outstanding ear can be remedied by wearing an appropriate bandage for

FIG. 26.—MALFORMATION OF THE EXTERNAL EAR. (Specimen from the collection of the Göttingen Women's Clinic.)

a few weeks, or by the excision of portions of skin along the insertion of the auricle.

Congenital Fistula of the Ear

This is a fistula in the region of the ear, which may be regarded as the remains of the first branchial cleft. The fistula is situated most commonly in front of the ear above the tragus, near the malar bone, or in the lobe of the ear. It may be unilateral or bilateral, and, at times, combined with fistulæ in the neck.

These fistulæ most commonly end blindly; but they may open, in rare cases, in the region of the opening of the Eustachian tube, into the middle ear, or into the pharynx.[1] According to Ahlfeld the malformations described as double meati must be considered as of this nature. These fistulæ give exit at times, spontaneously or on pressure, to a

FIG. 27.—MALFORMATION OF THE EXTERNAL EAR. (Specimen from the collection of the Göttingen Women's Clinic.)

yellowish-white fluid. If the opening becomes closed a small fluctuating tumour forms. The fistula, whenever possible, should be extirpated; or, if this is not feasible, it should be laid open as far as possible and

[1] It is probable that two conditions are included under the term 'congenital fistula of the neck.' The first is due to persistence of some part of the first branchial cleft, and this may be regarded as the explanation of such a case as that described by Virchow, in which it was possible to pass a probe through the fistula into the pharynx, while the second variety is due to imperfect union of the auricular tubercles taking part in the formation of the auricle. The latter is the less important and more common form.

cauterised. In the pathological collection of the Göttingen Women's Clinic there is a specimen (No. 167) of this nature. It is that of an eight-months-old fœtus with bilateral malformation of the ears (Figs. 26 and 27). On the right side there are three fistulæ with well-marked defect of the auricle and atresia of the meatus; on the left side, deformity of the auricle, an auricular appendage, and atresia of the external meatus.

Malformation of the External Auditory Meatus

The meatus may be entirely wanting (Atresia meatus auditorii externi). This is associated as a rule with other deformities—namely, of the auricle and of the tympanic cavity; while the tympanic membrane is nearly always absent also. The cause of these malformations lies in anomalies of the soft parts[1] or of the bones. In some cases there is only a membranous closure. In association with these complete atresiæ, narrowing of the external meatus also occurs. Operative treatment gives the best results in true cases of fleshy adhesions of the external meatus. In cases of stenosis, attempts may be made to carry out slow dilatation with laminaria tents (for further details, consult text-books on Otology).

The tympanic membrane may be congenitally completely absent, in which case defects of the external meatus are usually also present. Partial congenital defects of the tympanic membrane, in the form of fissure formations (foramen of Rivini),[2] are uncommon.

The position and inclination of the membrane may also be congenitally altered, as well as its form and size. In complete absence of this structure, the introduction of an artificial membrane should be advised (see Moldenhauer).

The auditory ossicles, as well as the tympanic cavity, may be deformed; in this case there is more or less marked difficulty in hearing.

The Eustachian tube may be completely absent; but here, too, other malformations are usually present. It may also be congenitally obliterated, or the pharyngeal end of the tube may alone be closed. Finally, congenital defect of the labyrinth, its complete absence or imperfect development, is always associated with other defects of the ear. The result, in every case, is incurable deafness. The auditory nerve may be completely wanting, or it may be pathologically altered: thus its consistence may be too hard or too soft (for further details, see Moldenhauer).

[1] Failure of the solid mass of cells to break down, from which the inner part of the meatus is developed, is no doubt the cause in some cases.

[2] Marx has not been able to find any authentic case, in the literature, of complete absence of the membrana tympani, and he regards the so-called foramen of Rivini as an acquired, and not a congenital, defect.

Injuries of the ears during delivery are of the greatest rarity. They may occur in the form of excoriations of the skin and crushing and separation of the soft parts—especially in forceps operations in contracted pelves. Whether a hæmatoma of the ear can occur as a result of birth-trauma, I do not know; but tearing off of the ear from the slipping of the forceps has been observed.

Malformations of the Nose

There is little to be said about malformations of the nose. Von Wutzel has described the so-called dog-nose—in which there is a nasal furrow, with separation of the two nostrils, usually associated with a median fissure of the upper lip. Rarely, a complete absence of the nose has been observed in cases of cyclops; usually, however, it is present as a movable snout-shaped appendage above the eyes. Congenital atresia and stenosis of the nostrils have also been met with; they are generally of a bony—less commonly, of a membranous—nature. Atresia of the posterior nares has also frequently been seen. Children born with this condition, as a rule, die soon after birth from asphyxia; while those which remain alive exhibit the disturbances described below.

Lastly, anomalies of the nasal framework, spurs and deviations of the septum, must be mentioned.

The malformations of the nose, above described, result in many disturbances of function. In the first place, the main function of the nose—the ability to smell—may be interfered with. In the same way, the other functions—the warming, cleansing, and moistening of the air—may also be performed imperfectly. Moreover, sucking by the infant is almost completely prevented by obstruction of the nose. Treatment consists in the removal of the closure by operative measures, and afterwards keeping the passages open by tubes, etc.

Lastly, the view held by some authors may be mentioned—namely, that the formation of adenoid growths in the naso-pharynx is a congenital affection.

Injuries to the nose during delivery are uncommon. They have been observed as injuries to the soft parts and fracture of the nasal bones—especially in forceps delivery, when the forceps has been applied in contracted pelves to the head lying above the pelvic brim in the transverse diameter. Olshausen has also observed a case of fracture of the nose in a normal labour.

Literature

Leser, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34; Finkelstein, *loc. cit.*, p. 301; Birnbaum, *loc. cit.*, p. 191; Moldenhauer, *Die Missbildungen des menschlichen Ohres.*, *Handb. d. Ohrenheilkunde von Schwartze*, vol. i., p. 154.

CHAPTER VII

Malformations and Congenital Diseases of the Neck

Congenital Fistula of the Neck

Congenital cervical fistulæ are to be considered as malformations due to arrest of development, as they are the result of want of closure of the embryonic branchial clefts and recesses.[1]

Heredity undoubtedly plays an important rôle in these cases; thus Ascherson found this malformation in eight members of the same family (quoted by Ahlfeld). These fistulæ form canals lined with mucous membrane into which only a fine sound or bristle can be introduced with difficulty, and which open externally or internally, or internally and externally. They are, accordingly, distinguished as complete cervical fistulæ, incomplete external and incomplete internal cervical fistulæ. The external opening is placed, most commonly, in the neighbourhood of the sterno-clavicular joint,[2] at the inner or outer margin of the sterno-cleido-mastoid muscle; or, more medianly, in the neighbourhood of the larynx. The inner orifice may open into the pharynx, larynx, or the trachea. Symptoms, as the result of the presence of the fistula, are seldom present.

According to the richness of the fistula in mucous glands, a larger or smaller quantity of clear colourless and odourless viscid fluid is secreted. If the fistula is a complete one, portions of food—especially of a fluid character—may escape externally. If the external opening of the fistula becomes contracted or blocked, a cyst may form which projects under the skin. In the case of an internal fistula, the internal opening may become dilated into a diverticulum. If the diverticulum is situated in the pharynx or in the œsophagus, marked difficulty in swallowing may occur; while if it is situated in the trachea (tracheocele) it may lead to the formation of a large sac, situated in the anterior part of the neck, which fills with air (see Kaufmann).

[1] The various clefts and fissures in the neck are to be explained by the persistence of some of the branchial clefts, or of the cervical sinus, of the whole or a part of the thyroglossal tract, and, in the case of a complete fistula, by the disappearance also of a cleft membrane.

[2] They occur most commonly at the sides of the neck, below the level of the hyoid bone, and nearly always open into the pharynx.

Treatment in the majority of cases is not necessary, as the symptoms are, as a rule, of no importance. In those cases requiring treatment, surgical measures are best—namely, excision of the whole canal. The injection also of a few drops of tincture of iodine may lead to the healing of these fistulæ. Of a different character are the true tracheal fistulæ which penetrate the trachea and end on the anterior surface of the neck; these occur as the result of incomplete closure of the fore-gut.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Leser, *loc. cit.*, p. 34; Tittmanns, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34.

Congenital Malformations of the Neck

Tumours in the neck of the new-born child—excluding goitre—are partly of a cystic, partly of a solid, nature.

The cystic tumours develop, in the majority of cases, from the embryonic clefts. In their classification, I shall follow Leser's clear table and, in accordance with this, distinguish endothelial and epithelial tumours.

To the endothelial cervical tumours belong:—

1. Lymphangiomata: also known as congenital multilocular cysts, congenital cystic hygroma of the neck, or congenital lymphangiectasis of the neck. A lymphangioma is a cystic tumour which is situated in the submaxillary region, or above the clavicle, and takes its origin from the lymphatic vessels of the neck. In their anatomical structure these tumours consist of numerous—in part closed, and in part dilated and enlarged—lymphatic vessels, communicating with one another. After birth, they often grow very rapidly and form marked swellings, which grow upwards, and can be observed in the floor of the mouth. The cystic dilatations show under the microscope an endothelial lining, and usually contain a light yellow coagulable fluid. The over-lying skin is either smooth, or thickened—as in cases of elephantiasis—or wrinkled. Large tumours of this kind may lead to a fatal issue, as the result of interference with the respiration[1] or the circulation. In rare cases, spontaneous healing occurs after rupture of the sac.

The radical extirpation of these cysts is usually impossible unless they are quite small. In the great majority of cases, treatment must be confined to multiple punctures and injection with iodine. In a few cases, simple incision and plugging with iodoform gauze leads to a cure. (For further details, see text-books on surgery.)

2. Cervical hæmatocele. This is a very rare condition, and occurs

[1] In their growth, they tend to follow the deep fascia towards the trachea, under the clavicle into the axilla, or into the anterior mediastinum.

as the result of cavernous enlargement of the veins. Clinically, it presents the appearance of a cystic tumour of the neck. Closed cysts must be distinguished from those communicating with the lumen of the vessel of origin. Surgical treatment—such as puncture and compression, or extirpation—should only be undertaken in the case of closed cysts. Grossmann has reported a case of such a cyst operated upon with success in a boy only one day old.

To the epithelial tumours belong :—

1. Hydrocele of the neck.[1] This is a unilocular cyst (Leser), lined by many layers of cylindrical epithelium, showing its origin from the embryonic visceral clefts. These tumours are usually thin-walled, and their contents are serous or sero-mucinous. As they are retention-cysts, their size may vary very considerably: they may, for example, attain the size of a child's head. The most common position of these tumours is the lateral region of the neck, between the mastoid process and the hyoid bone, or at the inner margin of the sterno-cleido-mastoid muscle, in the supra-clavicular fossa, and in the region of the interclavicular notch. According to Leser, puncture and injection with iodine is especially suitable for these tumours, and should be carried out before extirpation is considered.

Fig. 28.—Congenital Cystic Hygroma of the Neck. (After Kaufmann.)

2. The so-called deep atheromata or dermoids of the neck also take origin from the branchial clefts. They are dermoid tumours, which arise as the result of infoldings of the epidermis during the embryonic period in the neighbourhood of the branchial clefts. Their wall, therefore, exhibits the characters of skin, and is covered with stratified pavement epithelium. Their contents are a mass of shed epithelium, fat, and cholesterine. They are generally not of large size, and increase very slowly after birth ; their favourite position is the region [2] below the angle of the jaw, or the supra-clavicular fossa, where they are often in part covered by the sterno-cleido-mastoid muscle. Any surgical interference is contraindicated.

[1] The reader must bear in mind that many English writers describe a hydrocele of the neck as due to the congenital distension of connective tissue spaces. The hydroceles of the neck here described are better termed branchial cysts, as they are probably formed from the epithelium of the branchial clefts.

[2] They often also arise in the middle line of the neck beneath the tongue, or between the lower jaw and the hyoid bone. They may bulge up beneath the floor of the mouth.

Here it may be noted that (according to v. Winckel) the majority of lymphangiomata arise in a mechanical manner, and as a result of pressure by amniotic bands and the conditions resulting therefrom.[1] According to the same author, the observation that these tumours often show signs of healing is an argument against the widespread assumption that they are new growths.

From the obstetrical point of view, these tumours are nearly always grouped together under the not altogether appropriate name of cystic hygromata (see Kleinhans). They very seldom give rise to any difficulty during delivery. A correct diagnosis (as Kleinhans rightly points out) is very seldom possible before labour. In cases of delay in the delivery of the child, it is usually necessary to introduce the half or the whole hand, under anæsthesia, to determine the cause of the dystocia. If the tumours are situated in large part beneath the lower jaw, they can—as in the case of a congenital thyroid tumour—give rise to more or less marked extension of the head, and so to a primary brow or face presentation. When situated in the lateral region of the neck they can give rise to an asynclitic position of the head, and so to an anterior or posterior parietal presentation.

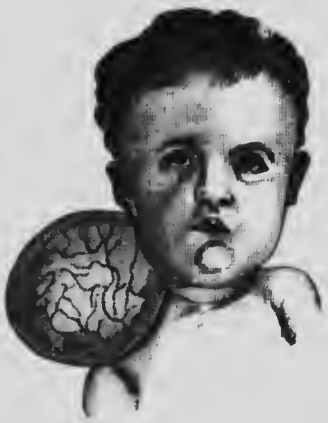


Fig. 29.—Blood-cyst of the Neck. (After Tillmanns.)

If the child presents by the breech, as a result of the separation of the chin from the chest, it will be very difficult to reach the mouth in carrying out the Smellie-Veit method of delivery. In the treatment of this variety of dystocia, the fact must be carefully borne in mind that these children can remain alive, if the tumours are carefully lessened in bulk by puncture during delivery (see Kleinhans).

Definite rules for the treatment of these cases can hardly be given, and each case must be treated on its own merits. In cephalic presentations and in selected cases the forceps may be employed, provided that the head is not too small. Hohl recommends, on the suggestion of Kilian, to use the large space, which the cavity of the sacrum affords in a well-formed pelvis, for the accommodation of the tumour—that is, to cause the tumour to descend into the cavity of the sacrum. This rotation, carried out manually or by the use of the forceps, will only succeed in head presentations when the head is situated above the pelvic brim; in breech presentations, however, it succeeds more

[1] That is to say, venous congestion from dragging on, and changes in, the vessels —conditions very similar to those met with in the limbs in cases of elephantiasis.

readily, as here there is a suitable hand-hold. On this account some writers recommend, especially in multiparæ, the performance of version. If the delivery of the child does not succeed, or if the child is dead, then puncture or removal of the tumour in question must be undertaken.

Lymphangiectatic tumours in other parts of the body may also be

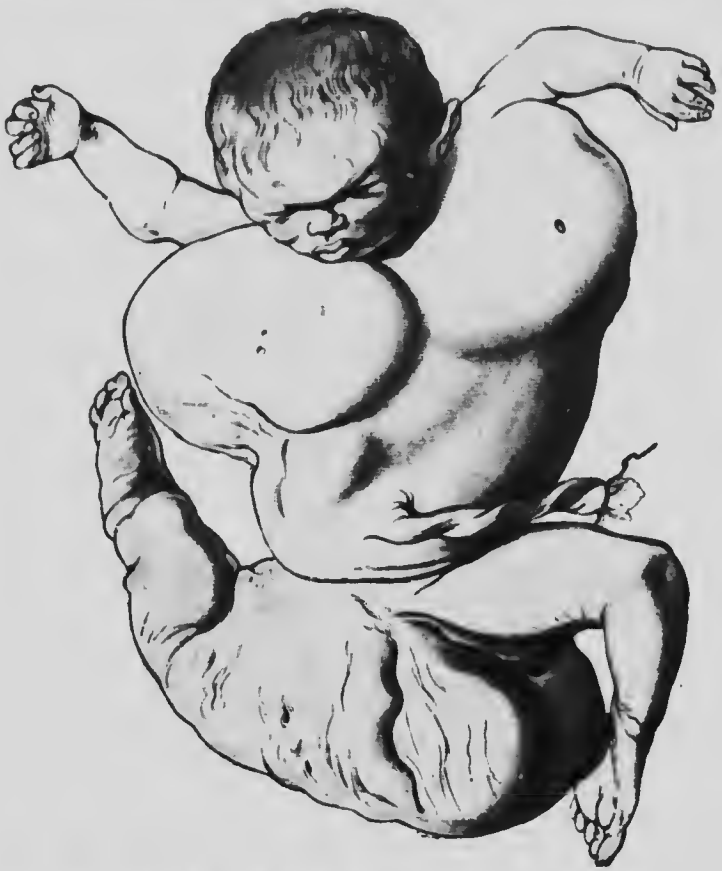


Fig. 30.—Diffuse Lymphangiomata causing Obstruction to Delivery. (After Ahlfeld.)

described here. Lymphangiomata may occur in all those regions of the body which, during embryonic development, exhibit clefts (fissural lymphangiomata). They occur especially in the axillary region, the chest, shoulder, abdomen, the sacral region, and the groin. Ahlfeld figures in his 'Text-book of Midwifery,' a fœtus with enormous lymphangiomata in various parts of the body in which birth took place normally (see Fig. 30). Steinwirker has described, under the name of Elephantiasis congenita cystica, the case of a

fœtus with well-marked dilatation of the lymphatics in the region of the head (figured in 'Text-book of Midwifery,' by Olshausen and Veit, p. 735).

Malcolm McLean recorded a large lymphangioma on the left upper arm which led to difficulty in delivery. As the birth was arrested in a cephalic presentation the right arm was brought down and the further course of the delivery followed with spontaneous evolution, the left arm with the tumour being born last. E. and A. Martin observed cases of multiple cystic formation in the jugular and axillary regions (quoted by Kleinhans).

V. Wörz described a lymphangioma, the size of a child's head, in the region of the axilla, which rendered delivery very difficult. On account of prolapse of the hand and the umbilical cord, he performed version; but the extraction of the child, as far as the umbilicus, was attended with great difficulty, and, on examination with the whole hand, the cause of the obstruction was recognised. The further extraction of the child was finally accomplished, though with great difficulty. Its weight was 3630 grm. Kleinhans records two additional cases of Eberhart and Barone. In the last case, 'an angioma cavernosum' was situated in the region of the upper third of the thigh and rendered the birth of the breech very difficult. Bonnaire and Bose exhibited to the Obstetrical Society of Paris a child with a congenital cystic lymphangioma on the right shoulder. About two years ago, I saw a lymphangioma, about the size of the fist, in the axilla of a child born spontaneously. Heil described a cystic lymphangioma (about the size of the double fist) of the left mamma, which had rendered decapitation necessary, in a child weighing 4 kg.

LITERATURE

Kaufmann, *loc. cit.*, p. 34; Ziegler, *loc. cit.*, p. 15; Tillmanns, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Kleinhans, *v. Winckel, loc. cit.*, p. 34; Hohl, *loc. cit.*, p. 15; v. Winckel, *Über menschliche Missbildungen, Volkmanns Samml. klin. Vortr.*, Nos. 373–374; Ahlfeld, *loc. cit.*, pp. 24 and 34; Küstner, *Müller, loc. cit.*, p. 15; Steinwirker, *Diss. Halle* (1872); v. Wörz, *Zentralbl. f. Gynäk.* (1894), No. 5; Ballantyne, *Diseases and Deformities of the Fœtus*; Grossmann, *Diss. Leipzig* (1903); Bonnaire and Bose, *Ref. Zentrabl. f. Gynak.* (1903), p. 669; Heil, *Gynak. Rundschau* (1908), vol. ix.

Congenital Goitre

In congenital tumours of the thyroid gland, two classes of cases must be distinguished from one another. The first group includes these cases of acute swelling of the thyroid which occur during labour in face presentations with marked lordosis of the cervical vertebræ.

Venous hyperæmia and œdema of the thyroid gland may occur as the result of some obstruction to the outflow of the blood in the veins, and very often at the same time smaller or larger effusions of blood in the tissues of the gland are present. This acute swelling of the gland is a condition which quickly disappears, is quite harmless, and has nothing to do with true goitre. A few cases have, however, been recorded in which the death of the child from asphyxia neonatorum has taken place as a result of such an acute swelling of the thyroid (Hecker, and others). As a consequence of this acute swelling, hoarseness of the new-born child occurs at times, which is to be attributed to the pressure of the enlarged gland on the recurrent laryngeal nerve (v. Winckel, quoted by Küstner).

The second group includes true congenital goitre, which may be observed at the same time in the mother and the child. This condition is found, primarily, in regions[1] in which endemic goitre is met with.

Thus Kaufmann, in Basle, has seen, in post-mortem examinations on children, thyroid glands weighing 35 grm.; the normal weight being 4·85 grm. In well-marked cases of congenital struma, thyroid glands have been described with a weight of as much as 100 grm. An enlargement of the thymus gland is comparatively frequently met with at the same time. Pathologically, these tumours assume an hyperplastic follicular form most frequently. Cysts in the gland, and enlargements of a mixed variety—namely: colloid, fibrous, or cystic enlargements—are also met with. In rare cases, the cause of the enlargement has been described as a dermoid tumour. Not infrequently, cartilage is found in the tumours.[2] In a recent work, Fabre and Thevenot distinguish five varieties of congenital goitre—vascular, simple hypertrophy, cystadenomata, fibromata, and cystomata.[3]

[1] In Switzerland, for instance, Demme (see Literature) reported 642 cases, and fifty-three of these were congenital. Of the fifty-three cases, there were fourteen in which both parents were goitrous, twenty-three in which the mother alone was affected, and sixteen in which both parents were free from the disease.

[2] Bone and transversely striped muscle fibre may also occur.

[3] Clifford White has recorded a unique case of congenital exophthalmic goitre. The mother, a primipara, developed symptoms of the disease at the fifth month of her pregnancy, and they steadily progressed in severity. The fœtus was delivered at full term by forceps, and presented at birth all the typical symptoms—namely: exophthalmos, tachycardia, tremors, and enlargement of the thyroid gland. The fœtal heart-rate was noted to be over 200 per minute during the first and second stages of labour, and the possibility of the fœtus being affected with the disease was discussed. The tachycardia continued, and the child died of cerebral hæmorrhage thirty-six hours after delivery. An autopsy showed that the thoracic and abdominal organs were normal. The pituitary body was also normal. The thyroid gland was uniformly enlarged, and measured 3·5 by 2·9 by 2 cm. Microscopic examination showed its structure to resemble that found in cases of exophthalmic goitre.

In its position, the anatomical relations of a congenital goitre are much the same as in the goitres of adults; but it must be pointed out that, as a result of the high position of the thyroid gland in the new-born child, a retro-sternal goitre is very seldom met with. Demme gives some particulars with regard to the frequency of congenital goitre; thus among 642 children with goitre, in thirty-seven cases it was of the congenital variety (cited by Ewald). A fact of much interest, is that transposition of the viscera is not uncommonly associated with congenital goitre.

Children affected with this condition are either born dead or asphyxiated; or they exhibit, soon after birth, signs of dyspnœa; or present a clinical picture of the so-called goitre asthma—that is to say, attacks of suffocation occurring from time to time. A congenital goitre can, moreover, from compression of the soft trachea or of the vagus nerve, lead to speedy death. If the child lives, and the goitre continues to grow, it may give rise to the well-known bow-like deformity of the soft trachea. The chief symptom of this narrowing, is dyspnœic long-drawn-out inspiratory and expiratory breathing, associated with a hissing sound (Baginsky). From the pressure of the goitre on the veins of the neck, venous stasis, with its sequelæ, may occur in the brain. In many cases a marked enlargement, or diminution in the size, of the tumour occurs during the months following birth.

B. Kamann has recorded a case where death occurred soon after birth. The mother also had a goitre. The child was born with very rattling respiration, and died seven hours after its birth, with increasing cyanosis. At the post-mortem examination, the trachea and larynx were found to be markedly compressed, anteriorly and laterally, by the parenchymatous thyroid tumour. The jugular veins were markedly enlarged and pushed over to one side, so that they regularly encircled the goitre. Macdonald has also reported a case of marked compression of the trachea by a congenital goitre. The child died before tracheotomy could be performed (see p. 120). Ballantyne, Simpson, and others reported similar cases. The observation, constantly made, that the mothers of the affected children had taken, for one reason or another, iodide of potassium during their pregnancy, does not appear to me to be of any importance in considering the ætiology of this condition.

As regards treatment, Finkelstein has rightly pointed out that the period in which the most serious accidents occur, in cases of struma, is within the first few hours or days after birth. In these cases, energetic treatment must be undertaken: stimulation of the breathing, the application of cold, or even iced, compresses to the region of the goitre, the promotion of marked extension of the neck, and, when sucking is difficult or impossible, feeding with a spoon. By these means the venous

congestion, which is always present, may be lessened. In sucklings with congenital goitre, thyroid preparations have been given to the nursing mothers with good results. During pregnancy, mothers, who have already borne several children, should be recommended to move to a region free from goitre.

The later treatment consists in the internal or external use of iodine preparations, which are especially recommended in those cases in which syphilis is an ætiological factor; further, in the internal administration of iodothyrin, injections of alcohol into the goitre, or even in operative interference. The removal of the whole goitre is no longer undertaken at the present day, as the condition of so-called cachexia strumipriva, with tetany, myxœdema, and mental hebetude, is very likely to follow this procedure. Operative treatment should only be undertaken when there is a danger of suffocation. Tracheotomy should especially be avoided, as the danger of secondary pneumonia, after this procedure, is very great.

Goitre has also, in many ways, a purely obstetrical interest. It can, in the first place, give rise to a primary face presentation; and this position of extension of the head (as Küstner has rightly pointed out) is to be regarded as beneficial for these fœtuses. The normal position of flexion, on the other hand, is dangerous: since by the flexion of the chin on the sternum the enlarged and degenerated thyroid, and with it the large vessels of the neck—especially the jugular veins—may be compressed. As a result of this, some obstruction to the outflow of blood from the skull may occur, and the children may die from an accumulation of carbon dioxide in the blood.

These tumours can, although very rarely, give rise to difficulty during delivery from their size (see Kleinhans). Kleinhans has collected five cases of this kind: those of Hofbauer, Vonwiller, Burghagen, Billig, and Dentler. In one of these cases (Hofbauer) a goitre one and a half times the size of a child's head was present; in another (Vonwiller), an enchondromatous goitre,[1] twice the size of a child's head; in the third case (Burghagen)—a colloid tumour—the size of a man's fist, and containing fat and cartilaginous tissue; in the fourth case (Billig), the tumour was a mixed one, the size of a double fist, a colloid and cystic goitre, containing bony and cartilaginous deposits: while in the last case, that of Dentler, a papillary cysto-adeno-chrondro-sarcomatous goitre, was present, the size of a child's head. All the children were born spontaneously, but after very long and difficult labours. On three occasions the presentation was a breech, and in one case marked hydramnios was present (10 litres). Seitz demonstrated to the Gynæcological Society of Munich a cystic goitre larger

[1] This case is figured in v. Winckel's *Lehrb. d. Geburtsh.* (1892), Fig. 102, p. 400. The child was born spontaneously, but died before the arrival of the doctor.

than a child's head, which extended from the chin to the ensiform process. The child was delivered spontaneously.

The diagnosis, as a rule, is made for the first time when an obstruction to delivery arises. The presence of marked extension of the head will be of value in coming to a conclusion. In the treatment or removal of the obstruction, the most important rule to be observed is delivery with the greatest degree of safety to the mother. When possible—and more especially in the cystic form of congenital struma—puncture of the tumour should be carried out, for the reason that such children may survive.

Injuries to the neck during delivery are not common. Injuries to the cervical vertebræ especially separation of single vertebræ has been observed in difficult delivery of the after-coming head, in cases of contracted pelvis and hydrocephalus. These injuries are particularly likely to occur when, in the attempt to deliver the head, traction is made not in the long axis of the neck, but at an angle thereto. The most frequent cause, at one time, was the now practically obsolete Prague method of delivery, in which the neck is bent at an angle. Children with these injuries naturally succumb during delivery or soon afterwards. Under injuries of the soft parts are to be mentioned the so-called marks of stretching on the neck, and injuries to the neck muscles, especially to the sterno-cleido-mastoid.

The marks of stretching are fine tears of the skin, with red marks running transversely across the front of the neck (Kaltenbach). Injury to the sterno-cleido-mastoid muscle occurs almost always in manual extraction with pelvic presentations;[1] less commonly in forceps deliveries, and still more rarely in spontaneous cephalic, and breech deliveries. In forceps deliveries, the effusion of blood, as a rule, only occurs when the apices of the blades of the forceps reach up on to the neck. As a result of tearing of the muscle fibres, there occurs a unilateral or bilateral effusion of blood, which is usually first recognised in the second week of life.

The prognosis of this condition is good, as in the majority of cases the hæmatoma becomes absorbed very quickly; in a few cases, however, a wry-neck may result (see Küstner, Stumpf, and Birnbaum).[2]

Seitz mentions a very interesting case of incurable asphyxia of the child, due to an exostosis of the cervical vertebræ, which

[1] Of 115 cases collected by Stumpf, in 72 (or 62 per cent.) there was a breech presentation—a frequency eighteen times greater than the normal.

[2] Apparently, in some cases, such an injury is predisposed to by an abnormal shortness of the muscle. The exact relation of the hæmatoma to wry-neck is not quite clear. The condition of the muscle in cases of torticollis is not such as might be expected to follow a hæmorrhage, and many authorities attribute the deformity to an interstitial myositis, possibly of an ischæmic character.

closed the entrance to the larynx. (For injuries to the larynx, see p. 125.)

Literature

Kaufmann, *loc. cit.*, p. 24; Kleinhans, *loc. cit.*, p. 34; Ewald, *Nothnagel, loc. cit.*, p. 53, vol. xxii.; Baginsky, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Küstner, *loc. cit.*, p. 15; Hecker, *Monatsschr. f. Geburtskunde*, vol. xxxiv., p. 307; Ballantyne, *loc. cit.*, p. 15; Seitz, *Zentralbl. f. Gynäk.* (1904), p. 875; Kamann, *Wiener klin. Rundschau* (1903), No. 46; Macdonald, *ref. Zentralbl. f. Gynäk.* (1903), p. 1220; Planchu and Richard, *Gazette des hôpitaux* (1902), No. 54; Ahlfeld, *loc. cit.*, p. 34; Richard, *Diss. Lyon* (1906); Mekler, *Diss. Lausanne* (1906); Fabre and Thevenot, *Lyon méd.* (Dec. 8, 1907), and *Revue de Chirurgie* (June, 1908); Finkelstein, *loc. cit.*, p. 101; Fabre and Bourrel, *Säuglingskrankheiten*; Commandeur, *Geb. Gesellsch. Lyon* (Feb. 19, 1908), *Zentralbl. f. Gynäk.* (1908), p. 1151; Stumpf, *loc. cit.*, p. 44; Birnbaum, *loc. cit.*, p. 101; Seitz, *loc. cit.*, p. 15, vol. iii., pt. 3, p. 87; Hofbauer, *Württemb. Zeitschr. Chirurgie u. Geburtskunde* (1851), No. 1; Demme and Eberhardt, *Handb. der Kinderkrankheiten*, vol. iii., pt. 2, p. 388 (1878); Clifford White, *Proc. Roy. Soc. Med., London* (March, 1912).

Thymus

The thymus gland only attains a considerable size during the fœtal period, and for the first two years of life. Later, it remains stationary at the size attained, and about the tenth year undergoes atrophy. There is very little to be said about the malformations and congenital affections of this gland. First of all, it must be mentioned that, as in cases of goitre, separate accessory portions of the gland may be present in the neighbourhood of the main gland.

Very rarely absence of the whole gland has been observed in normal children (see Hoffmann); more frequently this defect occurs in malformed children, especially in association with hemicephalus.[1] Hæmorrhages into the tissues of the gland occur not infrequently after difficult deliveries, and especially in asphyxiated children. Further, in congenital syphilis, multiple small abscesses are found in the tissue of the gland. As appears to be proved by Hoffmann in his monograph, these in reality are not true abscesses, but multiple cysts, the contents of which consist of a purulent fluid. Microscopically, epithelioid cells with large nuclei, and lymphoid cells with small nuclei, are found; and the wall of the spaces is lined with epithelium many layers in thickness. Congenital hypertrophy of the thymus, which has in some cases been held responsible for the sudden death of otherwise apparently healthy children, has lately come very much into prominence. These fatal cases have occurred, on the one hand, as the

[1] And also in acephalic twin fœtuses. Ballantyne has, however, been struck with the constancy of the thymus gland in the dissection of all kinds of monstrous fœtuses.

result of mechanical compression of the trachea,[1] including the vagus or the recurrent laryngeal nerves by the gland; and, on the other hand, have also occurred with normal-sized or only moderately large thymus glands.

In such cases, the so-called 'status thymicus' has very often been recognised (Paltauf) namely: on post-mortem examination, there is found, most commonly, an enlarged thymus, enlargement of the lymph glands, of the tonsils, of the intestinal lymph follicles, of the spleen, and narrowness of the aorta. On the question of the relationship of hypertrophy of the thymus to sudden death, and on the importance of the status lymphaticus, the reader should consult the detailed observations of Finkelstein. He is inclined to regard most of the cases of death, supposed to be due to the thymus, as the result of intestinal intoxication.

A case of this kind occurred a short time ago in the Göttingen Women's Clinic (Jr. No. 24634). The case was that of a child, without any sign of disease, delivered rapidly, and very lively at the time of its birth. Twelve hours later, the child died quite suddenly, without the possibility of any mistake or accident being the cause. A post-mortem examination showed the presence of status thymicus, hypertrophy of the thymus, hæmorrhages into the lungs and the pericardium, congestion of the organs, œdema of both legs, and of the scrotum, but nothing else of any moment.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Finkelstein, *loc. cit.*, p. 101; Tillmanns, *loc. cit.*, p. 34; Hoffmann, *loc. cit.*, p. 53, vol. xxiii.; Boxberger, *Diss. Kiel* (1903); Ahlfeld, *loc. cit.*, p. 24; Hauser, *Deutsche med. Wochenschr.* (1890), No. 28; Hennig, *Gerhardt, loc. cit.* (Nachtrag, 1893), vol. iii.

Larynx

Congenital malformations of the larynx are very uncommon. Single cartilages may be completely absent, or only imperfectly developed. (For congenital fistulæ in the region of the larynx, especially tracheal fistulæ, see p. 112.) The larynx, in exceptional cases, may be very small; and, associated with this, there is, in most instances, a very high-pitched voice. On the epiglottis, notches not uncommonly occur which may penetrate it so deeply that the condition may be described as a reduplication of the epiglottis. Congenital curvatures of the

[1] Most writers do not at the present time believe that death is caused by the pressure of the enlarged thymus, but agree with Paltauf in his view that the fatal syncope is due to some constitutional weakness associated with the status lymphaticus. In all these cases the thymus, which normally weighs about 6 grm., is above the normal size, and may weigh 10 to 50 grm.

cartilages, and the deviations of the epiglottis, are of practical interest, as in this way congenital stenoses may be brought about.

Congenital stenoses of the trachea are, as a rule, incompatible with the survival of the child. Congenital atresia of the larynx is still less common. A so-called congenital diaphragm of the larynx has been described in several cases; this consists of a membranous formation, the origin of which is to be found in epithelial adhesions in embryonic life. This membrane is most commonly stretched between the two vocal cords; less commonly, beneath them.[1] It is situated almost always in the region of the anterior commissure of the vocal cords, extends a variable distance backwards, and ends in a semi-lunar free margin which partly obstructs the glottis. The symptoms usually described—namely: hoarseness, aphonia, and asphyxia—may, however, be completely absent. These membranous formations very seldom occur on the posterior wall. The treatment consists, in well-marked cases, in endolaryngeal division of the membrane, or its excision, after preliminary splitting of the larynx. Other writers recommend bloodless dilatation with bougies. The ventricles of the larynx may be abnormally deep, and become enlarged so as to form air-sacs—laryngoceles (Kaufmann).[2] (For details regarding compression of the trachea by an enlarged thyroid or thymus, see p. 119.) Congenital tumours of the larynx are very rare, and are nearly always papillomata.

Congenital larynx stridor—inspiratory stridor—of the new-born (Laryngismus stridulus) is a congenital condition of not infrequent occurrence. In some cases, immediately after birth, in other cases not until a short time later, a loud-toned inspiratory stridor is observed with free expiration (Finkelstein). Finkelstein, very suitably, compares the noise to the clucking of a hen or the note of a hiccup.

The voice in this condition is clear, and the breathing shows only a slight inspiratory effort. In any case, a marked inspiratory movement in the pit of the stomach or in the costal arches is absent. Attacks of asphyxia are very uncommon, but slight dyspnœic attacks have been observed during sleep. The cause is obscure; but from the literature

[1] The rudiment of the larynx appears in the form of two lateral swellings which lie behind the fourth visceral pouches. At an early stage, after the separation of the trachea and œsophagus, the slit-like cavity between the swellings is for a time partly obliterated by the cohesion of the opposed epithelial surfaces. The persistence of this adhesion is no doubt the cause of the web-like membrane described. Another congenital defect is the incomplete union of the ala of the thyroid cartilage in front they are formed by the expansion and amalgamation of the skeletal bases of the fourth and fifth visceral arches, in the form of two lateral plates, which are united ventrally by an intermediate nodule of cartilage.

[2] These sacs are normally developed in anthropoids after birth, and attain in them considerable dimensions (Keith).

it is clear that a number of very varying conditions have been confused together. For example, the following have been put forward as *etiological factors*: enlargement of the thyroid, adenoids, and coryza. Lately—probably, correctly—the cause has been sought in certain peculiarities of the child's larynx (see Finkelstein), lateral narrowing of the epiglottis, rolling inwards of the same organ, or marked narrowing of the aryepiglottidean folds. To these must be added the marked softness of the child's larynx. As a result of this, a sucking together of the wall of the larynx occurs during respiration, and this produces the stridor. The above-mentioned anomalies are described by some authors as primary malformations.

The stridor is usually without any further result, and disappears spontaneously—most commonly, in the first or second year of life. Children affected with this condition are, according to Finkelstein, only exposed to danger when inflammatory lung affections occur. In such cases, tracheotomy may become necessary as a result of well-marked symptoms of stenosis. Without doubt, some of these cases of congenital laryngeal stridor, which have been noticed soon after birth, have been wrongly diagnosed and subjected to tracheotomy, ostensibly as cases of stenosis and congenital tumours. In all cases where there are symptoms of congenital stenosis of the larynx, it is well to follow the advice of Finkelstein, and to examine the base of the tongue and the entrance to the larynx with the finger; for congenital tumours, cysts of the thyro-glottidean duct, and dermoids occur which, clinically, very closely resemble stridor neonatorum in their symptoms.

Congenital hoarseness, from paralysis of the recurrent laryngeal nerve and from the pressure of a cor bovinum on the nerve, has been observed (Hauser).

Injuries to the larynx during delivery are uncommon. Interruption of its continuity with entrance of air into the cellular tissue of the neck, subcutaneous emphysema, attempts at intra-uterine inspiration, and œdema of the glottis have been observed, especially in forceps deliveries when the forceps have reached on to the neck.

CHAPTER VIII

Malformations and Congenital Diseases of the Thorax, Bronchi, Lungs, and Heart; including those of the Diaphragm

Bronchi and Lungs

In the bronchi, the following malformations have been observed: congenital diverticula, obliteration of the bronchi, bronchiectasis two eparterial lateral bronchi in place of three, enlargement of the bronchi (see Hoffmann and Ahlfeld).

Congenital malformations of the lungs are uncommon, and no special importance is to be attributed to them. Abnormal lobulation is the most common; thus the left lung may have three lobes, the right two or four, or even more. The accessory lobes are attached almost always by small bronchi to the larger lobes, and not infrequently they are situated at the base of the lung. In transposition of the viscera, the right lung has two lobes, and the left lung three.

Very rarely, congenital bronchiectasis (already alluded to) occurs —so-called cystic disease of the lungs. Cases of this kind have been described by Grawitz, Kaufmann, Meyer, Couvelaire, and others (see also Hoffmann). The lung—generally only one lung is affected —shows a reticulated spongy appearance. In place of the normal lung parenchyma, there are present smooth-walled cystic spaces with serous contents. Universal bronchiectasis and telangiectatic bronchiectasis should be distinguished from one another. In the first, the cysts open into a general cavity, the widened primary bronchus; in the second, a cystic dilatation of the bronchi of the third or fourth degree occurs, which may be so marked that the condition resembles an ovarian cyst (see Finkelstein). The cause is not very evident. According to some authors, it is a manifestation of congenital syphilis; others—for example, Kaufmann—regard it as due to a want of development (agenesia), or to a dilatation—the result of retention of secretion behind a stenosis of the bronchi, due to inflammation.[1]

[1] Heller has described an atelectatic form in which there is an abnormal growth of the bronchial cartilages, with remnants of unexpanded lung tissue; the lining epithelium being of the pavement type.

In a case described by Convelaire, difficulty of breathing occurred suddenly on the fifth day after birth, with marked cyanosis. On the sixth day, with an increase of these symptoms, death occurred. Post-mortem examination showed cystic degeneration of the middle lobe of the right lung. The air-passages leading thereto formed, according to the report of Fricklinger, an intercommunicating system from the hilus to the alveoli, of which very few remained normal. In their place, there were cystic spaces, into which canals of varying calibre opened. According to Convelaire, the condition was really one of an embryological malformation: the cause of which lay in an abnormal increase in the epithelium of the canals.

Congenital hernia of the lungs have also been described. Macé has recorded a case in which the hernia of the lung was placed close to the sternum. It protruded during expiration from the chest wall, and during inspiration formed a depression. The hernial aperture was the result of a defect in the thoracic wall. According to Ahlfeld, such a hernia is situated most commonly below the axilla, and, ætiologically, is due to an anomaly of the amnion.

Congenital hypophasia and maldevelopment of some of the lobes of a lung, or of the whole lung, is very rare;[1] and the children affected generally die of asphyxia at birth, as a result of these conditions. If they survive, the opposite lung undergoes compensatory hypertrophy and undertakes the function of the undeveloped lung. At times, a dropsy *e vacuo* occurs (see the very interesting case recorded by Finkelstein). It is evident that individuals with only one lung run a very severe risk when disease of this organ occurs—such as pneumonia or tubercle.

Among congenital diseases of the lungs, septic pneumonia and pleurisy must be mentioned, which may occur as the result of placental infection, through the aspiration of infected liquor amnii, or the septic secretions of the mother's genital tract—especially in cases of contracted pelves, premature rupture of the membranes, and inefficient antiseptic precautions during the progress of labour (see Finkelstein and Runge).[2]

Of a similar character is the so-called pneumonia alba,[3] met with in congenital syphilis. This condition is a very frequent one, and is characterised by the occurrence of a cellular exudation into the alveoli of the lungs. Besides this alveolar pneumonic form, interstitial

[1] Absence of both lungs appears only to occur in acardiac or acephalic twins.

[2] Septic pneumonia does not occur in these cases as often as might be expected, as a very abundant secretion usually takes place from the air-tubes in children born partially asphyxiated, which washes away the aspirated particles.

[3] This condition is found only in the lungs of stillborn, usually premature, children, or of those who have survived but a short time after birth.

infiltration and circumscribed gummata have also been observed. Numerous spirochætes are usually found in the cellular exudation described.

Lastly, I must mention so-called congenital atelectasis; in this condition, isolated portions of the lungs, after birth, remain in the fœtal condition of atelectasis—that is to say, the affected parts are devoid of air, and the alveoli shrunk together. On post-mortem examination, small scattered patches are seen mostly in the lower lobes of the lungs, posteriorly. These areas are dark coloured, and situated below the level of the surrounding portions of the lungs. The cut surfaces are flat, and not granular as in pneumonic processes. These portions of the lungs can be inflated with a tube from the bronchus. The cause of this atelectasis is to be found, usually, in asphyxia occurring during delivery. The children are born asphyxiated, and have not been sufficiently resuscitated, and especially have not been induced to cry loudly. Changes of this kind in the lungs are nearly always found in the post-mortem examination of premature children.[1] The common diagnosis of congenital debility (debilitas vitæ congenita) and the frequently resulting fatal issue are explained by these changes. A condition very frequently accompanying atelectasis is marked œdema (atelectatic œdema). Treatment consists in the vigorous treatment of asphyxia neonatorum, and in the appropriate management of the children when premature (see the detailed consideration of this subject by Runge).

Injuries of the thorax of the child during delivery have been described on several occasions. With strong labour pains, and especially with rough handling of the thorax in breech presentations, fracture of the sternum and of the ribs[2] has been observed. Further, after difficult extractions or versions, and also after spontaneous deliveries, effusions of blood may occur into the cavity of the thorax. They are usually the result of tearing of the delicate venous plexuses in the neighbourhood of the spine which, in asphyxiated children, are engorged with blood.

Injuries to the lungs are found, primarily, in cases where there are fractures of the ribs and clavicles; some authors throw the responsibility for these on Schultze's swinging method, when this has been employed. This is, however, strenuously denied by others (see Runge).

1 In such children, the condition is predisposed to by the soft and yielding nature of the thoracic walls, so that the movements of the thorax are incapable of bringing about the inflation of the undilated lungs; and especially is this the case when the air-passages are partly blocked with aspirated particles.

2 Stumpf has recorded a case of fracture of all the true ribs, and of the horizontal ramus of the pubis on the right side, after a difficult version in an oblique presentation.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; M. Runge, *Krankheiten der ersten Lebenstage*, 1906; Ahlfeld, *loc. cit.*, p. 34; Hoffmann, *loc. cit.*, p. 53, vol. xiii.; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Finkelstein, *loc. cit.*, p. 101; Macé, *Geburtsh. Ges. Paris* (June 16, 1905), *ref. Zentralbl.* (1906), p. 531; Gouiron and Couvelaire, *Geburtsh. Ges. Paris*, *ref. Zentralbl.* (1904), pp. 984 and 1174; also *Monatsschr. f. Kinderheilk.* (1907), vol. vi., p. 123; Birnbaum, *loc. cit.*, p. 101; Fritsch, *loc. cit.*, p. 44; Stumpf, *loc. cit.*, p. 44; Smith, 'Larynx Stridor,' *Brit. Med. Journ.* (Aug. 20, 1907); Mathieu, 'Larynx Stridor,' *Diss. Nancy* (1903); Boulard, 'Larynx Stridor,' *Diss. Paris* (1904); Semon and Watson Williams, *Clifford Allbutt, loc. cit.*, p. 89, vol. iv., pt. 2, p. 185; Keith, *loc. cit.*, p. 96.

The Heart

The heart, as the principal organ of the circulatory system, is not uncommonly the seat of congenital malformations. E. Lewy found among 4800 children 137—that is, 2·8 per cent.—with congenital abnormalities of the heart. Further details may be found in the very excellent contribution by Vierordt, in 'Nothnagel's Handbook of Pathology and Treatment,' which gives a good account of the congenital diseases of the heart.

While, in later life, disease of the heart mainly affects the left half, as a rule the right half is the seat of congenital affections. As Fischl rightly points out, the cause is to be found in the fact that while, on the one hand, fœtuses with faults of development in the left heart die early, on the other hand inflammatory processes are commoner in the right heart, as more strain is placed upon this half owing to the special conditions of the fœtal circulation.[1]

Congenital malformations of the heart may occur at various stages of its development. The most serious consequences ensue when the malformation becomes established at a very early stage in the development of the heart, as then the primary abnormality may give rise to other defects (Keller). On the other hand, anomalies of a less serious nature can also occur when the development of the heart is nearly completed.[2]

As regards the ætiology of congenital diseases of the heart, we must (according to Vierordt) distinguish special and general causes. If we consider the general causes, the frequent association of other malformations

[1] According to Rauchfuss, this is only so in association with malformations; otherwise, the left heart is as frequently affected.

[2] It must be remembered that the development of the heart at the end of the second month of fœtal life is as complete as at birth; but that after birth, in association with the changes in the circulation occurring at that time, certain post-natal changes take place—namely: the closure of the foramen ovale and of the ductus arteriosus—and defects in these may be the cause of malformations. There are, therefore, two periods during which congenital cardiac malformations may originate: in the two first months of intra-uterine life, or after birth.

with those of the heart at once attracts our attention: for example—acephalus, hare-lip, cleft palate, cyclops, henicephalus, transposition of the viscera, fissures of the abdominal wall, defects of the diaphragm, anomalies of the intestinal tract, horse-shoe kidney, cystic kidneys, umbilical hernia, and anomalies of the genital organs. We are compelled, with Vierordt, to regard this coincidence as not merely accidental, for no less than 10 per cent. of all malformations of the heart are associated with other malformations.[1]

Heredity also plays a part; and Vierordt has collected a considerable amount of material, bearing on this, from the literature.[2]

The following are also given as ætiological factors: venereal disease, blood relationship of the parents, tubercle, and typhus; while even chills and fright on the part of the mother have been put forward as causal factors. Vierordt considers these, together with fœtal endocarditis and arrests of development, as predisposing causes.

Fœtal endocarditis undoubtedly plays a very important part in the production of congenital anomalies of the heart, as Rokitansky has pointed out. Congenital anomalies of the heart, therefore, may be divided into two groups: to the first group belong all those anomalies which are to be considered as due to errors of development; to the second group, those produced by fœtal endocarditis.[3]

So sharp a division cannot, however, be made to apply to all cases; since it is a well-known fact that fœtal endocarditis can readily develop in the walls of a malformed heart. On the question of the mutual dependence of the various malformations of the heart, I must refer the reader to the details on the subject in Vierordt's monograph.

As regards the frequency of congenital malformations in the two sexes, it is clear from all the statistics that the male sex is in a majority; whereas in other malformations, as is well-known, the female sex—especially in the case of double monsters—is in a majority.[4]

[1] In fourteen of twenty-three malformed fœtuses examined by Keith, the heart was the seat of some malformation.

[2] De la Campe demonstrated with the Röntgen rays a patent condition of the ductus Botalli in six members of the same family.

[3] As the fœtal heart is as well developed at the end of the second month as it is at full term, it is difficult to conceive how fœtal endocarditis can play any large part in the production of cardiac malformations. If it, however, occurs early enough, we may suppose that it might interfere, to some extent, with the final stages of development; and that at a later date its occurrence might predispose to, or even bring about, anomalies in the changes in the heart and in the circulation which should occur after birth. The influence of heredity, the association of other malformations with those of the heart, and the fact that in the early stages of embryonic life the heart is on the surface of the body, are all in favour of the view that its malformations are due to defects of development, and possibly owe their origin—in, at any rate, some cases—to some external influence.

[4] Vierordt collected 383 cases, and found 248 male and 135 female.

In the following review of congenital anomalies of the heart, I have adopted, on the whole, the division given by Fischl—based on the works of Rokitansky, Bamberger, Rauchfuss, Hochsinger, Flatow, and Pott.

1. Acardia—Absence of the Heart

Complete absence of the heart, or the presence of only functionless rudiments, is found only in monsters—primarily, in twins—from a single ovum. In this case, the fœtus is either well developed or more or less maldeveloped. Most frequently, its head is lacking (see the chapter on Acardiaci, p. 351). The heart of the sound twin carries on the circulation in the other malformed twin as well as its own.

2. Abnormal Situation of the Heart

(*a*) *Dextrocardia.*—Transposition of the heart to the right, so that its impulse is to be felt to the right of the sternum, occurs in cases of transposition of the viscera (Situs inversus viscerum totalis regularis): when the liver, the spleen, and the stomach are also transferred to the opposite side—very rarely, however, as a true dextrocardia. According to Vierordt, a case of undoubted true dextrocardia has not yet been observed—or, at any rate, has not been confirmed by an autopsy.[1]

(*b*) *Ectopia.*—Most frequently the heart protrudes through a congenital defect of the sternum, or of the thoracic wall (Ectopia cordis pectoralis). In such cases, it is either surrounded by the pericardium, or is completely uncovered. Such malformed children are not capable of living. In all the known cases, the children have died, usually in a few hours; or, very rarely, on the second day after birth. Nevertheless, an attempt should always be made to cover the heart with skin-flaps by a rapidly carried out operation (Ahlfeld). Lannelongue, according to Fischl, cured a defect (the size of a shilling-piece) in a female child six days old, through which the apex of the heart protruded, by freshening the edges, incision, and suture. The condition was probably the result of ulceration, and not a congenital defect of the chest wall. The wound healed, by second intention, in twenty days. Greiffenberg has recorded briefly a very interesting case of ectopia cordis. The

[1] An explanation of this occurrence (put forward by v. Baer) is that there is inversion of the normal cardiac curve, and that this is due to an abnormal position of the embryo on the embryonic area. Dareste suggested that there was unequal growth of the two halves of the heart; and that while, normally, the right half is the larger, in these cases the left half is the larger, and the curve is inverted. Whatever the cause may be, it must come into play at a very early period of development.

child lived five hours after birth. The mother suffered from atresia ani and anus vulvaris. Hydramnios was also present, to the extent of five litres. In the child, the following condition was found: Ascites, with marked œdema of the face and neck, which had pressed upon the trachea and given rise to difficulty in breathing. At the lower end of the united costal arches, the protruded heart hung, attached to the vessels. On its posterior aspect, a Simonart's ligament passed from the auriculo-ventricular junction to the umbilical cord, just beyond the navel. A single dilated and hypertrophied ventricle was present in the heart, and two auricles, imperfectly separated from one another, and also from the single ventricle. The vena cava superior, the pulmonary artery and veins, and the aorta were present; but there was nothing to be seen of the vena cava inferior. The condition was, therefore, one of arrested development, which had occurred during the second week of embryonic life. The amnion had acquired an adhesion to the heart, and interfered with the closure of the thoracic cavity. As a result of the drag produced by the amniotic band, the heart was subsequently drawn out of the closing thorax.[1]

FIG. 31.—ECTOPIA CORDIS WITH FISSURE OF THE STERNUM. (Specimen from the collection of the Göttingen Institute of Anatomy.)

[1] This case is strong evidence in favour of the view that ectopia cordis is brought about by the influence of the amnion, acting in the very early stages of development, when the heart is exposed on the surface of the body. The rarity of the condition may be explained by the comparatively short time during which the heart is exposed to such influences.

Rieländer has described a similar case; here also there was an amniotic fold which had brought about ectopia of the heart; anomalies of the large vessels (aorta, etc.) were also present.[1]

In ectopia cordis abdominalis, the heart—as a result of defect or fissure formation in the diaphragm—is placed in the abdominal cavity. In the very rare case recorded by Arndt, the heart was displaced into a coexisting umbilical hernia. In the case described by Koller and Aeby, there was a congenital diverticulum of the heart in a hernia of the umbilical cord.

In ectopia cordis cervicalis, the heart is displaced towards the palate (Kaufmann and Ahlfeld). Observations on the heart,[2] in these cases of ectopia cordis, have given us very valuable information as to the functions of the heart—more especially as to its changes in size.

3. Abnormal Form and Size of the Heart

To this class belong congenital hypoplasia (smallness of the heart) and congenital hypertrophy of the heart. Hypoplasia of the heart and of the large vessels predisposes, according to some authors, to chlorosis and tubercle (Virchow).

Congenital hypertrophy (Cor bovinum) is found frequently in cases of patency of the fœtal blood-channels, in hypertrophy of the thymus, in congenital stenosis and atresia (with secondary compensatory hypertrophy of the heart), in ectopia cordis, and acardiacus. A congenital idiopathic cor bovinum occurs also, although very rarely. New-born children, affected with this condition, exhibit feebleness, rapid breathing, pallor, meteorism, and engorgement of the liver and spleen (Finkelstein). According to Finkelstein, these children die, at the latest, in the third year of life.

To variations in form belong the cylindrical heart, the round heart, and the heart bifurcated at the apex.[3]

Duplicity of the heart occurs chiefly in double monsters, where the

[1] Specimen No. 826, Univ. Coll. Hosp. Med. School Museum, is a good example of ectopia cordis. In a fœtus between the fourth and fifth month of utero-gestation, there is an oval opening in the abdominal parietes, three-quarters of an inch in length, through which the stomach, liver, and some of the intestines, protrude. Immediately above, and to the left of these, is the heart, with its apex pointing to the left shoulder; a narrow band of membrane separating it from the liver. The aorta passes into the chest by a small round opening; the pulmonary artery cannot be seen externally.

[2] This represents the persistence of the original subpharyngeal position of the heart (which in mammals is lost, as the result of the development of the neck), in the second month of fœtal life, forcing forwards the head and pharyngeal region (Keith).

[3] Like the heart of a dugong.

doubling affects the upper half of the body. The two hearts lie either separate, each in its own pericardium, or are contained in the same pericardial sac. A double heart in a single individual is very rare[1] (see Ahlfeld).

4. Abnormal Disposition of the Various Parts of the Heart

Under this heading are included abnormalities of the septa, of the trunks of the vessels, of the openings, and of the valves.

(1) If the septum is completely absent, only one ventricle and one auricle are present (cor biloculare). If the septum is present only in the region of the auricle or of the ventricle, then the condition is distinguished as a cor triloculare. Most commonly, two auricles and one ventricle are present. Further, the left auricle may communicate with the right ventricle by an obliquely placed opening, and in a similar manner the right auricle with the left ventricle. Finally, with gross defects of the septum, abnormalities in the origins of the large vessels may also occur. Thus the inferior vena cava may open into the left auricle instead of into the right, and the aorta may take origin from the right side of the heart, as in a case observed by me in the Göttingen Women's Clinic.[2]

(2) Abnormalities of the trunks of the vessels are mainly due to defective development or faulty insertion of the septum. The most frequent anomalies are :—

(*a*) The pulmonary artery may be completely absent, or it may be abnormally narrow at its site of origin, and become widened for the first time after its union with the ductus Botalli, which conveys blood to it from the aorta. Blood which is held back as a result of a pulmonary stenosis can also flow into the abnormally inserted aorta, as a result of a defect in the inter-ventricular septum (see p. 142). Further, the aorta may take over completely the rôle of the pulmonary artery, when only one chamber of the heart is present.[3]

[1] A suggested explanation of such an occurrence is the want of union of the originally double heart-tubes, which begin as the continuation of a vitelline vein.

[2] The reader must remember that malformations of the auricles are rendered very complex by the presence of the foramen ovale (which fails to close, in 25 per cent. of people, within the first year); and the fact that the inter-auricular septum is made up of three portions—namely, two endocardial cushions which fuse, and the septum primum and the septum secundum. In the same way, although normally it contains no persistent foramen, the inter-ventricular septum is made up of several parts—the ventricular septum proper—and the endocardial cushions formed in the auricular canal, and the bulbus arteriosus. Between these three, is found the inter-ventricular foramen, the abnormal persistence of which is one of the commonest of all cardiac malformations.

[3] The stenosis may also affect the bulbus arteriosus, and, according to Keith, the majority of cases of pulmonary stenosis are in reality due to arrested development of this part of the heart, which, he considers, takes part in the formation of the infundibulum of the right ventricle.

(*b*) The same conditions may obtain in the aorta as those just described in the case of the pulmonary artery.

(*c*) The fœtal type of the distribution of the blood may persist in extra-uterine life, so that the aorta supplies the upper half of the body, and the pulmonary artery, through the ductus Botalli, the lower half of the body.

(*d*) Transposition of the large vessels may occur: so that the aorta takes origin from the right ventricle, the pulmonary artery from the left ventricle. This transposition can come about as the result of an abnormal disposition of the intervascular partition, which divides the common arterial trunk into aorta and pulmonary artery.[1]

(*e*) Both vessels may take origin from one chamber.

(*f*) The aorta passes along the defective ventricular septum and takes origin in two equal or unequal portions, with one division springing from the left ventricle, the other from the right.

(*g*) The bulbus arteriosus may become enormously enlarged, and form a third ventricle.[2]

(*h*) The ductus Botalli may remain patent, especially in cases of stenosis and atresia, so that by this means the resulting deficiency of blood, distally, is made good.

(*i*) The aorta may be obliterated at the opening of the ductus Botalli (Rokitansky and Ranchfuss). In well-marked cases, when the ductus Botalli remains open, the fœtal type of circulation may persist at the same time.

(*k*) A transposition of the openings of the veins in the auricles may occur, while the pulmonary veins and the venæ cavæ may also open into one auricle.

(3) Congenital abnormalities of the openings and valves :—

(*a*) Stenosis of the pulmonary artery or of the aorta. Stenosis of the pulmonary artery is the most frequent cause of congenital cyanosis. Usually, in such cases, the foramen ovale remains open, or there is imperfect development of the ventricular septum.

(*b*) As a result of alterations in the valves, insufficiency, atresia, or stenosis may result. The tricuspid and semilunar valves may be completely absent in certain conditions.

[1] Any failure or abnormality in the bulbus arteriosus coming into its due relation with the anterior part of the inter-ventricular septum will be the direct cause of the transposition of these vessels. It must be remembered that the bulbus arteriosus originally communicates with the right ventricle only, and at one stage both the aorta and pulmonary artery take origin from the right ventricle.

[2] In the fish, the bulbus arteriosus forms one of the cavities of the four-chambered heart. Keith maintains that in the human heart, the musculature of the bulbus arteriosus becomes limited to the pulmonary part of the aorta, and ultimately fuses with the infundibulum of the right ventricle.

(*c*) The Eustachian valve (the valve of the foramen ovale) may be completely absent, prematurely closed, or imperfectly developed.

In his monograph Vierordt gives another classification of the congenital malformations of the heart, arranged from the point of view of their anatomical development.

With reference to their prognosis, these malformations of the heart may, according to Bamberger, be divided into three groups.

The first group includes those cardiac malformations which render life impossible. To this group belong the monstrosities, ectopia cordis, with fissure of the sternum and defect of the skin, the heart consisting only of a single chamber, and especially cases of transposition of the large vessels. Children with such malformations cannot respond to the demands which are placed upon the heart after the fœtal circulation has been altered. They perish during pregnancy, during delivery, or soon after delivery, with the symptoms of increasing heart weakness or asphyxia. In cases of transposition of the large vessels (see the case from the Göttingen Women's Clinic)—a very rare condition—very marked cyanosis occurs. The children, when crying, appear almost blue-black; the cyanosis being absent in only very exceptional cases. The temperature, corresponding to the marked cyanosis, is in most cases subnormal. Hæmorrhages into the skin (from the nose, mouth, and intestine), into the brain, and from the lungs, have also been observed; together with dyspnœa, accompanied by attacks of suffocation, Cheyne Stokes's respiration, atelectasis of the lungs, and convulsions. The course of the circulation of the blood in cases of true transposition is very curious. The aorta conveys the venous blood to the body, and the pulmonary artery the arterial blood to the lungs. The body contains only a minimal amount of arterialised blood, which, as a result of the intercommunication of the terminal branches of the pulmonary arteries with the bronchial arteries and with the bronchial veins, passes over into the latter.

Physical examination does not show any uniform result in the recorded cases. There is, however, almost always some increase in the cardiac dullness, and murmurs are sometimes present. The prognosis in transposition of the vessels, as has been mentioned, is bad. According to a table given by Taruffi and Vierordt, most of the children (77·3 per cent.) die in the first year of life, and the majority of these die in the course of the first six months. Only very few reach a later age.

To the second group belong, according to Bamberger, those varieties of malformation in which the children may remain alive, and—at any rate, to begin with—may develop more or less normally. In the course of years such disturbances of the circulation occur, however, that the children die at latest about the commencement of the second

decennium. Under this group are included congenital narrowings at the origin of the pulmonary artery or aorta, large communications between the ventricles or the auricles, or of one ventricle with its corresponding auricle, the origin of the aorta from both ventricles, and patency of the ductus Botalli (see Group 3).

To the third group belong the unimportant anomalies in which the circulation of the blood is not altered, and, as a result, the affected individuals are not hindered in their development. This includes, for example, alterations in the external form of the heart, the cylindrical heart, the round heart, or the heart bifid at its apex. Further, dextrocardia with coexistent situs inversus, patency of the foramen ovale, and, under certain conditions, defects in the ventricular septum, and patency of the ductus Botalli.

As regards the time of the first appearance of symptoms, in the majority of cases they make their appearance in connection with birth. If the disturbances of the circulation increase only gradually, or if the symptoms of the congenital heart disease become marked only after some years, then they are to be attributed to the persistence of a fœtal endocarditis, or to its revival after a long period of latency.

The best known and most conspicuous symptom is congenital cyanosis (morbus cœruleus or *maladie bleue*). The cyanosis shows itself most strikingly in those cases in which there is marked passive congestion of the blood in the veins of the body. It occurs most frequently on the lips, upper eyelids, ears, hands, feet, alæ of the nose, pharynx, glans penis, and the folds of the vulva. All possible shades of colour may be present, from a light blue to a deep, bluish black. The cyanosis is markedly increased by exertion—namely: by crying, coughing, vomiting, and laughing—and, further, by excitement and alterations in the temperature. After complete disappearance of these excitants, it soon diminishes again to its normal degree.

In rare cases, where passive congestion of the peripheral veins is not present, cyanosis may also be absent. In these cases, in place of cyanosis, the face may present a palish, grey colour.

At one time the view was held that cyanosis was the result of an admixture of arterial and venous blood. This view is at the present day given up as erroneous. Thus pulmonary stenosis and defects of the ventricle may be present without any accompanying cyanosis. If the former view were correct, as Vierordt points out, the embryo, which always has mixed blood in it, would constantly exhibit cyanosis. The true cause of the cyanosis lies rather in a deficient decarbonisation of the blood in the lungs, with or without stasis in the peripheral venous system (Fischl). This condition would be produced by some obstruction to the flow of blood in the left ventricle, with consecutive

engorgement in the pulmonary veins; by some stenosis in the right side of the heart, or a persistent patency of the ductus Botalli.

The cyanosis can, at times, be simulated in new-born children through unnecessary tight swaddling. Vierordt has recorded an interesting case, from the literature, of the simulation of cyanosis (Rayner's case). Rayner observed in several new-born children, simultaneously, a cyanosis lasting some three or four days, but otherwise harmless, which, apparently, was due to absorption of aniline chloride, contained in the ink which had been used for marking the swaddling clothes.

A chemical analysis of the blood, in cases of cyanosis, shows it to contain a considerable amount of carbonic acid; but, curiously enough, almost a normal amount of oxygen. The blood is remarkably concentrated, the specific gravity increased, as is also the number of the red blood-cells and the amount of hæmoglobin.[1] The alkalinity is about the normal. All these changes are, however, by no means characteristic of cyanosis (for further details on these points, see Vierordt). An accompanying or following symptom of the cyanosis is a decrease of the body temperature, often by 1·5° to 3·5°; as a result of this, the peripheral parts of the body, especially the arms and legs, feel damp and cold.

The maintenance of the body temperature, in cases of congenital heart disease, is of some prognostic value. If the temperature of the new-born child gradually rises to the normal after birth, the prognosis is good, and vice versa. The temperature may fall as low as 29° C. (84° F.), or even lower. A more or less marked dyspnœa often accompanies the cyanosis, especially on exertion. The dyspnœa may be so severe as to lead to attacks of suffocation, and may even be accompanied by epileptiform seizures. Besides the dyspnœa, palpitations of the heart occur which may give rise to very disagreeable feelings of oppression. In the course of time, children who survive develop the so-called club-fingers—the well-known knob-like swelling of the ungual phalanges of the fingers with claw-like bent nails.

The condition of children with congenital heart disease, Keller pictures in a very clear manner. 'Such children,' he says, 'lie very quiet and sleep a great deal; the voice is hoarse and feeble, their movements are not as lively as usual, but are slow and phlegmatic. The action of sucking both at the breast and with the bottle takes place very slowly, and with long pauses. One has the impression that these children are suffering from want of air and that they become tired very quickly.' Later on, the dyspnœic and asthmatic attacks, which have been

[1] The cyanosis is, no doubt, mainly due to imperfect aeration of the blood in the lungs and venous congestion, when it is increased by exertion; but it may be due partly to hyperglobinæmia, when it is constant.

described, become evident, and a disposition to bleeding from nose, and hæmoptysis occurs with hæmorrhages from other parts of the body—for instance, from the gums; catarrh of the air passages (especially in wet weather and with a low barometer), tubercle of the lungs, affections of the stomach and intestinal tract. The bad prognosis, as regards the life of these children, is evident from all these conditions. Besides the failure of development of their body, they are generally thin, with very little fat; the mind also fails to develop; even idiocy having been observed. The eruption of the teeth may be delayed, and also the onset of puberty.

I have already dealt with the expectation of life of children with congenital heart disease, and, as I have remarked, the majority of them die in the first year of life, while only a very few survive the first decennium or to a still later period.[1] Death ensues from the complications described, or from the secondary phenomena which result, especially failure of compensation, dropsy and cardiac weakness, tubercle of the lungs—especially in those cases of heart disease in which the blood supply to the lungs is hindered or rendered more difficult: for example, in pulmonary stenosis, and insufficiency and stenosis of the tricuspid valve.

Physical examination in cases of congenital heart disease is especially difficult. Too much stress must not be laid upon enlargement of the heart as this, although usually present in such cases, even without any other malformation, may be entirely absent, owing to a concomitant defect of development. More important than percussion is auscultation, especially in new-born children. At this time, the accidental or hæmic murmurs, which are so frequently present later on, are absent. According to Keller, the murmurs of congenital heart disease are nearly always systolic in time, and heard best at the base, not at the apex. The changes found on percussion are, according to him, slight or wanting; while the heart's impulse is not strengthened but rather weakened.

Fischl, as the result of his experiences at the Prague Findelanstalt, expresses himself very sceptically as to the possibility of a clinical differential diagnosis between the different forms of heart disease. The children in question, show all the symptoms of the so-called 'blue disease,' more or less marked increase of the area of cardiac dullness, sometimes sounds, sometimes murmurs at the different

[1] Abbott, from an analysis of 400 cases of congenital heart disease, concludes 'that the duration of life is relatively longer in uncomplicated defects of the interauricular septum in patent ductus arteriosus, coarctation of the aorta, and in pulmonary stenosis with closed interventricular septum; while in pulmonary stenosis, with defect of the septa, the duration of life is much shorter (quoted by Humphry, *Clifford Allbutt's System of Medicine*, vol. vi., p. 307 (1909).

orifices. It is possible in such cases, no doubt, to determine the presence of some variety of congenital heart disease, but not the actual kind. Fiehl hopes to derive some assistance from radiography, which has recently been used in the diagnosis of heart diseases.

On the diagnosis of fœtal heart disease intra-partum, Hölme makes several observations of importance. On the basis of a case of heart disease carefully observed and diagnosed intra-partum, he gives the following as diagnostic factors of importance: The rough character of a murmur not synchronous with the mother's pulse, its constancy as regards duration and strength, the continuing absence most commonly of both heart-sounds, less commonly of one only (the first), and the wide distribution of the murmur over the abdomen, with its greatest intensity corresponding to the region of the fœtal heart. Hölme maintains (quite rightly) that one should not, after having made a certain diagnosis of the presence of a fœtal heart disease, yield to the temptation to undertake any procedure which might entail any risk to the life of the mother, since experience has shown that the duration of the life of the child and its vitality are very limited in these cases.

A case in point has been recorded by Wetterill and Hall. The murmur heard during pregnancy was also heard in the new-born child, and a diagnosis of pulmonary stenosis arrived at.

Démelin and Gondert, on the ground of an analogous case observed during pregnancy, and especially on account of the greatly reduced frequency of the fœtal heart-rate—namely, fifty per minute—made the diagnosis of an intraventricular communication. The post-mortem examination confirmed the diagnosis; while transposition of the large vessels and a patent ductus Botalli was also found.

The treatment of congenital heart disease is not a very thankful task and corresponds, on the whole, to the treatment of chronic heart disease occurring in later life. Rest of the mind, and especially of the body, is essential. As the children grow older, they will very soon notice how very injurious exertion is for them. For new-born children, careful feeding is of the utmost importance, and the furnishing of a sufficient amount of warmth if necessary by means of a couveuse. On account of the danger of complicating stomach and intestinal diseases, the feeding, the preparation of the milk, and especially the care of the utensils necessary for the feeding—namely: the bottles and tubes—must be particularly carefully carried out. Further, to avoid the occurrence of affections of the upper air passages, so dangerous to children with heart disease, they must be kept very warm. Warm clothing, especially under-clothing (a flannel gown) is most necessary. Care must be taken to supply pure air; and of great importance is a rational care of the skin and a sufficient but mild hydrotherapy. Any constipation must be prevented, or efficiently

treated ; because of the straining associated therewith. On account of the danger of infection, children with pulmonary stenosis should be kept as far as possible from all forms of tubercle. In feeding, alcoholic and stimulating drinks should be avoided, and they should only be employed in conditions of collapse. In cases with failure of compensation, or conditions of extreme weakness, camphor, strophanthus, and digitalis may be tried. In very marked cyanosis an attempt may be made to improve the condition by the inhalation of oxygen.

As regards the obstetrical importance of congenital heart disease, we may remark that premature labour in such cases, even when the condition is complicated and severe, is rare. The conditions of the fœtal circulation apparently do not evoke any lasting damage, as the obstruction to the circulation, with its sequelæ, first begins at the commencement of extra-uterine life.

In many cases such children are born asphyxiated ; more frequently, however, the asphyxia first shows itself after birth. Treatment of the asphyxia, in such cases, can rarely be of much avail. (On the obstetrical importance of acardiac monsters, see p. 359.)

In obstetrical literature there is not much to be found on the subject of congenital heart disease. Two interesting and similar cases have lately been described by Lemaire. The first case was that of a child which died with increasing cyanosis on the eighteenth day after birth. At the post-mortem examination there was found doubling of the ductus Botalli, marked hypertrophy of the left side of the heart, the aorta arising from the right ventricle, and a communication between the ventricles and also between the auricles. The second case was a similar one. Bonchacourt and Condret describe two cases, one of which presented the following abnormalities : one ventricle with two vessels arising from it, transposition of the heart and liver, and absence of the spleen. The second fœtus had one normal and one poorly developed auricle and ventricle, and occlusion and atrophy of the pulmonary artery was also present. Devraigne demonstrated a case in which the ductus Botalli had persisted. The child died six hours after birth. Winkler has described a very interesting case. The case was that of a four-weeks-old child whose mother suffered from severe pulmonary tuberculosis, and who had acquired gonorrhœa during her pregnancy. The heart was almost entirely formed of the enormously hypertrophied right ventricle, and weighed 52 grm. The left ventricle formed only a small projection, from the left side of which there passed transversely across it a band, 2 mm. in thickness, forming a permanent obstacle to its growth. Pulstinger has described a unique case. In a four-weeks-old child, which had from birth suffered from asthma and cyanosis, there was found (at the autopsy) a

large defect of the septum, a rudimentary condition of the aorta which did not communicate with the left ventricle, and replacement of this by the pulmonary artery. Eggel has recorded two interesting cases. The first was that of a full-term, well-developed male child which became rapidly cyanosed and died on the seventh day after delivery. The somewhat enlarged heart consisted of only one ventricle and one auricle, between which was situated a tricuspid valve. The four pulmonary veins, the vena cava inferior and two superior venæ cavæ opened into the auricle, while the aorta sprang from the ventricle and gave off the two pulmonary arteries.

The second case was that of a strong boy, who died of asphyxia five hours after birth. The heart showed a faulty arrangement of the large vessels. The aorta sprang from the right ventricle, and joined the pulmonary artery, which sprang from the left ventricle, by means of a well-developed ductus Botalli. There was a defect in the ventricular septum. Von. Konstantinowitsch described, a short time ago, a case of a bilocular heart; the new-born child collapsed, suddenly, two days after birth, and died with symptoms of dyspnœa. Besides other cardiac malformations, a defect of the ventricular and auricular septa, a defect of the openings of the veins of the right side of the heart, and atresia of the opening of the aorta were present. Lastly, reference must be made to the case reported by Arnold of almost complete defect of the septum. The affected individual was some forty-two years old, and died as the result of an attack of pneumonia with heart symptoms. Since his earliest youth, cyanosis, dyspnœa, and clubbed fingers had been present.

In association with these observations, I must mention a case, reported by Phänomenoff, of a fœtal aortic aneurism which occurred between the origin of the renal arteries and the common iliac arteries, and which led to difficulty in delivery. The case must be quite unique in the literature on this subject.

In the Göttingen Clinic, during the last twenty years, among 4200 births, four cases of congenital heart disease have been observed.

In the first case (Jr. No. 17787), a circular defect, of about 3 cm. in diameter, was present in the ventricular septum. The aorta and pulmonary artery were normal. As a result of the defect, the valves were not separated, the anterior mitral valve being united with the anterior tricuspid, and the posterior mitral with the posterior tricuspid. The auricular septum was completely formed, and a foramen ovale was present. The child weighed at its birth—which took place spontaneously with a very short second stage—3070 grm., and was 47 cm. long. It cried immediately on being percussed. Very soon, however, a marked bluish discoloration of the whole body, and a jerky rattling breathing became evident. After about three hours' treatment with baths, hot and cold

alternately, Schultze's swinging method, and other measures, the marked cyanosis was apparently got rid of. The child died on the fifth day of sepsis, arising from some ulcers on the soft palate, as a result of which metastatic abscesses formed.

In the second case, Jr. No. 16298, the condition was one of obliteration of the right arterial opening. The child, which weighed 2110 grm. and measured 50 cm. in length, developed (soon after birth in a vertex presentation, and with a second stage lasting twenty-five hours) marked cyanosis with superficial breathing and inability to suck. The next morning, the condition remained the same; but the temperature was 34·8° C. in a warm bath. Brandy and sugar-and-water were administered; but death occurred that afternoon. The post-mortem examination, undertaken fifty-three hours after birth, showed marked cyanosis, especially in the region of the forehead and ears. The heart was enormously enlarged, the enlargement being mainly due to the marked dilatation of the right auricle. As the result of the enlargement, the remainder of the heart was displaced entirely into the left half of the thorax. Both ventricles also appeared to be considerably thinned. The apex was rounded, and formed mainly by the right ventricle. The right auricle was filled up with a dark-red clot, about the size of a hen's egg, and some fluid blood. The right ventricle, when laid open, appeared somewhat dilated. The right arterial opening was completely closed, and was situated at the level at which the pulmonary valves are generally placed. Beyond the closed portion was found the narrow pulmonary artery in which there was blood-clot. The foramen ovale and ductus arteriosus were wide open. The wall of the right ventricle had an average thickness of 2 mm.; and that of the right auricle appeared thickened relatively to its enormous dilatation. Further than this, there was venous hyperæmia of all the mucous membranes, of the spleen and of the kidneys, and hæmorrhages into the pleura and the capsule of the liver.

In the third case, Jr. No. 15424, the condition was one of transposition of the large vessels. The child, weighing 3240 grm. and measuring 50 cm., was born at full term in a footling presentation. The pelvis was generally contracted and liberation of the arms with extraction of the head was required. A depression was present on the left parietal bone, and the child was born deeply asphyxiated. Fifteen to twenty hours after birth, its condition became worse: it refused its food, and died twenty-four hours after delivery. An autopsy showed the following condition: the vena cava superior and inferior opened into the right auricle; from the right ventricle an arterial trunk took origin, at the commencement of which, corresponding to the position of the two porterior valves, were the origins of two coronary vessels, while the vessel itself continued as the aorta and gave off on the right side the carotid

and subclavian arteries; from the left side a carotid and subclavian artery also took origin; into the left auricle opened the pulmonary veins; and from the left ventricle sprang the pulmonary artery with its branches and the ductus Botalli. There was present on the right and left sides a valve with two and three flaps respectively.

In the fourth case, Jr. No. 19517, a healthy male child weighing 3230 grm. and measuring 52 cm. in length, was born spontaneously after a second stage lasting only half an hour. The child cried strongly immediately after its birth, showed on the following day no anomalies, and took its food well. Cyanosis was observed for the first time thirty hours after birth; this, however, disappeared, and reappeared several times in the course of the day. In the evening, the cyanosis increased, food was refused, and convulsions occurred, accompanied with a shrill cry; forcible inspiratory movements were also observed. The child died the following morning fifty-two hours after birth. During life, a marked systolic murmur was heard on auscultation over almost the whole cardiac area. This sign, together with the clinical appearances, suggested the diagnosis of some congenital malformation of the heart. Post-mortem examination showed that the ductus Botalli was widened and formed an open channel about the thickness of a pencil (3 mm. in diameter). No other malformation of the heart could be discovered. The wall of the right ventricle was, however, especially thick (5 to 6 mm.). On the mitral and tricuspid valves were small hæmatomata, there were also subpericardial hæmorrhages and hæmorrhages into the thymus with marked congestion of all the organs of the abdomen.

Literature

Kaufmann, *loc. cit.*, p. 24; Biedert and Fischl, *loc. cit.*, p. 34; Vierordt, *loc. cit.*, p. 53, vol. xv.; Keller, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, pp. 24 and 34; Lemaire, *Geb. Gesellsch.* (Paris, Dec. 18, 1902), *ref. Zentralbl. f. Gynäk.* (1903), p. 673; Höhne, *Arch. f. Gynäk.*, vol. lxix., pt. 1; Bouchacourt and Coudret, *Geb. Gesellsch.* (Paris, Jan. 21, 1904), *ref. Zentralbl. f. Gynäk.* (1904, p. 1142; Winkler, *Verhandl. d. deutsch. path. Gesellsch.* (Kassel, 1903; Jena, 1904); Wetterill and Hall, *Ref. Zentralbl. f. Gynäk.* (1905), p. 275; Pulstinger, *Diss. München* (1904); Eggel, *Münchn. gynäk. Gesellsch.* (May 15, 1904), *ref. Zentralbl. f. Gnyäk.* (1905), p. 1504; Démelin and Goudert, *Geb. Gesellsch.* (Paris, Nov. 17, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 1384; Von Konstantinowitsch, *Prager med. Wochenschr.* (1906), No. 49; Phänomenoff, *Arch. f. Gynäk.*, vol. xvii., p. 133; Koller and Aeby, *Arch. f. Gynäk.*, vol. lxxxii.; Finkelstein, *loc. cit.* p. 101; Greiffenberg, *Gynäk. Gesellsch. in Breslau* (March 17, 1908), *ref. Zentralbl f. Gynäk.* (1909), p. 31, *Zeitschr. f. Gynäk.*, vol. lxii.; Bonnabel, *Paris*, 1906 · Stevenal (*Dextrokardie*), *Diss. Bordeaux* (1906); Arnold, *Virchows Arch.*, vol. li. Rieländer, *Arch. f. Gynäk.*, vol. lxxxviii., p. 1; Kermauner, *loc. cit.*, p. 15, pt. 3. sect. 1 (1909); Herxheimer, *loc. cit.*, p. 15, pt. 3, sect. 2 (1910); Keith, *Studies in*

Path., Aberdeen Quarcent. Celebrat. (Aberdeen, 1906), p. 55; Humphry, *Clifford Allbutt. loc. cit.* p. 89, vol. vi., p. 276 (1909); Abbott, *Journ. Med. Res.* (Boston, 1908), vol. xix., p. 77.

For malformations of the œsophagus see p. 150.

The Diaphragm

The diaphragm is very seldom entirely absent. When this is the case, the prognosis as regards the viability of the child is very poor; as it is also when larger or smaller portions of the diaphragm are missing. Nevertheless, Riedinger (Tillmanns) mentions a case where a boy with a complete congenital defect of the diaphragm lived to be seven years old.[1] The abdominal organs were placed partly in the thorax. In some cases, nearly the whole of the abdominal organs may be displaced into the thorax (congenital diphragmatic hernia). This is the commonest variety of the internal herniæ.

In these cases, the abdominal organs pass through the defect in the diaphragm, into the thorax; less commonly, the thoracic organs, the lungs or heart, into the abdomen. As a rule—at least, in the congenital diaphragmatic hernia—the condition is almost always a malformation: that is to say, a congenital defect of the diaphragm. In the majority of cases, the abdominal organs simply pass through an opening in the diaphragm: not constituting a hernia, in the true sense of the word, but a spurious diaphragmatic hernia. These herniæ are examples of ectopia of the abdominal organs, and are most commonly situated on the left side in the membranous portion of the diaphragm, as their occurrence on the right side is hindered by the presence of the liver in this position. Of forty-two diaphragmatic herniæ, according to Popp, thirty-seven were on the left side and only five on the right (Tillmanns). Leichtenstern records (quoted by Ahlfeld) sixty-five cases of left-sided and only twelve of right-sided hernia. The spurious variety occurs, according to Liepmann, before the separation of the abdominal and thoracic cavities;[2] while the true hernia—hernia diaphragmatica vera, in which the abdominal contents push the peritoneum before them—occurs

[1] In most cases of congenital diaphragmatic herniæ, the children die soon after birth; but in other cases, the hernia has only been discovered some years after birth,—in one case, at the age of sixty-seven.

[2] The development of the diaphragm is very complicated; but it consists of five main portions: the central tendon (formed from the septum transversum), two ventro-lateral and two dorsal portions. The ventro-lateral parts are derived from the ventral longitudinal muscular sheets which give rise to the rectus abdominis, and the dorsal parts from the part of the transversalis sheet of muscle forming the subvertebral musculature (Keith). The pleuro-peritoneal openings lie between the dorsal and ventro-lateral parts of the diaphragm, and their persistence is, no doubt, the explanation of one variety of congenital diaphragmatic hernia.

after the separation (see Kaufmann). In eighty cases of congenital hernia collected by Bohn (Tillmanns), in only fourteen was a hernial sac present. The true diaphragmatic herniæ, as Tillmanns has especially pointed out, project into the pleural cavity through spaces in the muscle covered only by peritoneum and pleura. Such spaces are to be found between the origin of the diaphragm from the spinal column and the ribs, and also between its costal and sternal portions directly behind the sternum (see Tillmanns). Moreover, small openings, which may serve for the development of diaphragmatic herniæ, may be found where normally openings are present for the œsophagus, aorta, vena cava, the splanchnic and sympathetic nerves. The most important of these is the œsophageal opening.

As regards the organs which can pass from the abdominal cavity into the thorax, the frequency of their displacement occurs (according to Kaufmann) in the following order: stomach, transverse colon, omentum, small intestine, spleen (usually with the accessory spleen), liver, pancreas, and kidneys. In the majority of cases, two or more organs are displaced at the same time. If only one organ, this is most commonly the stomach; less commonly, the transverse colon, the small intestine, or the omentum. These organs pass either into the left or right pleural cavity, or into the mediastinum.

The clinical signs of a congenital hernia may be completely absent, so that the abnormality may be first found, as an accidental discovery, at a post-mortem examination.

Keller's statement is therefore not quite correct—namely: that 'congenital defects of the diaphragm render inspiratory expansion of the lungs impossible, as the entrance of the neighbouring abdominal organs follows the necessary muscular contractions, and the child at once dies post-partum.' In other cases, colic, stomach symptoms—especially after eating—dyspnœa, and heart symptoms, occur during life; the abdominal cavity appears abnormally empty, the abdominal wall retracted, and a post-mortem examination first makes clear the condition. In other cases, again, symptoms dangerous to life may appear; as signs of strangulation of the intestine, with its sequelæ, may be present—namely: marked pain, vomiting, collapse, symptoms of ileus and peritonitis, gangrene, and escape of the intestinal contents into the thoracic cavity.

As has been explained above, the diagnosis of a congenital hernia of the diaphragm is very difficult. Sometimes, a circumscribed pneumothorax may be recognised by diminished expansion of the affected portion of the thorax—usually the left side—with bulging, displacement of the heart, imperfect vocal fremitus, and failure of the normal breath-sounds; and a deep loud, tympanitic at times or non-tympanitic, percussion-note—a metallic sound (the bell sound or *bruit*

d'airain), may be heard on simultaneous auscultation and percussion (Nothnagel). But while these symptoms remain fairly constant for some time in a true pneumothorax, in a case of diaphragmatic hernia marked changes may occur in the percutatory and auscultatory conditions, dependent upon whether the abdominal organs present in the pleural cavity are more or less filled with air or partly with fluid contents. Attention must be paid to the lungs, the heart, and disturbances of digestion. According to Leichtenstern, by artificially filling the stomach (through the mouth) and the colon (through the rectum) with water or air, the portions of the organs concerned in the hernia can be determined. Surgical treatment of a diaphragmatic hernia should only be considered if it becomes strangulated (see Tillmanns).

In conclusion, a few observations of importance, from the obstetrical literature relating to the subject, may be quoted. Becker demonstrated a seven-months-old fœtus with a spurious left-sided diaphragmatic hernia, the result of a defect on the left side. The child died two hours after birth. The left half of the thoracic cavity was filled with coils of small intestine, above which lay the appendix, and below the ascending and transverse colon. To the right was situated the stomach, which had undergone rotation to the extent of ninety degrees on its cardiac extremity, with the large curvature directed upwards. Under the stomach lay the spleen, and close to this the compressed lung. The heart with the pericardium was displaced to the right, and lay over the right lung. The heart-beat could be felt clearly during life on the right side. In the normal position of the heart, lay a portion of the left lobe of the liver, which was separated from the remainder of this lobe by a part of the diaphragm stretching from the spinal column to the sternum, and along the anterior thoracic wall following the course of the eighth rib. A second portion of the diaphragm, forming a muscle-bundle 2 mm. thick, passed from the lumbar vertebræ to the posterior wall of the thorax and formed the œsophageal foramen; otherwise, on the left side, the diaphragm was almost completely defective.

Vrendenberg observed a similar case. In a female child, twenty-four hours old, he found marked dyspnœa, the sternum very markedly bulged, and the whole of the posterior part of the left half of the thorax dull. The heart-sounds were best heard on the right side; the breath-sounds were very weak on the left side, but stronger on the right. From the second day, onwards, attacks of cyanosis occurred, and the child died on the fifth day. On a post-mortem examination, the left pleura was found full of intestine—especially, large bowel. The diaphragm on the left was present only as a band some $1\frac{1}{2}$ to 2 cm. broad; otherwise, it was entirely wanting on that side. On the left side, the lungs were also rudimentary, but the heart was normal.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; v. Langenbecks, *Arch. f. klin. Chir.*, vol. xxvii., pt. 1; Keller, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Becker, *Gynäk. Gesellsch. in Breslau* (Feb. 23, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 1519; Vrendenberg, *ref. Zentralbl. f. Gynäk.* (1905), p. 966; Nothnagel, *loc. cit.*, p. 53; vol. xvii., p. 327; Ahlfeld, *loc. cit.*, p. 34; Gaillard, *Diss. Paris* (1907); Schreiber, *Geb. Gesellsch., Paris, ref. Zentralbl. f. Gynäk.* (1908), p. 1532; Gminder, *Fränk. Gesellsch. f. Geburtsh. u. Gynäk.* (May 28, 1905), *ref. Zentralbl. f. Gynäk.* (1906) p. 185; Planchau, *Presse méd.* (1904), No. 70; Marchand, *Geb. Gesellsch.* (Leipzig, Feb. 15, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 585; Liepmann, *Arch. f. Gynäk.*, vol. lxviii., pt. 3; Hirsch, *Münch. med. Wochenschr.* (1903).

CHAPTER IX

Umbilical Hernia—Hernia Funiculi Umbilicalis—Exomphalus—Congenital Omphalocele

An acquired umbilical hernia (Hernia umbilicalis acquisita ; acquired omphalocele) must be carefully distinguished from a congenital umbilical hernia. These two anomalies, which have nothing to do with one another, are sometimes confused. The acquired, in contra-distinction to the congenital, hernia first makes its appearance some weeks or months after birth. Its occurrence is favoured by a deficiency of elastic fibres in the region of the umbilicus (Herzog), and, as it owes its origin, primarily, to a want of fat in the abdominal walls and to an increase in the intra-abdominal pressure, it is very likely to occur in children who are overfed and suffering from flatulence, colic, and meteorism, or who cry a great deal or suffer from cough (Biedert and Fischl). According to Monti, improper swaddling of the children is also an important factor; while congenital phimosis has been given as an explanation. The umbilical ring yields, and there protrudes from it a larger or smaller tumour consisting of coils of intestine : less commonly, of omentum. (For further details, see Biedert and Fischl.)

A congenital umbilical hernia is something quite different. The condition is one of want of development of the ventral wall of the abdomen, and occurs most commonly in the umbilical area—as the abdominal cavity is closed last at the umbilicus. If the abdominal coverings are completely undeveloped, then the abdominal cavity is closed only by the peritoneum and the covering of the cord—the amnion. By the concurrent displacement of intestine, the umbilical area is more or less widely stretched. The development of this malformation occurs in the following way : the abdominal coverings of the embryo grow into the blastoderm and surround a cavity—the future abdominal cavity—in which a portion of the vesicle is cut off (Biedert and Fischl). The cut-off portion of the vesicle becomes the intestinal canal and communicates with the part of the blastoderm lying outside the abdominal cavity (the yolk-sac) by a canal—the omphalo-intestinal or vitello-intestinal duct (see Seitz). The margins of the abdominal plates surrounding this canal form the skin portion of the navel. As a

result of these conditions—especially in consequence of the stretching of the vitello-intestinal duct—normally, up to the third month of embryonic life, a loop of intestine, the so-called umbilical loop, lies in the commencement of the umbilical cord (Bolk). Usually, however, complete separation of the duct occurs, so that the loop of intestine recedes again into the abdominal cavity; if separation, however, does not occur, then a congenital umbilical hernia develops.[1] This mode of development has been confirmed by Ahlfeld by the recognition of the rudimentary ducts, described in cases of congenital umbilical hernia. Küstner has drawn attention to the fact that by the constant drag of the vitello-intestinal duct on the bowel, even the rectum may be markedly displaced upwards.[2] In this way, in certain circumstances, the union of the rectum with the proctodæum growing towards it from the skin may be hindered. In this way, too, Küstner explains the occurrence, so frequently observed, of atresia ani with a congenital umbilical hernia. Seitz has described such a case, where an open Meckel's diverticulum was also present.[3]

A further result of this traction of the omphalo-intestinal duct on the intestine or its mesentery is an alteration in the normal attitude of kyphosis of the fœtal spinal column, and its transformation into a lordosis or even a hyperlordosis. If the abdominal cleft is so wide that almost the whole of the intestinal contents are eventrated, then the lordosis can be so marked that the occiput of the fœtus nearly touches its sacral region. In opposition to Küstner, Höhl believes that the bending of the spinal column is the primary condition, and that the thoracic or abdominal clefts are secondary. Besides this—no doubt the most

[1] In the early human ovum, the yolk-sac is formed from the entoderm and mesoderm. The former becomes separated off from the embryonic cell-mass, and receives a covering from the primitive mesenchyme. There is probably no such ingrowth as the author describes. The primitive gut is developed by an infolding of the wall of the yolk sac (the splanchnopleure), and at the third week consists of a fore-gut, a hind-gut, and a mid-gut; the latter continuous with the cavity of the umbilical vesicle or yolk-sac. The communication between the two, ultimately becomes the vitello-intestinal canal; and Meckel's diverticulum is formed by the persistence of the intra-abdominal part of the canal. Finally, the yolk-sac by the closure of the umbilicus and formation of the cord, comes to lie on the placenta, and its neck is represented within the cord by the remains of the vitello-intestinal duct. The mid-gut forms the U-shaped loop of the intestinal canal, and this is at first really extra-abdominal, being situated in a portion of the cœlom (the umbilical cœlom), within the umbilical end of the cord (Keith). The U-shaped loop, instead of retreating within the abdomen at the end of the second month, may remain within this umbilical funnel and so give rise to a congenital umbilical hernia.

[2] Taruffi urges against this view that, even when the liver is displaced, the duodenum usually occupies its normal position.

[3] In order to account for the frequent association of these defects, Bryce suggests defective development of the allantoic stalk and hind-gut, which may interfere with the incurving of the tail end of the embryo, and so maintain the attitude of retrollexion of the trunk.

frequent cause of a congenital umbilical hernia, as determined by Ahlfeld—other ætiological factors must also be taken into account: such as abnormal shortness of the umbilical cord with resulting permanent traction, abnormal enlargement of the abdominal viscera (especially of the liver), abnormal retraction or contraction of the muscles of the abdomen, and adhesion of the intestines to the funis.[1] Seitz admits that in some cases these factors may be important; but for the great majority of cases they do not, according to him, come into play. The large herniæ, in which an abdominal cleft is present and where marked eventration occurs, are to be regarded as instances of failure of development, in which the union of the abdominal parietes is faulty.[2] Such failures of development, according to Ahlfeld and v. Winckel, are produced by amniotic bands. In favour of this view is the fact that, besides the umbilical hernia, other malformations of amniotic origin are often present—such as hemicephalus, spina bifida, and spontaneous amputations. Aschoff, in a very complete work, brings forward

FIG. 32.—ECTOPIA ABDOMINALIS. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] Cases have been recorded in which union of the abdominal walls has occurred in spite of the prolapse of some of the abdominal organs, as in the case described by Ahlfeld, in which a portion of the jejunum in a hernia had become entirely separated.

[2] In most cases, however, the recti muscles can be recognised in the margins of the cleft.

arguments against this view of Ahlfeld. According to his observations, which are of very considerable value, the liver, in an umbilical hernia is not secondarily displaced into the hernial sac, but is situated there from the very first. The so-called 'adhesion' of the liver to the wall of the sac is not the result of inflammatory action, but is due to the primary union of the liver with the abdominal wall, and to the fact that the separation of the two has been imperfect.[1] According to Aschoff, the condition is not one of extreme width of the sheath of the umbilical cord, in the strict sense of the word, since the hernial sac is in reality formed by the whole of the anterior abdominal wall. The lordosis, so frequently present, is, according to the same author, a primary condition: while the abnormal position of the base of the pericardial cavity is due to an unusual position of the primitive diaphragm.[2]

Only the small umbilical hernia have any great practical interest, as children with large umbilical hernia, and with marked eventration of the abdominal contents, are incapable of living: such children are either born macerated, or die during, or soon after, birth. Very small umbilical hernia have not uncommonly been overlooked—a circumstance which may prove disastrous to the child; as in tying the umbilical cord, loops of intestine present in the umbilical sac may very readily be included in the ligature, thereby leading to the formation of an umbilical faecal fistula. Small cylindrical hernia are most readily overlooked, since they may be regarded simply as thickened portions of the cord. It is important, therefore, to specially examine the umbilical end of the cord before tying it, and to remember the possibility of an umbilical hernia—especially when the union of the cord with the skin has a broad triangular form.

In a large umbilical hernia, the hernial sac may burst *in utero*, so that the child is born, with the contents of the sac, lying free on the abdominal walls. In a case recently observed by me, the sac, almost the size of the fist (and containing the whole of the liver, the small intestine, the stomach, and the spleen), burst at the moment

[1] The constant presence of the liver in the sac, and its union with the walls, are readily explained by its anatomical relations in the fœtus and its mode of development. The primitive liver-bud grows out from the junction of the fore-gut and the yolk-sac, and develops in the ventral mesentery. According to Aschoff, the condition of adhesion is really one of failure of the liver to become separated from the septum transversum.

[2] Kermauner regards all varieties of congenital umbilical hernia as due to failure of development of the primitive myotomes, and as originating about the third week of embryonic life. All other changes he regards as secondary. If the development of the muscle myotomes on one side of the body is defective, then the defect in the abdominal walls will be mainly on this side, and at the same time the vertebral column will present a concavity towards the same side. The position and extent of the defects present, he regards as dependent upon the number of the segments involved, and the degree of the maldevelopment.

it was being divided. Delivery occurred with a vertex presentation. In the Göttingen collection, there is a specimen (Fig. 33), where the hernial sac burst *in utero*, and the liver and intestine lay exposed. An erroneous diagnosis may be made as the result of such a rupture, as in a case recorded by Neugbauer. In a case in which a hernial sac burst *in utero*, the medical man felt coils of intestine in the uterus, which he regarded as maternal, and so made a diagnosis of rupture of the uterus. On performing an exploratory laparatomy, the uterus was found intact.

As regards the frequency of an umbilical hernia, it has been met with about once in every 2000 to 5000 births.

On examination, there is found a round or oval tumour of very varying size (from a cherry to larger than a child's head). The great majority of such hernie are approximately the size of an apple.[1] The umbilical cord passes directly into the tumour.

FIG. 33.—UMBILICAL HERNIA WHICH RUPTURED *in utero*. (Specimen from the collection of the Göttingen Women's Clinic.)

The sac of the hernia consists from without of the following: (1) *The Amnion*. This is attached firmly to the skin of the abdominal wall, at the base of the sac—just as in normal circumstances it is attached at the insertion of the umbilical cord to the skin. In a few cases, the skin, with a few muscle fibres, passes on to the wall of the sac; while in other cases, again, it does not even reach the base of the sac. The sheath of amnion in the region of the sac, like the rest of the amnion, is free from any vessels. It must therefore, later, be affected by any mummification or other process of decay that may occur. (2) *Wharton's jelly* in fairly thick layer. (3) *The Peritoneum*. Tillmanns's statement, that the

[1] In the great majority of cases, the tumour is asymmetrical and not in the middle line.

extruded abdominal contents lie in the cord, without any peritoneal covering, I have not found confirmed elsewhere. These portions of the wall of the sac are almost always attached, more or less closely, to one another.[1]

The hernial aperture is formed by the margins of the defect in the abdominal wall and varies very greatly in size. The contents of the hernia are various.

Very frequently, only coils of intestine are found; less frequently, the stomach, a portion or the whole of the liver. The latter is very often altered in form, and is united with the peritoneum of the hernial sac (see Aschoff). Sometimes a tongue-like portion of the liver is found in the hernia. According to Biedert and Fischl, the displacement of the liver takes place readily, and is in large part brought about by the traction of the umbilical veins.[2] Besides the organs mentioned, the pancreas, the kidneys, the spleen, the female genital organs, and even part or the whole of the heart, may be found in the sac. Such a case has been described by Arndt. He found the heart which had slipped into the abdominal cavity through a defect in the diaphragm, pulsating in the hernial sac without any pericardium. An operation was unsuccessful, as the organ was too damaged by compression to recover after the operation had been performed.

Kolly and Aeby have also described a case of a congenital diverticulum of the heart, in an umbilical hernia. In a female fœtus of seven months, a knob-like tumour, measuring 1 cm. in diameter, was seen pulsating, during the three hours the child lived, above an umbilical hernia, which was about the size of a spectacle-glass. The tumour was recognised as the apex of the heart, formed by the extremity of the left ventricle. In rare cases, separated-off portions of the intestine, with corresponding defects of the bowel, have been found in the hernial sac. The vessels of the umbilical cord pass from the navel, spreading out between the amnion and the peritoneum over the sac, to the margins of the aperture; the veins going to the liver, and the arteries to the region of the bladder. The umbilical cord is inserted, sometimes, on the summit of the tumour; more commonly, laterally. Anomalies of the vessels (for example, absence of an umbilical

[1] The coverings of the sac are often so fused together that it is difficult to say what they are composed of, and the presence or absence of the peritoneum may be impossible to determine.

[2] The liver is present more often than the stomach. In twenty-nine cases collected by Kramer, in twenty-two the liver was present; in ten, the stomach. Abnormalities in the anatomical relations of the veins of the liver, and also shortness of them, have been supposed to play a part in the displacement of the liver; but as these changes occur after the period at which an umbilical hernia originates, they are in all probability secondary.

artery) are not uncommon.[1] If an umbilical hernia is seen soon after birth, before the amnion has become dry, the transparent green intestine, filled with meconium, can at times be recognised (see Seitz).

In the conditions described, the diagnosis can nearly always be readily made; difficulty only arising, as a rule, with small umbilical herniæ. The presence of coils of intestine can be determined by palpation, and by the recognition of intestinal sounds.

If such hernie are left to themselves, either the sac bursts or the amnion gives way as the result of inflammation and suppuration; the peritoneum becoming covered with granulation tissue. In rare cases, spontaneous healing may take place by the formation of granulation tissue, so that the defect in the abdominal wall gradually shrinks and finally closes. More frequently, however, the whole sac sloughs, and inflammation of the umbilical artery, general sepsis, and peritonitis result with their sequelæ.

The prognosis of an umbilical hernia, if it is not too large, is more favourable than it was formerly; frequently, however, other concomitant malformations render the prognosis a bad one. In very large herniæ, with marked eventration, operative treatment is not of much avail, as in these cases it is usually impossible to overcome the marked protrusion of the abdominal viscera (see p. 156, under treatment). In the smaller and medium-sized herniæ, the prognosis at the present time is better than it was formerly; as all such cases can be treated with abdominal bandages sometimes, even with success.

Fischl observed a successful result of this kind in the case of a hernia, measuring some 5 cm. in diameter, treated by means of a simple bandage. See also Fleischmann and others (Baginsky).

According to the experience of the last few years, operative treatment, when carried out as soon as possible after birth, gives the best results (Landfors, Krukenberg, Knoop, Olshausen, Klausner, Runge, Buschan, Karewski, and others; see the literature quoted by Seitz). Thus Knoop was able to report, in the year 1903, thirty-five cases operated upon during ten years, of which twenty-five (or 71 per cent.) were cured. Watravens (Seitz) found that 80 per cent. were cured when operated upon in the first twenty-four hours; but only 33 per cent. when operated upon after forty-eight hours. Gutzeit operated upon an umbilical hernia, successfully, on the seventh day after birth, when gangrene of the sac had already occurred.

The operation is performed in the following manner: The sac is laid

[1] This occurs in about a third of all cases, and is most commonly seen on the side on which the defect in the abdominal wall is most marked. The most frequent anomalies of the veins are the persistence of the right umbilical vein and the separation of the inferior vena cava from the liver.

open and removed as widely as possible; any adhesions between its walls and its contents being separated, and the umbilical vessels tied with catgut. Careful ligature of the vessels is especially necessary: as a child, operated upon by Borelius, bled to death after a successful operation, from an unligatured vessel in the abdominal wall. The margins of the hernial aperture, after reposition of the contents, are freshened and the various layers—peritoneum, fascia, muscle, and skin—are very carefully sewn up, separately. On account of the excessive tension which is so frequently present, deep sutures of silkworm gut or of silver wire should be employed. Where the tension is very great, lateral incisions to relieve it can be made in the long axis of the body. On the conclusion of the operation, the child is placed in a warm bath and fed, as far as possible, with its mother's milk. A serious difficulty may arise, in the course of the operation, when the aperture of the sac is narrow and large organs—for example, the liver—lie in it. In these circumstances, the best plan is to divide the abdominal walls upwards and downwards. Küstner resected a portion of the liver, which he could not replace, with Paquelin's cautery, with a good result. The child died on the twenty-first day from other causes.[1] In the same way, Olshausen (among others) operated upon a case (recorded by Zillmer), resecting the liver, with a good result. In a case of umbilical hernia, the size of an apple (described by Borelius), the liver could not be replaced and had to be left outside the abdominal cavity; the child dying. Apparently, Borelius had omitted the useful division of the abdominal walls above and below.

A modification of this operation is suggested by Olshausen. He recommends that the amnion should be split and removed, together with Wharton's jelly—but the peritoneum should not be opened (extra-peritoneal method)—and then the freshened edges of the abdominal walls sewn together. This procedure is naturally only possible when the hernia is reducible and there are no adhesions present between the amnion and the peritoneum. It has the disadvantage that the contents of the hernia are not seen, and adhesions which may be present between the coils of intestine cannot be separated, and it is not available when the opening in the abdomen is very large. For such cases, Ahlfeld employs an antiseptic bandage, with alcohol compresses put on under anæsthesia; and especially recommends this method, on account of its simplicity, for general practice. It has, according to the author, the advantage that at a later period in life, under suitable conditions, as good results can be obtained as immediately after birth. The method is carried out as follows: 'The child is lightly anæsthetised, and then the abdominal wall and the covering of the hernia are thoroughly washed with soft soap and

[1] With hæmatemesis and melæna—possibly, septic in origin.

hot water and soft pads. The remains of the umbilical cord are firmly tied and cut off—all but 1 cm. The sac and the surrounding skin are then washed, for some minutes, with 96 per cent. alcohol, and the contents, as is now possible under the anæsthetic, replaced in the abdominal cavity. The abdomen is covered with flat compresses soaked in alcohol, and, finally, the whole encircled with a broad bandage, so that—and this is the most important point—even with the most forcible crying, none of the abdominal contents can protrude. The abdominal opening very quickly becomes smaller under this treatment. After three or four days, the dressing is changed again (under anæsthesia): the compresses, by the use of hot sterile water, carefully loosened from the sac (to which they are usually somewhat adherent), and reapplied as before. The markedly retracted sac becomes covered with skin from the periphery, and, after a fortnight, a simple bandaging with some wool and an abdominal binder suffices. In two cases of large umbilical herniæ, of which the one certainly, the other possibly, could not have been cured by operative treatment, the result was most satisfactory. In the first case, the circular scar, of the size of a five-shilling piece, was cut out, and at the same time two inguinal herniæ operated upon.' I have described this method fully, as it appears to me to be a most valuable one for those large umbilical herniæ which cannot be operated upon. In other cases it is well to operate as soon as possible after birth, as the experience of the last few years has shown.

The obstetrical importance of umbilical herniæ, strictly speaking, is not very great. As a result of the yielding nature of the contents, even in the case of the large herniæ, no obstruction to delivery usually arises. Only when the displaced organs—for instance, the liver—are markedly enlarged, from inflammatory changes or from tumour formation, may operative procedures be necessary for the termination of the delivery. B. Költsch records a case in which the liver, situated in an umbilical hernia, formed a hindrance to delivery. In these cases, the child is born as far as the umbilical region and then its further advance is delayed. The diagnosis can readily be made by the introduction of the whole or half-hand. In instances of definite obstruction to labour, embryotomy alone should be considered as a means of treatment. In such cases, it is important to remember that children, even with large umbilical herniæ, may remain alive with appropriate treatment. According to Hohl, in cases of fissure of the anterior surface of the body, difficulty during labour usually arises because the presentation is faulty. Thus the children may present, by the prolapsed abdominal contents, in a transverse or other presentation. Of thirteen cases of cleft of the anterior abdominal wall, Hohl found in eight a faulty presentation, of which there was five in an abdominal presentation—one presented by the breech with a prolapsed arm, and four were head presentations. Hohl

himself (in one case, where the enlarged liver with a portion of the bowel was covered only by the peritoneum) was compelled to perform version, on account of the presentation of this part; and, from his experience of this case, maintains that the recognition of such fissure malformations may be exceedingly difficult. He insists (rightly) on the fact that in the performance of version and extraction great care must be taken not to burst the hernial sac.

A further obstetrical factor in umbilical herniæ lies in the fact that abnormal shortness of the umbilical cord is often observed in association therewith. According to Küstner, the majority of cases of tearing of the cord during labour have occurred with abdominal fissures. A further sign of the presence of one of these herniæ occurs when hyperlordosis of the spinal column (which we have already described) is present. Küstner records a case in which version, indicated on account of a transverse presentation, was rendered very difficult for this reason.[1] Attempts to bring down the foot over the abdominal surface of the child—in normal circumstances, the correct procedure—failed; and only after prolonged efforts was it possible to bring down the feet over its back and so to extract the malformed fœtus.

Literature

Ziegler, *loc. cit.*, p. 15; Kaufmann, *loc. cit.*, p. 24; Spiegelberg, *loc. cit.*, p. 15; Tillmanns, *loc. cit.*, p. 34; Keller, *loc. cit.*, p. 34; Küstner, *loc. cit.*, p. 15; Leser, *loc. cit.*, p. 34; Baginsky and Biedert, *loc. cit.*, p. 34; Fischl, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, pp. 24 and 34. Hohl, *loc. cit.*, p. 15; Runge, *loc. cit.*, pp. 24 and 129; Monti, *Kinderheilkunde*, No. 20; Borelius, *Ref. Zentralbl. f. Gynäk.* (1903), p. 223; Lotheisen, *Wiener Klin. Wochenschr.* (1903), No. 42; Brodführer, *Med. Woche.* (1903), No. 31; Osterloh, *Gynäk. Ges. Dresden* (October 15, 1903); Zillner, *Zeitschr. f. Geburtsh. u. Gynäk.* vol. li., pt. 2; Fournier, *Geb. Ges. Paris* (March 17, 1904), *ref Zentralbl.* (1904), p. 1144; Gutzeit, *Deutsche Zeitschr. f. Chirurgie*, vol. lxxiii., pts. 1–3; Lindner, *Geb. Ges. Wien* (June 14, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 378; v. Neugebauer, *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xx., pt. 6; Ahlfeld, *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxii., pt. 2; Kindl, *Diss. Leipzig* (1904); Kannegiesser, *Gynäk. Ges. Dresden* (Dec. 22, 1905), *ref. Zentralbl. f. Gynäk.* (1906), p. 552; Küstner, *Gynäk. Ges. Breslau* (March 20, 1906), *ref. Zentralbl. f. Gynäk.* (1906), p. 1257; Olshausen, *Arch. f. Gynäk.*, vol. xxix., p. 443; Küstner, *Zentralbl. f. Gynäk.* (1901), No. 1; Arndt, *Zentralbl. f. Gynäk.* (1896), No. 24; Lindfors, *ref. Zentralbl. f. Gynäk.* (1884), pp. 255 and 472 (1889) and *Samml. klin. Vortr.* (1893), p. 482, No. 63; Krukenberg, *Arch. f. Gynäk.*, vol. xx., p. 269; Knoop, *Samml. klin. Vortr.*, No. 348; Klausner, *Münchner med. Wochenschr.* (1889); Karewski, *Die chirurgischen Krankheiten des Kindesalters* (Stuttgart, 1894); Aschoff, *Virchows Arch.* (1896), vol. cxliv., p. 144; Finkelstein, *loc. cit.*, p. 101; Paquy and Esmonet, *ref. Zentralbl. f. Gynäk.* (1903), p. 955; Költsch,

[1] Backward displacement of the lower limbs may accompany the lordosis of the spine, and this—as no doubt, occurred in the case recorded—may render version very difficult.

Monatsschr. f. Geburtsh. u. Gynäk., vol. x, p. 13; Bolk, *ref. Zentralbl. f. Gynäk.* (1908), p. 54; Fiedler, *Deutsche med. Wochenschr.* (1907), No. 3; Ringel, *Münchner med. Wochenschr.* (1907), No. 34; Weck, *Diss. Erlangen* (1907); Durlacher, *Münchner med. Wochenschr.* (1908), No. 11; Küstner, *Gynäk. Ges. Breslau* (June 30, 1908), *ref. Zentralbl. f. Gynäk.* (1909), p. 108; Ballantyne, *loc. cit.*, p. 44 (1904); Keith, *loc. cit.*, p. 96 (1910); Kermauner, *Schwalbe, loc. cit.*, p. 15; vol. iii., pt. 1, p. 48 (1909); Kraemer, *Zeitschr. f. Rationelle med.*, vol. iii. (1853); Seitz, *loc. cit.*, p. 15, vol. iii., pt. 3, p. 184 (1907).

CHAPTER X

Malformations and Congenital Diseases of the Organs of Digestion

For malformations and congenital diseases of the mouth, see p. 96.

The Œsophagus

I have followed here, on the whole, the detailed monograph by Kraus in 'Nothnagels Handbuch.'

Congenital malformations of the œsophagus are, in the majority of cases, so serious that the children die soon after birth, as only in anomalies of the slighter degrees can they reach any later age.

The following defects have been observed in the œsophagus:—

1. Total Absence of Œsophagus and Pharynx

This very rare malformation has been observed only in acardiacus, and other malformations of a serious nature of the upper half of the body. In these cases the mouth and the cardiac end of the stomach end blindly, being closed by a thin muscular membrane.

2. Complete or Partial Doubling of Œsophagus—Digesophagia

In this very rare malformation the œsophagus opens at its cardiac extremity by two openings into the stomach.

3. True Œsophago-tracheal Fistulæ

These abnormal communications between the trachea and the œsophagus are easily understood when we recall the mode of development of the parts. The whole intestinal canal, from the mouth to the anus, is formed in three portions: the stomodæum, the true intestine or primary rudiment of the intestine; and the proctodæum. The true intestine is again composed of three portions: the anterior, middle, and posterior gut. From the anterior portion develop the pharynx and the œsophagus. The stomodæum grows towards the fore-gut as a depression from the exterior, and the thin membrane separating the two gradually

disappears. Its remains[1] form, at the junction of the stomodæum and the fore-gut, the palatine arches and the uvula. In the earliest stage of development of the embryo, the pharynx and œsophagus are in relation with the respiratory apparatus.[2] The division, between the two, forms later; and is already present in Man at the beginning of the second month. After these few short remarks on the history of the development, we must return to a consideration of the true œsophago-tracheal fistulæ. They are fistulous communications between the otherwise well-developed œsophagus and the trachea, and are almost always situated at the bifurcation of the trachea, so the conclusion may be drawn that the membrane separating the œsophagus and the trachea closes in this position last. If the fistulous opening is very small, or is covered by a fold of mucous membrane, then the affected persons may present no symptoms during life, and the condition may be discovered accidentally at an autopsy. If such favourable conditions are not present, the children die; in the cases quoted by Kraus, this occurred from the fifth day to the seventh week; in the latter case, from pneumonia.

The cysts (about the size of a walnut, lined with ciliated epithelium, and with mucoid contents), found in the region between the bifurcation of the trachea and the œsophagus, are regarded by Kraus as the degenerated remains of the communication between the trachea and the œsophagus, after a closure of it at both ends has taken place.

4. Partial Obliteration

(*a*) *Simple blind ending of the œsophagus.*—Kraus has described a typical case of this kind of congenital atresia of the œsophagus with an instructive illustration. In this case, the œsophagus ended 6 cm. above the larynx;[3] the pharynx and trachea being perfectly formed.

Between this œsophageal rudiment and the stomach (which was adherent to the diaphragm), a thin muscular bundle, closely connected with the trachea, was found at the autopsy. The mucous membrane of the œsophagus was normal. In other cases, in addition to these or similar malformations, imperfect development of the brain, of the lower

[1] The remains of this septum are sometimes called the primitive velum; but the septum has nothing whatever to do with the formation of the permanent velum palati, or with the isthmus of the fauces (Bryce).

[2] The lung rudiment begins to develop from the ventral part of the pharynx at its junction with the œsophagus, in the form of an elongated vertical diverticulum of the fore-gut, in the third week. The trachea and larynx are formed by a separation from the œsophagus of the original median diverticulum, the separation commencing below, and leaving a relatively small connection between the two tubes above—the rudimentary glottis.

[3] These cases may be explained by the fact that, at one stage of development, the œsophagus in its cervical portion is filled with epithelium and closed.

jaw (agnathia and synotia), of the trachea, of the larynx, absence of the stomach, or its division into two or three portions, and atresia ani have been observed.

(*b*) *The œsophagus ends blindly, as in the cases just described; but a communication exists between its lower remaining portion and the trachea—œsophago-tracheal fistula, in a broad sense.*[1]—This is the most frequent variety of malformation. Usually, in this case, the upper part of the œsophagus which ends blindly is widened, sometimes to such an extent that the trachea is compressed from behind forwards. The lower part is usually pointed and funnel-shaped, and opens into the posterior wall of the trachea at or just above the bifurcation. Both parts of the œsophagus are often connected by a solid muscle-bundle.

(*c*) *The œsophagus is completely obliterated at, or close below, the bifurcation of the trachea; while the upper portion is normal and patent.*—This is a very rare malformation.

Children with any of the malformations enumerated are naturally incapable of living; the less so, as frequently other malformations, such as have been mentioned, are also present. They die soon after birth from gradually increasing inanition, especially from want of water or oxygen, or from septic pneumonia. They either cannot swallow at all, or the food swallowed regurgitates unaltered—even when only small quantities are taken—and escapes from the nose or from the mouth. Frequently, attacks of suffocation of such a character occur as to lead to the conclusion that a communication exists between the food and air passages (Finkelstein). The abdomen is retracted; and it is an interesting fact that new-born children, with closure of the œsophagus, are usually only moderately developed. This fact, indeed, has been put forward by Ahlfeld as an argument for the view that the liquor amnii supplies nutritive material to the fœtus. The development of the children may, however, remain completely unaffected (two cases recorded by Schwalbe). The diagnosis is made by the passage of a bougie, which impinges against a resistance placed at a higher or lower level, as the case may be. The operative treatment of these cases usually fails, owing to the increasing weakness of the child

[1] This form of fistula is somewhat difficult to explain. Shattock regards it as of secondary origin, and attributes it to a malformation, due in part to the growth of the diverticulum from the anterior wall of the œsophagus, which gives rise to the lower part of the trachea and the rudiments of the lungs. As a result of this outgrowth, the posterior wall of the primitive œsophagus is drawn forwards, and, when the upper part of the trachea is subsequently formed, the whole of the unduly contracted lumen, immediately above the diverticulum, becomes involved in the formation of the air passages; while the original communication may or may not remain between the lower part of the œsophagus and the lower part of the trachea. Keith and Spicer regard it as due to a faulty development of the œsophago-tracheal septum, which becomes attached to the posterior wall of the fore-gut and so leads to obliteration of the œsophagus.

(which soon becomes evident), and on account of the bad prognosis attending all operative procedures on the new-born.

The performance of gastrostomy and feeding by the stomach fistula so formed must be considered. Hoffmann has recorded such a case for Helferich. Later, the possibility of removing the solid membrane, which exists between the blind ends, might be considered (Kraus). In the numerous cases, where a communication with the trachea is present, this would still remain; so that the ultimate result would be very questionable.

5. Membranous and Valve-like Strictures

According to Kraus, these are very rare; the membrane is, as a rule, formed by a ring-like prolapsed fold of mucous membrane. In a case recorded by Brenner, an œsophago-tracheal fistula was present under such a fold. The condition was completely cured by an external œsophagotomy.

6. Simple Congenital Stricture

These are also very uncommon and are to be regarded as congenital, as the tissues of the œsophagus outside the stenosis are quite normal and the symptoms date from the time of birth. The narrowed portion is either circular, and confined to only a small area; or it exists in the form of a more or less long, narrow canal. Kraus quotes such a case.

The patient was a female child, nine years old, who, even when a suckling, brought back its food in a marked manner. After weaning, the child was unable to take solid food, which came back after a few minutes. On this account, she lived for nine years on milk and beef-tea. Examination showed a stricture at the lower end of the œsophagus, which was cured by the systematic passage of bougies. The symptoms of such stenoses consist mainly of dysphagia—that is to say, difficulty in the passage of the food into the stomach. The food, generally, is arrested above the site of the stricture and is regurgitated; or it is gradually passed through the constricted area. As a result of the stagnation of the food, more or less marked dilatation of the œsophagus usually occurs above the stricture. If the stenosis is not treated, the affected child may die of inanition; more rarely, of perforation of the wall of the œsophagus from ulcerative processes. The diagnosis may be made by the passage of bougies, by œsophagoscopy, or by a Röntgen-rays examination. The treatment consists either in gradual slow dilatation, rapid dilatation, or division of the stricture (internal or external œsophagotomy). (For further details, see text-books on surgery—Leser and Tillmanns.)

7. Congenital Dilatation of the Œsophagus

This is general or partial. If only a circumscribed portion of the wall bulges, then it is called an œsophageal diverticulum. Dilatation occurs, as has been pointed out, very often as a secondary condition following stricture.[1] Congenital dilatation may occur to such an extent that the whole œsophagus is widened to the size of an arm. In some cases, the dilatation affects only a circumscribed portion; either just above the diaphragm—fore-stomach (Luschka) or just beneath the diaphragm—antrum cardiacum (Luschka).[2] Both these anomalies (according to Riegel) are very uncommon, and frequently occur without any symptoms. In other cases, large portions of food lodge either in the fore-stomach or in the antrum, and give rise to severe pain. The clinical signs of such dilatations are not uniform. If the primary condition is a stricture, then the symptoms already described will be the most prominent. In pure dilatation, symptoms occur only in the most marked cases: primarily, marked disturbances of nutrition, as the food remains lodged in the œsophagus, and is regurgitated after a longer or shorter time. If such a sac—which, in certain conditions, may be felt from without—becomes completely filled with food, then difficulties in respiration may be set up from pressure on the trachea or on the lungs. The treatment consists in suitable feeding; while in cases of primary stricture, treatment might be adopted on the lines indicated above.

Completely circumscribed dilatations—true diverticula of the œsophagus or of the lower wall of the pharynx (Pharyngocele)—are, according to some authors, frequently congenital (Ribbert, Klebs, König, and v. Bergmann; see Kaufmann). It has been suggested that they are the remains of single internal branchial clefts.[3] These diverticula are often present without any symptoms; but, as the result of the decomposition of the food and its results, the formation of a tumour cellulitis, and perforation may occur. The symptoms consist of regurgitation of the food, the occurrence of a swelling in the neck, and signs of pressure on

[1] This is denied by some writers, who state that food does not, as a rule, lodge in the œsophagus, but is almost at once rejected. If dilatation does occur, the point of constriction is usually near the diaphragm.

[2] The portion of the œsophagus passing through the diaphragm is usually of its normal size and presents no sign of any stricture. In most cases of dilatation, hypertrophy of the muscular wall is present; and this condition is difficult to explain. It has been suggested that it is due to inhibition and atrophy of the external longitudinal muscle-coat, which, by leaving the cardiac orifice closed, would lead to obstruction at the lower end of the œsophagus.

[3] These pouches usually begin in the middle line, posteriorly, just at the junction of the pharynx and the œsophagus. In this situation, the anterior wall is formed by the cricoid cartilage, and the posterior muscular wall is thinner and weaker than elsewhere. There is thus a congenital predisposition to the formation of a hernial protrusion of the œsophageal wall as a result of pressure.

the air passages. The diagnosis can be made by the passage of a sound or by an X-ray examination. The treatment consists in careful feeling or in extirpation of the sac.[1]

Without alluding in greater detail to illustrative cases of anomalies of the œsophagus, I might mention, briefly, an interesting case from the recent literature. Eustache demonstrated to the Obstetrical Society of Paris a child born at the eighth month of pregnancy with atresia ani and recti, obliteration of the upper end of the œsophagus, and a communication of the lower end with the trachea. The child died sixteen hours after birth, after an unsuccessful attempt had been made to remove the obstruction by an operation.

LITERATURE

Kraus, *Nothnagel, loc. cit.*, p. 53; vol. xvi., pt. 2; Kaufmann, *loc. cit.*, p. 24; Schmaus, *loc. cit.*, p. 15; Lewer, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Keller, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, pp. 24 and 34; Hoffmann (Helferich), *Diss. Greifswald* (1899); Mekus, *Zentralbl. f. Gynäk.* (1888), p. 686; Eustache, *Gaz. Obs. Paris* (Dec. 17, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 1087; Riegel, *Nothnagel, loc. cit.*, p. 53; vol. xvi., pt. 2; Finkelstein, *loc. cit.*, p. 101; Schwalbe, *loc. cit.*, p. 15; Rolleston, H. D., *Allbutt, loc. cit.*, p. 89; (1907), vol. iii., p. 330; Shattock, S. G., *Trans. Path. Soc. London* (1899), vol. xli., p. 87; Keith and Spicer, *Journ. Anat. and Phys.* (1906), vol. xli., p. 52; *Zenus, Deutsche Zeitschr. f. Chir.* (1906), vol. lxxxii.; Bryce, *Quain's Anatomy*, vol. i., p. 157 (1908).

The Stomach

Complete absence of the stomach (Agastria) is very uncommon, as is also extreme smallness of that organ (Microgastria).

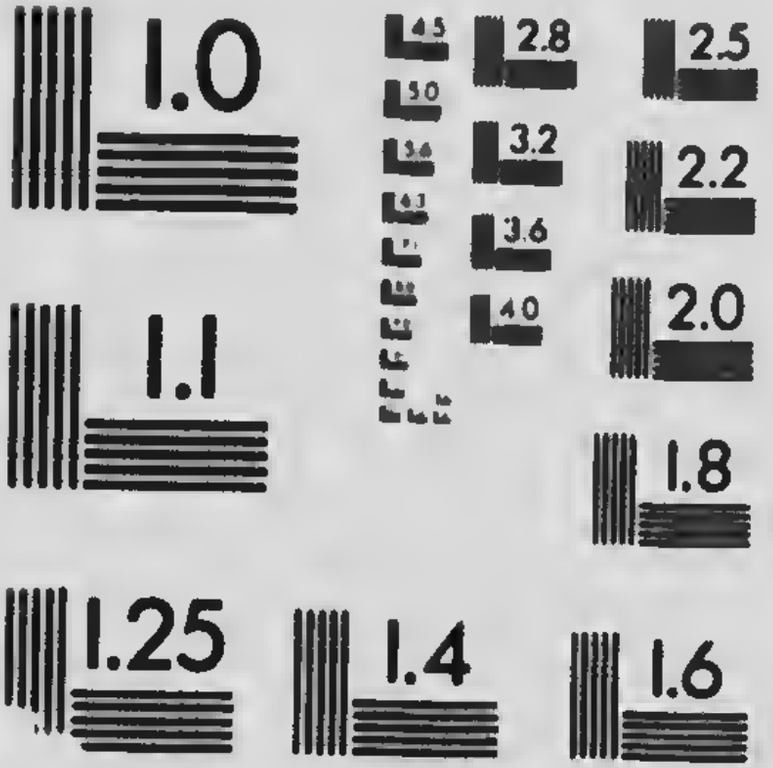
Duplication of the stomach has been described on several occasions; it is in reality due to the presence of diverticula which, when they are large, may be regarded as duplications (see also under 'Hour-glass' Stomach).

Congenital displacement of the stomach is more frequently met with. Thus, in a diaphragmatic hernia, the stomach may be displaced into the thorax (see p. 146); and in abdominal fissures, especially in umbilical hernia, it may be situated outside the abdomen (see p. 154). An interesting case of congenital prolapse of the stomach mucous membrane through the navel has been described by Tillmanns. The case was that of a boy, thirteen years of age, in whom a pedunculated tumour, the size of a walnut and covered with mucous membrane, was present at the navel. The examination of the digestive properties of the secretion from the mucous membrane, and the microscopical examination of the latter, proved that the condition was one of congenital prolapse of the mucous membrane of the stomach. No communication with the stomach itself could be found. The secretion from the tumour was

[1] Of twenty-three cases of extirpation collected by Veiel, eighteen recovered.

MICROCOPY RESOLUTION TEST CHART

(ANSI and ISO TEST CHART No 2)



APPLIED IMAGE Inc

1653 East Main Street
Rochester, New York 14609 USA
(716) 482 - 0300 - Phone
(716) 288 - 5989 - Fax

fairly profuse. Tillmanns explains the case in this way: namely, that originally, an umbilical hernia was present containing a diverticulum of the stomach: after the birth of the child, the diverticulum was ligatured with the umbilical cord, and thus a prolapse of the mucous membrane of the diverticulum formed—open towards the umbilicus, but closed towards the stomach.

Situs inversus of the stomach has been observed on one occasion. The persistence of the fœtal vertical position of the stomach has been distinguished as situs sagittalis. (For an account of the fore-stomach, see under the Anomalies of the Œsophagus, p. 164.)

In rare cases, a congenital hour-glass stomach is found.[1] Tillmanns observed in a new-born child an hour-glass stomach which was completely closed towards the duodenum. The stomach end of the duodenum ended blindly, so that there was no communication between the two. No analogous case has been reported in the literature. The cause of the congenital hour-glass stomach is the contraction of a ring of muscle tissue between the cardiac end and the pylorus. The stomach has the form of a double sac and is divided into two portions—a cardiac and pyloric portion. Both parts may be of the same size, or may differ in size. The slighter degrees of this anomaly often occur without any symptoms. Disturbances can, however, arise from the presence of an hour-glass stomach; for, as Riegel has pointed out, the food has to pass, as it were, over a hillock, owing to the drawing-up of the larger curvature of the stomach. The diagnosis is generally made by distending the stomach with air or carbonic acid gas. A further diagnostic feature of importance is the recognition of bubbling sounds in the two stomachs; while at the same time, it is not possible to completely evacuate the stomach contents with a tube. A cure can be obtained, in severe cases, only by operative procedures—such as gastro-anastomosis or resection of the stomach. The formation of congenital septa may also be associated with true hour-glass contraction.

Congenital closure of the stomach, generally of the pylorus or of the cardiac extremity, is very rare (see the case of Tillmanns, mentioned above). Congenital stenosis of the pylorus has been more frequently observed, and is almost always accompanied by some muscular hypertrophy at the pylorus. As to the nature, cause, and anatomical relations of this interesting anomaly, widely differing views are held. In some cases, organic narrowing; in others, spastic contraction (hence the designation pyloric spasm)—possibly, the result of a congenital local tenderness with general heightened nervous disposition; a tumour-like

[1] In an experience of considerably over 1000 operations on the upper abdomen, in which the stomach could be seen and examined, over 500 of which have been on the stomach itself, Mayo Robson has not met with a single instance in which he could say that the case was one of congenital hour-glass contraction.

hypertrophy (Hypertrophia muscularis, myoma (?)) of the circular muscle; or a thickening of the mucosa;—all these have been suggested[1] (see Kaufmann, Biedert and Fischl, and Finkelstein). A secondary dilatation of the stomach is usually not present. The clinical symptoms are of a dyspeptic character: Uncontrollable vomiting of large quantities of food, severe pains, refusal of the bottle or of the breast with screaming, distension of the stomach region, obstinate constipation alternating at times with diarrhœa, scanty urine, and progressive and noticeable wasting. Sometimes the thickened pylorus can be felt.[2] According to Biedert, the length of life of such children, if nothing is done, is from three weeks to six months. Heubner considers the prognosis as rather better than this; Finkelstein, as rather worse. Treatment consists in suitable feeding (if possible, with the mother's milk), the administration of Carlsbad Mühlbrunnen, regular washing out of the stomach, and sedative medicines (see Finkelstein). In hopeless cases, surgical measures must be considered (gastro-enterostomy or pyloroplasty).[3]

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34; Keller, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Riegel, *Nothnagel, loc. cit.*, p. 53; Tillmanns, *Deutsche Zeitscher. f. Chirurgie*, vol. xviii.; Savonat, *Diss. Lyon* (1905); Finkelstein, *loc. cit.*, p. 101; Champetier, Guinon, Frelet, *Geb. Ges. Paris* (Jan. 13, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 1097; Still, G. F., *Allbutt, loc. cit.*, p. 89; vol. iii., p. 515; Mayo Robson, *Keen, loc. cit.*, p. 53; vol. iii., p. 53

The Intestines

Situs inversus of the intestine may occur alone; but more frequently it occurs as part of a general situs inversus. Abnormal positions of the intestines are also observed congenitally: for example, as a result of abnormal shortness of the colon, the cæcum may be situated in the region of the umbilicus or even higher. In some cases the whole of the large intestine lies on the left side, so that the ascending colon

[1] Anatomically, there is an excess of the circular muscle fibres of the pylorus. Two explanations of this have been put forward: the first, that the condition is congenital and a pure hyperplasia; the second, that it is a secondary hypertrophy, the result of excessive action due to spasm of the pylorus. The hyperplasia and the spasm may begin *in utero*. In any case it is certain that the spasm is not due to any permanent narrowing of the lumen. The variation in the degree of obstruction, and the fact that permanent and complete recovery may follow medicinal measures, show that the obstruction is spasmodic in character.

[2] The two most important physical signs are visible peristalsis of the stomach, and the recognition of the thickened pylorus.

[3] The most successful medicinal measure is systematic lavage; and of the operative measures, forcible dilatation of the pylorus seems to hold out most hope: this method succeeded in eight out of eleven cases recorded by Still.

passes over into the descending colon by a sharp angle in the region of the spleen.[1]

On several occasions, congenital herniæ have been observed in the inguinal and crural regions (see Ahlfeld). The cause is to be found in a patent condition of the processus vaginalis. The coils of intestine present in the sac may become adherent to one another; but these adhesions are secondary and are not to be considered as the cause of the herniæ. In the great majority of the cases these herniæ have been observed in male children.

For hernia of the uterus and the ovaries, see p. 231.

Doubling of one portion of the intestine occurs very rarely. Ahlfeld cites a case (recorded by Schreiber) in which there was a partial duplication of the ascending colon. Ahlfeld considers it possible that these cases of doubling of the intestine are to be regarded as examples of separated cysts of the bowel.

For an account of the anatomical relations of the intestine in double monsters, see p. 946.

Complete absence of the intestine occurs only with extreme rarity in the most marked forms of acardiaci; while grave defects of the intestine have nearly always been observed with other severe malformations. Clogg has recently recorded such a case, without any other malformation except an umbilical hernia. The child was operated upon six days after birth. The small intestine ended blindly in the hernial sac; the large intestine was entirely absent, with the exception of a hollow cord of small diameter which was found in the position of the transverse colon. The child died soon after the operation; in which the intestine was opened and a drainage tube inserted.

Partial defects of the intestine are more frequently met with. Thus the vermiform appendix may be absent;[2] or the continuity of the intestine, in some places, may be interrupted for longer or shorter distances. David has recently described such a case. The child (weighing 2060 grm.)

[1] About the sixth week, a diverticulum is seen to grow out from the posterior limb of the primitive U-shaped loop of bowel. This forms the cæcum and appendix. The part of the loop above this forms the ascending and transverse colon, and the anterior part develops into the coils of the jejunum and ileum. At the seventh week, a rotation of the loop occurs so that the splenic flexure comes to lie against the spleen, and the cæcum in front of the right kidney, near the gall-bladder. At about the time of birth, the cæcum and ascending colon migrate towards the right iliac fossa. The majority of cases of displacement of the large intestine can, therefore, be explained by some failure in these changes of position during development.

[2] Up to the third month, the cæcal diverticulum is undifferentiated; but after this, the appendix can be distinguished. Probably, its absence is a persistence of the original condition. It is a lymphoid diverticulum of the cæcal apex (R. J. Berry). Although only seen in Man, the anthropoids, lemur, and opossum, yet a corresponding lymphoid structure is present in mammals. The appendix is, therefore, not to be regarded as a vestigial structure (Keith).

died three days after birth with the symptoms of an acute ileus. At the post-mortem examination, a perforation of the greatly distended colon in the region of the cæcum was found, and a complete occlusion of the descending colon. The latter ended in a blind sac, and the intestine was absent over an area of 1 cm., although the mesentery was well formed. To this portion was joined the normally developed sigmoid flexure with the rectum. David rejects, as an explanation of the cause of this malformation, the generally accepted explanation of fœtal peritonitis with the formation of adhesions, and also fœtal enteritis with secondary shrinkage. He suggests, as more probable, a primary imperfect development of the blood-vessels.

In rare cases, completely separated portions of intestine have been found in umbilical herniæ or at the umbilicus, with corresponding defects in the bowel. Tillmanns quotes a case observed by Ahlfeld,[1] in which (in a new-born, almost full-term, well-developed child) a prominent tumour, about the size of an apple, was found at the side of the navel, and connected with the latter by a fine stalk. The tumour was composed of a separated convolution of intestine and was formed, as shown by an autopsy, of the lower part of the ileum, the cæcum, and ascending colon, together with the corresponding portion of mesentery.

Congenital contractions, stenoses, occlusions, and atresiæ of the intestine, with the exception of atresia ani (to be mentioned farther on), are very uncommon. In the small intestine, they are commoner than in the large, and they are most frequent in the duodenum,[2] where the common bile duct and the duct of Wirsung enter, or above or below their point of entry, or at the union of the duodenum with the jejunum.[3] In the ileum, they are situated most commonly just above the ileocæcal valve, or at a point corresponding to the origin of the omphalo-mesenteric duct. The occlusion, sometimes complete and at other times incomplete, is generally of a membranous septum-like character;

[1] An illustration of this case appears in Schwalbe, *Die Morphol. d. Missbild.*, vol. i., pt. 3, p. 52.

[2] Congenital occlusion of the duodenum is commoner than stenosis; in fifty-seven cases collected by Cordes there were forty-eight of occlusion and only nine of stenosis. Shattock puts forward the same explanation for this anomaly as for atresia of the œsophagus: namely, that the outgrowth of the liver diverticulum produces a kink on the opposite side of the intestinal tube, and that this leads to secondary closure of the bowel. In view of the fact that the liver diverticulum and one pancreatic bud grow out from the ventral side of the bowel, and the second pancreatic bud from the dorsal side, it is a little difficult to see how such a kink can be produced. The occlusion occurs, usually, at the line of junction of the fore-gut and the mid-gut, and, according to Tandler, is due to proliferation of the epithelium, lining this portion of the gut after the outgrowth of the liver and pancreatic buds.

[3] It has been suggested that these cases are due to pressure on the bowel by the arrangement of the mesentery at the duodeno-jejunal juncture. Dueros has collected three cases of stenosis, five of obliteration of the lumen, and three of complete interruption of the intestine at this point.

less commonly, circular and scar-like. It can be brought about, as Tillmanns points out, by an axial rotation of the gut, the result of the traction of the umbilical cord.

Voron has, among others, recorded a case of congenital atresia of the duodenum. A few minutes after beginning to suck, the child invariably vomited, unaltered, non-coagulated milk, without any admixture of bile. It died on the fourth day; its weight having decreased from 2570 grm. to 1800 grm. The post-mortem examination showed occlusion of the duodenum. The lower end of the upper, somewhat dilated, blind sac became lost in the tissues at the head of the pancreas, as did also the lower end. Between the two, there was a thin connecting band of tissue without any lumen. The bile and the secretion of the pancreas emptied themselves into the lower sac.

Kuliga records, in his very detailed and critical work, a case in which there was an hour-glass stomach and multiple stenoses of the small intestine; the formation of an artificial anus did not prevent a fatal result.

Clogg has recorded a case of atresia of the intestine in the region of the ileum.[1] The child, which had very marked distension of the abdomen, was operated upon on the fourth day of life. The lumen of the bowel was closed in the region of the lower part of the ileum, where there was present only a solid band. The condition of the child did not permit of any further operation, such as an anastomosis, and it died shortly afterwards. Another case of occlusion of the ileum has been described by Nijhoff. In this case, perforation of the intestine and escape of the meconium into the abdominal cavity had occurred before birth, so that delivery was rendered difficult owing to the marked distension of the child's abdomen.

Congenital stenoses and atresiæ of the large intestine are most commonly situated in the region of the sigmoid flexure. They may occur as a result of failure of development through rotation of the bowel, resulting from inflammation of the mesentery occurring during fœtal life. Brindeau and Moncany demonstrated in a new-born child a stenosis of the transverse colon 15 cm. in length. In consequence of the appearance of the symptoms of ileus an artificial anus was made, but the child died forty-eight hours after the operation. Jeannin and Cathala showed a perforation of the bowel in a new-born child below a complete atresia of the colon, the intestines forming a convoluted mass which could not be unravelled.

As causes of these anomalies, the most various conditions have been suggested—namely: fœtal peritonitis (in which case it is often impossible

[1] It has been suggested that the most likely cause of stenosis and atresia of the ileum is, that the process of atrophy and involution of the vitelline duct has become excessive and involved the intestine at this point.

to say whether it is primary or secondary), fœtal enteritis, occlusion of the mesenteric arteries, axial rotation or invagination of the intestine, congenital intestinal cysts, anomalies of the vitelline duct, or of Meckel's diverticulum, imperfect formation of the lumen of the bowel as a primary condition, or compression by congenital tumours as well as by the enlarged head of the pancreas.[1] The septum-like formations are generally considered as excessively developed valvulæ conniventes.[2] Bonnaire has recently described a case of occlusion of the intestine from a large congenital kidney tumour. The case died in spite of the formation of an artificial anus. On the whole, with reference to the ætiology, Kuliga is no doubt right, when he attributes the great majority of these anomalies to errors of development.

At a post-mortem examination of such cases, besides the stenosis or atresia, the intestine just above the site of the stenosis is usually found to be markedly dilated by the liquor amnii which has been swallowed. The musculature is usually also thickened, and, as a result of these changes, ulceration and perforation not uncommonly occur after birth; only exceptionally, however, does the perforation occur *in utero*. The portion of the bowel below the narrowed or occluded portion is collapsed, and the wall atrophied; while the lumen may finally become entirely obliterated.

In several cases, well-marked congenital dilatation of the intestine has been recorded without any stenosis. Thus Torkel has described a congenital cylindrical dilatation of a portion of the jejunum, without any obstruction being present in the distally situated portions; nor was any anomaly found in the structure of the dilated bowel wall. Torkel regarded these changes as due to some defect of development.

The diagnosis of these malformations of the intestines is usually attended with considerable difficulties, especially in the case of strictures. The abdomen is distended by the dilated portion of bowel lying above the stenosis. The percussion note is tympanitic; while meconium may be passed, in cases of stenosis or atresia of the upper part of the intestinal tract. In such a case, the absence of fæculent stools would first call attention to the obstruction. As watery motions can pass well-marked strictures, even for a considerable time, strictures of the small intestine remain latent longer than those of the large intestine. Stenoses of the large intestine and of the rectum, produce primarily marked constipation. In complete atresia, the symptoms are usually so acute and marked that the diagnosis of intestinal obstruction can at least be made. Of importance are the condition of the bowels, flatulent distension, the early onset of fæcal vomiting, distension of the abdomen, and colicky pains.

[1] The head of the pancreas may entirely surround the second part of the duodenum.

[2] The valvulæ conniventes, however, are, as a rule, but little developed during fœtal life.

When the occlusion is high up,[1] the course of the case is a very acute one, and, according to Jaffé, it is accompanied with marked indicanuria. In stenosis and atresia of the duodenum, below the point of entrance of the common bile duct and the pancreatic duct, marked biliary vomiting is present. The frequent and continued overflow of bile into the stomach is an important symptom in cases of stenosis low down in the duodenum; while in such cases, the stools are free from bile. In cases where the obstruction is high up, microscopical examination of the meconium may aid in the diagnosis (Finkelstein).

As nothing is carried down by the swallowed liquor amnii, no lanugo is found in the intestinal contents of the new-born child affected with atresia.

The prognosis of these anomalies of the intestine is usually bad. The majority of the cases, especially of atresia, die from inanition or from septic peritonitis, in the first or second week; less commonly, in the third or fourth week of life. Even in cases of stenosis, the children generally die, as the retained intestinal contents very soon undergo decomposition and give rise to a severe form of intestinal catarrh. Operative measures can very seldom produce any result, especially in view of the bad outlook attending such procedures in new-born children, and they are the less likely to do so because the children, when the diagnosis is made, are usually almost moribund. Finkelstein has found only one case cured by operation in the whole of the literature. This was a case of ileus, due to the compression of a peritoneal band, which was readily loosened.

Congenital Dilatation of the Descending Colon—Dilatatio Coli Congenita—Congenital Megacolon—Hirschsprung's Disease [2]

This congenital anomaly, first described by Hirschsprung and, later, observed by many other authors, consists of an abnormal elongation and widening of the descending colon and of the sigmoid flexure, up to as much as 25 cm. and more in diameter. In the majority of cases, this congenital dilatation and widening of the large intestine is a primary condition, and may be regarded as an example of partial gigantism. It can, however, also occur as a secondary condition, the result of a valve-like occlusion.[3] (Kaufmann).

These cases exhibit the clinical picture of marked constipation and distension (meteorism) of the abdomen—symptoms which become evident very soon after birth. A movement of the bowels may be

[1] If situated above the entrance of the bile-duct, there will be no bile in the vomit, which will be at first clear and then of coffee-ground colour. Jaundice is uncommon.

[2] Sometimes called neuropathic dilatation and hypertrophy of the colon.

[3] Or there may be a congenital abnormal development of the colon, so that it hangs over into the pelvis and forms a loop in which fæces and gas collect, leading to distension and even colitis and ulceration.

absent for ten or twelve days and even longer, and the discharge of meconium may be absent for a very long time. As a result of these anomalies, the children usually die of disturbances of nutrition. If the condition lasts a long time, ulcerative processes readily occur in the mucous membrane, and, following on this, perforation of the wall takes place. Treatment, whether medical or surgical—as may be seen from the literature—has not produced any good results in these conditions.[1]

Resection of the descending colon or of the sigmoid flexure, and the formation of an artificial anus may be considered as possible surgical measures to be resorted to.

Congenital Atresia Ani, and Similar Malformations

In this malformation, a congenital closure of the rectum is present so that defæcation cannot take place. Such anomalies are very frequently combined with other malformations. Thus Bürger demonstrated to the Vienna Obstetrical Society, the case of a child, eleven days old, with a congenital defect of the right lower jaw, partial stunting of the upper extremities, atresia ani, retained testes, phimosis, and hydromyelia. Maygrier and Farvy showed a fœtus with atresia of the urethra, with resulting retention of urine, absence of the right ureter, atrophy of the corresponding kidney, cystic degeneration of the opposite left kidney, atresia of the rectum with opening of the large intestine into the posterior wall of the bladder, atrophy and malformation of the extremities. Deficiency of the liquor amnii was suggested as a cause in this case. Orthmann demonstrated a child with fœtal peritonitis, double uterus and vagina, congenital hydrometra and hydrocolpos, atresia of the anus and vagina. Eustache showed a child which exhibited, besides atresia ani and recti, an obliteration of the upper end of the œsophagus. The lower end of the œsophagus communicated with the trachea. R. Freund demonstrated a seven-month fœtus with uterus bipartitus, atresia of the bladder, of the vagina, and of the anus. Freund regarded the case as one of persistence of the embryonic condition, representing the state of union of the allantois and the rectum, without the formation of any cloaca.[2]

[1] Successful cases of resection of the distended portion of intestine have, however, been recorded.

[2] The cloaca in the human embryo is formed from a posterior diverticulum of the primitive gut which grows out into the body stalk. When the posterior end of the body is formed, the hind-gut is carried with it and forms a Y-shaped cavity (the Y placed horizontally), the ventral limb of the Y forming the allantois, the dorsal the rectum, and the stem, at their junction, the cloaca. A thickening of the epiblast of the primitive streak forms an invagination or depression—the proctodæum—the floor of which is the cloacal membrane, and when this breaks down, in the fifth month, the cloaca opens on the surface. The allantois, and the Müllerain and Wolffian ducts—ventrally to the rectum—open into the cloaca (Keith).

The anomalies belonging to this series can readily be understood when the normal development of the rectum is recalled (see Kaufmann, Hertwig, Biedert and Fischl, and Tillmanns). The rectum is formed from the originally blind lower portion of the embryonic intestine, from the end or hind-gut. The anal opening itself is developed from an ingrowth from the exterior, which begins about the fourth week. The lower end of the bowel and the depression become united together and form an open canal. At this time, the gut is still in open communication with the lower end of the allantois (urachus and bladder), and also with the Wolffian ducts.

A cloaca is also present: that is to say, a communication between the urinary and genital passages and the hind-gut.[1] Usually, about the

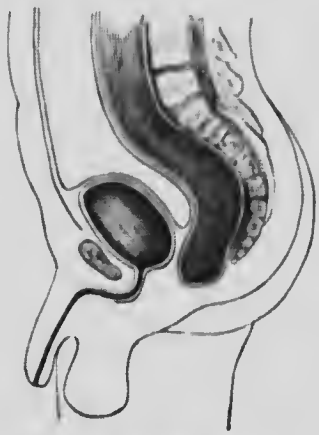


Fig. 34.—Atresia Ani. (After Leser.)

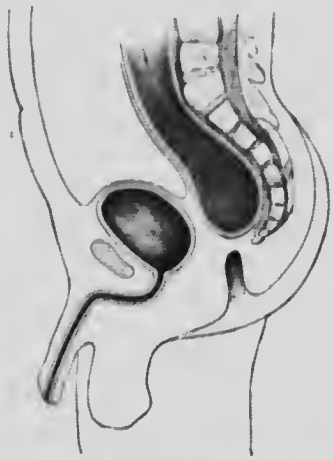


Fig. 35.—Atresia Recti. (After Leser.)

tenth week, the various parts are separated from one another. If the process described does not occur in a normal manner, it leads to the formation of an atresia ani, either with or without the formation of a cloaca. The causes of this failure of development are not very clear. According to Ahlfeld, excessive or long-continued traction exerted upon the bowel by the omphalo-mesenteric duct, may play an important part. As a result of this traction, the union of the hind-gut with the proctodæum is hindered.

The various forms of Atresia ani are as follows:—

[1] About the seventh week, the cloacal orifice of the rectum closes, and the dorsal limb of the Y-shaped hind-gut, the so-called sub-caudal gut, grows out behind the cloacal opening of the rectum towards the epiblastic invagination, forming the anal depression. The floor of this (the anal membrane) now breaks down, and the rectum opens on the surface of the body behind the perineum, which is formed by the tissues lying in front of the rectum and anal depression (Keith).

1. Atresia Ani Simplex—True Atresia Ani

The anal aperture[1] is absent; the occluded rectum does not reach the skin surface. The occlusion is due, in some cases, to an epithelial layer,[2] readily broken down by the finger; in other cases, to a more or less thick layer of tissue. In rare cases, the occlusion of the bowel is not produced by adhesions, but by a fibrinous epithelial plug or 'concretion' (see Finkelstein).

2. Atresia Ani et Recti

Rectum and anus are absent. The rectum or the colon ends blindly above; while between the colon and the perineum there is a more

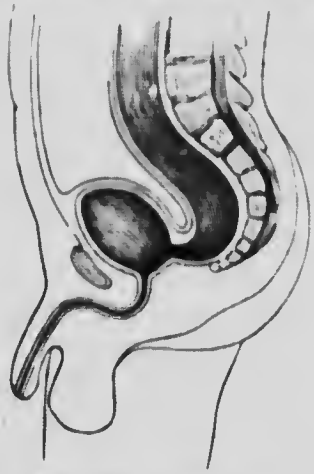


Fig. 36.—Atresia Ani Vesicalis. (After Leser.)

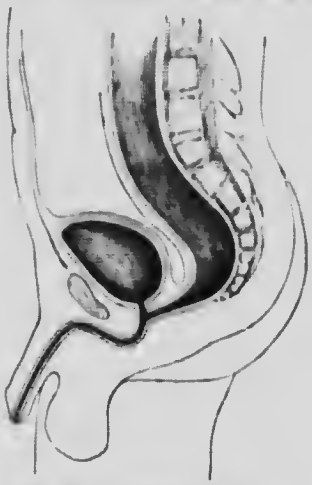


Fig. 37.—Atresia Ani Urethralis. (After Leser.)

or less wide space. The anal depression is absent, and in its place there is a flat smooth surface. In the position of the rectum there is a solid band of tissue.

3. Atresia Recti Simplex

The anal aperture is present, but ends blindly (anal depression). The rectum, which is present, is closed and reaches more or less close to the anal depression.

4. Congenital Formation of a Cloaca, with Atresia Ani

In this malformation, the anus is absent and the rectum communicates with the bladder or the urethra, in the male; with the vagina, in the female. Under this heading may be distinguished:—

(*a*) *Atresia Ani Vaginalis, seu atresia ani et communicatio recti cum*

[1] That is, the anal depression. [2] That is, the anal membrane.

vagina.—Where with closure of the anus the rectum opens into the vagina.[1] In some cases, the anal depression opens also, near the cloacal rectum, into the vagina. Atresia ani uterina is a very rare condition, in which the rectum is in communication with the uterus.

(*b*) *Atresia Ani Vesicalis, seu atresia ani et communicatio recti cum vesica urinaria.*—The anus is closed, the rectum opens into the bladder, very rarely into the ureter.[2]

(*c*) *Atresia Ani Urethralis* (*Prostatica*), *seu atresia ani et communicatio recti cum parte prostatica urethræ.*—The anus is closed, the rectum opens into the prostatic portion of the urethra,[3] or at the glans penis. A case recorded by Kafferata was of this nature. The apparently healthy child showed nothing abnormal during the first two days of life. Vomiting then set in, and in a short time meconium was brought up by the mouth. The anus was absent, and the scanty passage of meconium, which had taken place so far, was found to proceed from the urethra. At the autopsy, the sigmoid flexure was found to be enormously dilated, and to end in a blind sac resembling the rectum. A small opening, about 2 mm. in diameter, formed the communication between it and the posterior part of the urethra. The urine in the bladder was clear, but the urethra was filled with meconium.

5. Atresia Ani (seu Recti), with Formation of a Fistula.

This is not an instance of a true defect of development, but the result of pathological processes. The fistulous opening is due to an abnormal escape of the meconium, and the wall of the fistula is frequently scar-tissue, and only partly covered by epithelium. The opening may be situated in the scrotum (*Atresia ani cum fistula scrotali*); in the raphe of the penis or urethra (*Atresia ani cum fistula suburethrali*); in the vestibule of the vagina or in the region of the labia pudendi (*Atresia ani cum fistula vestibulari*); in the raphe of the perineum or of the sacral region (*Atresia ani cum fistula perineali*).

6. Absence of Rectum and Part of Colon, with Artificial Anus.

The rectum and a portion of the colon are entirely absent, but there is present an artificial anus at the navel or elsewhere.

The clinical symptoms of these anomalies do not correspond accurately to this classification. If the anus is completely absent and a cloaca or

[1] That is to say, there is a persistence of the cloacal condition.

[2] The first appearance of the kidney is a bud-like diverticulum from the dorsal surface of the Wolffian duct just above its opening into the cloaca. From this diverticulum are formed the ureter, the pelvis of the kidney, the calyces, and the collecting tubules. The persistence of the primitive anatomical relations will explain the opening of the rectum into the ureters.

[3] The primitive cloacal opening of the rectum is situated in the prostatic portion of the urethra.

fistulous opening is not present, the child, unless treated, dies with the well-known symptoms of ileus, after two to four days; seldom later. The most important symptom is the failure to pass meconium. An inspection of the anus leads at once to a diagnosis when the anus alone is closed; but if the external anal depression is normally developed (as in atresia ani simplex),[1] the same anatomical relations are found, as regards the anus, as in a normally formed child. Only after some time, when no meconium is passed, the abdomen becomes more and more distended, and the condition of the child appears unfavourable—will not take the breast and becomes restless—will the diagnosis be made; then, on the administration of an enema, an obstruction is met with and the whole of the fluid is returned.

An examination with a probe or with the little finger clears up the condition of things. According to Jakubowitsch, there is total absence of the rectum, or only a cord-like rudiment of it present, when the pelvis is very small and narrow and the perineum does not project. When the rectum opens into the bladder or the urethra, faecal impaction—with its results—very frequently occurs. Disagreeable and, in certain circumstances, dangerous secondary complications only arise if the communication is large enough, from the occasional retention of faecal masses by the action of the sphincters—for example: cystitis, pyelitis, nephritis, and abscesses in the kidney. The diagnosis in such cases can readily be made; as on passing a catheter, urine escapes mixed with meconium. The most favourable cases are those of communication of the rectum with the vagina. In these cases, the faeces, unhindered by any sphincter action, escape continually; and although the condition of the child is very disagreable, smelling as it does of faeces, and the thighs and genitalia constantly being soiled with them, yet a more advanced age may be reached (cases of Pupke and Kobrich). Tillmanns records a case in which a mother brought to him a child three years old in whom an atresia ani vaginalis had been noticed, for the first time, at two years of age. In a case, too, of atresia ani vaginalis, marked constipation may be present, as we observed in a girl fifteen years of age.

During labour, a case of atresia ani without the formation of a fistula or cloaca is of interest; inasmuch as meconium will not escape when the child is becoming asphyxiated or in a breech presentation.

The treatment of these anomalies, setting aside the cases of epithelial occlusion, which can readily be broken down, is purely surgical. The results of surgical treatment are only too often questionable, as the midwife does not readily recognise the state of things; so that the children are usually in a hopeless condition when first brought to the surgeon. The prognosis is best, in cases of membranous and epithelial

[1] This should be atresia recti simplex: see No. 3, p. 175.

occlusion of the anus. In these cases, the obstruction can readily be overcome with a trocar, a pointed bistoury, or even with the finger.

If only a thin layer of tissue intervenes between the anus and the ...ed rectum, the latter can be opened and its mucous membrane ...atured to the skin. In other cases, proctoplasty must be performed —that is, the formation of a new anus (see Tillmanns). If a successful result is not obtained in this way, then a deep dissection must be carried out from the perineum: the peritoneum opened, and the nearest coil of bowel drawn down to the perineum and opened, or a colotomy must be performed, and an artificial anus made in the sigmoid flexure. The prognosis of these operations is generally poor on account of the feeble condition of the children, which, as we have mentioned, is generally the case. As a result of this, they very readily die of peritonitis, cellulitis, or asthenia, after the operation. According to the observations of Baginsky, the operation of Callisen (that is to say, lumbar colotomy) gives bad results; while Littre's operation, with opening of the abdominal cavity in the left iliac region, is attended with nearly 30 per cent. of cures. According to Finkelstein, later statistics give on an average some 37 per cent. of cures.

Congenital Meckel's Diverticulum

Meckel's Diverticulum, which is met with fairly frequently, is the remains of what is known in embryology as the omphalo-mesenteric duct (vitelline duct). As is well known, the umbilical cord of the embryo contains, besides the two umbilical arteries and the umbilical vein, two other ducts—the omphalo-mesenteric duct and the urachus. The first passes from the navel to the lowest part of the ileum. On this portion of the bowel there develops, as the result of the traction[1] exerted by the vitelline duct, a cylindrical or button-shaped swelling, which is known as Meckel's diverticulum. The point of origin of the duct, from the intestine, is situated on its convex free border, opposite to the mesenteric insertion, about one-half to one metre above the ileo-cæcal valve, in adults, and about 3 to 4 cm. in new-born children. Its length reaches about 3 to 10 cm.; its diameter is 1·25 to 5 cm. In rare cases, it may attain a maximum length of 25 cm.; while, when least developed, it forms merely a slightly protuberant swelling on the ileum. In rare cases, it possesses a rudimentary mesentery. The structure of its walls is the same as that of the intestine; while the diameter of its lumen is almost always less than that of the bowel. That it is to be regarded as a drawn-out

[1] It is probable that a Meckel's diverticulum is due to the failure of the intra-abdominal portion of the vitello-intestinal canal to close completely, rather than to the action of any tractive force.

portion of the intestine is shown by the absence of folds in its mucous membrane, and the poor development of its muscular layers[1] (Ahlfeld). At the point of opening of the diverticulum into the bowel, a valve-like formation is sometimes observed. As has already been mentioned, Meckel's diverticulum occurs not infrequently, and is found about once in every fifty autopsies. Occasionally, a persistent umbilical vein is found as well as the diverticulum.

In rare cases, the omphalo-mesenteric duct remains completely open, and then a fæcal fistula, which leads to the ileum, is present at the navel of the fœtus (open Meckel's diverticulum). This fistulous opening becomes evident either after the separation of the cord, or if the omphalo-mesenteric duct passes into the cord at the time it is ligatured. Seitz calls attention to the fact that, in some instances, some time after the separation of the remains of the cord, the blind end of the fistula, may burst open at the umbilicus, as a result of suppuration or from increased intra-abdominal pressure (crying or straining). This condition is termed an umbilical fæcal fistula, or a diverticulum fistula of the umbilicus. These intestino-umbilical fistulæ heal very readily when the communication with the bowel is a narrow one. The wall of such a cylindrical formation shows the structure of the ileum. Lieberkühn's glands are found and, at times, also Peyer's patches. The mucous membrane of this cylindrical duct may prolapse at the umbilicus (prolapse of Meckel's diverticulum at the umbilicus), and may bleed very readily. Frequently, also, it forms a little granulation tumour on the umbilical projection (Ahlfeld). Cases have also been observed in which large portions of the small intestine have prolapsed through the umbilical opening,[2] so that symptoms of obstruction were produced. To this class, no doubt, belongs a case (imperfectly recorded by Jaswitzki in the 'Zentralbl. f. Gynäk.') in which, in a new-born child, the cæcum and the vermiform appendix were found at the umbilical ring. The further history of the case remains unknown, as the child was not brought up again by its parents to the doctor.

From the wall of these patent ducts, in later life, large tumours may develop (Kleinhans and Kaufmann). In Kleinhans' case, a cystic tumour with adenomatous proliferation was present.[3] In Kaufmann's case, a spindle-celled sarcoma, the size of a hen's egg, occurred in a woman seventy-two years of age.

In some cases, the duct has no connection with the intestine, but ends

[1] This may be equally well explained on the assumption of imperfect development.

[2] When the diverticulum alone prolapses, there is only a single orifice; but when the prolapse is more complete, the upper and lower ends of the ileum appear.

[3] So-called entero-teratomata may occur. These are small pedunculated tumours, not unlike a raspberry, which, on section, are found to be formed of Lieberkühn's crypts on a muscularis mucosa. Columnar-celled carcinoma has also been observed.

blindly. In other cases, again, the communication with the intestine remains open, but at the umbilical end it closes. Then, in some instances, tumours form, which contain meconium and which gradually increasing in size burst externally. The umbilical faecal fistulæ, just described, must not be confounded with the faecal fistulæ which are found in cases of umbilical herniæ (see p. 152). These occur, as has been pointed out, through careless ligation of the cord, so that a loop of intestine lying within the cord is included in the ligature.

In the majority of cases, the distal end of Meckel's diverticulum, is obliterated. The band so formed may be adherent to the navel, or it may subsequently become loosened and lie free in the peritoneal cavity. These bands may give rise to ileus, from strangulation (internal strangulation).[1]

Brehm has reported several such cases of ileus from Meckel's diverticulum, and has operated upon two, of which one died. The free end of the diverticulum may further become adherent in any position (mesentery, small intestine, cæcum, colon, inguinal rings, omentum, the pelvic viscera, and the abdominal wall). In other cases, an ampulliform swelling of the free end occurs, by which strangulation of the intestine with ileus may readily occur. Finally, the diverticulum may become invaginated into the lumen of the bowel (intussusception of Meckel's diverticulum). In a few rare cases, the bowel, below the point of entrance of the diverticulum, is stenosed or even atresic, so that the further prognosis is far from favourable. For the surgical treatment of these anomalies, the reader should consult the text-books on surgery. On the relation between the omphalo-mesenteric duct and the ætiology of umbilical hernia, see p. 150.

Occasionally, an aberrant pancreas, the size of a lentil or pea, is found in the diverticulum (Kaufmann).

In concluding this chapter, a few words may be said on the obstetrical importance of these anomalies. Hennig has recorded a case of a cyst of the intestinal tract associated with ascites, leading to obstruction to delivery, which was only overcome when he perforated the abdominal cavity and let out three litres of fluid. At the autopsy, he found a lax cyst in the abdomen, between the two layers of the mesentery, without any communication with the ileum. Its contents were mucoid and of a red colour; the inner wall was covered with cylindrical epithelium, and intestinal glands were found in it. Hohl mentions a further case, possibly of this nature, quoted also by Kleinhans (Peter Frank). Unfortunately, one cannot form a very clear picture from the somewhat confused description: 'Peter

[1] Meckel's diverticulum caused intestinal obstruction twenty-one times among 669 cases, in thirteen years, at the London Hospital; and of these cases no less than sixteen (or 76·2 per cent.) died.

Frank describes the case of the delivery of a child with fluid ascites (?). As the dead child, although in a good position and division (?), was not born, and the presence of ascites was suspected, paracentesis was performed with a penknife. As little water, however, escaped, delivery with some difficulty had to be completed by extraction. The reason why so little fluid escaped, appeared to be because it was collected in sacs (?) which lay among or in the intestines, especially in the large intestine (?)'.

The occurrence of obstetric injuries to the intestine—especially to the large intestine—as the result of too strong pressure on the abdomen (improper position of the hand in extraction by the breech), has been denied by various authors (see Stumpf). Such injuries (generally, multiple perforations of the intestine) may occur during spontaneous delivery, and are to be explained, probably, as the result of over-distension of the intestine with meconium, or as a consequence of malformations.[1]

Injuries to the anus, from rough examinations in the diagnosis of the presentation of the child, have been observed on several occasions (Stumpf). In some cases, the anus has been mistaken for the rigid external os; and, as the result of manual dilatation carried out on this supposition, extensive injuries to it have occurred.

Literature

Kaufmann, *loc. cit.*, p. 24; Ahlfeld, *loc. cit.*, p. 34; Keller, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Finkelstein, *loc. cit.*, p. 101; Seitz, *v. Winckel, loc. cit.*, p. 15; vol. iii., p. 180; Nothnagel, *loc. cit.*, p. 53; vol. xvii.; Leichtenstern, *v. Ziemssens Handb. d. spez., Path. u. Ther.*, vol. vii.; Hirschsprung, *Jahrb. f. Kinderheilk* (1888); Monti, *Allg. Wiener med. Zeitschr.* (1894), No. 35; Jakubowitsch. *Arch. f. Kinderheilk*, vol. vii.; Bürger, *Geb. Ges. Wien* (April 22, 1902), *ref. Zentralbl. f. Gynäk.* (1903), pp. 51 and 923; Boulounie, *Diss. Nancy* (1903); Vonon, *Lyon méd.* (April 10, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 158; Kuliga, *Diss. Heidelberg* (1903); Clogg, *Lancet* (1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 944; Nijoff, *ref. Zentralbl. f. Gynäk.* (1905), p. 1072; Jaswitzki, *ref. Zentralbl. f. Gynäk.* (1903), p. 474; Brindeau and Moucany, *Geb. Ges. Paris* (March 16, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 1387; Jeannin and Cathala, *Geb. Ges. Paris* (May 18, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 1388; Torkel, *Deutsche med. Wochenschr.* (1905), No. 9; Funck and Brentano, and Deroide, '*Hämatemesis infolge Duodenalatresie*,' *Geb. Ges. Paris.* (November 11, 1907), *ref. Zentralbl. f. Gynäk.* (1908), p. 539; Kleinhans, *v. Winckel, loc. cit.*, p. 15 and *Prager*

[1] Spencer has, however, recorded a case of hæmorrhage into the wall of the cæcum, bursting through the mucous membrane and filling the cæcum and adjacent part of the ileum with blood. The effusion caused intestinal obstruction, from which the child died on the fourth day after birth. The mother of the child was a multipara, and the labour lasted forty-eight hours, the membranes having ruptured, many hours before delivery. There had, however, not been any interference. He has also met with cases of slight hæmorrhage into the mucous membrane of the duodenum, jejunum, and ileum.

med. Wochenschr. (1907), No. 25; R. Freund, *Geb. Ges. Leipzig* (April 18, 1908), *ref. Zentralbl. f. Gyn.* (1908), p. 1147; Broca, *ref. Zentralbl. f. Gynäk.* (1908), p. 1357; Chrobak and Rosthorn, *Nothnagel, loc. cit.*, p. 53; vol. xx.; Bonnaire, *Geb. Ges. Paris* (March 19, 1708), *ref. Zentralbl. f. Gynäk.* (1908), p. 1152; Pupke, *Diss. Leipzig* (1903); Köbrich, *Diss. Halle* (1903); Cafferata, *Arch. de méd. des enfants* (1904), No. 12; Brehm, *Petersb. med. Wochenschr.* (1904), No. 24; Maygrier and Faroy, *Geb. Ges. Paris*, (March 15, 1906), *ref. Zentralbl. f. Gynäk.* (1906), p. 1137; Clochard, *Diss. Bordeaux* (1901); Orthmann, *Geb. Ges. Berlin* (Dec. 11, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 146; Eustache, *Geb. Ges. Paris* (Dec. 17, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 1087; Bauereisen, *Fränk. Ges. f. Geburtsh. u. Gynäk.* (Jan. 28, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 469; Hyde, *Ref. Zentralbl. f. Gynäk.* (1905), p. 1070; Ihl, *Festschr. f. Olshausen, Stuttgart* (1905); Hennig, *Zentralbl. f. Gynäk.* (1880), p. 398; Hohl, *loc. cit.*, p. 15; Kersten, *Berliner klin. Wochenschr.* (1907), No. 43; David, *Geb. Ges. Paris* (March 21, (1907), *Zentralbl. f. Gynäk.* 1907), p. 944 (atresia of intestine and peritonitis with a defect of the colon); Stumpf, *v. Winckel, loc. cit.*, p. 15; Fuhrmann, *Angeborene Darmatresie, Med. Klinik* (1907), No. 46; Unger, '*Fötale Peritonitis,*' *Monatschr. f. Geburtsh. u. Gynäk.* vol. xxix., p. 5.; Ballantyne, J. W., *loc. cit.*, p. 44; Keith, A., *loc. cit.*, p. 96; Spencer, H., *Trans. Obstet. Soc. London*, vol. xxxiii., p. 203 (1892); Rolleston, H. D., *Clifford Allbutt, loc. cit.*, p. 89, vol. iii., p. 564 (1907); Barnard, H. L., *Clifford Allbutt, loc. cit.*, p. 89; vol. iii., p. 704 (1907).

CHAPTER XI

Malformations and Congenital Diseases of the Liver, Spleen, and Pancreas

The Liver

Congenital alterations in the form of the liver occur which are distinguished under the name of lobed liver. In these cases, abnormal lobulation of the liver is present, due to clefts in the free margin; or a small portion is pedunculated, like a polypus, and the size of a bean or hazel-nut. As a result of these[1] constrictions and divisions, so-called accessory livers are formed. Definite duplication of the liver is extremely rare. On the other hand, the normal division of the liver into lobes may be absent. Further alterations in its form are met with—such as a triangular, quadrate, flat, broad, or round shape. In monsters, total absence of the liver has been observed. As regards alteration in the position of the liver, in cases of congenital transposition of the viscera (situs inversus), it may be situated on the left side of the body. In congenital right-sided fissures of the diaphragm (see p. 146), the liver may be displaced into the right pleural cavity. In umbilical herniæ, sometimes, a part of the liver, or even the whole, and often a considerably enlarged liver, may be found in the sac; most commonly, however, it is merely a tongue-shaped process. As Ahlfeld points out, the liver in its development tends to fill up every free space or gap.

Congenital syphilis of the liver is relatively common. In these cases, either a small-celled infiltration is present, or localised disease in the form of definite gummata. As a result, the liver is often two to three times its normal size, and its consistence is hard. In rare cases, as a result of syphilis, the liver becomes diminished in size, in which

[1] In the human fœtus, the size and prominence of the Spigelian and caudate lobes is remarkable, and the latter is often separated from the rest of the right lobe by a fissure, the caudate fissure, which may persist in the adult. When this fissure is particularly well marked, the condition resembles that found in the liver of a dog. The presence of obliquely placed fissures on the under surface of the right lobe, points to a persistence of the condition seen in the gorilla (Thomson).

case it has a more nodulated appearance.[1] This diminution in size occurs as a result of the contraction of the newly formed connective tissue. Still more uncommonly, in congenital syphilis, large gummata are found.

This variety of syphilis of the liver may occur without any marked symptoms. In other cases, however, as a result of the localisation of the process in the periportal connective tissue, the circulation through the portal vein is interfered with; as a consequence of the syphilitic shrinkage of the tissues, a large number of the branches of the portal vein are obliterated, and portal congestion with ascites and enlargement of the spleen then follows. Sometimes the enlargement of the liver is so marked that difficulty in the delivery of the child ensues, and this may be so extreme as to require operative interference for its treatment (puncture or perforation).

Congenital tumours also have been met with in the liver. The most important of these is a cavernous angioma. Thus Kaufmann observed in a new-born child an area on the right edge of the liver outside the middle line, the size of a sixpenny-piece, on which were present a large number of injected vessels. On making a section through it, a well-defined mass was found, 2 cm. broad and 1 cm. deep, which appeared partly reddish-brown, and cavernous; partly lighter, whitish-red, and compact. Microscopically, it was formed, like a cavernous angioma, of wide blood-spaces lined with endothelium; in many places communicating with one another, and elsewhere separated from one another by connective tissue septa. At the autopsy, macroscopically, it showed but little, as it scarcely projected above the upper surface of the liver; while on incision, it collapsed. Steffen has described an isolated angioma the size of an apple. Hammer recorded a case in which a child died rapidly, seven days after birth, with the symptoms of dyspepsia, icterus, and a distended abdomen. On a post-mortem examination, three cavernous angiomata were found in the liver, varying from a hazel-nut to a florin in size, of which one had ruptured. As a result of this, a fatal hæmorrhage had occurred into the peritoneal cavity.

[1] The so-called flint liver (*foie silex*), or flint liver with semolina grains (*foie silex avec grains de semoule* of the French writers). The two conditions are often combined in the same liver. The small celled infiltration is characterised by its extreme diffuseness, the whole liver being packed with formative cells, and developing fibrous tissue, or with fully developed fibrous tissue, so that the arrangement of the liver cells themselves is entirely lost. Its consistence is often compared to that of rubber (for a very good coloured drawing of such a liver, see Ballantyne, *The Fœtus*, plate 10, p. 233). By appropriate stains, the treponema pallidum may be found in large numbers. Under the microscope, the condition is one of a diffuse hepatitis ending in a pericellular cirrhosis. The gummata, probably, represent a later stage of development of the miliary syphilomata of Wagner, or the so-called semolina grains.

Congenital cysts of the liver have been described on several occasions, in some instances of so large a size as to interfere with delivery. According to Hoppe-Seyler, two forms are to be distinguished: the simple (frequently, solitary) cyst, and multiple cysts which may affect the whole liver (so-called cystic liver, cystadenoma of the liver, cystic degeneration of the liver). Both varieties have been observed congenitally. In many instances, cystic degeneration of the kidneys (see p. 197), of the ovaries, the broad ligaments, and other organs, is present at the same time. The origin of these cysts is generally to be referred to some defect of development in the intra-hepatic system of bile-ducts[1] (Kaufmann). The inner wall of the larger cysts is lined by flattened, cylindrical, or ciliated epithelium. Frequently, in cases of cysts of the liver, ascites, and swelling of the spleen, the results of the portal congestion, have been observed (For further details see Kaufmann.)

For the obstetrical importance of these tumours, see p. 187.

As a result of the unskilful employment of obstetrical procedures—especially in extraction by the breech—ruptures of the liver have been observed on several occasions. In severe ruptures of the liver,[2] death from internal hæmorrhage is unavoidable. Some obstetricians have met with such injuries to the liver after the employment of Schultze's method of treating asphyxia neonatorum. With regard to all such assertions, as Runge points out, one must be very sceptical. The effusions of blood, not infrequently observed under the peritoneal covering of the liver,[3] are of a different nature to the hæmorrhages due to rupture of the liver. In these cases, the parenchyma of the liver is uninjured. They are not, as Stumpf has rightly pointed out, to be laid to the blame of the obstetrician, but occur as the result of marked venous congestion from asphyxia of the child.

Lastly, I will mention very briefly the changes met with in the fœtal organs in eclampsia of the mother. On the whole, the same pathological

[1] In 19 per cent. of all cases of cystic kidney, the liver is also cystic. The single cyst is, as a rule, only found in women; and it has been suggested that it is due to dilatation of the bile-ducts, with their subsequent fusion, to form a single cyst. In other cases, large numbers of these cysts may be present, and the liver may be greatly enlarged by them. In cystic degeneration of the liver, the cysts are said to be always associated with congenitally aberrant bile-ducts.

[2] This is an exceedingly rare injury and only occurs, as a rule, with fracture of the ribs or of the clavicles.

[3] Spencer regards these hæmorrhages as due to pressure on the abdomen, and points out that they do not depend upon the variety of the presentation, but are especially liable to occur when the liver is pressed upon from some cause—such as a large size of the abdomen, or small size of the cervical canal, or in an impacted breech. He further shows that the hæmorrhage often occurs in the area of the liver least supported, rather than at the point of greatest pressure.

changes are met with as in the mother—especially in the central nervous system, the liver, and the kidneys.[1]

The enlargements of the liver, mentioned above, have at times an obstetrical interest, since, as a result of their degree of development, or of the ascites so often present at the same time, the delivery of the child may be rendered difficult or impossible without artificial aid.

Sänger and Klopp have recorded a very interesting case: that of a premature female child which was delivered rapidly in a vertex presentation as far as the shoulders, but then stuck fast. The medical man, called in after some hours, delivered the child; but, from the vigorous traction exerted during this procedure, the abdominal walls burst and a large amount of fluid escaped. The post-mortem examination showed total situs inversus of the organs of the abdominal and thoracic cavities; the circumference of the abdomen, measured above the umbilicus, was 49 cm.; an umbilical hernia, sixteen separate spleens, and some large cysts (the largest, the size of a child's head) were present in the right side of the abdomen. Examination of the cysts showed that they were composed of cystic formations of the intestines and of the accessory bile-ducts. For further details, I must refer the reader to the original work.

Witzel described a case of hemicephalus with large cysts of the liver and of the kidney, and other malformations, in which, after delivery of the hemicephalic head, the labour came to a standstill. By puncture of the enormously distended abdomen with the scissors perforator, Witzel let out three litres of yellow fluid. The cystically enlarged liver almost filled the abdominal cavity; it was larger than the liver of an adult, and had an irregular knobby surface. Each lobe of the liver was transformed into a large cavity. Each kidney was the size of a man's fist, and was beset with a number of small cysts—the largest of which did not, on an average, exceed the size of a pea.

Only a few isolated cases of difficulty in delivery, from tumours of the liver, are to be found in the literature. Hohl describes a case, published by Seulen, in which the liver was enlarged and general dropsy was present, with resulting difficulty in delivery. He quotes, further, a case recorded by Haase, in which the child, in an oblique presentation with a prolapse of the cord, and a liver enlarged by 'Physokonia,'[2] was delivered by version, extraction, and eventration. Hohl also describes the following case. Müller made an incision in

[1] Degeneration of the liver-cells and necrotic foci have been described, with congestion of Henle's tubules and extravasation of blood into the kidneys, and hæmorrhages into the brain and spinal cord. All these changes, however, may be found in fœtuses dying during delivery from other causes.

[2] Physconia, from *φύσκων* ('big-bellied'), is usually employed to mean an enlargement of the abdomen due to a slow-growing tumour.

the side of a dead child with the head, shoulders, arms, and thorax born, and which would not advance any farther. From the incision, a large quantity of fluid escaped; after which, the extraction was readily accomplished. Besides the collection of fluid, he found a lymphoma of the liver, weighing 4 lbs. He further records a case (described by Schlesinger) of a child with the head born as far as the neck: while the abdomen of the mother was still of enormous circumference, although the liquor amnii had escaped. On account of dropsy of the abdomen of the child, Schlesinger performed paracentesis, in the absence of a trocar, with the blade of a pair of long scissors. A large quantity of serous fluid escaped, after which the child was instantaneously born. The feet of the child were œdematous. Schlesinger found the liver and the kidneys unusually large. Kleinhans has also recorded other cases from the literature. Nöggerath's case: Difficulty in delivery, due to a tumour of the liver, weighing 2½ lb., which, microscopically, appeared to be a carcinoma. W. S. Bagot's case: The mother was syphilitic, and the labour was ended by puncture of the abdomen of the fœtus below the xiphoid cartilage. The autopsy showed a cyst in the left lobe of the liver, containing 1½ litres of fluid. Porak and Convelaire's case was that of a child with a cyst[1] in the liver, giving rise to difficulty in delivery, together with cystic kidneys, exencephalus, achondroplasia, and polydactyly.

General rules for the conduct of the labour in such conditions of the liver, naturally, cannot be laid down. Hohl's optimistic remarks on this point have hardly any practical value. The principal points are these: In no case should force be employed when there is delay in the delivery of the child; an examination should be carried out with the whole or half-hand, under an anæsthetic, and an attempt made to remove the obstruction by the rotation of the abdomen of the child into the roomy hollow of the sacrum; and, lastly, whatever operative procedures may be indicated should be carried out (puncture or evisceration).

As an appendix, I will mention, briefly, some cases of congenital obliteration of the bile-ducts, and some cases of atresia of the common bile-duct. The first case is one recorded by Emanuel. The child, born of healthy parents, became jaundiced on the fourth day after a normal delivery, and passed watery stools without any bile, with resulting loss of weight. The liver and spleen were enlarged and hard. The anti-syphilitic treatment prescribed had no result, and the child died at the fourth month. The autopsy showed complete obliteration of the bile-ducts, while the gall-bladder was rudimentary. In the liver, marked

[1] The cyst wall showed two layers: an inner of connective tissue, and an outer of liver tissue. Most of these single cysts are covered by a thin layer of liver substance.

biliary cirrhosis was found with marked atrophy of the parenchyma. The pancreas and spleen showed extensive fibrosis.[1] Another very interesting case of total atresia of the common bile-duct is recorded by Würtz. The child, a boy weighing 6¼ lb., was born of healthy parents. During labour, the child passed white fatty stools. The fœtal heart-sounds were very good during the second stage. In spite of this, after birth, it was only by the employment of marked external stimuli that the child could be made to cry and take deep breaths. The skin was slightly jaundiced. After the third day, the stools were quite white, the urine dark, and the icterus much more marked. The cord separated, and the stump healed, without any disturbance. The temperature was normal; but the child exhibited slight stupor. On the eighth day, marked eructations occurred. During the following night, small hæmorrhages took place from the mouth, and there were small ecchymoses on the tip of the tongue and on the gums. In the course of the day, the hæmorrhage increased; and towards evening, melæna set in. The child was definitely wasting; the jaundice[2] markedly increased, and hæmatemesis occurred. On the tenth day, there were more black stools, and the urine contained albumen and bile pigments. The umbilicus (which hitherto had been dry) was found to be damp with a blood-stained discharge. There was no fever. Severe bleeding, which could only be arrested by the cautery—gelatine producing no result—followed the taking of blood from the lobe of the ear for bacteriological investigation. With increasing stupor and marked jaundice—almost of a bronze tinge—severe umbilical hæmorrhage occurred on the eleventh and twelfth days, which could not be arrested in spite of the employment of acupressure, plaster of Paris,[3] adrenalin, and compression. The hæmatemesis and melæna ceased; but shortly before death, on the twelfth day, diffuse hæmorrhages[4] occurred under the skin in various parts of the body. A post-mortem examination

[1] In the majority of these cases, congenital syphilis is not an essential element in the causation. The sequence of events in this condition is probably as follows: A congenital narrowing of the bile-duct of developmental origin produces stagnation of the bile in the liver, with resulting retention of irritating metabolic products. The elimination of these toxic bodies leads to inflammatory changes in the ducts and their final obliteration, with necrosis and cirrhosis of the liver. After birth, the liver is consequently unable to carry out its protective functions properly; toxic bodies accumulate, and the child dies from toxic poisoning. In the same way, the fibrosis of the pancreas and spleen is explained (Thomson).

[2] Jaundice is always a very marked symptom, but varies in the time of its onset. It may be present at birth or may not be noticed until the second or third day of life; or, in some cases, not until the end of the second week.

[3] Hæmorrhage from the navel very often occurs in these cases after separation of the cord, and is always very difficult to stop.

[4] Thomson points out that, in those cases in which more than one member of a family is affected, the tendency to hæmorrhage is particularly marked and the child rarely survives the first fortnight.

revealed complete atresia of the common bile-duct in an otherwise normal child. Würtz was able to collect from the literature ninety-five similar cases. He considers, with Heneke, that the atresia of the large bile-ducts is a malformation. Perhaps, the condition is one of 'constriction from internal causes.'

Chivie demonstrated the liver of a new-born child which died on the seventh day after birth. It had never passed meconium, and the stools which were passed later were colourless. Death ensued with increasing icterus. At the autopsy, all the tissues were found infiltrated with bile pigment, and the liver was coloured green.[1] The common bile-duct was permeable only as far as its entrance into the hepatic duct. The latter and the cystic ducts were obliterated. The gall-bladder was dilated and filled with mucoid colourless contents. Kaufmann, in a new-born child, observed absence of all the bile-ducts, from the hilus of the liver to the duodenum.

Further anomalies of the bile-ducts are: abnormal opening of the papilla—namely, too big an opening in the region of the pylorus; doubling of the orifice; doubling of the common bile-duct. Total absence[2] of the gall-bladder has been observed (see Kaufmann: two cases in adults), and also duplication of it. In this case, the bile-duct may be double or it may be single, and then the two gall-bladders may be united to one another by a separate duct. Ahlfeld has described a very interesting case of this kind. In a full-term child a cyst, the size of an ostrich's egg, was found in a large umbilical hernia, which had rendered delivery difficult. The cyst was found, on examination, to be a second—or, at any rate, a separated—portion of the gall-bladder. The nature of the contents was in favour of this view, as well as the fact that the liver substance spread out over the cyst in an arborescent-like manner. On the upper aspect of the cyst, there was present a second diaphragm enclosed in a trefoil manner by the liver tissue. Besides this, situs inversus of the heart, liver, and spleen was present.

Literature

Kaufmann, *loc. cit.*, p. 24; Ahlfeld, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Strümpell, *loc. cit.*, p. 34, vol. ii., p. 276; Quincke, and Hoppe-Seyler, *Nothnagel, loc. cit.*, p. 53; vol. xviii.; Steffen, *Jahrbuch für Kinderkrankheiten* (1883), vol. xix., p. 348; Witzel, *Zentralbl. f. Gynäk.* (1880); Sänger and Klopp, *Arch. f. Gynak.* (1880), vol. xvi., p. 415; Kleinhans, *v. Winckel, loc. cit.*, p. 44;

[1] The green colour is due to innumerable small plugs of inspissated bile, which are found to distend the smaller bile-ducts in many places.

[2] This may occur with congenital obliteration of the bile-ducts, and is the normal condition in some animals.

Hohl, *loc. cit.*, p. 15; Gessner, *Diss. Halle* (1888); Hammer, *Zeitschr. f. Geburtsh. Gynak.*, vol. iii.; Paul Bar, *Lehrbüchrigne Jahrg.*, vol. viii., pt. 4; Chirie, *Arch. de Paris* (Dec. 10, 1896), *ref. Zentralbl. f. Gynak.* (1896), p. 1423; Runge, *loc. cit.* p. 129; Stumpf, v. Winckel, *loc. cit.*, p. 44; Birnbaum, *loc. cit.*, p. 101; Emanuel *Brit. Med. Journ.* (Aug. 17, 1907); Würtz, *Med. Klinik.* (1908), No. 52 (with literature); Thomson, A., *Journ. Anat. and Phys.* (N.S., 1899), vol. xiii., p. 546; Spencer, H., *loc. cit.*, p. 89, vol. xxiii., p. 272 (1892); Thomson, J., *Clifford Allbutt, loc. cit.*, p. 80; (1908), vol. iv., pt. 1, p. 103.

The Spleen

Aplenia—congenital absence of the spleen—has been very rarely observed. Either it is absent in monsters—such as the true acephali, where, generally, at the same time other organs are absent (liver, stomach, pancreas, and peritoneum)—or it is absent[1] (a very rare condition) in otherwise completely normal individuals (Lemery, Schenk and Graffenburg, Sternburg, and others). Abnormal smallness of the spleen occurs, primarily, in immature fœtuses. An abnormally large spleen is occasionally observed in monsters. Naturally, I do not include here enlargement of the spleen from syphilis, septic infection, or portal congestion. Numerous notches may be found (Crenæ lienales), or division into several parts; while the shape, according to Litten, may be round, tongue-like, disk-like, cylindrical, crescentic, triangular, or quadrate. It may also be double as broad below as above (as in most of the quadrupeds). Double spleens, of very similar shape and form, have also been described. In many instances, there are present at the same time other malformations (absence of the pancreas, or hour-glass stomach). The so-called accessory spleens are distinguished as Lien succenturiatus, Lien accessorius, or multiple formation of the spleen.[2] Generally, they are small structures of varying size; most commonly, round, of similar shape to the spleen, and of the same consistence and colour. They occur in the ligamentum gastro-lienalis, in the mesentery, and the head of the pancreas. Albrecht found, in one case, 400 accessory spleens scattered all over the peritoneum (see Kaufmann). In some malformations, for example, in diaphragmatic hernia—it is almost the normal condition to find several accessory spleens (Ahlfeld). In disease of the spleen itself, the accessory spleens are also sympathetically affected (swelling in leukæmia, and typhoid).

[1] In a case of absence of the spleen, lymphatic hyperplasia elsewhere has been described (Hodenpyl, quoted by Rolleston).

[2] There are two possible explanations of these accessory spleens: they may be separated-off portions of the spleen connected by blood-vessels only, or isolated fragments of the mesoblastic tissues which form the main organ—comparable, therefore, to suprarenal rests. They may become embedded, secondarily, in other organs; and it has been suggested that cavernous angiomata of the liver are, in reality, accessory spleens embedded in that organ.

Congenital displacement of the spleen has been frequently observed. By ectopia of the spleen is understood its displacement in congenital diaphragmatic hernia, into the pleural cavity: and into the hernial sac, in umbilical hernia[1] and in large clefts of the abdominal wall. At an autopsy, on a case of congenital hydrocephalus, I found the spleen lying close to the spinal column in the left pleural cavity. Litten quotes a very remarkable case observed by Preus. The latter, after opening the perfectly normal stomach, found the spleen small but normally developed free in the cavity, attached only by its vessels to the mucous membrane. The so-called 'wander spleens' have also been observed as a congenital condition.[2]

In congenital syphilis, the spleen is relatively frequently affected. The disease, as in other parenchymatous organs, occurs in a diffuse or, less frequently, in a circumscribed form. Perisplenitis has also been observed in these cases. The weight of the spleen may be very remarkably increased in consequence. The normal weight, according to Kaufmann being 9 grm.; in congenital syphilis, it may weigh 20 to 40 grm. and even as much as 100 grm.[3] Tumours of the spleen have also been observed, according to Mannaberg, in several instances, as a congenital condition when malaria from the mother has affected the child *in utero*. Such intra-uterine infection is, however, of very rare occurrence.[4]

In very rare cases, tumours of the spleen have given rise to difficulty in delivery. Such cases have been [illegible] by Spiegelberg and Kleinhans (Petit-Maugin, Francis, and W[illegible]). Obstetric injuries of the spleen—rupture of the spleen, with haemorrhage—are very uncommon.[5] They may occur from the unskilful and forcible extraction of the child in pelvic presentations (case of Ballantyne; see Stumpf and Wyder).

[1] Of 330 cases of diaphragmatic hernia collected by Rochard, the spleen was involved in seventy-eight.

[2] Rolleston quotes a most remarkable case in which a long process of the spleen, bound down to the posterior wall of the abdomen by the peritoneum, passed down into the scrotum; no doubt carried down in the descent of the testis.

[3] Hereditary syphilis is no doubt the commonest cause of enlargement of the spleen in the infant. Thus in 348 children, under twelve years of age, with splenomegaly, syphilis was present in 147, or 42·2 per cent. (Carpenter).

[4] The possibility of such transmission is still a disputed point. In those cases in which the organisms of malaria have been found in the blood of the new-born child, in all of them a sufficient length of time had elapsed after birth to allow of a fresh infection. On the other hand, in a number of recorded cases of infants born of malarious mothers, examination of the blood has not shown any evidence of fœtal infection; although parasites were on one occasion demonstrated in the maternal part of the placenta.

[5] Spencer found hæmorrhage into the spleen only three times among 130 still-born children. He explains this infrequency by the small size, deep position, mobility and distensibility of the organ.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, p. 34; Litten, *Nothnagel, loc. cit.*, p. 53; vol. viii.; Wyder, *v. Winckel, loc. cit.*, p. 15; Stumpf, *v. Winckel, loc. cit.*, p. 44; Kleinhans, *loc. cit.*, p. 34; Petit and Maugin, *Gaz. méd.* (Paris, 1833); Francis, *Med. Press. and Circ., ref. in Zentralbl. f. Gynäk.* (1884), p. 253; Webber, *Med. Press and Circ.* (London, 1883); Marfan, *Revue mens. des malad. de l'enfance* (1903), *ref. Zentralbl. f. Gynäk.* (1903), p. 1520; Mannaberg, *Nothnagel, loc. cit.*, p. 53; vol. ii.; Hodenpyl, *Proc. Path. Soc.* (New York, 1899), p. 185; Rolleston, H. D., *Clifford Allbutt, loc. cit.*, p. 89; vol. iv., pt. 1, p. 443 (1908); Rochard, *Les Hernies*, p. 382 (Paris, 1904); Carpenter, *Brit. Med. Journ.*, vol. ii., p. 463 (1903); Spencer, H. R., *loc. cit.*, p. 89; vol. xxxiii., p. 284 (1892).

The Pancreas

In rare cases, pancreatic tissue is found in atypical places: for example, on the stomach, the duodenum, the jejunum, and the ileum[1] (accessory pancreatic gland, pancreas accessorius). These structures are composed of portions of the gland about the size of a shilling. In rare cases, the combination of a Meckel's diverticulum with an accessory pancreas is found. Actual anomalies of the pancreas have also been observed. Of this character, are the separation of the head or of the tail of the gland, or the division of the pancreas into two equal or unequal parts, or a collar-like arrangement of the pancreas round the descending portion of the duodenum (see Oser). Situated at the head of the pancreas, an appendage is at times found which has been distinguished as the pancreas minus (Ahlfeld).[2]

Not uncommonly, variations of the excretory duct have been observed.[3] Congenital absence of the pancreas is very infrequent and occurs only in malformations—most frequently with malformations of the intestinal tract. Very rarely, small accessory spleens are found embedded in the substance of the pancreas. Congenital displacements of the pancreas are uncommon, but have been observed in diaphragmatic and umbilical herniæ.

In congenital syphilis, the pancreas at times is found indurated. Microscopically, it shows (as in other parenchymatous organs) a diffuse small-celled infiltration, or a circumscribed affection in the form of gummata.[4]

[1] In these cases, the nodules of glandular tissue may be situated on the summit of intestinal diverticula—apparently, the result of traction exerted by the rudiment.

[2] This may be due to the persistence of the left of the two diverticula of the ventral pancreas which springs from the base of the bile-duct. According to some authorities, only the right ventral diverticulum usually persists (Helly).

[3] The opening of the duct of Santorini most commonly becomes obliterated; the presence of a third duct is, however, not uncommon.

[4] Pancreatic lesions are much more common in inherited than in acquired syphilis; and, most probably, this organ is always affected when the liver is.

Cystic degeneration of the pancreas has frequently (Beuker) been described together with cystic disease of the kidneys and other malformations. Glaug has described a case in which, on account of dystocia, eventration had to be performed.

Small disseminated hæmorrhages into the tissues of the pancreas are not uncommonly met with in new-born children who have died, during birth, from suffocation. Ipsen has observed a very extensive pancreatic hæmorrhage; and in this case he found, at the autopsy, that there was a large subcapsular and interstitial hæmatoma. Other changes—such as might have been due to syphilis—were not recognisable. Ipsen explained the hæmorrhage as caused by severe pressure on the child—the result of clumsy manipulation at the time of the birth. Death, in this case, occurred (in my opinion) in the same manner as in Goltz's tapping experiment: namely, as a result of reflex failure of the heart's action through the solar plexus situated over the pancreas.

Literature

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; Beuker, *Diss. Erlangen* (1902); Glaug, *Diss. Leipzig* (1904); Ipsen, *Verhandl. deutsch. Naturforscher u. Ärtze* (Dresden, 1907), *ref. Zentralbl. f. Gynäk.* (1907), p. 1358; Oser, *Nothnagel, loc. cit.*, p. 53; vol. xviii.; Ahlfeld, *loc. cit.*, p. 34; Helly, *Arch. f. Micros. Anat.*, vol. lxxvii. (1905).

CHAPTER XII

Malformations and Congenital Diseases of Kidneys, Suprarenals, Bladder, and Urethra

The Kidneys

Malformations of the kidneys occur relatively frequently; and, as a result of the close connection between Müller's ducts and the urinary ducts, malformations of the genital organs are often found at the same time.

Complete absence of both kidneys, or the presence of only an undeveloped rudiment, is very rare and occurs only in monsters incapable of living. Moulon, indeed, has recorded (Senator), in a girl fourteen years old, complete absence of both kidneys, of the ureters, and the bladder; but this observation must have been founded on some gross error.[1] Absence of one kidney (agenesia or aplasia) or its maldevelopment (hypoplasia) has been met with not infrequently.[2]

Thus, recently, I saw in an otherwise normally developed child—still-born as the result of a placenta prævia—complete absence of the left kidney and suprarenal body, together with the ureter and renal vessels. The right kidney was not enlarged, and there was only one ureter present. In congenital defective[3] development of one kidney, the urinary tubules and glomeruli may be entirely absent. When one kidney alone is defective, the other kidney almost always takes on its functions and becomes markedly hypertrophied. The defect is almost always left-sided, and the male sex is affected twice as often[4] as the female. The cause of this unilateral kidney defect

1 Probably, as Ballantyne suggests, the kidneys were misplaced and not really absent.

2 Winter gives a total of 237 cases. In 98 of these, the kidney was absent on the right side, and in 129 on the left side.

3 When only one ureter is present, it may open in the middle line. When only one renal mass is found it may be: (1) a normal kidney, the second being entirely absent; (2) a fused kidney; (3) a normal kidney, the other being represented by a small mass of tissue—perhaps, presenting some traces of renal structure (Morris).

4 This may be only apparent, and due to the greater frequency of autopsies in men.

has been explained by the hypothesis that the outgrowth of the primary kidney from the Wolffian duct is in some way hindered.[1] The ureter is usually absent in these cases as well as the renal arteries and veins. Less commonly, two ureters take origin from one kidney and pass separately to the bladder. Occasionally, it happens that while two ureters pass from the bladder towards the kidney region, only one of them reaches the kidney, the other ending blindly.[2] While in the presence of a well-developed kidney no disadvantage results, yet the defect may be of great practical importance if this kidney is diseased. If a cystoscopic examination has not been undertaken, and it is removed, a fatal result is naturally unavoidable. Undoubtedly, this occurs more often than is evident from the literature.

In other cases, the condition is that, apparently, only one kidney is present. This anomaly arises from more or less extensive union of the two kidneys. This union can readily occur when the primitive rudiments of the kidneys lie too near together, or the ends of the renal ducts growing up come into union with one another too soon. All possible gradations of this union (Ren concretus), which is often associated with a displacement (Dystopia renis), may be observed. The horse-shoe kidney (Ren arcuatus) is most frequently met with. In this condition, the lower poles of the usually slender and elongated kidneys lie close together, and are united either by connective tissue or by kidney substance: the two kidneys forming a curve, with its concavity upwards. The ureters are, usually, normal; but all possible variations may occur (see Kaufmann). Such horse-shoe kidneys are frequently displaced to the region of the sacr[1] promontory, and, as a result, errors in diagnosis may occur (confusion with ovarian tumours, etc.). Less commonly, the upper poles are united;[3] and less commonly still, the median borders are completely fused with one another (Cake kidney). Finally, the kidneys can also be placed behind and over one another.

By dystopia of the kidney is understood, what we have already shortly alluded to—namely, the displacement of one or both kidneys. It occurs especially often when the two kidneys are united into the

[1] The first appearance of the kidney in the human embryo is seen at the beginning of the second month, in the form of a bud-like diverticulum, which grows out from the dorso-mesial aspect of the Wolffian duct, immediately in front of its opening into the cloaca. This diverticulum, together with a mass of mesoblast from the posterior segment of the intermediate cell-mass, forms the kidney. The ureter, pelvis, calyces, and collecting tubules, arise from the epithelium of the diverticulum; while the glomeruli, vessels, and connective tissue, arise in the mesoblast.

[2] No doubt, the result of the development of two diverticula instead of one, as the subdivision of the diverticulum, may lead to the formation of several ureters.

[3] The isthmus is placed above in only about 7 per cent.; and when this is the case, it is somewhat more commonly behind the aorta.

horse-shoe kidney. A displacement of both kidneys, when not united, is very uncommon. A unilateral (usually left-sided) displacement is most frequently met with, and more frequently in men than in women (20 to 9).

The displaced kidney is almost always situated in the region of the sacral promontory; sometimes, even lower down in the pelvis[1] (Pelvic kidney). In one case, it was found in the inguinal ring. The course and the situation of the vessels is variable (see Kaufmann); the suprarenal glands, however, are almost always placed in their normal position. Very frequently, a malformation of the genital organs is present; less frequently, one of the intestine. In the majority of cases, such displacements possess no clinical interest, as they are symptomless. Mistakes in diagnosis[2] and confusion in connection with tumours of the tubes and ovaries have occurred.

As a result of such errors in diagnosis, the removal of displaced kidneys has been carried out in cases of united kidneys, naturally, with fatal results. In the Göttingen Polyclinic, I was able recently, to diagnose such a horse-shoe kidney, which lay in the hollow of the sacrum behind the uterus, and yet closely resembled a retro-uterine tumour of the adnexæ, suitable for operation. It is evident that these tumours, especially when enlarged and fixed in the true pelvis, may give rise to difficulty in delivery.

As regards the external form of the kidneys in new-born children, so-called fœtal lobulation[3] and segmentation is met with. These divisions may, in certain cases, affect the whole thickness of the organ, so that completely separated portions of kidney may result.[4] Such so-called 'stray' or 'isolated' kidneys may, later, give rise to the formation of malignant kidney tumours. Kaufmann, in a man sixty-six years of age, saw a complete division of the kidney into two equal portions, with two calyces, but a single pelvis and one ureter. As a rule, the formation of supernumerary kidneys is of the rarest possible occurrence (Ahlfeld). Among the more frequent malformations of the pelvis of the kidney, is its duplicature or division into several portions with a single or several ureters. The ureters may be double on one side and

1 When the left kidney occupies the iliac fossa on that side, the descending colon generally crosses the middle line, and the rectum commences on the right side of the sacrum.

2 The diagnosis must be made by the form, unchanging size, and consistence, of the misplaced body, together with the absence of one or other kidney from the loin.

3 Up to the time of birth, the lobules remain distinct; and this is the normal condition in some mammals. Usually, after birth, the external signs of lobulation disappear; but they may persist.

4 The commonest of all variations from the normal size or shape is the so-called congenital atrophy. In these cases, the kidney may be lobulated as well as small; but always possesses an amount of secreting structure weighing from three-quarters of an ounce or less to one ounce and a half, or a little more (Morris).

open separately into the bladder. Such a condition, I have recognised, recently, on two occasions on making a cystoscopic examination. It may also happen that the ureters are double above and then unite; or one ureter may open in its normal position in the bladder and the other in an abnormal position into the urethra just below the bladder, into one vesicula seminalis, the vas deferens, on the colliculus seminalis, into the uterus, vagina, or vestibule.[1] Further, stenoses and atresia of the ureter, on one or both sides, have been observed with either the development of consecutive hydronephrosis to a greater or less degree, or else complete atrophy of the kidneys.

The vesical opening of one, or (as Kaufmann has also observed) of both ureters, may be so narrowed that the ureter forms a cyst,[2] bulging towards the bladder, and may ultimately lead to difficulty in the outflow of urine from the other ureter (see Kaufmann).

Finally, valvular formation from folds of the mucosa, abnormal kinking or torsion,[3] or insertion at too acute an angle into the pelvis of the kidney with resulting hydronephrosis, have also been observed in the ureters.

For obstruction to delivery from dilatation of the ureters, see p. 215.

Congenital Cystic Kidneys—Hydrops Renum Cysticus Congenitus

Congenital cystic degeneration of the kidneys is, as a rule, bilateral. Lejars, among sixty cases, only found one example of unilateral cystic degeneration (Senator). The cysts are of varying size, from that of a pea to a cherry; rarely the size of a fist. In this form of cystic degeneration, the shape of the kidney may remain unaltered as a whole, and a section then shows a honeycomb-like appearance; or the cysts may arise more or less from the superficial aspect of the kidney, and then there results a grape-like appearance, similar to that of the racemose ovarian cysts. The substance of the kidney (the parenchyma) is either entirely wanting, or persists only in a fragmentary condition in the form of islands or bands in the sometimes thin and sometimes thick septa. The size of a congenital cystic kidney may be such that, as we shall see later, it can give rise to difficulty in delivery. According to Kleinhans, the distension of the kidney, in marked cases, may reach up

[1] These anomalies are readily explicable when we remember the relation of the lower end of the ureter to the primitive cloaca.

[2] In some cases, congenital prolapse of the ureter occurs—the vesical end forming a pendulous pouch hanging into the interior of the bladder; and in the female, it may even project at the urethra. The orifice is often non-existent, or of pin-hole size, and the patient is very likely to suffer from septic nephritis.

[3] Congenital valves and congenital torsion of the ureter are both, usually, situated near the renal end of the duct.

to 10 to 20 cm., and its weight may be increased even up to 2250 grm. In such cases, the kidneys may reach to the linea innominata, or still farther downwards. The contents of the cysts are, most commonly, a clear serous fluid of a golden or citron-yellow colour, less commonly, mucoid, milky or colloid, and of a reddish-brown or chocolate colour, as a result of intra-cystic bleeding.[1]

The smell of the fluid is at times urinous, or ammoniacal; its reaction neutral or alkaline. On chemical examination, it is found to contain albumen, urea, uric acid, and oxalate of lime; microscopically, red and white blood corpuscles, fat, epithelial and granular debris are recognisable. The process may make its appearance even in the first months of embryonic life. Thus I have seen marked cystic degeneration of both kidneys in a fœtus expelled at the third month of pregnancy. The kidneys had the appearance of a sponge with fine pores, or a compact mass of sago—as Kaufmann has very aptly described it. The cyst wall was composed of fibrous connective tissue, which was covered on its inner surface, in many places, with a layer of flat polygonal epithelium. In some cysts, this epithelium resembles closely the epithelium of the urinary tubules (Senator). Congenital cysts of the liver are often found in association with cystic degeneration of the kidneys; and Kaufmann has found it also present at the same time in the pancreas. Other malformations are frequently present with cystic kidneys: thus atresia of the urethra, of the anus, obliteration of the ureters, defects of the bladder, double uterus or vagina, hernia of the brain, hare-lip, and clefts of the hard palate.

Fig. 38.—Cystic Kidney. (After Kaufmann.)

Freund demonstrated to the Leipzig Obstetrical Society the case of a child with bilateral cystic disease of the kidneys, cysts of the pancreas, and complete situs inversus—a very rare complication. There was serious difficulty in delivery. After the birth of the head, the delivery of the trunk did not follow, and the head tore off. Delivery was finally completed by version and extraction, after evisceration had been practised. Freund regarded a failure of development as the cause.

As to the mode of development of cystic kidneys, opinions are very

[1] Sometimes the contents of the cysts are solid, in which case they consist of particles of fat, epithelium, and crystals of cholesterine, uric acid or triple phosphates.

divergent. Klein, Rosenstein, Brigidi, and Severi (Senator) maintain that the first factor in the formation of the cysts is hæmorrhage into the glomeruli, by which the escape of the transuded urinary secretion is hindered, and the capsule stretched and converted into a cyst. This theory—a very possible one for the slighter degree of cystic formation—fails as regards the larger and more voluminous cystic kidneys. According to other views, there must be, somewhere, an obstruction to the escape of the urine. This obstruction cannot be present in the larger excretory ducts—from the pelvis of the kidney downwards—or it would lead to the development of a hydronephrosis. It must be sought higher up. According to Virchow, this obstacle is found in an atresia of the papillæ, so that the opening of the collecting tubules into the calyx of the kidney is in many places interfered with, or entirely closed. The atresia of the papillæ is produced by inflammatory conditions with their sequelæ; for instance, by small uric acid concretions—embryonic nephritis papillaris or pyelonephritis. Other authors (see Kaufmann) place the inflammatory changes between the lobules of the kidney in the straight arterioles; while primary fibrous nephritis has also been suggested, in some cases (Arnold and Kaufmann). A further group of authors see the cause in a malformation or some defect of development. In favour of this view, is the fact that other malformations are so often present with cystic kidneys, and also (as has been found in a number of cases) that cystic kidneys have been met with in several children of the same mother. A partial impermeability of the urinary tubules follows as a result of this failure of development,[1] and cystic dilatation of the urinary canals and of the glomeruli ensues. Of recent authorities, Kaufmann, among others, regards this view as correct for the majority of the cases. According to other authors (Ribbert and Hildebrand), an increase of connective tissue is present (the result of inflammatory changes), which brings about imperfect union between the urinary tubules and the glomeruli, so that

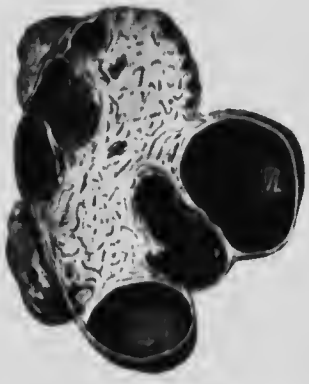


Fig. 39. — Section of a Cystic Kidney. (After Kaufmann.)

[1] Imperfect union between the portion of the kidney, developed from the diverticulum of the Wolffian duct and that formed by the intermediate cell-mass, may be the explanation of the congenital defect present. Shattock has suggested a combination of the mesonephros (Wolffian body) with the metanephros (true kidney) in these cases, and that the cysts arise from remnants of the mesonephros embedded in the true kidney.

the convoluted urinary tubules do not develop. Against the general acceptance of this view, however, is the fact observed by other authors that the convoluted tubules are present in many cases. (For further observations on the ætiology of cystic kidneys, see Kaufmann.)

It must here be noted that some authors regard cystic kidneys as tumours of new formation; and it cannot be denied that this explanation is correct for a small number of the cases. According to this hypothesis, the condition is one of an epithelial tumour—a multilocular cystadenoma of the kidney—which has developed as a result of atypical glandular proliferation from what was originally a normal or (at times) a malformed kidney (Nauwerck, Hufschmid, v. Kahlden, Singer, and Birch-Hirschfeld). According to these authors, the cysts of the liver which, as has been mentioned, so frequently occur at the same time, are cystadenomata also. Lastly, we may note that Nieberding, in a case he recorded, found the ductus Botalli completely wanting. He suggested, from this, that cystic disease of the kidneys might arise from venous congestion, as a result of a rise of pressure in the area supplied by the vena cava.

Even if children with well marked congenital cystic kidneys are born alive, they almost always die in a short time. As a result of the high position of the diaphragm and the compression of the lungs, asphyxia and suffocation occur; and, later, also interference with the heart's action. As regards the course of cystic kidneys, really congenital, but developing completely only later in life, I must refer the reader to the text-books on medicine (Senator, and others).

It is evident that such alterations in the kidneys will give rise to difficulties in delivery only if they attain an extraordinary size (see Freund's case mentioned on p. 198). Full information on this subject is to be found in Kleinhans and Hohl. Both authors, it must be admitted, bring forward many other causes for the distension of the abdomen; but there are also many interesting obstetrical histories of cases of cystic kidneys. In markedly cystic kidneys, the abdomen of the pregnant woman may be distended to much above the normal; so that, in some cases, the fundus of the uterus, even at the seventh or eighth month, reaches to the ensiform process. This excessive distension is especially marked when hydramnios is present at the same time—as it so often is in malformations. As a result of this over-distension of the uterus, a premature interruption of the pregnancy frequently occurs. According to Magenau (quoted by Kleinhans), delivery occurs most commonly in the seventh or eighth month: among forty-four cases twenty-five times; while in only six cases was the child at full term. A pelvic presentation occurs about ten times more frequently than usual. This fact is to be explained by the displacement of the centre of the body-weight downwards. Severe difficulties in delivery

are only found in the most marked cases of cystic kidneys; less marked disturbances are, however, often described. Magenau found that, among forty-five cases, in only six did delivery occur spontaneously: but these figures appear to me to be much too low.[1]

To diagnose this condition during pregnancy must almost always be a matter of impossibility. Hohl speaks as follows, in considering the special diagnosis during labour: 'If the enlargement of any one organ (kidneys, liver) is alone the cause of the increased circumference of the abdomen, then we find it even more tense than in ascites; further, the abdominal walls offer a greater resistance to pressure, and, on palpation, we impinge on a hard body. This will permit us, if we have regard to its position, to guess at the organ and possibly to recognise it when both kidneys are affected, as in this case the abdomen is especially distended at the sides.' He then continues: 'We shall not, with such an examination and inspection, so readily as J. Fr. Osiander, mistake an obstruction to delivery from enormous kidneys for ascites; nor open the abdomen, like the doctor in the case recorded by Höring, to let out the water, when in reality the kidneys are diseased.' As a rule, the general diagnosis of 'over-distension of the abdomen' is made when the birth of the child is suddenly arrested, and the recognised manœuvres usually practised for its delivery meet with no success. By the introduction of the whole or the half-hand, the medical attendant will be in a position to determine the nature of the distension of the abdomen.

Often, however, the true cause of the distension cannot be recognised during delivery if it is not possible to carry out a complete examination of the abdominal cavity of the fœtus after opening it.

The prognosis of these labours for the mothers is very good, as pathological over-distension of the lower uterine segment does not appear from the literature to occur.

The treatment or the removal of the obstruction consists in opening the abdomen, and making multiple punctures of the diseased kidneys when possible. If this leads to no result, then the enlarged kidneys must be removed in pieces, as well as possible, with the hand. When the child, in a cephalic presentation, is born as far as the thorax, then opening the thorax and perforation of the diaphragm may be necessary, so as to reach at the cystic kidneys. In less marked cases, especially in pelvic presentations, an attempt should be made to rotate the enlarged abdomen of the child into the roomy hollow of the sacrum. According to Hohl, by this manœuvre all other operative procedures may be avoided. Further, as the children may remain alive, this method should always be tried first, before any other manipulation endangering the life of the child is carried out.

[1] In three cases, recorded by v. Winckel, in which the children were small but the tumours of large size, spontaneous delivery occurred.

Here, we may mention, that congenital malignant tumours of the kidneys have been observed. Thus Stubinger, in 1903, collected three cases of congenital carcinoma, and nine cases of congenital sarcoma of the kidney, and gives in his dissertation full clinical histories. Congenital sarcomata of the kidney frequently contain enclosed glandular structures (Kaufmann), which in some portions may be so numerous as to remind one of an adenocarcinoma. Transversely striped muscle fibres, fatty tissue, cartilage, and bone, have also been found in them. Birch-Hirschfeld explains these as separated portions of the Wolffian body which have undergone further development after birth. (For further details, see Kaufmann.)

In congenital syphilis, according to Kaufmann, the kidneys are often affected. The process affects, principally, the cortex, but also the medulla. Small-celled, diffuse, or localised infiltration is present; also increase of the interstitial tissue, with vascular and perivascular infiltration, compression of the urinary tubules in the medulla, and resulting cystic formation; less seldom, induration with shrinking and large gummata.[1]

For obstetric injuries of the kidneys, see p. 206.

Uric Acid Concretions

In almost all new-born children, so-called uric acid concretions (first described by Virchow) are found in a section of the kidneys. These consist of light-yellow or brick-red streaks in the papillæ and the medulla, which converge in a tuft-like manner at the apex of the papilla. Microscopically, these streaks may be recognised as aggregations of uric acid and urates. If they are dissolved in hydrochloric acid, on evaporation plate-like crystals of uric acid form, and a clump of delicate clear albumen may be perceived as the scaffolding of this formation. According to the observations of Flensburg (quoted by Biedert and Fischl), this albuminous substance is separated, during fœtal life and shortly after birth, in the convoluted tubules; it is then washed away with the stream of urine, and deposited during the later stages of formation of the infarct, in which it forms the framework for the uric acid and urate precipitate, already described.

The uric acid infarct is a physiological condition. It reaches its highest point of development on the first or second day, and then gradually

[1] According to Karvonen (quoted by Ballantyne), in full-term fœtuses degeneration, more or less marked, of the epithelium is quite recognisable. Of eleven cases collected by Still, in some the symptoms dated from as early as five or six weeks after birth—a fact suggesting the possibility that the nephritis may have been of intra-uterine origin. A very interesting problem is the possible relationship between syphilitic nephritis in the fœtus and the chronic interstitial nephritis which is occasionally found, without any explanation, in children and young adults (Still).

disappears, so that after the sixth day it is only exceptionally met with. If the urinary secretion is very scanty—as may be the case in feeble, poorly nourished, and premature children—so that the flow is not strong enough to wash away the uric acid infarcts, then the retained masses may give rise to the formation of calculi in childhood.

Uric acid infarcts serve as an indication of the overloading of the blood with uric acid. Uric acid is found not uncommonly on the prepuce, or on the swaddling clothes, of the new-born child, in the form of a yellowish-red or brown powder. According to the latest researches (Senator), uric acid, like the so-called xanthin bases, is a derivative of nuclein.[1] It is formed when a destruction of nuclein-containing material occurs, especially of cells, in considerable quantities. That a very marked destruction of cells occurs in the fœtal body, shortly before and during birth, as a result of the multiple changes which take place, must be accepted as a proved fact.

The uric acid infarcts serve, further, as a proof of life, if we accept the view that they can only be recognised in children who have lived. This view is no longer, however, considered correct, at the present time; as the infarcts have been recognised both in living and still-born children.[2] (For further information on these facts see Stumpf.)

In some cases, in icteric children, so-called bilirubin infarcts are found, generally together with the uric acid infarcts. According to Kaufmann, a brilliant orange-red striation of the papillæ of the medulla can be seen macroscopically, from which one can express the pulpy coloured masses. Microscopically, the bilirubin is seen partly as a formless substance, light yellow in colour and granular; partly in the form of needles and tufts; partly as rhomboid crystals of a rusty red colour. The masses described are most commonly situated in the medullary substance of the kidney; less commonly, in the cortex.

The Suprarenals

Complete absence of the suprarenals, although very rare, has been described in perfectly normal subjects.[3] More commonly, these defects—absence of the suprarenals or maldevelopment of them (Hypoplasia)—are observed in monsters, and especially in those forms in which gross

[1] Uric acid arises from the oxidation of the nuclein bodies of the food and those of the body, and also partly from preformed nuclein bases—such as xanthin and hypoxanthin.

[2] As these infarcts are often absent, sometimes occur in still-born children, and at times only appear for the first time on the first or second day of life, they cannot be regarded as a certain sign that the child has lived after birth.

[3] In such cases, although no other defect may be present, yet the skin may be bronzed.

defects of the brain are present—such as hemicephalus, anencephalus, cyclopia, encephalocele, microcephalus, and syncephalus. On the other hand, hydrocephalus and defects of the posterior part of the brain have no influence on the development of the suprarenals.[1] Between the suprarenals and the genital organs, a very definite relationship can be established: for example, Marchand (Kaufmann) found in a case of rudimentary development of the ovaries and hermaphroditism, marked hyperplasia[2] of the suprarenals, and an accessory suprarenal in the broad ligament. Very rarely bilateral absence of a suprarenal occurs. Congenital hypoplasia of the chromaffin system, consisting of cells in the medulla which are coloured with chromic acid salts, points, according to some authors (see Kaufmann), to a diminished power of resistance on the part of the organism; and, as a result of this, death may occur from relatively slight causes.[3] In the chromaffin cells of the suprarenals is found the substance termed adrenalin.

Displacements of the suprarenals are very uncommon. As has already been mentioned, in considering the displacements of the kidneys, these are practically never associated with any displacement of the suprarenals. Neusser has described a case, in which a union of the two suprarenals occurred in the form of a horse-shoe suprarenal.

The so-called accessory adrenals possess a very considerable importance. They are small scattered suprarenal nuclei, the size of a pin's head, or at most the size of a bean, which are placed usually in the cortical substance, less commonly in the medulla, and are, according to Schmorl, very common, being found in 92 per cent. of all autopsies. They are also found in the immediate neighbourhood of the suprarenals (especially at the hilum), in the kidney region (on the solar plexus, or the coeliac ganglion), in and under the kidney capsule, in the substance of the kidney (especially in the cortex),[4] under the capsule of the right lobe

[1] The two parts of the suprarenals have a totally different origin: the cortex is derived from the mesothelium, covering the inner aspect of the Wolffian body; while the medulla is derived from the sympathetic, and is formed by an ingrowth of cell-groups on the mesial aspect of the gland. These cells-groups are not ganglia, but masses of indifferent cells, which develop into ganglion cells, and into the chromaffin cells of the gland. The medulla begins to be marked off from the cortex in the fourth month of fœtal life.

[2] Hyperplasia of the suprarenals is, however, sometimes associated with precocious sexual development; and this is of interest, in view of the fact that the same condition accompanies some cases of hypernephroma.

[3] It has been shown that, in some chronic exhausting diseases and in some acute diseases, the medulla is deficient in adrenalin; and it has been suggested that this is the cause of the muscular and cardio-vascular weakness in such conditions. Hypoplasia has also been suggested as the cause of the status lymphaticus and hæmophilia (Wiesel).

[4] When embedded in the substance of the kidney, liver, and pancreas, they are usually distinguished as suprarenal 'rests'; in the kidney, they may grow and give rise to innocent tumours—the so-called renal adenomata and lipomata.

of the liver, in the pancreas, and in the hepatico-duodenal ligament. They have also been observed, at a further distance from the suprarenals, on the sacro-iliac articulation, in the course of the internal spermatic vessels, and of the spermatic cord;[1] in the broad ligament near the parovarium, in the corpus Highmori of the testis, and in the retro-peritoneal tissue. Robert Meyer found that all three layers of the suprarenal cortex were present (Zona glomerulosa, Zona fasciculata, and Zona reticularis) in the accessory fœtal suprarenals—or, at least, two of them; while in adults, the division into layers could not be determined with certainty.

The importance of the accessory suprarenals lies, primarily, in the fact that they can take the place of the degenerated suprarenals, so that the occurrence of the injurious symptoms which might result from such degeneration is prevented; and, secondly, that they may become the starting-point of very malignant tumours (Grawitz: hypernephromata).[2]

Venous hyperæmia of the suprarenal bodies is frequently met with in new-born children, as a result of syphilis and asphyxia. Hæmorrhage into the substance of the suprarenals[3] is especially frequent in asphyxia occurring during delivery, and takes place almost always into the medulla. Various authors attribute this to the employment of Schultze's swinging method—undoubtedly the best method of restoring the apparently dead child. A large number of obstetricians, however, maintain, at the present day, that such hæmorrhages are not the result of this method of resuscitation; for such an occurrence, the suprarenals undoubtedly lie much too protected. As the hæmorrhage is very often bilateral, some central cause must be accepted, and this is to be found in the deep asphyxia (Stumpf). According to Stumpf, pressure on the abdomen plays but a secondary rôle. Piccoli (quoted by Stumpf) saw a case of a hæmatoma, the result of tearing of the suprarenals, from the application of forceps to the breech. Garipuy and Schreiber have recorded a case of hæmorrhage from the suprarenals in which death

[1] The presence of accessory suprarenals in the broad ligament and in connection with the spermatic cord, is, no doubt, to be explained by the descent of the Wolffian body, accompanying that of the testis or ovary; such supernumerary bodies being derived from the cortical portion of the gland developed within the Wolffian ridge.

[2] These tumours may originate either from the cortex or the medulla; those arising from the mesoblastic tissues of the cortex resemble, mainly, a perithelioma; while those arising from the neuro-ectodermal tissue of the medulla are allied to the glio-sarcomata. In view of their indeterminate character, it is best to term them malignant hypernephromata.

[3] In 105 still-born children, Spencer found either congestion or hæmorrhage in fifty-three cases; congestion alone being met with twenty-seven times. In twenty-four cases, hæmorrhage occurred into the medulla; and in three cases it had burst through the capsule. In half the cases, the bleeding was bilateral; and in a majority, the labour had been difficult and the presentation a pelvic one.

occurred. The child died on the third day after delivery, with the signs of internal hæmorrhage. At the autopsy, a large hæmorrhage was found, which had come from the right suprarenal body; the cause, however, could not be determined. The two authors suggest that the condition was one of a primary change in the suprarenal capsule: perhaps, excessive friability of its substance, possibly associated with an acute rise in the blood-pressure.

Obstetric injuries of the kidneys are very rare. They consist nearly always of rupture with more or less marked bleeding. Morbid enlargements of the kidneys, which occur not uncommonly from malformation, may be the explanation of the occurrence of the rupture (Stumpf).[1]

LITERATURE

Ziegler, *loc. cit.*, p. 15; Kaufman, *loc. cit.*, p. 24; Olshausen and Veit, *loc. cit.*, p. 44; Ahlfeld, *loc. cit.*, pp. 24 and 34; Keller, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Kleinhans, v. *Winckel, loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Senator, *Nothnagel, loc. cit.*, p. 53; vol. xix., p. 1; Neusser, *Nothnagel, loc. cit.*, p. 53; vol. xviii.; Runge, *loc. cit.* p. 129. Hohl, *loc. cit.*, p. 15; Nieberding, *Münchner med. Wochenschr.* (1887), p. 633; Theilhaber, *Monatsschr. f. Geburtsh. u. Gynäk.* vol. ix., p. 496; Stübinger, *Diss. Leipzig* (1903); Stumpf, v. *Winckel, loc. cit.*, p. 44; Hauch, *Geb. Ges. Paris*, (Feb. 19, 1908). *ref. Zentralbl. f. Gynäk.* (1908), p. 1151; Krull, *Gynäk. Ges. Dresden* (June 19, 1902). *ref. Zentralbl. f. Gynäk.* (1903), p. 561; Theuveny, *Geb. Ges. Paris* (June 19, 1902). *ref. Zentralbl. f. Gynäk.* (1903), p. 669; Seitz, *Münchner gynäk. Ges.* (Dec. 17, 1902). *ref. Zentralbl. f. Gynäk.* (1903), p. 837; Kirchner, *Magyar Orvosok Lapja* (1902), No. 31, *ref. Zentralbl. f. Gynäk.* (1903), p. 1446; H. Beucker, *Diss. Erlangen* (1902); Ehrenfreund, *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xvii. (supplementary number); Lafoeeade, *Diss. Lyon* (1903); M. Cadoré, *Diss. Lille* (1903); O. Orth, *Zentralbl. f. Gynäk.* (1905), p. 7; Zimdars, *Diss. Greifswald* (1903); Daniel, *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xx. (supplementary number); Engström, *Mitteilungen aus der gynäk. Klinik.*, vol. vi., p. 3; Sträter, *Niederl. gynäk. Ges.* (May 14, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 1304; Hill, *Bulletin of the Johns Hopkins Hospital* (1906), Nos. 181 and 185; Freund, *Geb. Ges. Leipzig* (Feb. 17, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 676; Garipuy and Schreiber, *Geb. Ges. Paris* (Jan. 15, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 1149; Robert Meyer, *Geb. Ges. Berlin* (April 3, 1908). *ref. Zentralbl. f. Gynäk.* (1908), p. 1453; Gilles, '*Zystenniere.*' *Geb. Ges. Paris, ref. Zentralbl. f. Gynäk.* (1908), p. 84; Potenko, '*Zystenniere.*' *Journ. f. Geburtsh. u. Gynäk.* (*russisch*) (1906), pp. 8 and 12; *ref. Zentralbl. f. Gynäk.* (1908), p. 351; Ballantyne, J. W., *loc. cit.*, pp. 15 and 44; Bryce, T. H., 'Embryology.' *Quain's Anatomy*, vol. i. (1908); Morris, H., *Clifford Allbutt, loc. cit.*, p. 89; vol. iv., pt. 1, p. 645 (1908); v. Winckel, *loc. cit.*, p. 44; Still, G. F., *Power and Murphy's System of Syphilis*, vol. i., p. 316 (1908); Wiesel, *Wien. med. Wochenschr.*, vol. lxxvii., p. 674 (1907); Rolleston, H. D., *Clifford Allbutt, loc. cit.*, p. 89, vol. iv., pt. 1, p. 423 *f.* (1908); Spencer, H. R., *loc. cit.*, p. 89; vol. xxxiii., p. 274 (1892); Winter, *Arch. Klin. Chir.*, vol. lx.ix., p. 614 (1903).

[1] In Spencer's cases, hæmorrhage into the kidneys occurred thirty-eight times. Of these, in twenty-two into the hilum; and in thirteen, beneath the kidney or beneath the capsule. His figures show that such hæmorrhage is favoured by breech or footling presentations.

The Bladder

Congenital malformations of the bladder are easy to understand in the light of embryology. In the fourth week of embryonic life, the navel closes; and the portion of the allantois hitherto lying anterior to the abdominal cavity, the primitive allantois, is cut off. Only the lowest part of it persists to form the bladder; the summit of which, remains in connection with the umbilicus through the urachus.[1] In normal conditions, the abdominal wall closes over the bladder; while the urachus becomes obliterated and converted into the ligamentum vesico-umbilicale-medium. If the closure of the urachus fails, then a fistula is formed, which leads from the apex of the bladder to the navel (fistula-vesico-umbilicalis—persistent urachus), so that the urine in part escapes by the umbilicus.

If the urachus remains only partly open, then so-called urachus cysts are formed, which may be multiple and of varying size. Large cysts of the urachus may project in the middle line of the abdomen and be mistaken for other tumours—such as ovarian cysts. The numerous ovarian cysts, described in the literature, without any pedicle, are possibly such cysts of the urachus[2] (see Tillmanns).

If the abdominal wall does not become closed in the normal manner, then there remains a fissure, with various resulting malformations. For instance, the lower half of the bladder, the urethra, and the genital apparatus may be closed normally, but the upper portion of the abdominal wall remains open, so that there is a defect of the anterior bladder wall; or, more frequently, the whole of the anterior abdominal and bladder walls may be absent, so that the posterior wall of the bladder is freely exposed. This malformation[3] is distinguished as fissure of the abdominal walls, ectopia or exstrophy of the bladder, and eversion of the bladder. According to Fischl, two forms may be distinguished—partial, and total, cleavage. In the partial form, there is present a well-developed navel, normal genitalia, and an opening in the anterior abdominal wall. In new-born children, there is present in the position of the bladder a space about the size of a florin, with sharp skin-edges and a dark-red base—the posterior bladder wall. As Fischl

[1] The bladder is developed mainly from the hypoplastic portion of the primitive cloaca, forming the urogenital sinus, and in part from the allantois. The two form a tubular canal in which a dilatation appears to form the bladder. The openings of the ureters and Wolffian ducts are at first common; but they become separated from one another by the growth of the intervening portion of the sinus, so that the ureters open into the bladder proper, and the ducts into the sinus. The trigone of the bladder is formed entirely from the sinus.

[2] A good many of these are so-called parasitic cysts in which the pedicle has undergone torsion and separation.

[3] This is a rare malformation, its frequency being about one case in 50,000 births; but it occurs six or seven times as frequently in males as in females (Ballantyne)

further points out, the bladder wall first becomes protruded in a tumour-like form after birth, as the result of straining during crying or during the passage of stools. The superficial surface of the tumour is moist and slippery, and on more careful examination, if the tumour is somewhat displaced upwards, the openings of the two ureters can be recognised as two fine button-like projections; from which at regular intervals the urine (which very quickly undergoes decomposition with the formation of carbonate of ammonia), escapes drop by drop. The

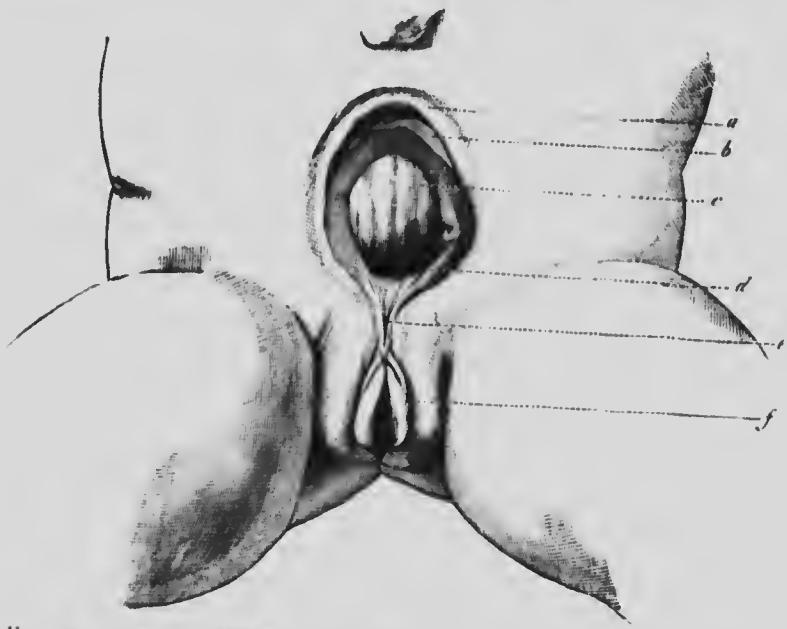


FIG. 40.—FISSURA ABDOMINIS ET VESICÆ URINÆ. *a*, Margin of skin; *b*, peritoneum; *c*, bladder; *d*, small cavity corresponding to the trigone of the bladder; *e*, gutter-shaped urethra; *f*, labia minora. (After Ziegler.)

prolapse becomes smaller in the course of years, while the skin is either drawn together or grows together more and more. The portion of the bladder mucous membrane, which is always prolapsed above the openings of the ureters, becomes dry; while that below, as the result of the continual moistening with ammoniacal urine, acquires a fungating appearance.

In total fissure of the bladder, the genitalia are affected at the same time. In the female, the clitoris is cleft, the urethra absent, and the vulva gaping; while the vagina may also be absent. The perineum is very short, and the anus lies immediately behind the genitalia, and can even open into the posterior bladder wall. In many cases, the uterus and vagina are double. Frequently, the union of the pubic rami

fails, and they may be separated from one another by a distance of several centimetres (split pelvis);[1] while, at the same time, a diastasis of the recti muscles is nearly always present.[2] In boys, associated with the fissure of the abdominal walls and of the bladder, the penis is rudimentary, with the glans split into two portions, below which the prepuce lies as a thick fold of skin—epispadias is generally present at the same time—so that the urethra is open above, and forms a shallow furrow on the dorsum of the penis. In extreme cases of fissure, the scrotum is also affected. As a result of this, errors in the determination of the sex may very readily arise, and the difficulty is increased by the fact that in such cases the testes very often remain in the abdominal cavity. Such errors often occur in practice in country districts. Inguinal herniæ are frequently present at the same time. Lastly, associated defects have been met with in the pelves of the kidneys, in the ureters:[3] atresia ani, absence of portions of the intestine (especially of the large intestine), an anal opening situated in the small intestine, and spina bifida. (For further details, see Ahlfeld.)

The results of such extensive abdominal fissures are naturally of a very distressing nature. The children affected, have a very disagreeable smell from the decomposing urine; and in the neighbourhood of the fissure a widespread weeping eczema occurs, with itching and scalding.

In the more marked forms of this malformation, the procreative capacity still remains. The bladder mucous membrane, as a result of its exposed position, is very disposed to hæmorrhage and ulceration. The great danger is that of infection of the kidneys, occurring through the ureters; and it is this infection which not infrequently brings about a fatal result. Yet cases have been recorded in which such individuals have lived to be more than seventy years old. Fischl quotes a case (recorded by Huxham) of a woman affected with a congenital prolapse of the bladder and a cloaca, who married in her twenty-third year, conceived and bore a child.

These fissures, in very rare cases, have undergone cicatrisation and healing *in utero* (see Kaufmann). On the walls of the extroverted bladder, malignant tumours, adenocarcinomata, and colloid cancer, may develop.

[1] The suggestion that the changes in the form of the split pelvis are due to the action of the body-weight is negatived by the fact that they are present at birth. They are, in reality, due to imperfect development, in varying degrees, of all the elements entering into the formation of the anterior half of the pelvic ring.

[2] The defect is usually broader transversely than vertically, and the umbilicus is situated at its upper margin. In some cases, however, there is an area of healthy skin between the upper end of the bladder and the umbilicus.

[3] The dilatation of the ureters, which is sometimes present, has been put forward as an argument in favour of the old view that the condition is secondary to over-distension of the bladder.

As to the æetiology of these malformation, opinions are widely divergent. According to one view (Ahlfeld and Rokitansky), retention of urine occurs, during the period of fœtal development, as a result of atresia[1] in the posterior part of the urethra, or following failure or retarded formation of the anterior part of the urethra. As a result, rupture of the anterior wall of the bladder occurs, after the abdominal walls have been pushed apart by the over-distended bladder. According to other authors (Meckel and P. Reichel), the condition is one of a defect of development, or of the survival of an early embryonic state. Such a fœtal stage of development (that is, the bladder split along its anterior wall) is, indeed, met with in mammals, but in Man has not yet been observed.[2]

Other hypotheses have been put forward by Bartels, Perls, Rolgans, and others (see Tillmanns). For the operative treatment of this disagreeable condition, the reader should consult text-books on surgery. Here, I will say only a few words.

A fistula of the urachus requires merely the freshening and suture of the edges. This method suffices also for partial fissures in the upper or lower part of the anterior bladder wall. In such cases, incisions for the relief of tension are often necessary. In the more marked forms, more complicated measures must be employed. Thiersch, Czerny, Trendelenburg, Schlange, Mikulicz, Sonnenburg, and others, have done good work in this difficult branch of surgery. The majority of the methods consist of a plastic operation, so as to cover the defect with skin, union of the freshened edges of the bladder, and procedures for the production of a greater degree of mobility: for example, partial division of the sacro-iliac articulation (Trendelenburg and Mikulicz);

[1] Such a condition is, however, practically never present. Ahlfeld's view, that traction by the vitelline duct plays a part in the production of the malformation by interfering with the normal development of the cloacal membrane, or of the urogenital septum, may be of importance (see p. 212).

[2] The hind-gut, together with the Wolffian ducts, opens (at the third week) into a dilated chamber—the cloaca entodermica. This is closed, on its ventral aspect, by a membrane—the cloacal membrane—composed of epiblast and hypoblast, derived from the persistent portion of the primitive streak. The primitive cloaca, as we have seen (p. 207), is divided later into two portions—a dorsal or rectal and a ventral or urogenital—by the formation of a septum between them. By the ingrowth of mesoderm, between the layers of the cloacal membrane, the body wall is formed, behind the umbilicus as far back as an angular projection—the genital papilla; behind this, the cloacal membrane persists until it ultimately breaks down, so that the urogenital opening is established. It is obvious, therefore, that this membrane takes a part in the formation of all the tissues of the body wall extending from the umbilicus to the tail-end of the embryo. By abnormalities in the development of the so-called urorectal septum or of the cloacal membrane, it is possible to explain nearly all the malformations met with in this region; and the failure of the mesoblastic elements of the membrane to develop, explains the maldevelopments of the body wall associated with exstrophy of the bladder.

extirpation of the everted bladder (Sonnenburg, Rein, Maydl, and v. Eiselsberg); and implantation of the ureters in some other position — the penile furrow, the rectum,[1] or the skin. These operations should first[2] be undertaken in later years — from the fourth to the tenth year. If the operation fails or is refused, then an apparatus devised by Earle must be worn. It consists of a hollow silver shield, from which a gum-elastic tube with a stop-cock leads, and which is pressed into position over the gap by a double-truss arrangement.

Other malformations of the bladder are very rare. Total absence of the bladder, with opening of the ureters into the urethra, has been described. Embryologically, this malformation is to be explained by the fact that the portion of the sinus urogenitalis, placed above the openings of the Wolffian ducts, has not developed.

The bladder may further be shut off from the urethra and the ureters. In the first case, the urachus may remain open; in other cases, hydronephrosis most commonly occurs. The bladder may, further, be abnormally small or abnormally large: the latter condition generally occurs in congenital closure of the urethra, and, as a result, the bladder may become so dilated that a marked obstruction to delivery is produced. The same thing may occur in cases of marked hydronephrosis. The excessive retention of urine may be overcome in this way: namely, the urachus again opens and the urine escapes by this channel. True ectopia of the closed bladder is present when it is situated in an abdominal or umbilical cleft.

Patency of the posterior bladder wall is very uncommon. In such a case there is a communication between the bladder and the peritoneum, or with the vagina (*fistula vesico-vaginalis congenita*), or with the rectum (*fistula recto-vaginalis cloaca*). At the same time, atresia ani and atresia urethræ may occur.

A bipartite bladder[3] (duplicity of the bladder) is also very uncommon. The septum may be placed either horizontally or sagittally. In a case described by Fürth (quoted by Zuckerkandl), the bladder was divided into two lateral halves by a septum; into each of these, a ureter opened. Nearly always, the two halves of the bladder are united to one urethra. The origin of this rare malformation has not yet been completely cleared up (see Zuckerkandl). According to Ahlfeld, it is possible that the hind-gut brings about the division of the allantois in this way—

[1] In some cases, where the cloacal septum is imperfect, the ureters open into the rectum.

[2] The impossibility of forming any kind of efficient sphincter, renders operations for the formation of a urinary reservoir very unsatisfactory; and the best results have been obtained by the implantation of the trigone of the bladder, including the openings of the ureters, into the sigmoid flexure.

[3] This occurs also in association with reduplication of other pelvic organs — such as double penis and vagina.

namely: through excessive distension, or by traction externally, it draws the posterior wall of the allantois against the anterior, and so divides the allantoic cavity, more or less, into two portions.

A bilocular bladder (unequal division of the bladder into two halves) is somewhat more frequent, although the condition is often really one of a diverticulum. Blasius (quoted by Tillmanns) has described a case where the bladder consisted of five separate cavities.

In very rare cases,[1] a cleft of the abdominal walls and bladder is associated with a cleft of the intestine (*Fissura abdominalis intestinalis seu vesico-intestinalis*). The fissure of the intestine affects, primarily, the cæcum[2] or the first part of the colon, or the portion where the omphalo-mesenteric duct is attached (see Ahlfeld). Like the bladder mucous membrane in a fissure of that organ, the mucous membrane of the bowel is extroverted in this condition (*Inversio seu ekstrophia intestini*). In this malformation (according to Ahlfeld), the ileum, the cæcum, and the large intestine are displaced, to a greater extent than usual, away from the spinal column by the traction exerted by the omphalo-mesenteric duct; so that in this region the mesentery is elongated. By the displacement of the bowel, a space is left between the spinal column and the intestine which is only in part occupied by the small intestine, kidneys, or liver. For this reason, the coccyx and the lower part of the sacrum are bent sharply forwards, and (as a result of this curvature of the lowest part of the spinal column) an enlargement of the spinal canal (at the bend) occurs, with accumulation of cerebro-spinal fluid, a cleft of the vertebræ, and a spina bifida. The typical pelvis resulting therefrom, Ahlfeld has named the pelvis inversa.[3]

These malformations are of no obstetrical interest with the exception of over-distension of the bladder. In a large number of cases the distension is such that delivery can only occur after puncture of the bladder. The cause of this excessive distension is to be found in atresia, or complete absence of the urethra. Very often obliteration

[1] Taruffi, however, gives notes of no less than forty-one cases of this kind.

[2] There may be one, two, or three, openings present, situated at the upper part or in the middle of the bladder area. On opening the abdomen, the small intestine is seen to be more or less normal; but its lower end, instead of ending in the cæcum, opens into the area of the everted bladder. The remainder of the large intestine is usually more or less completely atrophied, and ends blindly.

[3] Ahlfeld's view, that this combined deformity is due to traction exerted by the vitello-intestinal duct on the intestine, so that it is dragged out of the pelvic cavity, and interferes with the union of the anterior wall of the pelvis, and, further, if the traction is severe enough, leads to rupture of the allantois and of the bladder, is not generally accepted at the present time. It is, however, possible that some such physical explanation may account for the maldevelopment, which is the essential cause of the malformation. The time at which it occurs must be about the end of the third week of embryonic life.

of the meatus urinarius, or partial defect of the urethra, is present. Among a series of cases collected by Magenau, in thirty-two there was occlusion of the urethra; in twelve, complete absence, or merely a cellular cord; in the remainder, only partial (or in some cases, a membranous) occlusion. In seven cases, the urethra was permeable, but in three of them it was stenosed. In one of these cases (Hartmann), the stenosis was due to a retention cyst of a Littré's gland. In another case (Runge), the urethra was found, at the post-mortem examination, permeable; but the large intestine was atresic and distended with meconium, and had compressed and occluded the urethra. In other cases, more or less well-marked phimosis was present.

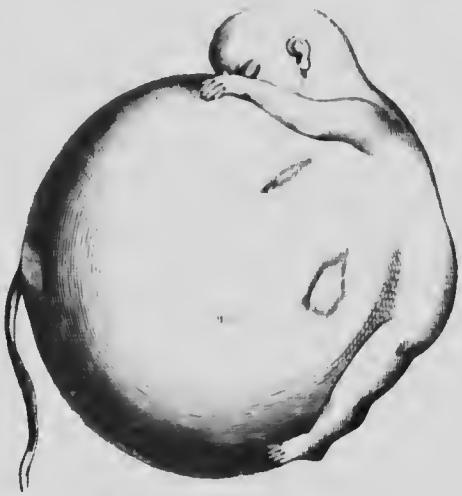


Fig. 41.—Marked Over-distension of the Fœtal Bladder. (After Hecker.)

Very often, besides complete absence of the urethra, other malformations are present at the same time: absence of the external genital organs; maldevelopment of the internal genitalia; atresia ani, together with a cloaca. Herbinet and Faix described a case with complete absence of the urethra and marked retention of urine, delivery only occurring after 1550 cm. of urine had been evacuated with a trocar. The post-mortem examination showed that besides this there was a communication of the base of the bladder with the large intestine. This rare malformation occurred in a fœtus four months old.

A very rare cause of retention of urine has been recorded by Rudaux due to compression of the urethra from a collection of mucus in the vagina.

In a case figured by Ziegler, the enormous dilatation of the bladder led to compression and stunting of the legs.

In a number of the cases, ascites has been present at the same time (Hohl and Kleinhans). The ureters are almost always more or less widened; in parts, also atresic. In some cases, the kidneys and ureters are absent on one side; in other rare cases, both ureters are absent; although both kidneys are present, but markedly altered. Kleinhans, quite correctly, explains the absence of the ureters by the premature atrophy of the kidneys as a result of the retention of

urine.[1] The changes found in the kidney were atrophy, hydronephrosis, and cystic degeneration, as the result of obliteration of the urinary tubules. The wall of the dilated bladder is frequently hypertrophied. I have already mentioned that at times the usually obliterated urachus is permeable when the bladder is dilated (p. 211). The distension of the bladder can occur so early that, even in a six months fœtus, it may lead to difficulty in delivery.

In the majority of cases, the children die, as a result of the retention of urine, in the seventh or eighth month. The diagnosis during pregnancy, and at the commencement of labour, cannot be made with any certainty. Further, the enlargement of the mother's abdomen, present, is found also in hydramnios and in all the other possible causes of enlargement of the child's body—for example, ascites, cystic kidneys, tumours of the liver, hydrocephalus, meningoceles, tumours of the coccyx, and fœtal inclusions.

The diagnosis is almost always first made at the birth of the child. It is best to explore with the whole or half-hand, under anæsthesia, when the progress of the delivery is delayed. Even then, the special diagnosis is not easy to make, as on the one hand ascites resembles very closely distension of the bladder, and on the other hand the enlargement of the abdomen may be due to a number of other anomalies. The detailed diagnostic points given by Hohl, for the differentiation of the anomalies which have to be taken into account, can hardly be considered as of practical value. Pathological over-distension of the lower uterine segment and rupture of the uterus appear to occur very seldom with these anomalies; at least, I have only found one such case in the literature. Treatment must be based upon the assumption that only in the least marked cases of distension of the bladder will delivery occur without its puncture. In these cases, the advice given by Hohl should be followed and an attempt made to bring the enlarged abdomen into the roomy hollow of the sacrum. In other cases, the delivery of the child only takes place when the ascites, so often present at the same time, has been got rid of by puncture. In all cases of marked dilatation, the bladder must be punctured by a trocar, perforator, or scissors. It is important in all cases first of all to make a trial of puncture, as it

[1] Ballantyne thinks that two different classes of cases should be distinguished: the first, in which the bladder is distended alone; and the second, in which it is hypertrophied as well as distended. Spicer, however, is not convinced that there is any necessity for such a distinction. He maintains that an imperforate urethra is not incompatible with fœtal life; that it is not necessarily accompanied by distension of the bladder, nor does it render dystocia inevitable; that it becomes dangerous only in the presence of a large secretion of fluid by the fœtal kidneys, and that such a secretion is pathological. He further holds that hypertrophy and dilatation of the bladder can only arise where there is an actual or virtual means of exit for the contained fluid.

is not unknown for such children to remain alive after the puncture of the abdomen. In very rare cases, opening of the bladder is avoided, as the urachus reopens, and this leads to the formation of an umbilical urachus fistula; or the contents of the bladder escape into the rectum, or externally through the abdominal walls.

In occlusion of the peripheral portion of the urethra, it may rupture and hypospadias or epispadias result (see p. 224). If in a cephalic presentation the bladder or the abdomen cannot be reached, then it will be necessary to proceed through the thorax; first, opening its cavity, then perforating the diaphragm, and then opening the bladder. In a case quoted by Kleinhans, as a result of useless attempts at extraction, rupture of the uterus occurred with a fatal result (case of Stevens, dilatation of the bladder, of the uterus and of the tubes).[1]

In very rare cases, obstruction to delivery may occur as a result of dilatation of the ureter only. Such cases have been described by Ahlfeld, Gervis,[2] Freund, Morris, Magenau, and others. In some of the cases the ureters were converted into cystic tumours; in others, the bladder was also involved (see Kleinhans).

In this place must be mentioned the well-known case (quite unique) recorded by Olshausen. There were present dilatation of the bladder, enlargement of the uterine cavity, and enormous distension of the large intestine.[3] The uterus was in communication, by a separate passage, with the bladder and rectum. As a result of the escape of urine from the tubes, chronic foetal peritonitis occurred. Olshausen, after the delivery of the head of the child, opened the body (without result) with the scissors perforator. The head was then removed, and delivery easily effected by podalic version and extraction.

Literature

Leser, *loc. cit.*, p. 34; Ziegler, *loc. cit.*, p. 15; Ahlfeld, *loc. cit.*, pp. 24 and 34; Baginsky, *loc. cit.*, p. 34; Biedert and Fiseld, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, 34; Kaufmann, *loc. cit.*, p. 24; Zuckerkandl, *Nothnagel*, *loc. cit.*, p. 53; Hohl, *loc. cit.*,

[1] This case was really recorded by Horrocks. The child was a female, and the urethra, vagina, and rectum were all obliterated: resulting in enormous distension of the bladder with urine, of uterus and tubes with fluid of some kind, and of the intestine with meconium. After delivery of the head and shoulders, the doctor in attendance found it impossible to deliver the body. As a result of traction the cervical spine was fractured and the pectoral muscles lacerated. Finally, the uterus ruptured and the patient died.

[2] This is an error. In Gervis's case, no obstruction to delivery occurred. The child died at the age of five weeks; both ureters and the pelvis of the left kidney were dilated, and the right kidney was considerably enlarged. There was no obstruction to micturition after birth.

[3] With urine.

p. 15; Olshausen and Veit, *loc. cit.*, p. 44; Müller, *loc. cit.*, p. 15; vol. ii.; Kleinhe... v. Winckel, *loc. cit.*, p. 34; Magenau, *Diss. Tubingen* (1902); Duparcque, *Jour. d'obstetrique* (1842); Depaul, *Gaz. hebdom.*, (1890), No. 20), and *Soc. de Biologie* (1861); Hecker, *Klin. d. Geburtsh.* (1861), vol. i., p. 122, and *Monatsschr. f. Geburtsh.*, vol. xviii., p. 373; M. B. Freund, *Breslauer Beitr.*, vol. ii., p. 240; Rose, *Monatsschr. f. Geburtsh.*, vol. xxv., p. 425; Kristeller and Hartmann, *ibid.*, vol. xxvii., pp. 165 and 271, Olshausen, *Arch. f. Gynak.*, vol. li., p. 280; Arnold, *Virchows Arch.*, vol. xlvii., p. 6; Whittaker, *Amer. Journ. Obstetr.*, vol. iii., p. 389; Duncan Carmichael, *Edinb. M. J. Journ.* (Aug. 1870), and *Edinb. Obstetr. Journ.*, vol. xi., p. 781; Comelli, *Wiener med. Wochenschr.* (1879), No. 37; Wolczynsky, *Wiener med. Presse* (1882), No. 36 ('Ascites and Hydronephrosis of left Kidney'); Robertson, *Glasgow Med. Journ.* (1889), p. 113 ('Imperforate Anus'); Ahlfeld, *Arch. f. Gynak.*, vol. lv., p. 161; Morris, *Med. Times* (1876), vol. i., p. 501; Czerwinsky, *Kronika Lekarska* (1900), p. 230, *Monatsschr. f. Geburtsh. u. Gynak.*, vol. xiv., p. 670; Debrunner, *Berichte und Erfahrungen, usw.* (Frauenfeld, 1910), p. 106; Devé, *Soc. obst. et gyn. de Paris* (Frommel's Jahresber., May 9, 1895), p. 674; Jilden, *Diss. Würzburg* (1890); Möller, *Arch. f. Gynak.*, vol. xlvii., p. 130; Neumann, *Monatsschr. f. Geburtsh. u. Gynak.*, vol. iv.; Michelmann, *Diss. Berlin* (1902); Schwyzer, *Arch. f. Gynak.*, vol. xlii., p. 333; Silbermann, *Wiener med. Presse* (1890), p. 332; Walther, *Zeitschr. f. Geburtsh. u. Gynak.*, vol. xxvii., p. 333; Wolff, *Arch. f. Gynak.* vol. lxv., p. 229; Henking, *Diss. Marburg* (1904); Chaigneau, *Diss. Bordeaux* (1904); Herbinet and Faix, *Gaz. Gesellsch. Paris* (Feb. 15, 1906), *ref. Zentralbl. f. Gynak.* (1906), p. 1136; Budaus, *Gaz. Gesellsch. Paris* (April 2, 1906), *ref. Zentralbl. f. Gynak.* (1906), p. 1378; Bryce, T. H., *Quain's Anatomy*, 'Embryology,' vol. i. (1908); Ballantyne, J. W., *loc. cit.*, p. 15; Kermauner, F., *Schmalhe, loc. cit.*, p. 15; vol. iii., pt. 1, p. 66 (1909); Maydl, *Wien. med. Wochenschr.*, vol. xlvii. (1896); Turnili, C., *loc. cit.*, p. 44, vol. vii., pt. 1, pp. 403–502; Spicer, J. E., *Proc. Roy. Soc. Med. Obstet and Gyn. Sect.*, vol. ii., pt. 2, p. 1 (1909); Horrocks, P., *Trans. Obstet. Soc. London*, vol. xxxvii., p. 6 (1895); Gervis, H., *Trans. Obstet. Soc. London*, vol. vi., p. 221 (1865).

The Urethra

(See also Malformations of the Bladder; for Epispadias and Hypospadias, see p. 224).

As has already been pointed out in considering dilatation of the bladder, congenital narrowing, obliteration, and defects of the urethra occur. Complete absence of the urethra has been observed in both sexes; very frequently in boys the penis is absent at the same time, and a cloaca is often present. The narrowing of the urethra may be partial or (less seldom) total.[1] In girls, the bladder may open into the vagina.

[1] The male urethra, between the sinus pocularis and the fossa navicularis, is formed from the urogenital sinus. The female urethra corresponds to the prostatic portion of the male urethra above the sinus pocularis. In the male, the internal genital folds meet to form the bulbous urethra, and are prolonged forwards on the ventral aspect of the genital papilla: first, as a solid epithelial ridge; then, from the shedding of the central cells, as a groove, which ultimately closes from behind forwards, forming the spongy portion of the urethra. In the glans, the urethra closes independently, so that the last part to close is at the junction of the glans and body. In the female, the groove remains open and forms the entrance to the vestibule; while the hypoblastic portion of the cloaca forms the urethra from the bladder to the opening of the vagina.

Stenosis of the urethra can also be brought about by the pressure of a retention cyst, through a stenosed large intestine distended with meconium, or by hypertrophic enlargement of the colliculus seminalis. Of the partial occlusions, the skin-like closure of the external urinary meatus forms the slightest grade (*Atresia membranacea orificii externi*). In the least marked cases, this consists of only an epithelial adhesion. In other cases, the whole of the glandular portion of the urethra is closed (*Imperforatio glandis*). Rarest of all, according to Kaufmann, is the closure of the internal opening of the urethra. It has been pointed out already that defects and stenoses of the urethra may lead to such marked retention of urine as to cause marked obstruction to delivery.

Cases of doubling of the urethra,[1] free from objection, apparently have never been described in the literature. Multiple openings of the urethra, as well as channels opening blindly in the glans in the neighbourhood of the urethra, have been frequently met with (see Tillmanns).

Congenital fistulæ of the penis must be distinguished from the similar diverticula of the urethra, consisting of depressions ending blindly which are in relation with Cowper's glands, the ejaculatory duct, or the prostate, but not with the urethra.

For the practitioner, the occlusions of the urethra (mentioned on p. 213) are the most important. It is of the utmost importance to recognise retention of urine as soon as possible after birth, as children so affected are quickly lost. The diagnosis of external occlusions of the urethra is easy. Deep occlusions are recognised generally by the failure to pass urine: the swaddling clothes remaining dry, and, eventually, by the gradually increasing distension of the abdomen. The treatment is easiest in epithelial or membranous occlusion of the external orifice; this may be broken down readily with a probe, or the membrane may be divided, if necessary, with a sharp knife. Deeply placed obstructions, too, can generally be overcome by the passage of a fine catheter; for they consist, as Ahlfeld has pointed out, nearly always (in viable well-formed children) merely of a stricture. In imperforation of the glans penis (according to Voillemier), one should push down, from the end of the glans to the presumed position of the anterior end of the urethra, a fine trocar or hollow needle, and then keep open the canal so created with a metal tube or a laminaria tent (see Tillmanns). If in the case of deeply placed occlusions the passage of a catheter does not lead to

[1] In a case recorded by Fordyce, of which Ballantyne gives an illustration, there was a penis-like body which contained two canals communicating not with the bladder, but with two vaginæ (?). Fordyce regarded the body as the united labia minora containing the irregularly developed clitoris. Ballantyne quotes Szymanowski's case of three urethræ (?) in one glans penis.

any result, or if the condition is one of extensive obliteration, then as a preparatory measure the urethra must be found. If it cannot be found under guidance of the fingers placed in the rectum, a trocar must be introduced into the bladder, and then a catheter inserted. This procedure is not without danger. It can be best done, as Kaufmann suggests, when the occlusion is situated in the membranous portion of the urethra—if this is separated from the bowel. Biedert operates upon these cases in this fashion: He opens the bladder from above, and then searches for the urethra from within. If in these occlusions the retention of urine is extreme, and rapid emptying of the bladder is required, then it must first be emptied by puncture. In other cases, the urachus remaining open above the symphysis; or reopening, as a result of the retention, permits of an intermittent escape of the urine. Obviously, in these circumstances, the urachus, after the closure of the urethra has been cured, should be closed again by freshening and suturing the edges.

As regards congenital narrowing of the urethra, narrowing of the external orifice has been most commonly observed. The result is more or less difficulty in passing water. The treatment is simple: splitting back the glans to the frænulum and suturing the margins to the mucous membrane of the urethra.

Other less common congenital narrowings of the urethra have been observed in the region of the fossa navicularis, and of the prostatic portion, in the form of valves or folds. The treatment consists in the use of bougies, catheters, laminaria tents, and external incisions with removal of the valvular folds.

Congenital widenings or diverticula of the urethra are very rare. Bokay, de Paoli, and others have described such cases. They consist, as a rule, of pouch-like bulgings, especially of the lower wall of the urethra (see Tillmanns). The pouches are flaccid, when empty, but fill up with urine during micturition, and may reach the size of a hen's egg. A catheter may either pass into them, become arrested in them, or glide over them. As a result of stagnation of urine, catarrhal processes and suppuration occur which may lead to a fatal result. The causes of the diverticulum are, according to Kaufmann, to be sought in retention of urine in the fœtus, which occurs as a result of the presence of membranous occlusions which may eventually give way, or from strictures, or folds of the mucous membrane. Treatment consists in the extirpation of the diverticulum, and the restoration of the original anatomical relations of the urethra. Any folds that may happen to be present should be removed at the same time, and a self-retaining catheter placed in the urethra at the end of the operation.

Literature

(See also Literature on Malformations of the Bladder.)

Kaufmann, *loc. cit.*, p. 24; Ziegler, *loc. cit.*, p. 15; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Tillmanns, *loc cit.*, p. 34; C. Kaufmann, *Deutsche Chirurgie*, pt. 59 (1880); Englisch, *Arch. f. Kinderheilk.* vol. ii. (1881), and *Zentralbl. f. Krankh. d. Harn- u. Sexualorgane*, vol. iii. (1892), vol. vi. (1895); Lindemann, *Diss. Jena* (1893); v. Bókay, *Jahrb. f. Kinderheilk*, vol. xxxviii., p. 138; Lhept, *Diss. Bordeaux* (1900); Foley, *Diss. Paris* (1895); Fordyce, W., *Teratologia*, vol. i., p. 61 (1894); Szymanowski, *Vrtljschr. f. d. prakt. Heilk.*, vol. lxxviii., p. 23 (1864).

CHAPTER XIII

Malformations and Congenital Diseases of the Genital Organs, including the Breasts

Malformations of the Male Genital Organs

In order to understand this and the following chapters, some short account must be given of the development of the urogenital organs. The genital glands, which, as is known, are derived from the germinal epithelium, develop either into testes or ovaries. In men, the Wolffian duct is of considerable importance, and plays an important part; while Müller's duct undergoes atrophy. In women, on the other hand, Müller's duct is of importance; while the Wolffian duct atrophies. The development of the male genitalia occurs in the following manner: The genital glands become the testes. The anterior cephalic portion of the Wolffian body forms the head of the epididymis—the Wolffian duct, the vas deferens of the testis. Its first part forms the tail of the epididymis. From the posterior portion of the Wolffian body are formed the vasa aberrantia epididymis—Giraldès' organ (Paradidymis).[1]

In men, as has been mentioned, from the atrophied Müller's duct is formed the hydatid[2] of the epididymis; while its point of opening into the sinus urogenitalis remains as the uterus masculinus or the prostatic sinus of the urethra. The seminal vesicles are simple outgrowths from the posterior wall of the vas deferens, which from this portion onwards is known as the ductus ejaculatorius. The prostate is composed of smooth muscle and glands, and is formed by downgrowths of mesenchyme from the urethral mucous membrane.[3]

[1] In the male, the Wolffian duct forms (Keith) the tube of the epididymis—the vas deferens and the common ejaculatory duct; while the vesiculæ seminales arise from it. The genital tubules of the Wolffian body become the rete testis, vasa efferentia, and coni vasculosi. The renal tubules form the vasa aberrantia found in the globus minor, and the paradidymis or organ of Giraldès.

[2] The stalked hydatid, seen at the upper end of the epididymis, is regarded by Keith as representing the most anterior (cephalic) end of the Wolffian formation; probably the pronephros—it certainly corresponds to the pronephric remnant found in the frog.

[3] The glandular tissue of the prostate develops as a series of epithelial downgrowths from the epithelium of the urogenital sinus, on either side of the openings of the Wolffian ducts. The muscular and connective tissues are derived from the mesenchyme of the genital cord.

The testes, which originally are placed on the posterior wall of the abdomen, gradually pass, during the further growth of the embryo, as a result of the pull of the gubernaculum Hunteri or testis, into the inguinal region. An outgrowth of the peritoneum (the processus vaginalis) then forms, which passes into a fold of skin in the region of the external genitalia (the ultimate scrotum). Into this, the testis [1] with the vas deferens passes.

The development of the female genital organs is as follows: The genital glands become the ovaries. The anterior portion of Müller's ducts becomes the Fallopian tube, while the lower portions are placed close to one another. Both,[2] finally, unite into a single tube, from the upper part of which the uterus develops; from the lower part, the vagina. From the upper part of the Wolffian body is formed the epo-öphoron (Paravarium); from the lower portion, the paro-öphoron. The Wolffian duct almost entirely disappears. At times, remains of its lowest portion are found in the neighbourhood of the cervix uteri. From the plica gubernatrix, the round ligament is formed.[3] In the female, a descent of the ovary occurs like that of the testis in the male; but the ovary remains in the true pelvis, and does not pass into the inguinal region.

[1] At the third month of fœtal life, the testis lies in the iliac fossa, attached by its fold of peritoneum (the mesorchium), and with the portion of the Wolffian duct which forms the epididymis on its outer side. This also has a mesenteric attachment, which fuses with that of the testis in the common urogenital mesentery. This is continued to the groin by a fold of peritoneum (the plica gubernatrix). In the fourth month, the subperitoneal layer of non-striated muscle tissue in the plica gubernatrix and the mesorchium takes on rapid growth, and forms the gubernaculum testis. This grows down into the scrotum, carrying with it a representative from each layer of the abdominal wall, including the peritoneum in the iliac fossa, to form the processus vaginalis. As the peritoneum is drawn down, the testis is dragged with it 'like a log on a sledge' (Keith). The subsequent atrophy of the gubernaculum pulls the testis completely into the scrotum. The fold of peritoneum forming the processus vaginalis becomes obliterated at the internal abdominal ring and just above the testis.

[2] The two Müllerian ducts, which at first lie on the outer side of the Wolffian ducts, as they pass into the genital cord cross over the latter and lie close together in the middle line. The anterior portions of the ducts form the Fallopian tubes; while the posterior portions fuse and form the uterus and vagina. The fusion begins near the lower end, and proceeds down and up. The septum thus formed, disappears first in the region of the cervix, then below this level, and finally at its upper end about the middle of the fifth month of fœtal life. The vagina is formed in the following way: the epithelial lining of the lower ends of the Müllerian ducts forms solid cords, the so-called vaginal cords, which proliferate in the tissue of the genital cord and fuse together; the cavity of the vagina being formed by the subsequent disappearance of their epithelial core after the fourth month. The hymen is probably developed in the epithelium in which the vaginal cords end.

[3] From the same layer of muscle tissue is formed also the muscular tissue in the utero-sacral ligaments and in the broad ligaments, and the outer layer of muscle tissue of the uterus.

A short description is now necessary of the development of the external genital organs. Originally the rectum opens, together with the allantois (sinus urogenitalis), externally, into the so-called cloaca. Later, a septum forms between the two and brings about their separation; this septum also forming the perineum. The external genitalia develop around the opening of the sinus urogenitalis and are composed of the following parts:—

1. The genital eminence:[1] a ring-like fold of skin surrounding the opening of the sinus urogenitalis.

2. The genital papilla: a projecting portion of the skin at the anterior part of the genital eminence.

3. The genital furrow: a furrow-like formation of the sinus urogenitalis on the posterior or under-face of the genital tubercle.

4. The genital folds: which bound the free margins of the genital cleft.

The development of the external genital organs proceeds in the following manner: The genital eminence[2] forms the labia majora; the genital tubercle, the clitoris; the genital furrow, the vestibule, and the genital folds,[3] the labia minora.

In the male sex, very marked changes occur. The genital eminence becomes very large and forms the scrotum, into which, as has been described, the testes pass from the abdomen. The genital papilla increases markedly in length and becomes the penis. The furrow placed on the under surface of the genital papilla also increases in length; and its free borders, the inner genital folds, grow together and unite to form a canal running along the under surface of the penis —the urethra[4] (Michælis and Hertwig).

Malformations of the External Genitalia

Complete absence of the external genitalia is found almost exclusively in the so-called sireniform monsters, or in other severe malformations of the whole body, such as fissures of the bladder or persistence of a cloaca. The internal genitalia are almost always malformed at the same time. Total absence of the penis (Aplasia penis), and its maldevelopment (Hypoplasia penis) is also rare. Total absence is almost always

[1] These terms are not used by the author in quite the same way as they are by most English writers.

[2] Namely, the outer genital folds.

[3] Namely, the internal genital folds.

[4] That is, the floor of the penile portion of the urethra. The roof is formed by the anterior part of the urogenital sinus, which is carried forward in the growth of the penis. The part of the urethra in the glans is formed from a solid ingrowth of epiblast which subsequently becomes canalised.

accompanied by severe malformations of the external genitalia.[1] In very rare cases, the testes are present. In other cases, the penis is only apparently absent, being placed under the skin of the scrotum. Defective development of the penis is generally accompanied by hypospadias or epispadias (see p. 224).

Still rarer, is doubling of the penis (Penis diphallus). As a rule, an incomplete duplicature of the lower end of the body—namely, three extremities[2]—is present at the same time. Tillmanns, however, observed a case of doubling of the penis in a perfectly normal child.

Congenital hypertrophy of the penis has been observed on several occasions, and in these cases the external genitalia are often developed to a striking degree. Sexual precocity is generally present.

Congenital narrowing of the prepuce (phimosis) is relatively frequent. In this condition, the prepuce—especially the inner layer—is so narrow that it cannot be drawn back over the glans. It may also be hypertrophied and elongated in a snout-like manner (hypertrophic phimosis). As Leser rightly says, in almost all male new-born children there is some physiological narrowing of the prepuce, and only in later life does the prepuce stretch so that it can be drawn back over the glans. Very frequently, in cases of phimosis, there are present epithelial adhesions between the prepuce and the glans, and in the epithelial masses epithelial pearls are sometimes found. As Bókay has pointed out, this epithelial adhesion is physiological in new-born children. The complete separation of the prepuce and the glans occurs in the first year of life, as Fischl maintains, from the formation of smegma. If the prepuce is so narrow that it cannot be drawn back over the glans, then the union of the two remains still longer. Complete atresia of the orifice of the prepuce, from inflammation, is found as the most marked form of phimosis in new-born children. The urine cannot be passed, and the prepuce becomes distended by it into a large bladder. More frequently, the orifice of the prepuce is narrower than the orifice of the urethra, so that it is easier for the urine to flow into the preputial sac than to flow out of it. In this case, a bladder-like distension of the prepuce naturally occurs. In the majority of cases of phimosis, the narrowing of the prepuce is easily overcome. The sequelæ of this anomaly may be balanitis (in severe cases, with ulceration), retention of urine from inflammatory processes, enuresis nocturna, a predisposition to masturbation, impossibility of coitus, increased liability to syphilis and

[1] Harris has recorded two cases, and collected five others. In all the cases, the testes were present in a scrotum, and the individual was definitely a male; and in five of them, the urine was passed into the rectum.

[2] For a collection of these curious cases, see Ballantyne, '*Teratologia*,' vol. ii., pp. 182–255 (1895). Taruffi regards the condition as really two pelves joined, so that each pelvis is represented by one ilium, and the sacrum is in reality double.

gonorrhœa, preputial stones developing from the smegma, carcinoma, epilepsy, hydrocele, inguinal and umbilical herniæ from straining, and death from uræmia following retention of urine. Baginsky, further, has recorded a case of death from pyæmia, as the result of a widespread eczema in a case of phimosis. In these circumstances—as even in sucklings severe troubles may arise—the treatment of the condition should be undertaken. It is important in new-born boys to examine the prepuce for such anomalies. Not uncommonly—even when there are more or less marked epithelial adhesions, between the prepuce and the glans—these can be separated with a probe. Occasionally, it is necessary to incise the prepuce in the middle line above, or undertake other operations for the condition.

Congenital paraphimosis also occurs, in rare cases. In this condition, there is a want of development of the prepuce,[1] so that it does not grow out round the glans, as usual, but is present merely in a rudimentary form united with it (Biedert and Fischl). Hypospadias is often present at the same time. Ammons' suggestion, that congenital phimosis is the result of the Jewish circumcision, Fischl does not consider as correct. We met with such a case of congenital paraphimosis, with accompanying hypospadias, in the Göttingen Women's Clinic, a short time ago. The child was otherwise well developed. The external meatus urinarius was closed except for two small openings.

The most frequent malformations of the urethra are hypospadias and epispadias. By epispadias, or fissura urethræ superior, is understood a want of development of the urethra or of the penis, in which the upper part of the penis together with the upper wall of the urethra is more or less split open, so that the urethra forms an open furrow. A slight and a severe form of this malformation are distinguished. In the first, the epispadias affects only the glans (glans epispadias), the opening of the urethra being situated behind the glans. In the more marked forms, the fissuring affects the whole penis, and this is often accompanied by ectopia vesicæ (see p. 208). The defects resulting from epispadias are very disagreeable. As a result of the continued moisture which occurs from the imperfect action, so often present, of the sphincter muscle of the bladder, widespread excoriation and ulceration take place. In the less common glans epispadias, incontinence is not present. The power of erection of the penis is usually retained; but it is only in glans epispadias, as a rule, that the power of fecundation is present, as in total epispadias the semen does not pass into the vagina. Treatment consists in plastic operations, for which I must refer the reader to text-books on surgery.

[1] The prepuce in both sexes is developed by the ingrowth of a solid ridge of ectoderm which, by separating into two lamellæ (Berry Hart), sets free a cutaneous fold as a cap to the glans.

Hypospadias, or fissura urethræ inferior, is most frequently met with. In this, the borders of the urethra fail to unite to a greater or less degree—the urethra forming a furrow open below, so that its opening is not at the summit, but on the under surface of the penis; in well-marked cases, even at the root of the penis or behind the scrotum in the perineum (Hypospadias perineo-scrotalis). In this case, too, there may be distinguished a glans hypospadias, a penile hypospadias, and the perineal hypospadias just described. In the severe forms, the penis is generally smaller than usual, and during erection is drawn down or to one side. Sometimes there is a union between the scrotum and the penis, especially in glans hypospadias. In the well-marked third form, the scrotum is divided into two completely separate portions by a furrow, and the urethral opening is on the perineum some 4 to 5 cm. from the anus. Very frequently, the penis is maldeveloped, bent downwards in a concave manner, and fixed. As a result of this, an error in the sex may easily occur. The determination of the sex is rendered the more difficult because in such individuals the testes are often undescended, so that the two empty halves of the scrotum may readily be mistaken for the labia majora. A case of marked hypospadias of this character might be termed pseudo-hermaphroditus masculinus externus (apparently, feminine). Persons affected in this way are often brought up as girls, and married as such. The error of sex is generally first recognised when the male peculiarities and characters make their appearance (sexual impulse, growth of the beard, and manly habits and voice). In many cases, the mistake is first recognised at marriage; in rare cases, not at all.

The interference with the functions of the organs in hypospadias is the same as in epispadias. Widespread ulceration and eczema of the penis and of its neighbourhood often occur. . orifice of the urethra is sometimes so small that difficulties are produced by this. The performance of coitus in marked bending of the penis is impossible; while the conditions as regards fecundation are the same as those we have already described in epispadias. Treatment for this condition, too, can only be of a surgical nature.

In the Gottingen Women's Clinic, only two cases of hypospadias of the penis were met with among 4200 births. In both cases, the mothers were multiparæ who had borne other normal children. No ætiological factor could be determined in the history. The children were well developed and throve well. (See also Malformations of the Urethra.)

Malformations of the Testes

By anorchia (Aplasia testis) is understood the rare malformation in which both testes are absent. In such persons, child-like habits are found, and abnormal smallness of the larynx is especially marked. In

apparent anorchia, it must always be remembered that a bilateral cryptorchism may be present. In monorchia, only one testis—which has usually undergone compensatory hypertrophy—is present. Here, too, an incomplete descent of the missing testicle must be thought of. By microrchia (Hypoplasia testis) is understood unilateral or bilateral incomplete development of the testis. Doubling of the testis[1] is extremely rare (Ahlfeld), as is also partial or complete defect of the epididymis with well-formed testes. The malformation which is distinguished as cryptorchism, or retention of the testes, occurs more frequently. The testes, which originally are placed in the abdomen, as has already been mentioned, descend[2] into the scrotum during fœtal life. Their descent is completed about the eighth month; but sometimes it occurs only during the first year of life. The later and often incomplete descent of the testes at puberty is distinguished as late descent of the testes. In retention of the testes, one or both may be arrested in its journey: either in the abdominal cavity (retentio testis abdominalis—abdominal testes), or in or near the inguinal canals (inguinal testes).

Of a different nature is ectopia testis (aberratio seu dystopia testis). In this case, the testis in its descent leaves the usual route and acquires a location in some other place: for example, in the neighbourhood of the perineum (ectopia testis perinealis); in the groin (ectopia testis cruralis); or between the scrotum and the thigh (ectopia testis scrotofemoralis).[3] Naturally, ectopia testis necessitates a retention of the testis. In dystopia transversalis, both testes lie in one side of the scrotum. When both testes are retained in the abdominal cavity it is possible, although very rare, for a union of the two to occur (Synorchidia).[4]

Retention of the testis has an obstetrical interest, in so far that in a bilateral case, and in a breech presentation, an error in regard to the sex may very easily occur during labour. The determination of the sex during labour in breech presentations is, of course, to some extent, more certain in male children. The retained testes, later on, may atrophy; but are very likely to be affected by inflammatory processes and especially to develop malignant tumours. Further than this,

[1] In almost all these cases of supposed supernumerary testis, the third body has turned out to be a tumour or a cyst.

[2] As Keith points out, in those mammals in which the testis leaves the abdomen, it does so to escape from the intra-abdominal pressure; for, from some cause at present unknown, a testis atrophies when subjected to this pressure. Further, a testis does not, as a rule, produce spermatozoa unless it reaches the scrotum.

[3] While the gubernaculum testis ends principally in the scrotum, bands pass from it to the root of the penis, the groin and the perineum. These bands may in some instances be sufficiently developed to influence the descent of the testis, and this is the explanation of its abnormal position.

[4] This may occur also in the scrotum, and is a condition normally found in some of the lower animals.

the formation of a hernia very often occurs coincidently. The causes of this malformation are various: abnormal formation of the genital organs, union of the testes with the intestines, narrowness of the inguinal canal; and, further, the passage to the inguinal canal may be hindered by a displaced kidney (Kaufmann).

From these considerations, it is clear that all such children should remain under observation. An improvement in the condition may be obtained by massage, suitable bandages, or by operation. Treatment of ectopia testis (described on p. 226) is the same as that for a retained testis.

Inversio testis is the rotation of the testis on its vertical, more frequently on its horizontal, axis; and, as a result, the epididymis very often is placed in front.[1]

Hydrocele of the testis occurs not uncommonly as a congenital condition. It is usually unilateral: most commonly, on the right side; more rarely, on both sides. In this condition the processus vaginalis may remain open in its whole length, or only in the region of the vas. The first is distinguished as a hydrocele of the processus vaginalis, or as a congenital communicating hydrocele; the last as a hydrocele[2] of the spermatic cord. Such hydroceles may give rise to diagnostic errors during birth. As congenital hydroceles may undergo spontaneous cure, they should be treated in an expectant manner. If spontaneous absorption does not occur, the hydrocele may be punctured with or without the injection of tincture of iodine, or alcohol; or, eventually, the question of an operation may be considered. For further particulars, I must refer the reader to text-books on surgery.

Demelin and Cathala have observed a case of congenital hæmatoma of the testis. Hæmorrhage was found in the substance of both testes in a child born in a cephalic presentation; the mother had albuminuria, but otherwise no probable cause was present.

Lastly, we must mention congenital tumours of the testis—a very rare condition. Microscopically, they may be placed among the teratomata: that is to say, tumours which are composed of very varying tissues, and which, according to their structure, may be distinguished as chrondrosarcoma, cystosarcoma, adenocystoma, adenomyosarcoma, cystocarcinoma. Such tumours have been described the size of a child's head. Ziegler gives in his text-book of general pathology a very good illustration of a congenital adenocystoma (teratoma of the testis) with pigmentation and formation of cartilage.

Congenital malformations of the prostate are very uncommon; with

[1] According to Keith, when the urogenital mesentery and mesorchium (see p. 221) persist, the testis is liable to become strangulated and twisted on its mesentery, or its upper end to fall downwards in front of the epididymis.

[2] Encysted hydrocele of the cord.

imperfect development of the testis, however, and with epispadias and fissure of the bladder, the prostate [1] may also be maldeveloped. Complete absence of the prostate occurs only in the highest degrees of malformations of the urogenital apparatus. Aberrant small prostatic glands have been met with in the neighbourhood of the bladder. Congenital retention cysts, through closure of the sinus prostaticus, have been described more commonly. Cysts [2] in the region of the prostate have also been described, arising from failures of development, and from the rests of the Wolffian and Müller's ducts (Dermoid cysts).

Malformations of the vesiculæ seminales are very rare. Absence of the vesicula seminalis, failure of the ejaculatory duct, and union of the vesicula seminalis and of the ejaculatory duct to form a single azygos structure have all been described.

Obstetrical injuries to the male genitalia are not very common. In pelvic presentations the swollen œdematous scrotum has been mistaken in some cases for the bag of membranes, and has been severely injured in attempts to rupture the membranes. The external genitalia more frequently have been injured in measures adopted for the termination of delivery, especially by the use of the blunt breech-hook. By the pressure of its point, the penis and testes may be badly crushed (Stumpf). Less severe injuries may occur from rough and unskilful examinations.[3]

Malformations of the Female Genital Organs

Atresia vulvæ (*congenital closure of the vulva*).—This, most commonly, takes the form of a so-called cellular atresia of the vulvar cleft: the labia minora being more or less united together, so that the vestibule of the vagina appears to be absent, while the labia majora may also be included in the union. The condition may give rise to difficulty in passing urine—even, indeed, to complete retention of urine.[1] According

[1] Arrested development of the prostate is especially associated with the condition of cryptorchidism, and cases of monorchidism have been described with maldevelopment of the corresponding lobe of the prostate, and of the vesicula seminalis on the same side.

[2] In seventy autopsies on new-born children, Englisch found this condition in over 7 per cent. These cysts may be large enough to encroach on the rectum, and their remains may be the explanation of some of the cases of enlarged sinus pocularis in adults.

[3] In 130 autopsies on still-born children, Spencer found congestion of the testis, scrotum, or spermatic cord, in thirty-seven cases; and hæmorrhage into the testis in fifteen cases (or 12·93 per cent.) of the male children. In nine of these, the bleeding occurred into the whole of both organs, and in three into the whole of one organ. Of the fifteen cases of hæmorrhage into the testis, in eight cases the child was delivered in a breech or footling presentation, and in six of these traction was made.

[1] This is uncommon, because a small opening is usually found near the root of the clitoris.

to Zweifel, it is due to an imperfect keratinisation of the superficial layers of the epithelium. In other cases, a very definite union of the labia, however, is present. The diagnosis is generally made soon after birth, from the pain on passing water. A cure can easily be obtained in the slighter cases by separation with a probe, or in the more severe cases by the use of the knife. Reunion may be prevented by the introduction of iodoform gauze into the vagina, or by suture of the raw surface.

Atresia hymenalis.—In this condition, in the place of the perforated hymen,[1] there is a thick imperforate membrane which prevents the escape of the secretion, and consequently at puberty leads to the retention of the menstrual blood in the vagina, the uterus, and the tubes (hæmatocolpos, hæmatometra, hæmatosalpinx). If small fine openings are present in the thick membrane, so that the menses can escape, yet if pregnancy occurs—and it must be remembered that pregnancy can undoubtedly occur without penetration of the penis—obstruction to delivery may result, as the membrane may present a resistance to the escape of the child's head. The treatment consists in incision or excision of the membrane.

Atresia ani vaginalis has already been mentioned with Atresia ani.

The further malformations of the female genital organs are easily understood in the light of their embryology (see p. 221).

I shall follow here the division which Kaufmann gives in his text-book of 'Special Pathological Anatomy.' He divides them into three main groups:—

Group I.—This comprises cases of incomplete coalescence, or incomplete fusion of parts which should be united. The result is doubling (duplication of the organs). Under this are included:—

1. *Uterus didelphys* or *Uterus duplex separatus.*[2]—In this condition, complete doubling of the uterus and vagina, internally and externally, is present. Very frequently, other malformations are found at the same time (formation of a cloaca; fissure of the abdomen and bladder).

2. *Uterus bicornis bicollis.*—The uterus is completely double, and the two sides of the body of the uterus diverge in a clavated manner;

[1] As has been mentioned (see note, p. 221), the hymen is developed in the epithelium in which the vaginal cords end, derived (according to Berry Hart) from the Wolffian ducts. It is covered on its outer surface by the epithelium of the uro-genital sinus, and on its inner surface by that of the vaginal cords. Usually, at several points, the epithelium fuses and breaks down; if it does not, then the hymen is imperforated. No doubt, in many instances, cases of so-called imperforate hymen are in reality due to atresia of the lower end of the vagina.

[2] No undoubted case of this kind has been recorded in a well-developed adult. The vagina is usually only developed in its upper part, and either ends blindly in the wall of the bladder, or in the bladder. A communication of the intestine with the bladder, between the two halves of the uterus, is frequently present.

while at the same time, the two portions of the cervix lie close to one another. The vagina is single or double.[1]

3. *Uterus bicornis unicollis.*—The conditions are similar to those in No. 2, but the cervix and vagina are single.

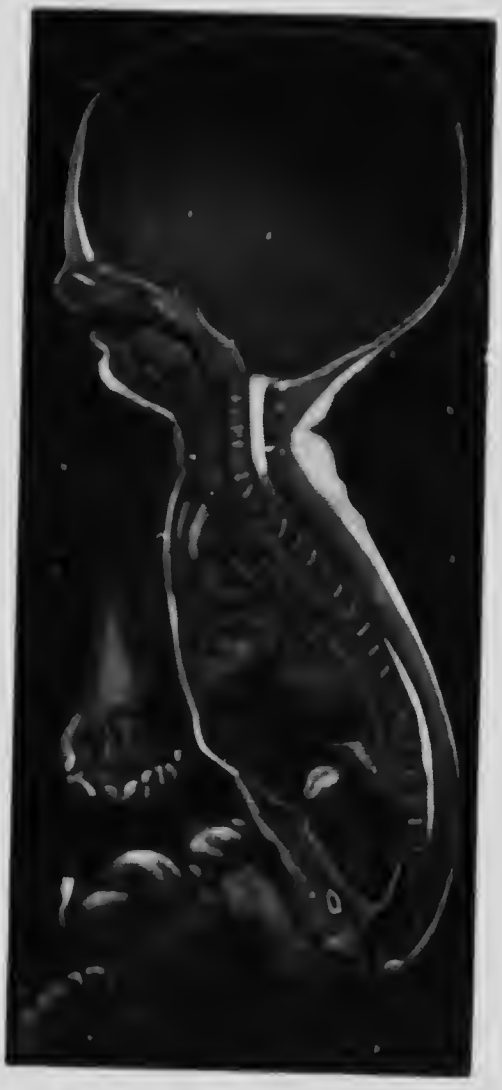
4. *Uterus arcuatus.*—A slight depression is present at the fundus, as the indication of the presence of two cornua. To this group belong, further, the malformations which occur from imperfect union of the two Müller's ducts—that is, externally, they are joined; but they do not form a single canal. There is thus a uterus which externally is single, but which, with the vagina, is divided by a septum into two halves. The uterus bilocularis, with a divided vagina, or uterus duplex septus, is of this nature. All possible gradations of the condition may occur. The uterus is double, but the vagina is single, or the uterine septum is incomplete (Uterus septus unicollis, or uterus subseptus). Such rudimentary septa may also be present in the vagina.[2]

Group II.—To this group belong the cases of aplasia (complete absence) or hypoplasia (rudimentary development) of the tubes, uterus, vagina, and ovaries. As regards aplasia, Müller's ducts may be completely stunted, or the stunting may only affect portions of the ducts symmetrically on the two sides, or one Müller's duct only may be undeveloped. In complete aplasia, the uterus, tubes, and vagina, are absent. The external genitalia then open into a short blind sac. If the aplasia is less marked, then the uterus may be absent, or completely rudimentary,

[1] The vagina is most commonly double, and even when a septum is present it is usually imperfect. In some cases, the two cervices are united by connective tissue only; and the loosening of this band of union, as a result of pregnancy, has led to the erroneous diagnosis of a uterus didelphys.

[2] The causation of these malformations is obscure. According to Thiersch, v. Winckel, and Frankl, they are due to abnormalities of the Wolffian body and duct. Frankl has elaborated this view, and holds that, as the result of excessive development of the round ligament, the Wolffian duct is drawn in a lateral and ventral direction and carries with it Müller's duct, so bringing about a separation of the two halves of the genital tract. Schwalbe, on the other hand, thinks that the development of Rathke's folds is at fault, and that these various deformities result from the failure of the two folds to meet one another accurately in the middle line, so as to form the urogenital septum. As the two Müller's ducts grow into these folds, any abnormality in their position will influence the union of the two ducts. He regards the recto-vesical ligament—a fold found in many of the cases running between the rectum and the bladder—as one of the remains of such a disturbance in the development of the urogenital septum. As we have already seen, however (see p. 210), many embryologists at the present time believe that the cloaca entodermica is not divided by two lateral folds as first suggested by Rathke. By an increase of the mesoderm in the tongue-like projection which intervenes between the allantois and hind-gut, the cloaca becomes separated into a dorsal or rectal tube and a ventral or urogenital tube; and by further growth of this septum, the ventral part of the chamber is displaced forwards, while the rectum is shifted backwards. Abnormalities in the development of the septum, however produced, will necessarily lead to corresponding abnormalities in the development and fusion of the two Müllerian ducts.

PLATE III.



A FEMALE FŒTUS WITH HYDROCEPHALUS, HARE-LIP, AND CLEFT PALATE, SEVEN DIGITS ON EACH EXTREMITY; THE FINGERS AND TOES SHOWING SYNDACTYLY.

The vagina is imperforate and distended with mucus; and between the urethra and the rectum is a solid fleshy septum. The legs are stunted.

(Specimen No. 855, University College Hospital Medical School Museum, Obstet. Sect. From a case under the care of Dr. F. R. Dodwell.)

or developed only in parts; while the vagina, external genitalia, tubes, and ovaries are present. The uterus unicornis[1] is a type of the marked asymmetrical aplasia, in which one Müller's duct [illegible] veloped, the other well developed, but somewhat displaced towards [illegible] side of its tube. Hypoplasia (that is, rudimentary development) may occur so that Müller's ducts are imperfectly developed in whole or in part. These changes may be present symmetrically or asymmetrically, and may extend to the uterus, tubes, or vagina. The uterus, for instance, may develop in the form of a solid muscular and connective tissue rudiment. If one horn only develops in a rudimentary manner, the so-called 'uterus unicornis with an accessory horn' results, which is usually not united with the well developed uterine cavity. Between the two, there is nearly always a bridge of solid connective tissue. The accessory horn may be solid, or, as in the majority of the cases, hollow. Pregnancy, too, may occur in the rudimentary accessory horn as the result of the external migration of the ovum; less commonly, from the external migration of the semen. In such cases, unless operative procedures have been undertaken, rupture of the gestation-sac occurs in the early months of pregnancy. If the mucous membrane of the rudimentary horn menstruates, then a hæmatometra forms. The tube on the undeveloped side may be normal, or converted into a solid cord. In the same way, the ovary may be developed perfectly, or more or less absent.[2]

By 'Uterus fœtalis,' is understood arrest of the development of the uterus at the fœtal stage.[3]

Group III.—The third group includes the cases of atresia which may affect the external os, the whole cervix, or the vagina. Cellular atresia and atresia hymenalis have already been considered. Atresia of the uterus and of the vagina may lead to hæmatocolpos, hæmatometra, and hæmatosalpinx.[4]

Displacements of the uterus have also been observed congenitally: for example, retroflexion of the uterus; prolapse of the

[1] According to Nagel, no case of a complete absence of the opposite Müller's duct in a case of uterus unicornis has been met with in a well-developed individual. It only occurs in cases of marked malformation.

[2] Associated malformations are often found. Thus Pallauf, in seventy-six cases, found rudimentary development or displacement of the kidneys in thirty-six. The bladder, too, may be developed on one side only.

[3] In these cases, the uterus and its appendages retain the anatomical characters they have at the time of birth.

[4] Nagel and Veit maintain that, in the majority of these cases the atresia or stenosis is acquired, and due to inflammatory conditions occurring after birth. According to Vagedes, such a cause is present in at least 61·5 per cent. of all the cases. Cases of this kind may vary in character, from complete absence of any trace of the vagina to simply a membranous obstruction, or perforated septum at one part of the canal.

uterus[1]; displacement of it into a hernia—for example, an inguinal hernia—(Hernia uteri inguinalis; see Birnbaum for literature on this subject); femoral hernia (hernia uteri cruralis), and umbilical hernia. Further, obliquity of the uterus (latero-position and extramedian position) has been observed.

Malformations of the Fallopian Tubes

Complete defect of the tube has been seen with absence of the uterus.[2] Accessory tubal ostia may be found, generally in the neighbourhood of the fimbriated extremity. So-called accessory tubes are not uncommon. They may be situated (according to Kaufmann) on the ala vespertilionis, on the isthmus of the tube, on the broad ligament, and between its layers; and may possess fringes, an opening into the abdomen, and folds of mucous membrane. Some are pedunculated, others sessile. In the first case, they are not in communication with the lumen of the main tube. Doubling of the tube is very rare; but in many instances, tubal diverticula have been described. Accessory tubes and tubal diverticula may predispose to the development of a tubal pregnancy, as the fertilised ovum may remain in these situations,[3] and (according to Freund) so may the persistence of the fœtal tubal tortuosities into extra-uterine life. Whether the hydatid of Morgagni,[4] or other small pedunculated cysts, which spring either from one of the fimbria of the tube, from the tube itself, or from its neighbourhood, are to be considered as malformations is not yet decided (they may represent remains of the Wolffian ducts, lymphangiectasis, cysts of the germinal epithelium, or cysts of the peritoneum). Complete congenital atresia of the tube is very rare.

Malformations of the Ovaries

The ovaries may, as has already been mentioned,[5] be absent on one or

[1] In nine of the twelve cases of congenital prolapse recorded by Ballantyne, a lumbo-sacral spina bifida was present, and in several of the cases a semiparetic condition of the lower limbs. He suggests, as other contributory factors—in view of the fact that a spina bifida has not always been present—narrowness of the false pelvis, enlargement of the pelvic inlet and outlet, and defective development of the connective tissue of the pelvis. [2] Unilateral defect is more common than bilateral.

[3] The embedding of the ovum in the wall of the tube has, no doubt, led in some instances to an erroneous diagnosis of the presence of a diverticulum.

[4] The hydatid of Morgagni must be distinguished from the pedunculated cyst usually attached to the mesosalpinx in the neighbourhood of the abdominal ostium, which is to be regarded, as we have seen, as probably representing the remains of the pronephros. The hydatid of Morgagni, on the other hand, is derived from the upper end of Müller's duct.

[5] Absence of both ovaries has only been seen in marked forms of malformation—such as the sireniform fœtus. Absence of one ovary has been established, but is usually accompanied by imperfect development of the corresponding half of the uterus, the tube, and the kidney, on the same side.

both sides, or be rudimentary. Cases have also been observed where a third or fourth ovary was present. In such a case,[1] a supernumerary tube is also found. More or less well marked division of the ovary has been described (Ovarium succenturiatum, Ovarium lobatum, or Ovarium partitum). Such divided ovaries, strictly speaking, cannot be considered true supernumerary ovaries. Lastly, functionating ovarian elements, separated portions of germinal epithelium in the form of primordial follicles, may be met with in the broad ligament. From such ovarian elements, various forms of tumour, especially teratomata, may take their origin (Chrobak and Rosthorn). One or both ovaries may be congenitally displaced into a hernial sac: for example, in an inguinal hernia (Hernia ovarii inguinalis); or in one of the labia majora (Hernia ovarii labialis).[2]

Malformations of the genital organs, naturally, but rarely give rise to any difficulty in delivery. Olshausen's case has already been described (p. 215), in which dilatation of the bladder and marked distension of the uterus and intestine, from retention of urine was present. The bladder was united with the rectum and the uterus, and, as a result of the entrance of urine into the abdominal cavity, chronic foetal peritonitis ensued. Gervis[3] and Davies have reported cases, in which difficulty in delivery arose as a result of the distension of the uterus with fluid, from closure of the cervix, or absence of it, or of the vagina. Rogers has also recorded a case in which the testes, converted into large fibrocystic tumours and retained in the abdomen, gave rise to some obstruction to delivery.

Obstetrical injuries to the female genitalia have been recorded, although very rarely. Stumpf cites several cases, in which through rough introduction or boring of the finger into the vagina, tearing of the recto-vaginal septum has occurred.

Hermaphroditism

Before I leave the section on malformations of the genital organs, I must consider, briefly, Hermaphroditism—a very important condition for the practitioner. A full consideration of this question will be found in the recent monographs of Chrobak and Rosthorn in 'Nothnagels Handbuch,' and of Stumpf in 'v. Winckels Handbuch.' I may also refer to the numerous accurate and detailed works of Neugebauer on this subject.

Two varieties of hermaphroditism are distinguished: true hermaphroditism (Hermaphroditismus verus—Androgynia)—that is, in the one

[1] It is doubtful whether there is any authenticated case of the presence of a true third ovary on record.

[2] Arrested descent of the ovary has been described in a number of cases, and Sellheim has collected forty-four instances of this kind.

[3] It has already been pointed out (see p. 215) that Gervis's case was not one of this character

individual a testis and ovary is found at the same time; and pseudo-hermaphroditism (or Hermaphroditismus spurius)—that is, bisexual development of the other sexual organs with unisexual development of the genital glands.

The best known division of the different forms of hermaphroditism is that given by Krebs. He distinguishes:—

1. Hermaphroditismus Verus

(Testis and ovary in one individual.)

(*a*) *Bilateralis:* that is, on both sides of the body a testis and an ovary.

(*b*) *Unilateralis:* that is, on one side of the body a testis and ovary, and on the other side only one sexual gland—a testis or ovary.

(*c*) *Lateralis:* that is, on one side an ovary, on the other side a testis.

As is well known, hermaphroditism played an important rôle in mythology. Further, its not uncommon appearance in the lower animal world must be recognised—thus, in the pig, true hermaphroditism has been seen on many occasions. As regards true hermaphroditism in Man, even at the present time, there are numerous authors who strongly deny its occurrence. Other writers admit the occurrence of hermaphroditismus lateralis, but do not recognise hermaphroditismus bilateralis or hermaphroditismus unilateralis. In recent times, at any rate, cases have been recorded which on histological grounds must be acknowledged as examples of hermaphroditismus verus (Salén, Garré-Simon, and Pick; see Chrobak and Rosthorn, and Stumpf). These authors found in their cases a hybrid gland—that is to say, an organ in which histologically testicular and ovarian substance could be recognised.[1]

2. Pseudo-hermaphroditism

A.—*Pseudo-hermaphroditismus masculinus:* testes only present.

(*a*) *Internus:* male type of the external genitalia with persistence of Müller's ducts.

(*b*) *Externus et Internus (completus):* female type of the external genitalia with persistence of Müller's ducts.

[1] For the description of a case of true unilateral hermaphroditism, and a criticism of all the cases recorded up to that date, see Blacker and Lawrence, *Trans. Obstet. Soc. London*, vol. xxxviii., pp. 265–317 (1896). If the term 'true hermaphroditism' is taken to mean merely the presence of two different sexual glands in one individual, without implying that their functions are present or perfect, then the condition has undoubtedly been met with in the human subject. According to Siegenbeek van Heukelom and Nagel, no case of true hermaphroditism in Man, so far, has been described; but, as an illustration of the difficulty of coming to a conclusion, it may be pointed out, that while the first regards Blacker and Lawrence's case as a male, the second regards it as a female (see Fig. 42).

(*c*) *Externus*: female type of external genitalia with no remains of Müller's ducts.

B.—*Pseudo-hermaphroditismus femininus*: ovaries only present.

(*a*) *Internus*: female type of external genitalia, with persistence of Wolffian duct.

(*b*) *Internus et Externus* (*completus*): male type of external genitalia with persistence of Wolffian duct.

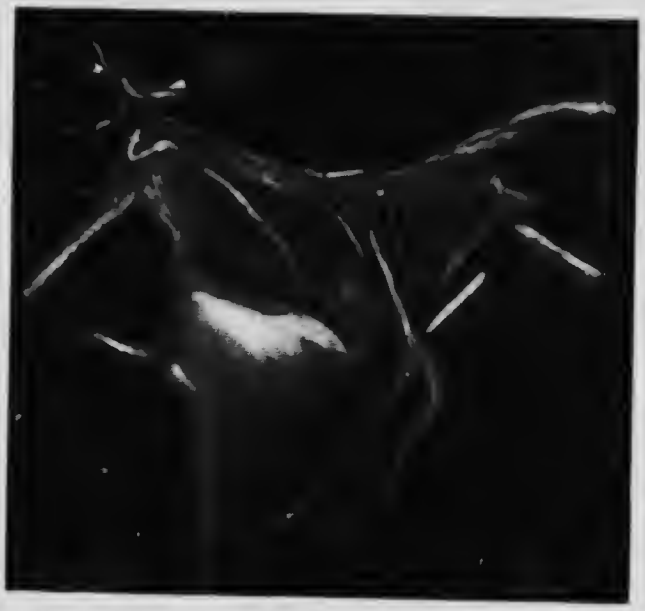


FIG. 42.—TRUE UNILATERAL HERMAPHRODITISM. A, Epididymis; B, fimbriated extremity of rudimentary Fallopian tube; C, ovarian portion of left genital gland; D, testicular portion of left genital gland; E, rudimentary Fallopian tube; F, uterus; R, combined gubernaculum testis and round ligament; S, processus vaginalis; T, left infundibulo-pelvic ligament; U, vas deferens. (University College Hospital Medical School Museum, Obstet. Sect., No. 851.)

(*c*) *Externus*: male type of external genitalia with no remains of Wolffian duct.

According to the observations recorded, in the majority of cases the condition is one of pseudo-hermaphroditism, and much more frequently of Pseudo-hermaphroditismus masculinus (Androgynia) than P. femininus (Gynandria); the numerical relation being 427 to 125.[1] As the classification of Klebs indicates, pseudo-hermaphroditismus masculinus

[1] In 910 cases of pseudo-hermaphroditism collected by Neugebauer, there were 722 instances of the masculine variety, and only 188 of the feminine variety; and 613 of the whole series were examples of pseudo-hermaphroditismus masculinus externus.

internus, besides the testes, shows typical external genital organs; while, internally, Müller's ducts are more or less developed into the uterus, tubes, and vagina. Pseudo-hermaphroditismus masculinus externus et internus (completus) shows—besides the testes and the vasa deferentia and the prostate—a female type of the external and internal organs; in the case of the latter, of a more or less developed form. Pseudo-hermaphroditismus masculinus externus shows, besides the testes, a female type of the external genitalia, while Müller's ducts have not undergone development.

In all these malformations, the sexual characters are naturally so confused that grave errors in the determination of the sex may readily occur; and, therefore, these anomalies have a considerable social and forensic interest. As Chrobak and Rosthorn point out, such errors are, primarily, to be attributed to the fact that an endeavour to determine the sex can generally, in the living individual, only be made by the general habits and the external marks of sex. Scientifically, therefore—especially in true hermaphroditism—only those cases are trustworthy in which a post-mortem examination, or an operation, has rendered a complete investigation of the internal genitalia possible, and in which a histological examination of the affected genital glands has been carried out. In pseudo-hermaphroditismus masculinus, the most striking feature is the resemblance of the external genitalia to those of the female sex. This is readily explained by the fact that parts, which in fœtal life should have become joined, remain separate. The penis is only partly developed (Penis clitoridien), and the scrotum is divided; so that, as cryptorchidism is frequently present, the similarity of the empty halves of the scrotum to the labia majora is, naturally, still more complete. Very frequently, also, hypospadias is present; in the higher grades of which the penis is undeveloped, like the clitoris. The sinus urogenitalis is wide open and deep. When such malformed individuals, further, lack the secondary male sexual characters, and also the habits of the male sex; and when, also, female breasts are present (Gynæcomastia), and there is an absence of beard, a soft female form, a high-pitched voice, and feeble sexual desire towards women, it is clear that such persons may readily be mistaken for females.[1]

Pseudo-hermaphroditismus femininus, as already explained, is much rarer. In this case, the female breasts are wanting (Andromastia), and the external genitalia may assume a male type throughout—that is, the clitoris is markedly enlarged and takes on a penis-like form (*clitoris d'aspect phalloide*); the vaginal canal is narrowed; and, as the labia are more or less extensively united, the urethra and vagina may open

[1] For a complete description of a most striking instance of this variety, see a paper by Tuffier and Lapointe, *Revue de Gynecologie et de Chir. Abdom.*, vol. xvi., No. 3, p. 209 (1911).

separately into the sinus urogenitalis. Confusion in these cases is even more likely when the ovaries are placed in the labia majora and so the appearance of a scrotum is produced. In these cases, too, the secondary female sex characters and the female habits may be absent; while sexual desire for the female sex, male form of body, large bone formation, well-developed muscles, a man's voice and growth of beard, may all be present. As Chrobak and Rosthorn point out, the inclination and habit of the affected individual should not be relied on too much in the determination of the sex; as in this case the educational factor will play too important a rôle. The authors mentioned give in tabular form a complete list of all the important facts, which have proved of value in the diagnosis of doubtful cases.

For the practitioner—especially for the obstetrician and the legal expert—the subject of hermaphroditism is of the greatest importance. As has been mentioned, it often happens that such malformed individuals (that is, of male type) are brought up, on the decision of a midwife, as of the female sex, and the error is only discovered much later, or even at times not at all, during life.[1]

The Prussian text-book for midwives certainly orders the midwife that she shall, in all such doubtful cases, consult the doctor.

It has further happened that an hermaphrodite of male sex has been rightly certified, soon after birth, as a boy; after a short time, on the erroneous diagnosis of a doctor, has been inscribed in the registry of births as a girl; and finally, on the decision of higher authorities, with much trouble has attained his proper civil rights. That the correction of a mistake in the register of births causes a great deal of trouble is well-known.

Such an error of sex undoubtedly occurs not infrequently;[2] and

[1] In view of the fact that in the great majority of the cases the individual is in reality of the male sex, in all doubtful cases the child should be brought up as a boy. In three-quarters of the cases, the ision come to will prove to be correct; and the inconveniences which may result if nistake has been made are much less in the case of a supposed boy than in that of a supposed girl. Further, such a decision will tend to markedly diminish the chance of the individual becoming the victim of a homosexual marriage. Tuffier and Lapointe are of opinion that in doubtful cases an incision is justifiable to determine the nature of a sexual gland, especially when it is situated in the inguinal region; and, indeed, they would go further and suggest, in view of the grave inconveniences that a wrong decision may cause, that an abdominal section for diagnostic purposes is even justifiable.

[2] When a medical man discovers, in the course of an examination or an operation, that an individual is not of the sex in which he has been brought up and is at the time assuming, the question as to whether he should divulge his knowledge or not is one of considerable difficulty. It must be remembered that many of these individuals are quite happy in their ignorance, and the grave inconveniences which may follow a change of sex on the part of an adult must be remembered. In cases where marriage is contemplated and the medical man is consulted on a question of doubtful sex, an operation may certainly be recommended if necessary to determine the point

Neugebauer has collected a number of marriages in which, sooner or later, it was determined that both parties were of the same sex. As a rule, the male individuals (according to their sexual glands) play the rôle of the woman. Such erroneously made marriages can very seldom pursue a happy course. Frequently they result in a demand for a divorce, in which the doctor, as an expert, plays a large part. In some cases, the giving of a definite decision is not possible; for cases occur, as the above observations show, where the pathologist or the operator is the first person in a position to determine the true sex. All the same, Neugebauer has recorded happy marriages in which one of the parties was an hermaphrodite. The observation of Neugebauer, that male hermaphrodites have played the part of prostitutes successfully, is a very interesting one. Lastly, cases have been known where the sexual feeling of a hermaphrodite has been of both kinds, and the individual has sought sexual satisfaction on both sides, and has found it. If such persons are concerned in criminal actions (criminal assaults, sexual perversity, Imperial Criminal Code, Par. 175), it is of the utmost importance to determine their sex. As hermaphrodites often exhibit signs of mental degeneration, these facts must be borne in mind by the medico-legal expert. Chrobak and Rosthorn give a sketch, at the end of their monograph, of the legal enactments which are of importance in connection with these malformations. I can enter here only very briefly on the legal aspect of the question (on the enactments of the earlier Prussian Common Law, see p. 61, 'Allg. Landrecht,' T. 1, Tit. 1, §§ 19–23). The new Common Law Code, like the Austrian, recognises neither the word nor the meaning of 'hermaphrodite.' It is the intention that any malformed individual should be recognised as of a definite sex on the decision of a medico-legal expert. The conception of the Common Law Code that, according to present-day ideas, an hermaphrodite no longer exists cannot be accepted as correct in view of the observations mentioned above. According to this view, an undoubted defect in the Code exists (see also under the Legal Relations of Monsters).[1]

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Ahlfeld, *loc. cit.*, p. 34; Chrobak and Rosthorn, *Nothnagel, loc. cit.*, p. 53; vot. xx.; Stumpf, c. *Winckel, loc. cit.*, p. 44; Ziegler, *loc. cit.*, p. 15; Olshausen and Veit, *loc. cit.*, p. 44; Kleinhans, c. *Winckel, loc. cit.*, p. 34; Michaelis, *Kompendium der Entwicklungsgeschichte*; Hertwig, *Entwicklungsgeschichte*; v. Frisch, *Nothnagel, loc. cit.*, p. 53; vot. xix.; Spiegelberg and Wiener, *loc. cit.*, p. 15; Tillmanns, *loc. cit.*, p. 34; Leser, *loc. cit.*, p. 34; Biedert and Fischl, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Gebhard, *Pathologische Anatomie der*

[1] In regard to the legal relations of such persons, if married, it must be remembered that the English law takes cognisance only of the capability for coitus, and that on these grounds, in the majority of such cases, quite apart from the proof of the exact sex, a divorce might be obtained.

weiblichen Sexualorgane (Leipzig, 1889); Gütschow, *Weibliche Epispadie w. Diss., Rostock* (1904); Mackenrodt, *Hypospadia femin. Geb. Gesellsch. Berlin* (March 10, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 554. (For malformations of the uterus, tubes, ovaries, and vagina, see the text-books of Veit, Hofmeier, Runge, Fritsch, and Küstner.) Stolper, *Zwitter. Aerztl. Sachverst.-Ztg.* (1905), No. 1; Neugebauer, *Samml. klin. Vorte., N. F.*, No. 393 (Leipzig); L. Pick, *Über Neubildungen am Genitale bei Zwittern, usw., Arch. f. Gynäk.*, vol. lxxvi., p. 2; Demelin and Carhala, *Kong. Hématokele des Hodens. Gynäk. Gesellsch.* (Paris, Dec. 21, 1905), *ref. Zentralbl. f. Gynak.* (1906), p. 640; Merkel, *Einseitige Hodenverdoppelung, usw., Beitr. z. path. Anat. u. allg. Path.*, vol. xxxii.; Beuttner, *Atresia hymen. cong., Beitr. z. Geburtsh. u. Gynäk.*, vol. vi., p. 3; Schickele, *Tubenmissbildungen., Beitr. z. Geburtsh. u. Gynäk.*, vol. xi., pts. 1 and 2; Keith, A., *Human Embryology and Morphology* (1910); Bryce, T. H., 'Embryology,' *Quain's Anatomy*, vol. i. (1908); Berry Hart, D., *Atlas of Female Pelvic Anatomy* (1884); Taruffi, *loc. cit.*, p. 44, vol. viii. (1894); Ballantyne, *loc. cit.*, pp. 15 and 44; Englisch, *Wien. med. Jahrbuch* (1873), vol. i.; Spencer, H. R., *loc. cit.*, p. 86; Kermauner, F., *Schwalbe, loc. cit.*, p. 15, vol. iii., pt. 2, p. 253; Thiersch, *Münch. illustr. med. Zeitung*, vol. i. (1852); Frankl. O., *Volkmanns Sammlung klin Vorträge*, No. 363, (1903); Nagel, *Veit, Handbuch der Gynäkologie*, vol. i., p. 569 (1897); Pahauf, A., *Med. Jahrb. der K. K. Ges. d. Aertze* (Wien, 1885); Vagedes, *Inaugural-Diss. Breslau* (1903); Siegenbeek van Heukelom, *Zieglers Beiträge. path. Anat.* (1898), vol. xxii., p. 144; v. Neugebauer, *Hermaphroditismus beim Menschen* (Leipzig, 1908).

The Breasts

In conjunction with the malformations of the genital organs, the malformations of the breasts must now be considered. Congenital absence of the breast (Amazia) as well as congenital smallness of the nipple, or of the whole gland (Micromastia) are both very rare. A congenital increase in the number of the breasts (Polymazia, hyperthelia, hypermastia,[1] polymastia, mamma succenturiata, mamma accessoria) is more frequent. Supernumerary nipples, polythelia or hyperthelia, have also been observed. Either a double nipple is present on a single areola; or they belong, when they are situated some distance from the areola, to a mamma succenturiata (Ahlfeld). Hug has recently described a case of division of the left nipple into three portions. Both these malformations may occur in boys, and even more commonly in girls,[2] and must be regarded as a reversion to ancestors with multiple breasts (Atavism). The accessory mammæ are situated usually in the course of two lines[3] which pass from

[1] The presence of accessory mammæ in the neighbourhood of the primary gland is easily explicable if we recall the fact, that when the primary budding of the epiblast takes place, from which the gland is developed, the subdermal mesoblast is shallow and of small extent. In the subsequent growth of the thorax, the tissue in which the mamma is developed becomes widely spread out.

[2] According to some authors, accessory nipples are commoner in the male sex than in the female.

[3] The mammary line is a slight ridge of epiblast which stretches from the axilla to the groin, and represents the first stage of development of the mammæ in the mammalia. The majority of supernumerary mammæ and nipples occur along this line; most commonly, above the level of the umbilicus but sometimes below it.

the axillæ to the inguinal region. Exceptionally, they are found also in the region of the axilla, on the acromion process, on the back, in the middle of the abdomen (in the region of the navel), on the thigh, and even on the labium majus.

The majority of the small accessory glands hypertrophy during pregnancy, and secrete milk. Ahlfeld quotes a case (recorded by Robert Magendie) in which a woman, besides two normal breasts, possessed a third, with a well-marked nipple, on the thigh. In addition to her own child, which was suckled for thirty months on the thigh mamma, she suckled three other children for quite a long time.

From the tissues of such accessory glands, carcinoma, and benign adenomata may develop.[1] The ætiology of these interesting malformations is not very clear. According to Ahlfeld, it is probable that, by the pressure of the amnion, portions of the original mamma are separated and (adhering to the amnion) are transplanted on to the superficial surface of the body. In many cases, the direct transmission of the anomaly from the mother to the child has been observed.[2] Congenital hypertrophy of the breast is rare, and, in persons so affected, sexual precocity usually occurs (early occurrence of the menses, and early development of hair).

By gynæcomastia is meant the development of female breasts in the male. In the majority of these cases, some malformation of the genitalia is present at the same time (Hermaphroditism, or atrophy of the testes). A very remarkable case of gynæcomastia is recorded by A. V. Humbold, in which a man, thirty-two years of age, suckled his child for five months after the death of his wife. According to Stieda, the tissue of the mammary glands in cases of gynæcomastia is definitely different from that of the female breasts.

Literature

Kaufmann, *loc. cit.*, p. 24 ; Ziegler, *loc. cit.*, p. 15 ; Tilmanns, *loc. cit.*, p. 34 ; Ahlfeld, *loc. cit.*, p. 34 ; Hug, *Diss. Strassburg* (1908) ; Finney, *Keen, loc. cit.*, p. 53 ; vol. iii., p. 565 (1911) ; Teizo Iwai, *Lancet*, vol. ii., p. 753 (1907) ; Bland-Sutton J., *Evolution and Disease* (1890) ; Schmidt, *Anat. Anzeig.*, vol. xi., p. 702 (1896).

[1] It is a curious fact that the occurrence of neoplastic changes in true supernumerary breasts is almost unknown, only one or two doubtful cases having been recorded (Finney).

[2] Three views have been put forward by different writers to explain the nature of polymastia—namely, that it is an example of atavism, or that it is an instance of dichotomy of a single gland, or lastly, that it is merely a spontaneous variation. The occurrence of these glands, in other positions than along the mammary line, is to be explained on the supposition that, in these cases, they are merely giant sebaceous glands.

CHAPTER XIV

Malformations of the Skeleton (including the Pelvis, Extremities, and Muscles)

For congenital malformations of the skull, see p. 35; for spina bifida, see p. 25.

The spinal column in the new-born child is almost straight, and its typical curves are only formed later as the result of sitting, standing, and walking, and by the action of the body-weight and of the muscles.[1]

Congenital curvatures of the spine are uncommon. In several cases, scoliosis—as well as lateral curvature and shortening of the spinal column—has been observed congenitally.[2] Most commonly, this occurs in non-viable children, with other severe malformations of the spinal column and central nervous system. Static scoliosis in children, with a congenitally shortened lower extremity, occurs gradually after birth.

For congenital lordosis of the spinal column in cases of umbilical hernia, see p. 150. The lordosis of the lumbar vertebræ, associated with congenital dislocation of the hip, occurs first after birth, as a result of the marked pelvic inclination which is present.

Hohl gives a very complete account of congenital curvature of the spinal column. Meckel observed in two cases the cervical and dorsal portions of the spine so markedly bent forward that the upper and lower halves were united with one another at an acute angle, and the head appeared to be situated in the lumbar region. Carus observed a congenital scoliosis in a child whose mother had suffered early in life from rickets. Similar cases have been seen by v. Froriep, Potthof,

[1] There is only one curve—an anterior concavity—until the third month. At the beginning of the fourth month, the sacro-vertebral angle forms between the lumbar and sacral regions. At birth, the cervical and sacral curves have appeared, but the sacral not to any marked extent. The lumbar curve forms as the child learns to walk, so as to allow the body to be brought vertically over the lower extremities. The sacral and cervical curves then also become more marked. The dorsal curvature and the sacro-vertebral angle are the primitive curves, and are present in all mammals (Keith).

[2] A few cases of congenital scoliosis have been described due to imperfect development of one-half of a vertebra, or to the presence of a supernumerary half-vertebra.

Siebenhaar, Burnett, Herrmann, Humby, Henot, Klein, Nivert (with hydrocephalus), Castelli (with hydrocephalus). Jörg described the case of a new-born child in whom the upper half of the back had been so far bent backwards[1] that the shoulders lay on the lumbar region and on the sacrum, and had become superficially united there by skin. Similar cases have been recorded by Montault and Breschet. In many of these cases, thoracic or abdominal fissures were also present. Curvature of the spine may have an influence upon the course of labour, and may render it more difficult. Thus Hohl describes in detail a case in which marked curvature of the spinal column and other malformations were present, so that labour had to be ended with the forceps. The extraction of the curved back gave rise to so much difficulty that the child was born dead. Marked changes were present in parts of the spinal column as a result of the curvature. The moulded head was in its antero-posterior diameter longer than the trunk, and the post-mortem examination showed a condition of marked hydrocephalus. Hohl thought that in this case the large head had pressed the trunk from above against the uterus, and on the one hand had interfered with the free development of the trunk, and on the other hand had produced the peculiar position of the lower extremities; so that, as a result of this and of the pressure, the curvature of the spinal column and the oblique position of the pelvis, which was also present, had been brought about.[2]

The recognition of a congenital curvature of the spinal column during labour, if some obstruction arises in the delivery of the child, may be very difficult. An attempt must be made by the introduction of the half or whole hand, under an anæsthetic, to carry out a complete examination of the child. In the case described above by Jörg, he was able to complete the delivery by the division of the skin adhesions. Very frequently, in consequence of a malpresentation, interference is necessary

[1] The most marked degree of this deformity occurs in the calf. In these cases the animal appears as if it had been turned inside out, its head and limbs being contained within an everted sac lined by hair; while attached to the outer surface are the various thoracic and abdominal viscera. There is marked lordosis of the spinal column, so that the head and the pelvis lie almost in contact. Knoop has described an early human ovum in which the head, neck, and back are adherent to the inner surface of the chorion, and which he regards as a very early stage of this so-called condition of schizosoma reflexum. As has already been mentioned (see p. 152), Kermauner regards the condition as due to imperfect development of the primitive myotomes, together with abnormal tendency of their growth; so that, while the layers of the splanchnopleure unite normally to form the intestinal tract, the layers of the somatopleure grow in a faulty outward direction, and give rise to thoracic and abdominal fissures.

[2] Specimen No. 824 in the University College Hospital Medical School Museum (Obstet. Section) is a good example of a fœtus retroflexus without any lateral curvature. The body of the fœtus is so bent backwards that the back of the knee and the shoulder of each side is in contact. There is a well-marked abdominal fissure, extroversion of the bladder, and a cloaca.

—especially the performance of version. According to Hohl, podalic version in these cases is only possible when the curvature is such that it allows of extension of the trunk. Otherwise the breech, or (in a posterio. curvature) the anterior surface of the pelvis of the child, must be drawn down into the inlet of the pelvis, and the child so extracted.

Defects of development in the spinal column and in the skeleton (see p. 241)—hypoplasia and hypertrophy—have been observed. In the case of defective formation, a portion of a bone may be completely absent, or its development may be hindered *in utero*: for example, portions of the vertebral arches may be absent. If a failure of continuity of the articular portion of the arch of the fifth lumbar vertebra is present, as the result of the defective union of the anterior and posterior centres of ossification of the vertebral arch (Congenital spondylolysis), then spondylolisthesis of the fifth lumbar vertebra occurs, and, as a result, a spondylolisthetic pelvis with narrowing of the antero-posterior diameter at the pelvic inlet. In these cases, the fifth lumbar vertebra, with the overlying portion[1] of the spinal column, slides downwards and forwards over the base of the sacrum, and at the same time a well-marked degree of lordosis of the spinal column occurs. The body of the fifth lumbar vertebra may be so displaced that its base lies directly on the anterior surface of the sacrum. The displacement downwards, described above, occurs first after birth; and in women, mainly during pregnancy.[2]

Besides these defects of development, accessory vertebræ in all parts of the spinal column have been occasionally observed. Further, so-called 'tail formations,' with elongation of the spinal column, have been described in the coccygeal region. True[3] and false tails may be distinguished: the first containing bones, and the latter neither bones nor cartilage. The last belong to the teratomata (see p. 251).

Supernumerary formations have also been described among the ribs at times—for example, the not uncommon cervical and lumbar ribs. More frequently, a bifurcation of the ribs occurs; most commonly, with

[1] The displacement of the last lumbar vertebra is rendered possible in one of two ways: (1) By a separation of the anterior and posterior halves of the vertebra by a want of union between the arch and the body; or (2) by a dislocation of the whole vertebra forwards upon the sacrum. According to Neugebauer, there may be first a dislocation, and, later, a separation, owing to the action of the body-weight.

[2] The change in the pelvis is usually produced gradually by the action of the body-weight. It is often accompanied by inflammatory changes in the bones, and the sacrum; and some of the lumbar vertebræ may become fused together into one mass.

[3] The true tails are due to the persistence of the condition which obtains in the fœtus in the second month, when some seven to eleven coccygeal vertebræ are present. The tail of the embryo at this period consists of two parts: a proximal portion, containing the vertebræ; and a terminal portion, the so-called caudal filament. In the course of development, the caudal filament disappears, and the proximal portion ceases to form a projection, but becomes incorporated with the surrounding structures, the supernumerary vertebræ disappearing by a process of fusion.

associated supernumerary vertebræ. In this place, too, should be mentioned the very rare condition of doubling of both frontal bones (see Ahlfeld).

Under this heading, cases of abnormally small increase in length (Hypoplasia of the skeleton) also belong; as a result of which, a striking shortness of the extremities occurs—micromelus; or, in the highest degrees, phocomelus[1] (so termed on account of the resemblance to a seal). In such cases, the soft parts are normally developed, and are therefore too long and too wide for the shortened bones. The skin, like a garment which is too large, lies in folds, and is either œdematous or very rich in fat. Sometimes, general dropsy or anasarca is present, which may lead to difficulty in delivery. As a result of the presence of the stunted extremities, the trunk, although really normal, appears unusually broad.

The majority of cases of micromelia are produced by the condition termed chondrodystrophia fœtalis.[2] We have to thank E. Kaufmann for his original work on these diseases or malformations of the bones, which have revealed the nature of the process. To Kaufmann belongs the credit of having brought clearness and system into the consideration of this subject, in which many different unrelated conditions had been jumbled together. All these cases were, formerly—and are, to some extent, even at the present time—erroneously termed fœtal rickets, or 'so-called fœtal rickets,' Müller's disease, and also micromelia chondromalacica. The French call the process 'Achondroplasia.' The most practical designation is 'Congenital fœtal rickets,' under which name, however, as has been mentioned, very different processes have been confused together. As Kaufmann has shown, the process is really one of faulty growth of the cartilage and premature cessation of the endochondral ossification (see Kaufmann, 'Textbook of Special Pathology,' pp. 703–708).[3] The diaphyses remain short,[4] although they

[1] The reader must note that some writers—Ballantyne, for instance—use the term 'Phocomelus' not as synonymous with 'Micromelus,' but as signifying a condition of arrested development of one or more of the proximal segments of the limbs.

[2] This condition was not recognised as a definite disease until 1860, when Müller differentiated it from cretinism and rickets, and showed that the failure of the long bones to develop was due to disturbance of the zone of proliferating cartilage at the epiphyseal line.

[3] A great degree of confusion exists upon the difficult subject of fœtal bone-disease. Kaufmann's classification is, on the whole, the simplest and most comprehensive. Under the general term chondrodystrophia fœtalis, he recognises four varieties of altered growth of the cartilage: a softening (Chondrodystrophia malacica), an arrest of its growth (C. hypoplastica), a growth unaccompanied by any increase in the length of the bones, and irregular growth (or C. hyperplastica).

[4] The long bones, besides being short, are much curved, and the ends of the shafts are cupped to receive the epiphyses. The skull-bones, however, show no marked deformity; but there is stunting of the length of the basis cranii, due to premature synostosis of the basi-sphenoid and basi-occipital.

often become (as the result of a relatively marked periosteal formation of bone) more or less sclerotic. As regards the appearance of the child, as a result of the process, either a marked depression of the root of the nose is present, so that the aspect is rather that of a cretin, or else the nasal region as a whole fails to project. The majority of the children are either born dead or die a few weeks after birth at latest. Only in very rare cases do they live to be thirty years old or older. Such individuals are then very small, plump, clumsy, and disproportionately short,[1] but usually of good intelligence. The pelvis assumes the form of the so-called dwarf pelvis (Pelvis nana). According to v. Franqué, the cause[2] of this bone-disease must be sought, primarily, in a want of room *in utero*, and this view is confirmed by the fact that frequently club feet and fissure formations, as well as defects of the extremities, have been observed. An interesting feature is that on many occasions such malformed children have been borne by the same mother, which favours the hypothesis that

Fig. 43. — Chondrodystrophia Fœtalis. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] The striking features are a form of dwarfism, characterised by disproportionate shortness of the extremities, a large head, pug nose, prominent nates and abdomen, and fingers diverging from one another like the spokes of a wheel (Poynton). The fingers are usually all of the same length, giving rise to the curious appearance called by Marie 'le main-en-trident.'

[2] This disease is met with in some breeds of dogs, such as dachshunds, and also in cattle. It has been suggested that it is a cretinous condition; but there is no evidence in favour of this, and, as a rule, there is no disease of the thyroid. Shattock thinks that it may be a paracretinous affection.

the cause lies in the maternal organism; for example, poor nutrition of the mother during her pregnancy has been suggested. The additional fact that, in several instances, hydramnios has been observed in such cases does not go for much: for, as is well known, hydramnios is a very frequent accompaniment of all possible fœtal malformations and diseases. The disease previously known as Müller's disease must also be included under this head (see Küstner, in 'Müller's Handbook of Obstetrics'). It presents congenital affections of the bones which, according to Müller, have a great resemblance to rickets; but which, as he points out, show very definite anatomical differences from this disease. The hypothesis, too, put forward with greater assurance by Winkler, that there are two forms of fœtal rickets can no longer be considered correct, in view of the observations of Kaufmann. Winkler distinguishes micromelica, shortly called micromelia (which he regards as past rickets, the result of which is micromelia), and rachitis annulans. In the second form, the rickets is in a progressive condition so that it has already led, *in utero*, to fractures of the bones. In other cases, again, the fractures occur first outside the uterus. Winkler has adopted the term 'rachitis annulans,' from the ring-like formations of callus lying one upon another on the bones of the extremities. We may certainly, at the present time, say that what he terms rachitis micromelia belongs to chondrosdystrophia, and rachitis annulans to osteopsathyrosis, the result of osteogenesis imperfecta.[1]

Chondrodystrophia is certainly not the only cause of micromelia which, as Kaufmann observes rightly, is not a disease *sui generis*, but only a symptom: for example, the so-called osteogenesis imperfecta leads to micromelia. According to the observations of Harbitz, Bnday, and Kaufmann, this condition is one of an imperfect formation of bone substance; and this not only on the part of the medullary, but also on the part of the periosteal, osteoblasts.

In contradistinction to chondrodystrophia, the endochondral ossification is practically normal[2] (see Kaufmann). As a result of these

[1] As Ballantyne points out, there is a good deal of conflict of evidence as to the frequency with which fœtal rickets occurs. Thus, while Schwarz found rachitic changes in some 80·6 per cent. of new-born children, Fede and Cacace, on the other hand, only found such evidence in one of 500 children born in Italy, and with their estimate Ballantyne is inclined to agree.

Cases have been recorded from time to time of children born with beading of the ribs, delayed ossification of the cranial bones, and green-stick fractures of the long bones, which appear to prove the possibility of fœtal rickets occurring.

[2] The layer of periosteal osteoblasts is thick, but the cells are spindle-shaped; and, instead of a continuous layer of dense laminated bone with stellate bone corpuscles and Haversian canals, plates of imperfectly laminated bone are formed with oval bone-cells and no Haversian canals. Even in the epiphyseal line ossification does not occur quite normally. The cartilage cells persist, and the newly formed trabeculæ of bone are developed by calcification of cartilage cells, and not through new bone formation by osteoblasts on a persistent cartilaginous matrix, as in normal ossific.

pathological processes, the bones are abnormally easily broken (osteopsathyrosis, osteoporosis congenita, and fragilitas ossium congenita). This osteopsathyrosis is undoubtedly one of the most frequent causes of intra-uterine and extra-uterine fractures.[1] Even during fœtal life, numerous complete and incomplete fractures may occur as a result of this pathological condition of the bones, especially in the ribs and the bones of the extremities. The fractures may already be healed at birth with marked formation of callus. During labour, too, such fractures may very easily occur (see also under intra-uterine fractures). Kaufmann gives two instructive illustrations of osteogenesis imperfecta with numerous fractures. In the Göttingen Women's Clinic, among 45,000 births, only one case of osteogenesis imperfecta was observed. A secundipara, twenty-six years old, was admitted into the clinic in labour. A female child, weighing 1050 grm. and 33 cm. long, was born in a footling presentation, and died one hour after birth. The extremities were short and markedly bent, so that the joints could not be distinguished; while the ribs were very soft, and almost all the bones gave crepitations. There was also abnormal mobility and crepitation of the lower jaw; while the skull-bones were very easily indented. When the bones were exposed, they were almost all imperfectly ossified, and in many places fractured. The thyroid gland also showed slight hypertrophy.

The bone-disease, formerly described as chronic parenchymatous osteitis (dul. Schmidt; see Küstner in 'Müller's Handbook of Obstetrics'), also belongs to this group. In this disease, recent and old intra-uterine fractures occur, and the slightest pressure leads to fractures and depressions of the bones.

Under this heading also belongs dwarf formation (Nasosomia, which has already been considered, see p. 23). In cretins, too, interference with the growth of the skeleton is frequently seen.

The obstetrical importance of chondrodystrophia consists in the fact that the general œdema, or dropsy of the cavities of the body, which is usually present, may lead to difficulty in the delivery of the child. For instance, Winkler records a case of difficult delivery from such a malformation. At the post-mortem examination of the child, effusions were found in all the serous cavities, a very thick panniculus adiposus, and œdema of the subcutaneous tissues. Ahlfeld pictures a very extreme case of chondrodystrophia fœtalis, which, as a result of the marked ascites present, led to difficulty in delivery. The illustration shows the typical nasal formation in this condition. In the Göttingen Women's Clinic, we had recently the

[1] Lueck has recorded a case of fracture of almost all the long bones in a child born spontaneously within the membranes, in a breech presentation. Julian Schmidt recorded a similar case, and Ridd observed fifty-four fractures in a child born spontaneously.

opportunity of observing the birth of such a deformity. The case was that of a strong, healthy woman who, three years previously, had given birth to a perfectly healthy child. The labour took place comparatively rapidly, with good pains, the child presenting by the breech. The head was delivered in Walcher's position with great difficulty. There was a considerable degree of hydramnios, and, besides the micromelia, a well-marked hydrocephalus, general slight œdema, and marked ascites were present. The child, a male, still-born, weighed 3500 grm.; its length was 39 cm., and the circumference of its head 36 cm. Another specimen of chondrodystrophia and hydrocephalus is in the pathological collection of the Göttingen Women's Clinic.

FIG. 44.—CHONDRODYSTROPHIA FŒTALIS. (Specimen from the collection of the Göttingen Women's Clinic.)

As the literature shows the delivery of such cases causes, as a rule, no great difficulty. In children with osteogenesis imperfecta, according to the observations on record, spontaneous fractures of the bones may occur very readily during labour, especially when any obstetrical manœuvres are undertaken—such as liberation of the arms or extraction—especially of the upper arm, of the thigh, and of the clavicle, for which the operator cannot be blamed.

In this place, also, must be mentioned gigantism (Macrosomia); although, strictly speaking, it does not belong here. In this malformation, a tendency to excessive growth is present, either of a partial or of a general nature. As Kaufmann points out, it is really a hypertrophy of congenital origin, which first becomes apparent after birth. The so-called giant children, which at their birth markedly exceed the normal weight, in the majority of instances have nothing to do with this condition (see p. 277).[1]

I must here, also, briefly refer to premature synostosis. When premature ossification occurs in the sutures or in the joints, the growth of the bones in these positions ceases completely. In the skull, a so-called microcephaly[2] occurs (see p. 35). In the pelvis (as the result of unilateral or bilateral premature synostosis of the sacroiliac articulation, with consequent defect of one or both ala of the sacrum), an obliquely contracted or Naegele pelvis occurs, or a transversely contracted or Robert's pelvis. Both of these pelves may lead to severe difficulty in delivery when the contraction of the oblique or of the transverse diameters is well marked.

Syphilis is a common congenital disease which, in the fœtus, especially attacks the bones. Congenital syphilis of the bones occurs rarely as periostitis ossificans, with or without osteochondritis syphilitica and gummatous osteomyelitis. Of more frequent occurrence, and well known since the time of Wegner, is osteochondritis syphilitica, which is especially met with at the line of union of the cartilage and the bone, in the long bones of the limbs (femur, tibia, and humerus). The recognition of osteochondritis syphilitica is proof of the diagnosis of inherited syphilis, and such recognition is possible even in macerated fœtuses. This pathological process in the bones can very readily be recognised when syphilitic changes in the other organs are not present. Indeed, a case is generally not diagnosed as congenital syphilis, if this change is not found. At the present day, we have (as is well known) in the recognition of the spirochæta pallida a more important and certain help in the diagnosis.[3] While the normal line of union between the diaphysis and epiphysis forms a fine straight line, in osteochondritis syphilitica a more or less dentated irregular broad line of a yellow or cheesy colour

[1] Ballantyne defines 'macrosomia' as 'monstrous largeness' of all the parts of the individual, and points out that the more characteristic cases are post-natal. In true gigantism (the 'neanio-gigantism' of Taruffi), the excessive growth of the individual usually begins at or after the fourteenth year.

[2] The reader must remember that in true microcephaly, premature ossification of the sutures is the exception and not the rule; and that in the microcephalic idiot, the skull is in reality adapted to a small undeveloped brain.

[3] It must be pointed out that such an authority as Still affirms positively that this condition—namely, osteochondritis—is not always present in syphilitic infants, and doubts whether it is usually present.

is found in this situation. Microscopically, the process consists in a small-celled infiltration, resulting in disintegration, and, later on, in suppuration. In severe cases, even *in utero*, as the result of the formation of granulation tissue, a separation of the epiphysis from the diaphysis may occur, which is distinguished as syphilitic loosening of the epiphysis, in distinction to the traumatic variety. The portions of bone thus separated can become united again *in utero*. It is evident, as a result of these bone changes, from the loosening which is present between the epiphysis and the diaphysis, that separation of the epiphysis may very readily occur during obstetrical operations—such as liberation of the arms, bringing down a leg, extraction in breech presentations—and that this must not be attributed to the fault of the obstetrician.

As regards obstetrical injuries to the skeleton, I must refer the reader to the remarks relating thereto in the different chapters. Here I will say only a few words on tearing of the spinal column. Tearing of the cervical vertebræ has been observed in very difficult and forcible delivery of the head—especially in contracted pelves when the force has not been exerted in the axis of the spinal column, or when the column has been to some extent twisted. The separation of the tissue in the vertebra occurs, as a rule, in the epiphyseal line[1] of the body of the vertebra; less commonly, in the intervertebral disk. Usually, hæmorrhage also occurs into the spinal canal; both stretching and tearing of the spinal cord and the nerves issuing therefrom, and hæmorrhage into the prevertebral connective tissue.

Maceration of the fœtus, as well as general œdema, favours these injuries to the vertebræ; the most frequent cause used to be the Prague method of delivery (at the present time almost given up), in which the cervical vertebral column was bent at an angle. Injuries to the thoracic and lumbar vertebræ are much less common, as the point of application of the force lies lower down.[2] In the pelvis, injury to the sacro-iliac articulation has been seen chiefly in extraction of the breech by the foot. According to Ruge, as a result of this injury, a so-called ankylotic obliquely contracted pelvis may occur. The injuries of the spinal column, described above, usually result in immediate death. Ahlfeld, however, saw a case in which the child lived for nine days after a fracture of the thoracic vertebræ.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Tillmanns, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Breus and Kolisko, *loc. cit.*, p. 24; Müller, *loc. cit.*, p. 15; v. Winckel, *loc. cit.*, p. 43; Biedert and Fischl, *loc. cit.*, p. 34; Ahlfeld, *loc cit.*, pp. 24 and 34;

[1] That is, at the line of union of the upper or lower epiphyseal plates with the body of the vertebra.

[2] And, therefore, the bending of the spinal column is much less.

Spiegelberg and Wiener, *loc. cit.*, p. 15; Hohl, *loc. cit.*, p. 15; Braun-Fernwald, *Geburtshilfe*; Stumpf, *v. Winckel, loc. cit.*, p. 44; Birnbaum, *loc. cit.*, p. 101; Wydner, *Geburtsh. Operationen, v. Winckel. loc. cit.*, p. 15; J. S. Fowler, 'Osteogenesis with numerous Fractures,' *Edinb. Med. Journ.* (Jan. 1906); Fleury, '*Angeborene Skoliose*,' *Diss. Paris* (1901); Knoop, '*Chondrodystrophia fetalis*,' *Versamml. deutsch. Naturf. u. Ärzte* (Köln, 1908); Bürger, '*Intraut. Osteoporose*,' *Geb. Gesellsch.* (Wien, June 3, 1902), *ref. Zentralbl. Gynäk.* (1903), p. 374; Michel, '*Osteogenesis imperf. mit kongenitalen Frakturen*,' *Virchows Arch.*, vol. clxxii.; Lindemann, '*Osteogenesis imperf.*,' *Diss. Berlin* (1903); Nau, '*Angeborene Skoliose*,' *Diss. Paris* (1904); Bar, '*Kongenitale Kyphose*,' *Geb. Gesellsch. Paris* (June 16, 1904), vol. i., p. 4; *ref. Zentralbl. f. Gynäk.* (1905), p. 1383; Schwalbe, *loc. cit.*, p. 15; vol. iii., pt. 1, p. 64; Knoop, *Hegars Beitrage*, vol. vii. (1903), p. 284; Neugebauer, *Spondylolisthesis et Spondylizeme* (1892); Ballantyne, *loc. cit.*, p. 44; Still, G. F., D'Arcy Power and Murphy, *loc. cit.*, p. 206; vol. i., p. 336 (1908); Poynton, F. J., *Clifford Allbutt, loc. cit.*, p. 89; vol. iii., p. 117.

Congenital Tumours of the Coccyx

I shall now consider the so-called congenital tumours of the coccyx; although, strictly speaking, according to their mode of origin they cannot, as a whole, be tabulated under one heading. All these tumours are situated either in the region of the sacrum, or of the coccyx, or of both. Under this name, as has already been pointed out, very different structures are included, and they may be divided into six classes as follows:—

Class I. The malformations already described, known as spina bifida (p. 25).

Class II. Tumours which arise from the coccygeal glands. (Some authors—Ahlfeld and Arnold—dispute this mode of origin.)[1]

Class III. Congenital lipomata and lymphangiomata, which mainly arise from the tissue between the sacrum and the rectum.

Class IV. Tail formations (see p. 243).

Class V. Tumours which are to be regarded as local defects of development, so that a displacement or separation of tissue has occurred within a single individual—monogerminal teratomata and teratomatous cysts. According to Ziegler, the multiplicity of the tissues in the region of the coccyx explains the fact that remains of the caudal gut, and of the embryonic medullary tube, as well as portions of the coccygeal vertebral column, and of the pelvis with muscle tissue, may take part in the structure of such a teratoma. These tumours are dermoid cysts containing various tissues, bones, cartilage, muscle, glands, hairs, sebum, etc.

Class VI. Teratomata, with rudimentary extremities, or various parts of the body, which must be considered as examples of a double monster—such as a rudimentary pygopagus, or as a dipygus parasiticus (see p. 325),

[1] The coccygeal glands are probably derived from the chromophil cells of the abdominal sympathetic system; and, while they have been found unaffected in cases of coccygeal tumours, yet recent observations have demonstrated that perithelio-mata may arise from them.

bigerminal teratomata, fœtus in fœtu (in which the well developed fœtus is distinguished as an autosite; the rudimentary twin as a parasite, more especially as an epipygus).[1]

Coccygeal tumours are divided into dorsal and ventral. I shall limit myself here mainly to a discussion of the dorsal tumours. According to v. Bergmann, 87 per cent. of the children with these tumours are either born dead, or die in the first few days after birth. They are most commonly of the female sex. The complete macroscopical diagnosis of such a tumour after the birth of the child is not always easy. Rectal examination is of great importance. If there is a cleft in the spinal canal, and if the tumour can be lessened by pressure, then it is a meningocele. Teratomata exhibit, in their interior, rudiments of organs; while, by the puncture of a dermoid, sebum can usually be recognised. The subcutaneous tissue of a teratoma is very often œdematous, and cysts may be observed at the same time, in the neighbourhood of the tumour, belonging to the autosite.

The nutrition of the tumour is usually carried on by branches of the middle sacral artery. As Ahlfeld points out, all coccygeal teratomata form under the gluteal muscles, and very frequently press the perineum and the rectum forwards.[2] In the same way, the anus and the genital organs are usually displaced forwards in such a way that it appears as if they were situated on the anterior upper surface of the teratoma. In very rare cases, movements have been recognised in sacral tumours which, probably, have been produced by the contraction of muscle fibres.[3] Diagnostic difficulties may occur, especially when the dermoid

[1] Classes V and VI, which correspond to the true congenital coccygeal tumours, may be conveniently divided, according to Schwalbe, into four divisions: (1) The cases of true parasites recognisable to the naked eye; (2) parasitic tumours containing parts of organs; (3) teratomata containing derivatives of all three germinal layers; (4) teratomata containing derivatives of two germinal layers.

[2] These points are well shown in Figs. 381, 382, 383, in Schwalbe's book; and, as the very excellent table which he quotes from Nakayama shows, every gradation of structure, from the simplest to the most complex, may be met with in these tumours. The more complicated the structure, the earlier is probably the period of development of the embryo at which it originates.

The terms 'monogerminal' and 'bigerminal' refer to the two views of their origin: namely, from the neurenteric canal or from a parasitic twin. The neurenteric canal passes from the primitive neural canal into the posterior end of the yolk-sac, and during the development of the neural folds should finally disappear. As it represents a canal derived from the ectoderm communicating with a cavity (the primitive hind-gut), lined by entoderm, and is surrounded by mesodermic tissue, it is easy to see how such a source of origin can explain the complicated structure of a teratoma. The tendency of many observers at the present day is to attribute all these tumours to the imperfect development of a second implanted ovum.

[3] The cases in which movements have been recognised in the tumour probably belong to division 2; and a good example of such a tumour is the sacral parasite, described by Schwalbe, in which a rudimentary extremity was present, and in which he was able to demonstrate muscle tissue.

suppurates, spontaneously or after an injury, with the formation of a fistula.

The size of the tumours varies between that of a walnut and that of a child's head.[1] The treatment of such formations is best carried out by the extirpation of the tumours. Great care is naturally necessary, especially if a spina bifida still communicates with the spinal canal. A large number of cases are recorded in the literature in which the extirpation of sacral tumours has been carried out successfully.[2]

FIG. 45.—COCCYGEAL TUMOUR. (Specimen from the collection of the Göttingen Women's Clinic.)

The obstetrical importance of congenital sacral tumours lies in the fact that with large tumours difficulty may arise in the delivery of the child. By far the larger number of the cases are born spontaneously without any difficulty; even although the pregnancy in most of them reaches its normal termination. This result does not depend upon the size of the tumour alone. The size of the child, the consistence and resistance of the tumour, the width of the pelvis, the character of the pains, and the mode of exit, all play an important rôle. Of forty cases which Hohl collected — among which some tumours in other parts of the body, certainly, were included— in eighteen cases artificial help was required (forceps, version, extraction, puncture, tearing of the sac, perforation of the sac, and the use of a sharp hook). In six further cases the labour was 'difficult.' Among the seventy-nine cases collected by Bramm, in twenty-two some difficulty in delivery occurred. It is a striking fact that such children commonly present by the skull, although the centre of gravity of the body is displaced towards the pelvic extremity. The

[1] Woolsey records a case in which the tumour weighed 10 lb.; and until removed prevented the patient, a man of thirty-four, from wearing trousers.

[2] In most instances, they can be readily enucleated; in the case of doubtful parasites any communication between the vertebral canal of the autosite and the parasite should be carefully looked for.

reason for this is that the breech and the sacral tumour together do not enter well into the lower uterine segment.

In many cases, however, malpositions and malpresentations have been observed. In the majority of cases, the hindrance to delivery first becomes evident when the head and the shoulders are born. The attendant is hardly ever in a position to make a certain diagnosis of such tumours by external examination; and even after the birth of the head and shoulders the exact diagnosis is not always easy. Conjoined twins, and especially the presence of a second child (impaction of twins, with simultaneous entry into the pelvis), will be thought of, or a second amniotic sac, when the tumour is cystic and the tumour wall very thin. Mistaking a sacral tumour for the bag of membranes is very easy when the tumour is the fore-coming part. Such mistakes, as appears from the literature, have been very frequently made. A spina bifida or a fœtal inclusion may also be thought of. The introduction of the half or whole hand, under anæsthesia, and a complete exploration of the child's body and its appendages is necessary. In order to determine the exact condition, it is important, by a preliminary external examination, whenever practicable, to exclude a twin pregnancy—a conclusion which, in such a case is, however, not always possible. Further, in cases of cephalic presentations, after the birth of the head, a complete examination of the thorax and the abdomen must be carried out, to determine whether the cause of the difficulty in delivery is to be sought in these regions. Hohl gives some delicate diagnostic signs, by which one may be in a position to differentiate the various tumours more certainly. While these various hints are certainly very valuable, they have mainly only a theoretical value, and can hardly lay claim to any practical importance.

The prognosis during delivery, in the case of such tumours, is good for the mother; but, as has been shown, is not so favourable for the child. It is important, however, to remember that a large number of these children with sacral tumours have been operated upon successfully. When no absolute obstruction to delivery exists it is important therefore to proceed carefully, as rupture of the uterus or other injuries of the mother's soft parts, may occur as the result of too forcible attempts at delivery. Rupture of the uterus may also occur when the obstruction is not recognised and the lower uterine segment becomes more and more stretched. As a means of delivery, the forceps must be considered, when the head is not too small; version; extraction by bringing down one or both feet; puncture or incision of the tumour.

A suggestion of great value is to draw the part causing the obstruction to delivery into the roomy cavity of the sacrum either by traction or by suitable manipulation on the lower extremities, which have been brought down, or on the trunk. In some cases, it is

possible, with or without puncture, to draw down the tumour with the hands, or an instrument, so that the further progress of the labour takes place readily. In other cases, the tumour bursts during forcible attempts at extraction. With dead children, or in absolute obstruction and danger to the mother, the difficulty should be overcome by the treatment of the tumour by puncture, perforation, incision, or crushing. The latter, according to Hohl, is better avoided as not a proper manœuvre. Here, as always in the exercise of the *ars obstetrica*, the primary axiom is 'the mother first, then the child.' In no circumstances is it right to endanger the mother's life for the sake of a child, whose continued existence is always very doubtful. In the collection of the Göttingen Women's Clinic there is a specimen of a fœtus with a sacral tumour (see Fig. 45), which has already been fully described (in 1827) in an anatomical journal.

Literature

Kaufmann, *loc. cit.* p. 24; Ziegler, *loc. cit.*, p. 15; Tillmanns, *loc. cit.*, p. 34; Baginsky, *loc. cit.*, p. 34; Beckert and Fischl, *loc. cit.*, p. 34; Müller, *loc. cit.*, p. 15; Kleinhans, *v. Winckel, loc. cit.*, p. 34; Spiegelberg and Wiener, *loc. cit.*, p. 15; Hohl, *loc. cit.*, p. 15; Brünn, *Diss., ref. Zentralbl. f. Gynäk.* (1903), p. 219; Gramm, *Diss. München* (1902); Véron, *Geb. Gesellsch. Paris* (June 19, 1902), *ref. Zentralbl. f. Gynäk.* (1903), p. 670; Peiser, *Vereinigung Breslauer Frauenärzte* (March 17, 1903), *ref. Zentralbl. f. Gynäk.* (1903), p. 908; Engelmann, *Arch. f. klin. Chirurgie*, vol. lxii., No. 4; Füth, *Geb. Gesellsch., Leipzig* (February 15, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 591; Uthmöller, *Vereinigung Breslauer Frauenärzte* (July 21, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 899; *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xix; Hoppe, *Diss. Breslau* (1903); Chiari, '*Histol. Untersuchungen von 13 Sakraltumoren*,' *Verhandl. d. deutsch. path. Gesellsch.* (1904), No. 2; Boissard, *Geb. Gesellsch. Paris* (July 7, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 1383; Frank, *Deutsch. Zeitschr. f. Chir.*, vol. lxxvii., Nos. 4–6; Kjelsberg, *ref. Zentralbl. f. Gynäk.* (1905), p. 1142; Fruhinsholz, *Ref. Zentralbl. f. Gynäk.* (1905), p. 1149; Hoffmann, *Diss. Leipzig* (1904); Thies, *Geb. Gesellsch., Leipzig* (Feb. 26, 1906), *ref. Zentralbl. f. Gynäk.* (1906), p. 932; Thaler, *Deutsche Zeitschr. f. Chir.*, vol. lxxix.; Ahlfeld, *loc. cit.*, p. 34; Braune, *Die Doppel-missbildungen und abnormen Geschwülste der Kreuzbeingegend* (Leipzig, 1862); R. Keller, *Arch. f. Gynäk.*, vol. lxxxv., No. 3; Schwalbe, *loc. cit.*, p. 15; vol. ii., pp. 359–374 (1907); Keen and Coplin, *Surg. Gynecol. and Obstet.* (Nov. 1906); Woolsey, *Keen, loc. cit.*, p. 53, vol. ii., p. 832 (1911).

The Shoulder Girdle

Congenital partial or total absence of the clavicle is very rare. Tillmanns figures a case of bilateral defect of the clavicles. The arms of the affected child could be brought completely into contact with one another over the chest, while no marked disturbance of function was present.[1]

[1] Ballantyne points out that defective development, as shown by absence or non-ossification of the acromial end of the bone, is commoner than absence.

A congenitally high position[1] of the right or left or of both shoulder-blades is more frequent. Tillmanns figures a case, observed by himself, in which, in an otherwise healthy female child, the right shoulder-blade was displaced upwards for 4 cm., without any functional impairment of the arm. According to him, the condition is to be attributed to an abnormal position of the child *in utero*, or to some disproportion between the size of the child and that of the uterine cavity. No definite treatment is required for this condition.

Congenital dislocations of the shoulder-joint have been observed on one or both sides. In such dislocations, the glenoid cavity on the scapula is either absent or markedly deformed, and at the same time a flail-like joint is present. Of the various dislocations possible, the most frequently observed is a congenital subcoracoid or infra-spinous, in which the head of the humerus is displaced, either under the coracoid process or under the spine of the scapula, and in these positions an abnormal joint-socket is formed in the majority of the cases. Treatment is not of much avail in these rare malformations. The abnormal displacement of the humerus which may occur during labour, especially in artificial delivery, from separation of the epiphysis or injury to the brachial plexus, is of a different nature from these foetal malformations of the shoulder girdle.

Traumatic dislocations of the shoulder girdle, according to Küstner, do not occur, as a result of trauma, during pregnancy or during delivery. When, as Küstner points out, such force is employed as would in an adult produce a dislocation, in the foetus separation occurs between the epiphysis and the diaphysis. However, some definite dislocations of the shoulder, the result of difficult delivery of the arms, have recently been recorded (Olshausen, luxatio subcoracoidea: see Stumpf).

Here we may remark that with a very marked shoulder girth—such as is met with in giant children and in cases of hemicephalus—difficulty in the delivery of the child may readily occur.

The obstetrical injuries which have been observed *intra partum* in the shoulder-girdle are fracture of the clavicle, separation of the epiphysis at the neck of the scapula, fracture of the neck of the scapula, breaking off of the acromion, and transverse fractures through the scapula. The most interesting and the most frequent are fractures of

[1] Congenital elevation of the scapula, or Sprengel's deformity. The scapula is not only elevated, but rotated so that the lower angle approaches the spine. Not uncommonly there are associated anomalies, such as absence of the radius, torticollis, or atresia ani. In support of the view that the deformity is due to an insufficiency of the liquor amnii, it is stated that the arms tend to become dorsally displaced after birth. Occasionally, there is a bridge of bone present connecting the scapula with the spine.

the clavicle. These may be met with, as Mums[1] from the result of his observations at the Copenhagen Clinic has remarked, and this has been confirmed by many authors, even in a spontaneous labour. Fractures of the clavicle, however, are most frequently the results of obstetrical operations. They may be caused either directly or indirectly. Directly, for example, by the use of Veit's grip, indirectly by the intermediation of the upper arm, or of the shoulder when the shoulder during extraction is drawn so far down that it becomes wedged in the pelvis; also in difficult cases of freeing of the arm when pressure is exerted on the scapula or on the clavicle. The fracture is generally situated at the junction of the middle and outer thirds of the clavicle. It is often overlooked, as it almost always heals spontaneously without any trouble. The diagnosis is made by palpation, by the recognition of the mobility of the broken bone, by the presence of crepitation, by the low position of the affected shoulder, and finally by the diminution in breadth of the corresponding half of the shoulder. The special symptom which is sometimes met with is that the child cannot move the arm of the affected side. The prognosis is good, and treatment is only required when marked displacement of the broken bone is present. The arm is simply bandaged to the thorax, which is protected by wadding. In asphyxiated children with fracture of the clavicle the use of Schultze's swinging method should be avoided, as by its employment injuries to the lungs have been met with, caused by the sharp ends of the broken bones. Still, such injuries (according to Schultze, Runge, and others) may be avoided by carefully carrying out the manœuvre.

LITERATURE

Tillmanns, *loc. cit.*, p. 34; Küstner, *Müller, loc. cit.*, p. 15; Birnbaum, *loc. cit.*, p. 101; Stumpf, v. *Winckel, loc. cit.*, p. 44; Wyder, v. *Winckel, loc. cit.*, p. 44; Spiegelberg and Wiener, *loc. cit.*, p. 15; Runge, *loc. cit.*, p. 128; Kirmisson, '*Schulterhochstand*,' *Geb. Gesellsch. Paris* (June 8, 1903), *ref. Zentralbl. f. Gynäk.* (1903), p. 1309; Mercier, '*Schulterhochstand*,' *Geb. Gesellsch. Paris* (March 17, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 1145; Kayser, '*Schulterhochstand*,' *Deutsche Zeitschr. f. Chir.*, vol. lxviii.; Haroutioun, '*Schulterhochstand*,' *Diss. Nancy* (1904); Serrès, '*Schulterhochstand*,' *Diss. Paris* (1905); Sprengel, *Archiv. f. Orth. Chir.* (1891), vol. xii.

The Upper Extremities

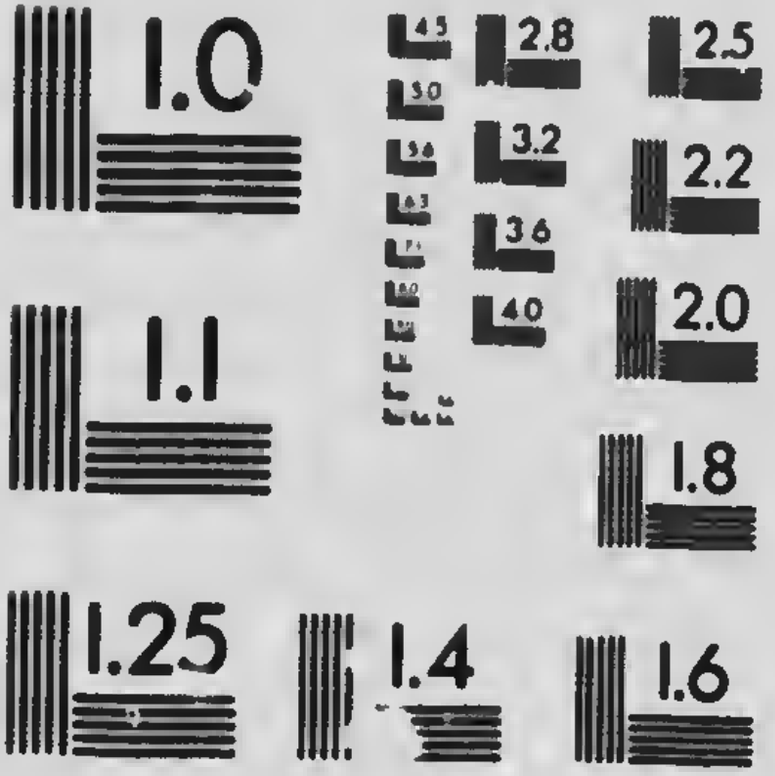
I will consider here briefly the question of so-called intra-uterine fractures of the bones. (On Intra-uterine amputations, see p. 9).

Children are born not infrequently with so-called intra-uterine

[1] He observed this accident on twenty-two occasions among 1600 children born in a cephalic presentation at the Copenhagen Clinic. In spontaneous deliveries, it appears to be caused by pressure of the anterior bone against the symphysis pubis, when marked traction is made on the posterior axilla.

MICROCOPY RESOLUTION TEST CHART

(ANSI and ISO TEST CHART No. 2)



APPLIED IMAGE Inc

1653 East Main Street
Rochester, New York 14609 USA
(716) 482 - 0300 - Phone
(716) 288 - 5989 - Fax

fractures. In such cases, bendings of the bones are found, chiefly of the long bones, especially of the tibia, which were formerly regarded (and are even at the present time, by a number of authors) as the result of badly healed fractures of the bones occurring *in utero*. In favour of this view appeared to be the fact that at the same time there was apparently some formation of callus, and scars were found on parts of the skin. It was assumed by Gurlt and others that these scars were due to traumata, which had affected the mother's abdomen during pregnancy, while the intervening soft parts had generally remained uninjured, just as in the adult severe internal injuries may occur as the result of contusions without externally any marked signs of the injury being recognisable. If there was nothing in the history to suggest the possibility of an accident, fall, or blow, then it was suggested that the uterine contractions during pregnancy could be invoked as an ætiological factor. (On intra-uterine fractures and intra-partum fractures as a result of osteogenesis imperfecta, see p. 247.) In more recent times, authorities, fortified by the microscopical examination (for which see Keller), are rather inclined to regard these bendings as the result of failures of development; and the more so as, very frequently, other similar defects—for example, of the fibula—may be present. Microscopical examination shows that nothing can be found which proves an old or recent fracture of the bone. The callus formation described is rather to be regarded as an attempt at the correction of the angular deformity of the bone. The microscopic examination of the skin further shows no signs of any formation of scar tissue, while the Röntgen rays give a negative result. As Keller remarks, the angular bending of the bones only occurs at a time when ossification has not yet begun, or at a time when the differentiation of the extremities of the bones is not yet completed. According to more recent conceptions, want of room *in utero* (the result of a deficiency of the liquor amnii), the presence of myomata, or of amniotic bands and folds must be taken into account as ætiological factors. The changes in the skin so frequently observed, the callus formation from periostitis, and the imperfect development of the portion of the extremity placed on the peripheral side of the bend, are all evidences in favour of its production by amniotic bands.[1]

In the upper extremity, the main interest lies in the obstetrical injuries, especially fractures of the humerus and separation of its upper epiphysis. Both these injuries occur practically only as the result of artificial delivery. According to Küstner, they are met with in almost equal frequency; but other statistics are in favour rather of the more frequent

[1] So-called intra-uterine fractures are, then, to be regarded as of two kinds: true fractures, the result of some excessive fragility of the bones—such as osteogenesis imperfecta—and occurring most commonly during delivery, but sometimes *in utero*; and pseudo-fractures, due to errors of development.

occurrence of fracture of the humerus. This fracture has also been observed occurring spontaneously. In this case, the condition is probably one of abnormal fragility of the bone during delivery, the result of osteogenesis imperfecta. In the majority of cases, the fracture occurs as the result of unskilful, improper, or very difficult liberation of the arms, especially when they are situated high up; or, further, when during extraction they are drawn too far backwards. The fracture is less frequently observed in drawing the arm down past the head to lessen the circumference of the shoulder. Cases also occur where the arm, wedged between the head and the pelvis, must be purposely broken, to deliver a living child. The mechanism of production of this fracture of the upper arm is, nearly always, that the obstetrician exerts the pull or pressure not on the bend of the elbow, but on the middle of the upper arm. Very often a definite crack is heard at the moment of the fracture. The most marked symptom of a complete fracture of the humerus is loss of function. The affected arm hangs motionless and limp, although the fingers can generally be moved. On examination the well-known signs of the fracture of a bone are found, abnormal mobility, crepitation, and pain. In doubtful cases, the aid of the Röntgen rays should be invoked. The prognosis is good even when, as occasionally happens, the fracture does not unite in a straight line, but at an angle opening outwards. In rare cases, too, paralysis of the radial nerve has been observed from compression by the callus thrown out. The treatment is very simple. The arm, bent at a right angle at the elbow, is bandaged to the thorax, the skin of which has been previously greased and covered with wadding. The hand and fingers must be left outside the bandage, so that they may not grow stiff (see Birnbaum).

Separation of the epiphysis at the upper end of the humerus was formerly often overlooked and regarded as paralysis of the nerves, or dislocation or fracture of the neck of the scapula. Küstner has pointed out the true nature of this injury. The epiphysis is rotated outwards to the maximum by the action of the external rotator muscle attached to it, while the diaphysis through the action of the latissimus dorsi and teres minor muscles is rotated markedly inwards.[1] If a correct diagnosis is not made and the proper treatment undertaken, healing occurs in this faulty position, and the arm as a result can be rotated neither outwards nor inwards. The injury almost always occurs when the pressure on the humerus, in freeing it, is exerted in the neighbourhood of the shoulder-joint; or when the freed arm is wrongly rotated, for example backwards. Treatment consists, after the replacement of the parts involved in the fracture, in the application of the necessary bandage (see Küstner, Stumpf, and Birnbaum).

[1] The shaft is usually displaced forwards and inwards. If no displacement is present, the diagnosis may be very difficult.

Another injury is the intentional amputation of a prolapsed arm (Brachiotomy), which at the present day should be avoided as a general rule (see a medico-legal case recorded by Stumpf).

Tearing of an arm, by rough traction on it when prolapsed, especially in transverse presentations, has been described. In such a case severe injuries may occur to the shoulder-joint muscles. Such injuries have also been met with in difficult version and freeing of the arms, i.e. tearing of the flexor carpi ulnaris from its attachment.

If the arm prolapses by the side of the head and becomes wedged in the pelvis, especially between the head and the promontory, signs of pressure and even gangrene of the arm may occur. Injuries to the arm may also occur when it is included in the grasp of the forceps during extraction.

Besides these injuries, paralyses of the upper extremity occur, the result of injuries to individual nerves or nerve plexuses. Paralyses of central origin also have been described from injuries of certain parts of the brain, hæmorrhages on to the surface of the brain, and injuries to the spinal cord. Paralysis may occur, further, from the affected nerve being damaged by the broken bones in a fracture (see Birnbaum). The most frequent and best known is the combined paralysis of the arm muscles first described by Duchenne and Erb (deltoid, biceps, brachialis anticus, and supinator longus); on the cause and the symptoms I will not enter here, but must refer the reader to the works of Finkelstein, Küstner, and Birnbaum.

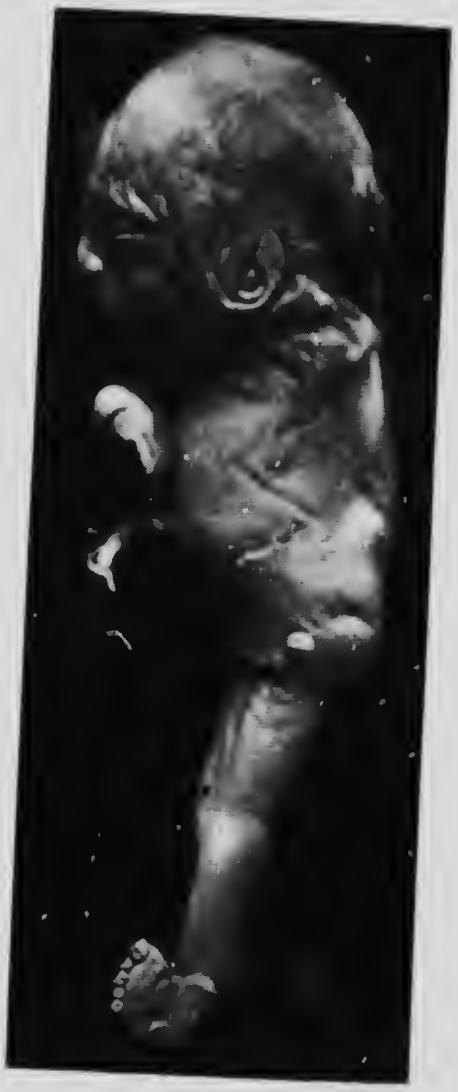
I may, however, point out that the prognosis of this form of paralysis is very good.[1]

In very rare cases, separation of the epiphysis at the lower end of the humerus has been observed. Other very rare injuries in this neighbourhood are tearing of the capsule of the elbow-joint, and dislocation of the head of the radius, generally forwards. Lastly it may be mentioned here that dislocations of the hand may follow obstetrical manipulations (see Stumpf).

Congenital dislocations of the elbow-joint are very rare, and are really to be regarded as malformations. Congenital dislocation

[1] This form of paralysis is a very important one from the obstetric point of view. Besides the muscles mentioned above, the supraspinatus, infraspinatus, teres minor and supinator brevis are all paralysed, so that the arm cannot be abducted or rotated out, and the forearm cannot be flexed or supinated. The limb remains in a position of adduction and rotation in with the forearm pronated and extended. The seat of the lesion is the fifth and sixth cervical nerve roots, or the cord formed by their junction. Such a brachial birth paralysis may occur, in either a vertex or breech presentation, when traction is exerted and the head is pulled away from the shoulders. Mild cases usually recover spontaneously and promptly; more severe cases may recover up to a certain point, but some paralysis persists if laceration of a nerve root has taken place. The only treatment for cases of permanent paralysis is the excision of the scar tissue, and the suture of the freshened nerve ends. Kennedy and A. S. Taylor have each reported seven cases with successful results.

PLATE IV.



A FULL-TERM FŒTUS, MEASURING ONE FOOT IN LENGTH, IN WHICH ONLY THE RIGHT LIMBS ARE DEVELOPED.

The lower limb is in a position of equino-varus, and there is a club hand with absence of the radius.

(Specimen No. 827, University College Hospital Medical School Museum, Obstet. Sect.)

of the radius backwards, behind the external condyle of the humerus, has been described on several occasions (see Tillmanns). Ronnenberg has collected a large number of various dislocations[1] of the elbow of a congenital nature. They may occur outwards, backwards, and forwards. (For the symptoms and treatment, the reader should consult text-books on surgery).

True fissures, duplicature of whole extremities, have not been observed with certainty. The conjoined extremities of double monsters naturally are of quite a different nature.

Malformations of the Forearm and Carpus

By abrachius is meant the absence of the upper extremities with well-formed lower extremities; by monobrachius is meant the absence of one upper extremity. This is either a true malformation or a so-called spontaneous amputation from an amniotic constriction or the pressure of an encircling cord. The forearm may be absent partially or totally. Abnormal shortness of the forearm with a normally developed hand (Peromelus, phocomelus) has been observed in chondrodystrophia (see p. 244). Abnormal shortness may result from an angular bend in the forearm: for example, in osteogenesis imperfecta. According to Tillmanns, primary obliteration of the blood-vessels and anomalies in the central nervous system play a rôle in the causation of these cases. Lastly, the bones of the forearm—most frequently the radius, less commonly the ulna—may be totally or partially absent. If the radius is absent, a so-called 'club hand' usually occurs, in which a defect of the thumb is frequently seen; while, in absence of the ulna, some of the fingers of the ulnar border of the hand are usually wanting.

Dislocation of the radio-carpal joint is very rare. The so-called 'club hand'[2] (Talipomanus), like the club foot—to be described presently—may result from want of liquor amnii and the pressure of the uterine wall. Very often the children are otherwise malformed (see Hohl).

Malformations of the Hand and Fingers

By syndactyly is understood the partial or total union of two neighbouring or of many fingers—a condition which at times is seen in all the members of the same family.[3] Generally, a finger with three phalanges

[1] It is very important in cases of difficult delivery of the child to carefully examine its whole body, so as not to overlook any of these various injuries.

[2] The name 'club hand' is applied to a congenital deviation of the hand from the axis of the forearm occurring at the wrist; with defect of the radius, the hand most commonly is deflected to the radial side.

[3] This condition represents the persistence of an embryonic state, since at the seventh week the fingers first appear as thickenings in the webbed hand.

is affected—most commonly the third and fourth fingers. The anomalies are to be considered as defects of development, in that the epithelial ingrowth which brings about the separation of the fingers is more or less absent. As a result of this malformation, adduction and abduction of fingers is prevented, and their bending is rendered more difficult.[1] Treatment is very simple, as the otherwise well-developed fingers are united only by a bridge of skin; but the formation of flaps is often necessary. (For details, see the text-books on surgery.)

In the split hand[2]—the opposite condition to syndactyly—only the outer fingers have developed. In this condition, malformations and defects in the region of the carpal and metacarpal bones are present (Perochirus). In rare cases also a double nail is observed on one finger. Achirus is complete absence of the hand, which occurs primarily from amniotic constrictions.

Macrodactylia (giant growth of the finger) is accompanied either by a proportionate increase in size of the bones and soft parts, or the condition is one of lipomatous or cavernous tumours of the soft parts. For this the appropriate treatment for such anomalies must be carried out. To macrodactylia belongs also the condition of increase in length of a finger from the presence of a supernumerary phalanx.

Brachydactylia indicates the shortening of a finger from the absence of a phalanx; and cetrodactylia is the absence of a whole finger.

Perodactylia is the shortening of a finger which primarily occurs from amniotic constrictions, so that the whole finger or a part of it may be cut off (spontaneous amputation, spontaneous dactylolysis).[3] Most commonly, stunting occurs in the finger—both in the distal portion and also in the part on the proximal side of the constricting band.

Polydactylia is the most frequent malformation in the hand. The condition is that of so-called supernumerary fingers, which may be attached to any part of the hand, but mainly on the ulnar or radial borders. Very frequently this malformation is handed down in some families through many generations. The cause lies in a duplication of the embryonic rudiment. Ahlfeld believes that disease of the amnion is also a very important ætiological factor. Tillmanns pictures in a very clear manner the various grades of polydactylia. The supernumerary fingers when they are very small may present the appearance of small tumours (Fibromata). In the majority of cases they consist of a finger-like outgrowth of skin: less commonly, there is a bony outgrowth like a phalanx. A well-formed supernumerary finger may be present.

[1] While this is true for the majority of cases, it must be remembered that sometimes the bond of union is a firm structure of skin and fascia; while in a few cases the union may be osseous, chiefly affecting the terminal phalanges, but even at times causing a fusion of all the bones of the two fingers.

[2] The middle finger, or the middle toe, is usually absent in this deformity.

[3] This term is often used for the disease (Ainhum).

inserted into a supernumerary metacarpal bone. The supernumerary finger may also be inserted into the head of the corresponding metacarpal bone of the ulnar or radial border of the hand. When the bones are double, in some cases the muscles, tendons, nerves, and vessels, appear to be also double, and the finger may be to some extent useful. (Ahlfeld). The least-developed form of polydactyly is the presence of two nails on one ungual phalanx. When such a supernumerary finger incommodes its bearer it should be removed. In a finger composed of skin, the connecting bridge is simply divided; while, in a well-formed finger disarticulation should be practised.[1] Even when the finger does not cause any inconvenience, at the wish of the mother it should be removed, as such children are subject to certain drawbacks during life. If the wish of the mother is not complied with, she will be discontented, and will find some other doctor who will consent to perform the operation. A short time ago, on the outer side of the right little finger of a new-born female child, I saw a tumour about the size of a cherry, attached by a very thin and fine stalk. After its removal, microscopical examination showed that it was a supernumerary finger, with well-marked œdema and increase of the embryonic connective tissue.

Clinodactylia is the abnormal displacement of the phalanges from their normal position into a dorsal, volar, or lateral, position.

Congenital contractures – primarily, flexions of the fingers at the phalangeal articulations – have been described.

Lastly, I will mention here a few well-known anomalies which have already been in part described:—

Amelus: that is, complete absence of the extremities, and in their place only warty or stump-like rudiments.

Peromelus: all the extremities are stunted.

Phocomelus (see p. 261): from *φώκη* and *μέλος*. The extremities are represented only by the hands and feet, which are attached immediately to the shoulders and pelvis.

Micromelus: abnormally small but well-developed extremities.

Congenital Malformations of the Hip-joints and Lower Extremities

Luxatio coxæ congenita (congenital dislocation of the hip-joint) is the most frequent of the congenital dislocations. As a rule, it is a dislocation of the head of the femur backwards (Luxatio iliaca); less commonly,

[1] Taruffi gives a very full description of this anomaly and quotes several series of cases; in the most marked of which there were as many as ten fingers on one hand. As Ballantyne points out, the ætiology of this condition is difficult to explain. It seems hardly possible that the excess of formation can be due to the action of amniotic bands, and he is inclined to regard it as 'a tendency to excessive formation existing in the germ—a sort of ill-regulated or uncontrolled gemmation of the distal extremities of the limbs.'

forwards (Luxatio pubica). Frequently, it is bilateral.[1] It is more frequent in girls than in boys (according to Tillmanns), because in the former the iliac fossæ are more perpendicular. According to Roser this dislocation is less frequently observed in boys, because by the presence of the external genital organs they are preserved from the marked adduction of the thighs which tends to bring about the dislocation, and which in girls there is nothing to prevent. In unilateral dislocation, lameness is present; in a bilateral dislocation, a waddling duck-like gait. The diseased condition is frequently first recognised when the children begin to run, or have even been running about for some time. On examination of the child standing up, the region above the acetabulum is filled up, and the trochanter lies above the Roser-Nelaton line.[2] In unilateral dislocation there is naturally shortening of the leg on the affected side. The other changes which are observed in older children (marked lordosis of the lumbar vertebræ, increase in the pelvic inclination, and a pendulous abdomen) occur at a later age, as the result of walking.

As to the ætiology of congenital dislocation of the hip, various hypotheses have been put forward. The condition is generally thought to be due to a defect of development, so that the acetabula, placed in their normal position, remain small, either as a result of premature ossification or of imperfect growth,[3] and the heads of the femora can no longer lodge in them. In the depth of the acetabulum there is a good deal of fat, the ligamentum teres is very long or may be entirely absent, and the capsule is long and pouch-like. The escape of the head of the bone from the acetabulum is favoured by the physiological position of flexion and adduction of the femur *in utero*, especially when the uterus is abnormally small, or there is too little liquor amnii present (Oligohydramnios). Inflammatory changes in the hip joint during fœtal life may further be considered as an ætiological or, at least, as a predisposing cause (effusions into the joint, with secondary distension and relaxation of the capsule).

[1] Krönlein, during a certain period at the Berlin Clinic, observed ninety cases of congenital dislocation of the hip, five cases of dislocation of the humerus, two of the radius, and one of the knee. In 1362 cases tabulated by Hoffa, there were 173 males and 1189 females; of these cases, 502 were double.

[2] That is, the line passing from the anterior superior iliac spine to the tuberosity of the ischium.

[3] In the third month, the margin of the articular surface for the hip grows out to form the acetabular cavity. When this outgrowth is arrested, congenital dislocation of the femur results. According to Keith, in this case human development is arrested at a reptilian stage. In unilateral cases, such a deficiency of the rim of the acetabulum may be demonstrated by the X-rays. The exact time at which the dislocation occurs is not known; it is possible that in some cases it takes place during delivery, as the result of traction on the thigh. In all probability the earlier the displacement occurs, the less likely is a good result to be obtained by treatment.

According to Tillmanns, congenital malformation of the head and neck of the femur plays a part in the development of congenital dislocation of the hip, as the head of the femur in such cases overgrows the acetabulum. In doubtful cases, an X-ray photograph should not be omitted. (For further details on these malformations, especially for their treatment, see text-books on surgery.)

Congenital coxa vara is a very rare congenital deformity of the neck of the femur in which the angle between the neck of the bone and its shaft is diminished (Kaufmann). The opposite condition (Coxa valga), in which the angle is increased, has also been observed congenitally.

I pass now to a consideration of obstetrical injuries to the thigh. In extraction in breech presentations, bruising of the soft parts is not uncommon, and the tissues are especially likely to be injured when the blunt hook or fillet is employed. The necrosed tissues separate in the first few days of life, and in such circumstances an extensive formation of scar tissue may result.

The occurrence of traumatic dislocation of the hip during delivery is not generally admitted: not, for example, by Kustner, who relies upon his experiments on the dead body. Some undoubted cases have, however, been recorded.

Fracture of the thigh is a relatively frequent accident. It occurs almost always during delivery as a result of some obstetrical operative procedure: not uncommonly during version and the bringing down of a foot, with the breech already fixed in the pelvis; more frequently in pelvic presentations, when the finger or an instrument (breech-hook or fillet)[1] slips from the groin on to the thigh, and the force is applied at this point. This fracture is also observed when the spontaneous descent of the limbs in a pelvic presentation is not waited for, but they are brought down by unskilful traction on the thigh. The bone nearly always breaks transversely in its upper third, at the place where it becomes suddenly markedly thinner.

In performing version, a fracture may occur if the limbs become crossed during their rotation, or if the operator applies his manipulations to the thigh instead of to the foot. The diagnosis of a complete fracture is easy, and can be made by the loss of function, the abnormal passive mobility, crepitation, and, lastly—in doubtful cases—by the Röntgen rays. (For treatment, see p. 266.)

Separation of the epiphysis of the femur is very rare. Like separation of the epiphysis of the tibia, it can only occur when, during extraction,

[1] This is most likely to occur when two fingers are used instead of one. The danger of fracture of the bone with the breech-hook is so great that many obstetricians only employ this instrument when the child is dead.

in place of a force applied in the long axis of the limb, rotatory movements or movements of over-extension are employed. In this way, the limbs may be rotated to such a degree that the toes point backwards (Kustner).

The best treatment of both these conditions consists in permanent extension after reposition, with the lower extremity brought up into a vertical position. Complete union, which usually occurs with marked formation of callus, takes generally about three weeks. Bad results are practically excluded by this mode of treatment; and the argument formerly brought against it, that the children cannot be fed naturally, is no longer true. By the use of light, easily transportable beds, the children may be brought to the mother, although it cannot be denied that natural feeding can be better and more easily carried out by the method to be described. In this procedure, according to the recommendation of Crede, the broken thigh is drawn up as high as possible on to the body and fixed here, either with a bandage or with a strip of adhesive plaster put on at the level of the popliteal space. According to some authors, by this means the displacement is not sufficiently reduced longitudinally, and the limb remains shortened. Other authors are very pleased with this certainly very simple method. I have personally observed, and also followed up for some time, three cases treated by Crede's method. In one there was a slight amount of shortening present, in the other two the fractures had healed faultlessly. In all cases, however, the treatment by extension is the more certain. Schultze's swinging method should not, of course, be employed when such an injury is present in a child partially asphyxiated.

I will consider too, here, obstetrical injuries in the remaining portions of the lower extremities: Separation of the epiphysis, at the upper or lower end of the tibia, has been observed as a rare injury; fractures of the tibia, from bending of the bone in a direction from behind forwards with the knee fixed; dislocations of the knee joint, from rotation of the leg; paraplegia of both lower extremities, as a result of crushing or tearing of the spinal cord, as after intrameningeal haemorrhage into the lumbar region of the cord—the latter occurs as the result of severe forcible rotation of the spinal column in difficult extractions. Paralyses, however, affecting the lower extremities are very rare; they occur, for example, as a result of injuries to the brain, as well as in spina bifida. The cerebral palsies are characterised as unilateral paralyses, in which, usually at the same time, the face and upper extremities are affected. The cause lies generally in abnormally difficult forceps extractions, especially in contracted pelves. The paralyses of spinal origin, as already mentioned in distinction to these, are bilateral in character.

Congenital defects of the femur are very rare. In a few cases, absence

of the whole femur has been described. Thus Ziegler records a case of absence of the femur and of the fibula.

Congenital dislocations of the knee-joint (more properly called dislocations of the tibia) are rare.[1] Most commonly they consist of a displacement of the tibia forwards; less commonly, outwards. The treatment (see text-books on surgery) consists in the reduction of the dislocation under an anæsthetic and the application of a plaster-of-Paris or extension bandage. In such cases, the patella may also be more or less imperfectly developed, so that the mobility of the knee-joint is markedly affected. Contractions of the knee-joint may also occur.

Rare, too, are congenital dislocations of the patella, of which two varieties may be distinguished—the permanent and the temporary. The patella is most commonly dislocated outwards;[2] if the condition is one of complete dislocation of the patella, a very marked disturbance in the functions of the knee-joint results.

In this place, I must mention also the very rare congenital contractions of the knee-joint, with which other malformations are commonly present. Flexion, extension, abduction (genu valgum), and adduction (genu varum) have been observed. The cause is to be found in an elongation of the muscles, or in a malformation of the joint, or in a deficient or excessive quantity of the liquor amnii. In other cases, especially associated with a deficiency of the liquor amnii, the contraction is the result of the formation of a wing-like membrane in the neighbourhood of the knee-joint.

Such wing-like membranes are seen occasionally also between the thorax and the upper arm. In such cases surgical interference gives good results.

Congenital curvatures of the tibia are observed, no doubt, more frequently than in any other bone, the curvature being generally forwards. Formerly (see p. 258), these curves were regarded as the results of badly healed fractures occurring *in utero*. At the present day these anomalies are referred, in the great majority of cases, to failures of development. Not infrequently, defects of the fibula and a valgus position of the foot are found together with these curvatures of the tibia. Less commonly, a diminished amount of room *in utero*, a deficiency of the liquor amnii, the presence of myomata, amniotic bands and folds, are concerned in the causation of these conditions which may be combined with true defects of

[1] Although rare, this is one of the more common of the congenital dislocations. Two forms may be recognised: the first, a congenital genu recurvatum, with hyperextension of the knee on the thigh; the second, a true dislocation.

[2] Of fifty-five cases collected by Potel, in fifty the dislocation was outwards, in four upwards, and in one inwards.

development. True healed or unhealed fractures, or green-stick fractures of bones are found only in cases of congenital fragility of the bones—such as occurs in osteogenesis imperfecta; in which case certain definite causes (see p. 247) may bring about a fracture.

These complete or incomplete fractures very often occur during delivery. The tibia and fibula[1] may also be absent. This condition in part is due to a defect of development; in part, to amniotic constrictions. In the first variety, deformities of the knee or ankle joint are often present at the same time. Tillmanns describes such a case of congenital defect of the whole tibia.

As regards congenital malformations of the foot, see p. 261 on the similar congenital malformations of the hand. In the foot micropus, apus, peropus, monopus, perodactylia, spaltfuss (split-foot), polydactylia, macrodactylia, syndactylia are observed as in the hand.

A malformation of the lower extremity, the so-called Siren formation—Symelia, sympus, monopus—is one of much interest to the obstetrician. The lower extremities are here united into a single fish-like structure (see Fig. 46 and Plate V).[2] In this condition, the limbs are rotated on their axes so that their external surfaces are united and the popliteal spaces look inwards. The bony pelvis may be absent, or very imperfectly developed: for example, it may be split. In the same way, the genitalia—frequently also the bladder, urethra, anus, and rectum—are almost always absent. Kuliga has recorded recently a complete autopsy on a case of siren formation. The anal and urogenital openings were absent, also the raphe of the perineum. In place of the external genitalia there was only a curved projection covered with skin. The internal female genital organs were rudimentary, but doubled. They were attached laterally to a large sac, recognised as the cloaca,[3] into which the large intestine and the left ureter also opened. The left kidney was undeveloped; the corresponding ureter and the pelvis of the kidney dilated. The right kidney and ureter were absent, also the rectum and the vermiform appendix. The jejunum, the ileum, the ascending colon, and the right portion of the transverse colon had a common mesentery. Besides this, there were a great many anomalies of the blood-vessels. In the skeleton, a slight

[1] Absence of the fibula is more common than that of the tibia, and occurs more often in boys than in girls.

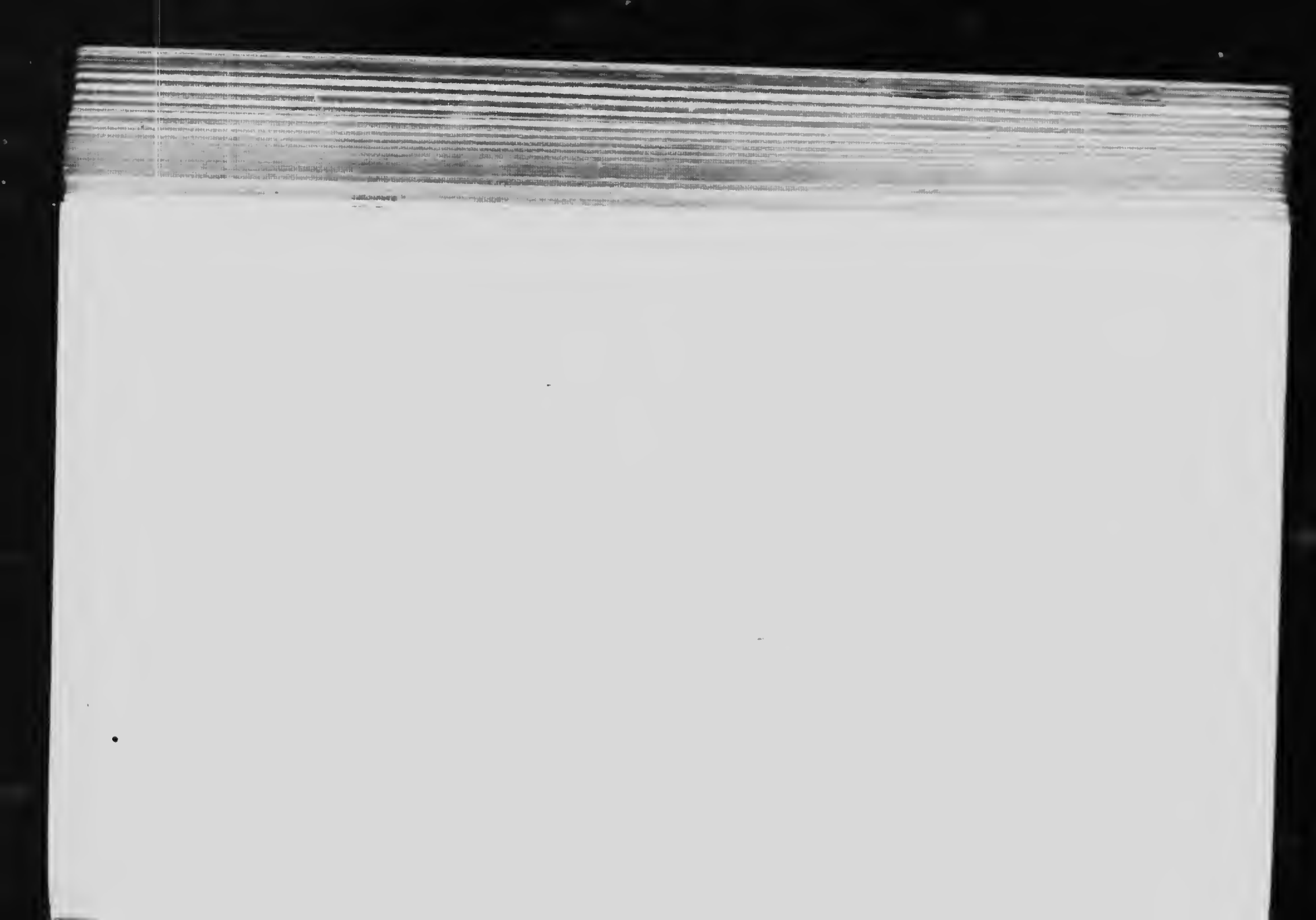
[2] Ballantyne points out that the term 'Symelia' refers merely to the fusion of limbs, which may be upper or lower; and that the fusion of upper limbs is only met with in cases of double terata. He therefore suggests the term 'Sympodia' as more accurate.

[3] There is no mention of the bladder in this case—and with a persistent cloaca it should be absent; this defect is particularly common in cases of sympodia: thus of fifty-two cases collected by Ballantyne, in forty, the bladder was declared to be wanting. The case illustrates very well the typical deformities which are usually present in this type of monster.

PLATE V.

SYMELIA.

(Specimen from the collection of the Göttingen Women's Clinic.)



degree of kyphoscoliosis, irregularities in the form and number of the lumbar and sacral vertebræ, and defects of the coccygeal vertebræ were present. The ilia showed abnormalities of their form, position, and union with the vertebral column. The ischia and the descending rami of the pubes were in contact and united by connective tissue. Kuliga believed that the period of development of the malformation must have corresponded to the third week of embryonic life. Etiologically, he attributed it to noxious influences working in a mechanical manner.

At the end of the merged lower extremities the legs may be entirely absent (Sympus apus), or some of the toes may be developed, or one or both feet may be present (Sympus monopus, or sympus dipus). In all monopods, according to Levy, only one umbilical artery is found (see Hohl).[1]

Delivery occurs in the siren formations, usually without difficulty, in a cephalic presentation. In other cases, version by the united legs and the extraction of a large body may be required.

Cichorius has recorded an interesting case of siren formation—the child living for one week after birth in the Leipzig Clinic. At the autopsy, completely normal conditions were found in the thorax, but the conditions in the abdomen were markedly abnormal. The bowel ended in a blind egg-shaped sac. In the place of the kidneys, a horse-shoe-shaped kidney was present, situated deep down in the small pelvis. The ovaries and tubes were present, but the uterus was entirely rudimentary. The pelvis was markedly altered, the femora displaced downwards and inwards. According to the author, the cause lay in some trauma inflicted during pregnancy, and an abnormally small quantity of liquor amnii.

An original account by a midwife of the birth of a siren formation appeared a short time ago in the 'Allgemein Deutsche Hebammenzeitung,' No. 17 (1908). The midwife found a vertex presentation and prolapse of the cord, as a result of which the child was still-born. She gave the following description of the malformation: 'The thighs, legs, and feet were united to a considerable degree, so that the form resembled the tail end of a large fish. The bones of the limbs could be felt, but the sex could not be determined.[2] At the head all was correct;

[1] In the variety known as sympus dipus there are two more or less complete lower extremities fused together, and in this case two acetabular cavities are present. In other cases, the two femora are united, and articulate with a single acetabular cavity. Owing to the rotation of the lower limbs, the knee-joints flex anteriorly. The fibulæ are often fused into a single bone. In the great majority of these malformations, only one umbilical artery is present, and, as Ballantyne points out, the single vessel usually takes origin from the descending aorta and probably represents a persistent vitelline vessel.

[2] In the majority of cases, the deformity of the pelvic extremity is such that the sex can only be determined by an examination of the sexual glands.

from the back the child looked like a wasted fish. I was so terrified at its appearance that I had to sit down. When I showed it to the sister and the mother they both fainted—a pretty business, as I was alone with them. It was night, and the heat such as Europeans seldom experience; so that one perspired on the least exertion. I had the child photographed on the next day, but it turned out a bad result. The father of the child is a queer fellow and suffers from syphilis.'

Hohl describes the siren formation very fully, and also gives a résumé of the cases recorded in the literature. According to him, it is possible that in monopods the movements of the child during pregnancy may be less felt than usual by the mother. He describes four cases of the birth of monopods.

In three of the cases a malpresentation was present, and the children had to be turned and extracted. In one case of twins, the healthy child was a head presentation; the second child, a monopus, was a malpresentation. Of the malpresentations mentioned, one was a shoulder presentation; in the other cases, the united lower extremities were placed obliquely across the inlet of the pelvis. In the first case, version was performed; in the other two, the united feet were drawn down—a procedure which, on account of the stiffness in the knee and hip-joints, was attended with some difficulty. According to Hohl, it is this stiffness in the lower extremities which gives rise to the malpresentations, since the child cannot assume the normal position. Great difficulty in the delivery does not as a rule occur, as such fœtuses are not usually of large size.[1]

[1] The possible ætiology of this curious malformation is a problem of the greatest interest. If we consider the parts, besides the lower limbs, which are mainly maldeveloped, it is evident that they are chiefly the structures developed in the caudal extremity of the embryo—that is to say, the rectum, the external genitalia, the bladder and, in part, the ureters.

The process of differentiation by which these various structures should become separated from one another is largely wanting. Further than this, the development of the urachus and allantoic vessels, or the inferior hypogastric arteries, is interfered with. In order to understand how the malposition of the lower limbs is brought about, it is necessary to remember that the primitive limb-buds are derivatives from the body wall, and that they have at first a dorsal or extensor surface, a ventral or flexor surface, and two borders—cephalic and caudal. In the growth of the limbs, the legs become rotated inwards and the arms rotated outwards, so that the extensor surface of the leg is placed anteriorly, and that of the arm is placed posteriorly. To explain the occurrence of a sympodial monster, it is necessary to assume that some mechanical agency—such as maldevelopment of the caudal part of the amnion—acts upon the embryo in or before the third week (either from the ventral or the dorsal aspect) in such a manner as to interfere with, or entirely prevent, the development of most of the structures lying between the two lower limb buds. In consequence of this, the two limb buds grow out in a line with the long axis of the embryo, instead of at right angles to it, and, coming in contact with their caudal borders which should ultimately form their external surfaces, fuse together in this position to a greater or a lesser degree.

Congenital abnormal positions of the feet are frequently seen. Under this are included—Pes equinus (fixation of the foot in plantar flexion), Pes calcaneus (contraction in dorsal flexion), Pes varus[1] (contraction in supination), Pes valgus (flat foot—contraction in pronation). All these deformities—especially congenital club foot—are very frequently associated with other malformations. They are, primarily, defects of formation due to interference with development; or, more commonly to mechanical conditions acting upon the growth of the extremities—such as deficiency of room in the uterus, deficiency of liquor amnii (already mentioned by Hippocrates), hydramnios; not uncommonly a combination of these factors is present. Sometimes pressure-marks can be recognised at the time of birth. In some cases, a pes varus of one foot and a pes valgus of the other has been observed from want of room in the uterus, and resulting compression of the feet.[2] A slight degree of pes varus and of pes calcaneus is physiological in new-born children. All these malformations may be unilateral or bilateral. Pes varus may occur from a defect of the tibia, or from paralysis, as a result of a congenital defect in the central nervous system.[3] An hereditary transmission of this

Fig. 46.—Radiogram of a Sympus Dipus.

[1] Pes varus, or more commonly pes equinovarus, in its most frequent and typical form is congenital in origin; the other three forms—pes calcaneus, pes equinus, and pes valgus—are more often acquired than congenital.

[2] Two varieties of congenital club foot must be distinguished: the first consisting of cases associated with some malformation of the bones of the foot, and the second, and much larger class of cases, occurring without any such defect.

[3] Such as anterior poliomyelitis, spastic or cerebral paralysis, and progressive muscular atrophy. The acquired form may also be due to improperly treated fractures of the ankle-joint, or to disease of that joint.

defect, as well as of pes valgus, has been seen. The latter may also occur from defect of the fibula, from synostoses in the joints of the foot, and from cerebral paralyses.[1] (For further information on these anomalies of position, especially as regards their treatment, the reader should consult text-books on surgery.)

From an inspection of the obstetrical notes of the Göttingen Clinic, I find that club foot, more often unilateral than bilateral, has been frequently observed. In most cases some deficiency of the liquor amnii has been present at the same time.

Lastly, a very rare defect must be mentioned: namely—congenital dislocation of the ankle-joint, which has been met with occurring both internally and externally, as the result of defective formation of one or other malleolus. This condition, as Tillmanns points out, is not strictly a dislocation, but rather a true malformation, or congenital contracture of the ankle-joint.[2]

Finally, I may mention here, congenital defects of the muscles, which at one time were considered to be very rare. The literature on the subject published during the last few years, has shown however that congenital defects of the muscles, or groups of muscles, are not so very rare. The best known is defect of the pectoral muscle. In 1902, Bing was able to report on over 102 cases of this defect. Defects have been observed of the trapezius muscle, eighteen times; of the serratus magnus, fourteen times; of the quadratus femoris, sixteen times; omohyoid, eight times; semimembranosus, seven times; four times of the deltoid, latissimus dorsi, abdominal muscles, and gastrocnemius; three times of the sterno-cleido-mastoid, rhomboids, supra- and infra-spinatus, and biceps flexor cubiti; on two occasions of the platysma, the extensor carpi ulnaris, the small hand-muscles, and quadriceps femoris; on one occasion of the face muscles, the stylohyoid, intercostals, longissimus dorsi, supra- and infra-spinatus, levator scapulæ, subscapularis, triceps extensor cubiti, brachialis anticus, supinator longus, extensor minimi digiti, flexor sublimis digitorum, and glutei. Defects of the muscles of the eye have also been described. Besides the defects of the muscles mentioned, other less common combinations have also been described (see the monograph of Lorenz in 'Nothnagel's Handbook'). With defects of the muscles, defects of the bones also occur in some cases. Very curious wing- or web-like membranous folds of skin also frequently occur. The views as to the ætiology of these malformations are very divergent. Pressure on the child *in utero*, amniotic adhesions, trauma inflicted on the pregnant uterus, imperfect embryonic rudiments,

[1] The common acquired form is the so-called static variety, due to a disproportion between the body weight and the sustaining power of the muscles and ligaments.

[2] A further congenital defect in this region is Volkmann's ankle deformity, in which there is a congenital defect of the bones of the leg, with a normal foot but obliquity of the ankle-joint.

and arrest of development have all been cited. According to some authors, they occur as the result of a dystrophy of the muscles occurring *in utero*, and remaining stationary at an early stage of development. (For the clinical symptoms and more complete anatomical details, see Lorenz. On obstetrical injuries to the muscles—Hæmatoma of the sterno-mastoid, tearing of the arm muscles—see p. 260.)

In conclusion, I will consider briefly the obstetrical importance of malformations of the extremities. (On the obstetrical importance of curvatures of the spinal column, see p. 242; on siren formation, see p. 268.)

Bent, ankylosed, and dislocated extremities, their union with the trunk, and the union of different portions of the body with one another, may have an influence upon the course of delivery since they may lead to difficulty in the expulsion of the child, or to difficulty in making a diagnosis. Hohl discusses in the most complete detail these various anomalies and their influence on labour; while other authors are content with short general remarks or do not consider them at all. He gives, too, a very interesting series of illustrative cases. Stiffness in the joints, marked curvatures of the extremities, club feet, union of various portions of the body with one another, can only be recognised on internal examination when one remembers the possibility of the occurrence of such abnormalities. During birth, ankylosed and bent extremities may give rise to trouble when they stand out from the body of the child. In this case, fractures may occur spontaneously, or may follow the operative measures necessary for the completion of the difficult delivery. Thus Hohl records a case in which, associated with ascites, the ankylosed lower extremities were flexed at the knee-joints, so that their anterior surfaces formed a concavity, and the legs were placed close to the abdomen. The presentation was a breech, and it was impossible to bring down the feet. Version and extraction may be extremely difficult in such circumstances. Hohl records, further, a case in which all the joints of the extremities were ankylosed. After the head had been delivered with the forceps, the birth of the trunk was delayed, and in the necessary artificial extraction the arm and thigh bones were broken. Obstruction to delivery from ankylosis has also been described with hydramnios (see p. 6).

Rigor mortis (intra-uterine rigor mortis) of the child gives rise at times to difficulty in delivery. I will take this opportunity of discussing briefly the meaning of this interesting phenomenon. At one time it was considered as very rare—Schwarz, for instance, denying its existence altogether.[1] At the present day its occurrence is certain, and a considerable number of cases have already been recorded, many of which were accompanied by dystocia. It has been contended that

[1] Casper, in his well-known *Text-book of Forensic Medicine*, stated that he had never seen an instance.

intra-uterine rigor mortis can only occur in fœtuses after the seventh month. L. Seitz, however, from ten cases he observed, asserts that it can occur in very young fœtuses—apparently, as early as the time of the appearance of transverse striation in the muscles.[1] The actual meaning of this condition is not certain, and the stimulus which produces it is still unknown (Dohrn). It depends, as in extra-uterine cases, on the coagulation of the albumen of the muscles. The majority of authors (Seitz, Lange, and Wolff) consider that every fœtus, soon after its death, is subject to the occurrence of rigor mortis; but that, as in extra-uterine cases, after a varying length of time, it again disappears. Seitz attributes—probably correctly—the relative infrequency of cases of intra-uterine rigor mortis to the fact that the children are seldom born during the comparatively short interval of time, rarely more than a few hours, in which the rigor mortis is present; and, further, that slight degrees of fœtal rigor mortis are easily overlooked. The last fact appears to me to be of considerable importance. In twin pregnancies, rigor mortis of one child, with the second child alive, has been observed. According to Wolff, rigor mortis attains its height of development in three to four hours, and is already disappearing, some four to five hours after the death of the child. In cases of eclampsia of the mother, fœtal rigor mortis has been seen especially frequently. In rare cases it is possible still to feel a definite cardiac impulse in children born with rigor mortis. Thus Dohrn, in a case of eclampsia of the mother, performed classical Cæsarean section, and delivered a child with definite rigor mortis, which still showed an active heart-beat. This fact is to be attributed to the automatic independence of the heart. A similar observation may be made comparatively frequently in cases of marked asphyxia. The heart still beats while the voluntary muscles fail to respond to attempts at resuscitation.[2] (For further details on the Physiology of Fœtal Rigor Mortis *in utero*, see Seitz, in 'v. Winckel's Handbook of Midwifery.') In a case recorded by Ahlfeld ('Text-book of Midwifery,' p. 423), death-spots[3] even were present. Fœtal rigor mortis is favoured by a high temperature in the mother, anæmia, and, as already mentioned, eclampsia. Wolff explains the frequency of cases with eclampsia by the fact that, on the one hand, there is the possibility that the poison of eclampsia promotes the occurrence of the rigor mortis, and on the other hand, that eclampsia offers the opportunity of delivering a child from the body of a patient practically moribund. The suggestion that the condition is not one of true rigor mortis, but of a tetanic spasm of the muscles, must be rejected; as in intra-uterine rigor mortis the normal position of the child can be demonstrated.

[1] It may be found in fœtuses from 18 to 31 cm. in length.

[2] This phenomenon has often been observed in still-born children without rigor mortis; Neugebauer has collected ten such cases.

[3] That is, areas of hypostatic congestion.

Difficulties in the delivery of the child, as has already been mentioned, may result from such intra-uterine rigor mortis. In some cases, liberation of the arms is rendered difficult (Martin and Schultze), or the delivery of the shoulders; or difficulty has occurred in other obstetrical operations (K. Das, difficult forceps delivery; Caruso, the same; v. Oordt, the same; Ulrich, difficult delivery of the shoulders; Jones, a severe tear of the perineum as a result of rigor mortis). Such obstetrical difficulties occur from easily understood reasons, especially if the operation is carried out at a time when the rigor mortis is at its maximum.

In the Göttingen Obstetric Polyclinic, the following case of rigor mortis with dystocia came under observation (Jan. 11, 1895). In a primipara, in whom the waters had ruptured fourteen days previously, forceps were applied on account of weakness of the uterine pains, with arrest of the labour for twelve hours. The fœtal heart-sounds could not be recognised with certainty. On applying the forceps, the delivery of the head was very difficult, and, after it had been born, it was not possible to produce any farther advance of the child. Traction with the fingers, introduced into the axilla, failed. After the blunt hook had been applied to the axilla, the shoulders and arms were delivered, but the trunk remained behind. An attempt to pass the hand into the uterus, to elicit the cause of the obstruction (locked twins, some sac-like appendage, dropsy, tumours—were all suspected), was prevented by the contracted cervix, which lay closely applied to the abdomen of the child. With the exercise of great force—by traction from below and pressure from above—a dead child was at length delivered, and, as a cause of the dystocia, intra-uterine rigor mortis was found to be present. The extremities were in the normal fœtal position. The recognition of fœtal rigor mortis is also of importance from the forensic point of view, as is illustrated by the case recorded by Parkinson (quoted by Seitz). A woman was accused of the murder of her child. On the examination of the child, rigor mortis was discovered. The expert called, gave it as his opinion that the child had had an extra-uterine existence, as in still-born children rigor mortis was not seen.[1] The woman in spite of this was acquitted.

Literature

On Intra-uterine Fractures, Malformation and Injuries of the Extremities, Siren Formation, and Defects of the Muscles

Kaufmann, *loc. cit.*, p. 24; Ziegler, *loc. cit.*, p. 15; Leser, *loc. cit.*, p. 34; Tillmanns, *loc. cit.*, p. 34; Ahlfeld, *loc. cit.*, pp. 24 and 34; Birnbaum, *loc. cit.*,

[1] From what has been said already, it will be clear that this opinion was quite wrong, and the presence of rigor mortis is of course no proof at all that the child has had an extra-uterine existence. Ulrich's case, in which a primipara was delivered spontaneously of twins—the first child being born dead in a condition of rigor mortis and the second child alive—is an interesting proof of this.

p. 191; Stumpf, *v. Winckel, loc. cit.*, p. 44; Wyder, *v. Winckel, loc. cit.*, p. 1..., Keller, *loc. cit.*, p. 34; Küstner, *Müller, loc. cit.*, p. 15; Olshausen and Veit, *loc. cit.*, p. 44; Braun v. Fernwald, *Lehrb. d. gesamten Gynäk.*, Wien, Braumüller ('*Sirenenmissbildungen*'); Spiegelberg and Wiener, *loc. cit.*, p. 15; Runge, *loc. cit.*, pp. 24 and 129; Ronnenberg, *Zeitschr. f. orth. Chir.*, vol. ii.; Lorenz, *Nothnagel, loc. cit.*, p. 53; vol. xi., pt. 3, p. 713; Finkelstein *loc. cit.*, p. 101; Bernard, *Kongenitale Radiusluxation*, *Diss. Lille* (1907); Bonnaire, *Kongenitale Radialluxation*, *Soc. Gesellsch. Paris* (March 19, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 1152; Riss, '*Kongenitale Radialluxation*,' *Diss. Paris* (1902); Springer, '*Peromelie*,' *Prag. med. Wochenschr.* (1902), *No.* 51; Holzapfel, '*Perocheirie*,' *Beitr. z. Geburtsh. u. Gynäk.*, vol. viii., 1; Fränkel, '*Kongenitale Kniegelenkluxation*,' *Gynäk. Gesellsch. Breslau* (November 17, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 1025; Keller, '*Klumpfuss Gelenkkontrakturen*,' *Arch. f. Gynäk.*, vol. lxvii., pt. 2; Peiser, '*Ätiologie des Klumpfusses*,' *Diss. Breslau* (1902); Cicherius, '*Sirenenmissbildungen*,' *Geb. Gesellsch. Leipzig* (Feb. 15, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 590; Kuliga, '*Sirenenmissbildungen*,' *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxvii., pt. 4; Olshausen, '*Traumatische Luxatio humeri*,' *Geb. Ges. Berlin* (July 14, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 1484; Dubrac, '*Fibuladefekt*,' *Diss. Paris*, 1904; Bolk, '*Polydaktylie*,' *Nederl. Tijdschr. v. Geneesk.* (1904), vol. i., *No.* 8; Levy, '*Perobrachius*,' *ref. Zentralbl. f. Gynäk.* (1905), p. 534; Champétier de Ribes, '*Kongenitale Hüftgelenksluxation*,' *Geb. Gesellsch. Paris* (November 14, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 1066; Vinzent, '*Kongenitale Kniescheibenluxation*,' *Diss. Paris*, 1904; Bauereisen, '*Sirenen*,' *Frank-Ges. f. Geburtsh.* (January 28, 1905), *ref. Zentralbl. f. Gynäk.* (1905), p. 469; Lemp, '*Missbildungen der Finger und Zehen*,' *Diss. München* (1905); Heming, '*Wachstum der weiblichen Pyramen*,' *Geb. Ges. Leipzig* (May 21, 1906), *ref. Zentralbl. f. Gynäk.* (1906), pp. 1122 and 1160; Meygrier and Lemeland, '*Kong. Geu. reversum, Klumpfuss*,' *Geb. Gesellsch. Paris* (June 15, 1905), *ref. Zentralbl. f. Gynäk* (1906), p. 530; Hirsch, '*Ätiologie der kongenitalen Fussverkrummungen*,' *Diss. München* (1905); Serrès, '*Kongenitale hintere Schulterluxation*,' *Diss. Paris* (1905); Kayser, '*Kongenitale Schultermuskeldefekte*,' *Deutsche Zeitschr. f. Chir.*, vol. lxviii.; Haroumoun, '*Kongenitale Luxation des Schulterblattes*,' *Diss. Nancy* (1904); Larue, '*Kongenitale Kontrakturen*,' *Diss. Paris* (1906); Lovett, R., *Keen, loc. cit.*, p. 53; vol. ii. (1911); Woolsey G., *Keen, loc. cit.*, p. 53; vol. ii., p. 732 (1911); Clark, Taylor, and Prout, *Amer. Journ. Med. Sci.*, Oct. 1905; Kennedy, *Brit. Med. Journ.*, Feb. 7, 1903; Tarnill, *loc. cit.*, p. 44; vol. iii., pt. 1, p. 422 (1886); Hoffa, *Orthop. Surgery*, p. 239; Keith, A., *loc. cit.*, p. 93; Potel, *Étude sur les Mal. congén du Genou* (Lille, 1897); Volkmann, *Deutsch. Zeit. Chir.*, vol. xcviii., p. 2.

Literature

On Intra-uterine Rigor Mortis

Seitz, *v. Winckel, loc. cit.*, p. 15.; vol. ii., pt. 2, p. 1269; Spiegelberg and Wiener, *loc. cit.*, p. 15; Runge, *loc. cit.*, p. 24; Ollmansen and Veit, *loc. cit.*, p. 14; Ahlfeld, *loc. cit.*, p. 24; Ballantyne, *loc. cit.*, p. 44; K. Das, *Journ. Obst. and Gyn. of the Brit. Empire*, Dec. 1903; Caruso, *Arch. di ost. e gin.*, April 1905; Van Oordt, *Nederl. Tijdschr. v. Geneesk.* (1906), vol. i., p. 1; Müller, *Korrespondenzbl. d. allg. ärztl. Vereins v. Thüringen* (1905), No. 8; Ulrich, *Zentralbl. f. Gynäk.* (1907), p. 1097; Jones, *Journ. of Obst. and Gyn. of the Brit. Empire* (1906), vol. x., *Nos.* 4 and 5; Feis, *Arch. f. Gynäk.*, vol. xlvi.; Lange, *Zentralbl. f. Gynäk.* (1894), p. 1217; Martin, *Zeitschr. f. Geb. u. Gynäk.*, vol. i., p. 55; Schultze, *Deutsche Klinik* (1887), No. 11; Seitz, *Samml. klin. Vortr.* (*Volkmann*), No. 343; Wolff, *Arch. f. Gynäk.* vol. lxviii., p. 3; Dohrn, *Geb. Gesellsch. Dresden* (Oct. 12, 1907), *ref. Zentralbl. f. Gynäk.* (1908), p. 530; Schwarz, *Die vorzeitigen Atembewegungen* (Leipzig, 1858), p. 230; Parkinson, *Brit. Med. Journ.*, Feb. 8, 1908.

CHAPTER XV

Fœtal Gigantism

I will discuss now the general excessive growth of tissue which is not uncommonly seen during the period of embryonic development, and which is known as fœtal gigantism. For an account of the gigantism—no doubt also dependent upon congenital conditions, but which first makes its appearance after birth—see p. 248. The subject now under discussion is that of fœtal gigantism, or, as the name implies, abnormally large children.

It is usual to define a certain limit as regards the weight, and the general custom is to reckon all children as giant children whose weight is over 10 lb. Among midwives, the term 'giant child' is much abused, since the children are simply estimated by the midwives as of some given size, without their weight being actually determined with the scales. A mother, too, is often inclined, with pardonable pride, to assume an excessive weight for her children. An over-estimation of the weight is especially likely when the delivery, for various reasons, has lasted a considerable length of time (premature rupture of the membranes, weak pains, rigid soft parts, contracted pelvis). In spite of this, however, a considerable number of undoubtedly giant children have been put on record.

It is important to lay stress on the fact, as Küstner points out, that the weight, the length, and the circumference of the head in these giant children usually increase in a definite relation to one another.[1] The following is a short table of some recorded cases of authenticated giant children:—

A. Martin: 7470 grm. (16·4 lb.), without brain, blood, and vault of cranium. Quoted by Olshausen and Veit.

Ortega: 11,300 grm. (24·8 lb.); length, 70 cm. Quoted by Kleinhans. This is the normal size of a child one year old.

Thoyer-Rozat: 9900 grm. (21·7 lb.); length, 61 cm.

Schwab: 10,260 grm. (22·4 lb.); length, 57 cm. Macerated.

Fuchs: 6105 grm. (13·4 lb.); length, 60 cm., and another, 7550 grm. (16·6 lb.); length, 65 cm.

[1] Varnier, however, has shown that the diameters of the head do not increase in the same ratio as the weight of the child.

Schubert: 6550 grm. (14·4 lb.); length, 64 cm.
Maclean: 8000 grm. (17·6 lb.).
Lichtenstein: 7000 grm. (15·4 lb.).

(See further the literature in 'Zentralbl. f. Gynäk.'; the tables given by Ahlfeld, 'Midwifery,' pp. 416–417, and Kustner in 'Müller's Text-book of Midwifery,' p. 680.)[1]

The majority of the children recognised as giants weigh over 4000 grm. (13·2 lb.). As to the causes of this abnormally marked intra-uterine growth, heredity is the most frequent. Thus in some cases the habitual production of giant children has been observed. In many cases, too, the children have been carried an excessive length of time *in utero*—partus serotinus (post-mature children). Kaufmann is of opinion, however, that the idea that, in such cases of gigantism, the cause is a too long duration of pregnancy (300 to 320 days instead of 280) cannot be entirely accepted; for such children are frequently born at the normal full term[2] that is, on the two hundred and eightieth day of the pregnancy. The cause which in such cases brings about the excessive fœtal development is not always a simple one. No doubt, all those factors play a part, which are of importance in determining the weight and length of the new-born child—such as the age of the mother,[3] the sex of the child, the size of the parents,[4] the conditions of the mother's nutrition during her pregnancy, and the number of the pregnancies. Further, it may also be said, that the weight of the child increases with the age of the mother and the number of the pregnancies; that boys are, on the average, larger than girls (the majority of giant children are boys); that very badly nourished mothers frequently bear thin children, and that very large mothers often also have very large children.[5]

The presentation of these children is generally a vertex; but a brow presentation, rare otherwise, is not infrequently met with in such cases.[6]

[1] Waller records the case of a child delivered alive by forceps and weighing 8570 grm. (18·9 lb.).

[2] As Ballantyne clearly points out, the post-mature infant is larger and further developed than the new-born child; the subject of gigantism, however, is larger than he ought to be for the age at which he has arrived, and he may be developed in proportion to his weight, or only in proportion to his age.

In 14–15 per cent. of all children weighing over 8·75 lb., the pregnancy has lasted over 300 days.

[3] The heaviest children are born when the mothers are between the ages of 25 and 35; but beyond the age of 35, or after the ninth pregnancy, the weight of the children tends again to diminish.

[4] In a case recorded by Beach, the father was 7 feet 7 inches in height, and the mother 7 feet 9 inches; the child weighed 23·75 lb., and had a length of 30 inches.

[5] According to Ettinghaus, four times more frequently than in normal-sized children.

In the delivery of these children, the size of the head and the breadth of the shoulders, in certain conditions, may give rise to the greatest difficulty. In such cases of dystocia not only the circumference of the head is a difficulty, but its bad conformation must also be taken into consideration, as the bones are very hard, and the sutures and fontanelles are very narrow. As a result of this, the head during labour is often very badly adapted to the pelvis, and, further, it must be remembered that the shoulders and thorax are less compressible than under normal conditions of weight. It is clear, then, that operative procedures—especially the employment of forceps and perforation, and the methods to be described below—are very frequently necessary for the delivery of the shoulders. In the face of these facts, the weight and length of the child, from an obstetric point of view, are equally important.

In the higher degrees of fœtal gigantism we have, according to these considerations—even in a norm[illegible]ondition of the pelvis practically often the same conditions or, at [illegible]y rate, the same difficulty during labour, as occurs in a generally contracted pelvis; and this, as regards the treatment, is of the greatest importance.[1] It accounts for the fact that in such deliveries the same difficulties arise as are so often observed in contracted pelves; for example—secondary uterine inertia from fatigue, thinning of the lower uterine segment, e[illegible]rupture of the uterus and bruising of the mother's soft parts. [illegible]hild, too, asphyxia occurs, or injuries especially in the extract[illegible]ne, fore-coming head: depressions of the bones of the skull es[illegible]ll, of the parietal bones: pressure on the brain (see also Kleinhans). If the difficulty in the delivery of the child is due to the breadth of the shoulders, then it may occur at the time when they are entering the pelvic brim. More frequently, however, the difficulty arises at the time of the exit of the shoulders from the pelvis. In the first case the head, already well engaged in spite of strong pains, is not born; so that a transverse presentation, a funnel-shaped pelvis, coiling of the cord or too short a cord, and resulting hindrance to the birth of the child are all suspected. In other cases, the shoulders do not follow the delivery of the head; so that locked twins, sac-like appendages, spina bifida, teratomata, umbilical hernia, ascites, over-distension of the bladder are thought of as possible complications. In the second case, when the shoulders, after the delivery of the head, are delayed for a long time in the pelvis, the child may be in danger of death by suffocation from pressure on the thorax and the attempts at inspiration thereby induced, as well as from compression of the cord. In pelvic presentations great difficulty is experienced in freeing the arms

[1] Fortunately, any contraction of the pelvic measurements is uncommon: and, indeed, not infrequently the pelvis is rather above the average in size.

and head. In transverse presentations, the increased circumference of the shoulders may render version very difficult; while their enormous breadth may also produce very severe tears of the mother's perineum after the birth of the head.

The diagnosis of fœtal gigantism during pregnancy and labour is generally prevented by its great difficulty. The abdomen, although neither hydramnios nor twins are present, is distended beyond the normal, and the head is unduly large; while the narrowness of the sutures and fontanelles, and the hardness of the bones of the skull, is striking. At times attention is arrested by the length of the sagittal suture, or the wide separation of the two fontanelles. An important fact is that the head, in spite of good labour pains, remains for an unduly long time movable above the brim, although neither contraction of the pelvis, nor a hydrocephalus, nor any other obstruction (Myoma, ovarian tumour) can be recognised. In foot or transverse presentations, with prolapse of the arm, the circumference of the palpable or visible portion of the child should strike the watchful observer.

The prognosis for the mother and child is evident from the foregoing remarks. In treatment, the forceps and perforation are of most importance. In well-marked forms of gigantism, pubiotomy is indicated, in selected cases, to avoid the necessity of perforating the living child.

If there is present in any patient a predisposition to the production of giant children, as has been described in the literature on several occasions, Prochownik's diet,[1] which undoubtedly is of value in contracted pelves, should be recommended. Still better in such cases is the induction of premature labour.

During labour the delivery of the shoulders as a rule causes the greatest difficulty. Küstner points out that even the practitioner with some experience often shows a want of method in his operative procedures in these cases. If the shoulders are still above the pelvic brim and hinder the delivery of the head, then it is best to follow the advice of Spiegelberg, and to exert a steady pressure above the anterior pelvic wall in a downward and backward direction, and so to move the shoulders forward, while steady traction is kept up on the head with the forceps. Ahlfeld recommends that four fingers should be passed behind the anterior shoulder so as to rotate the shoulders into the largest transverse diameter of the pelvis. In all cases with very large children this recommendation is hardly possible, as the head fixed in the outlet bars the way for the hand to reach the shoulders, and the

[1] Prochownik's special diet for the last six weeks of pregnancy consists mainly in the elimination as far as possible, of fats and carbohydrates and cutting down the amount of fluid drunk. In order to avoid the thirst which might, and does result to a considerable degree, such articles are allowed as contain a large quantity of water—especially green vegetables.

lateral incisions of the vulva, often employed, do not afford enough room. In such cases the child is almost always lost. The delivery of the child may succeed with very marked tearing of the neck. If this is not possible, perforation must be carried out so as to afford room to reach the arms, and to bring down one or both, so as to diminish the circumference of the thorax, and enable the child to be delivered by traction. In other cases, where the shoulders remain fixed in the pelvic outlet, the same treatment should be carried out as is done when the birth of the shoulders is delayed under normal conditions. The woman is requested to bear down strongly. When this is insufficient the head is seized between the two hands—the face being left free—and drawn downwards and backwards so that the anterior shoulder slips down behind the symphysis pubis and engages under the pubic arch. The head is then drawn upwards, and the posterior shoulder is allowed cautiously to glide over the perineum. If this method leads to no result, then an endeavour must be made to effect the delivery of the child by the shoulders (insertion of the two index fingers from the dorsum of the child into the two axillæ. Any coiling of the cord that may be present, naturally, must be loosened. According to Spiegelberg, the following method may also be employed. When there is very little room the anterior shoulder is pushed upwards behind the pubis so that the neck comes into the pubic arch. It is then possible to find room to seize the posterior shoulder, and to draw this down to the anterior margin of the perineum, and then to bring down the anterior shoulder, either by pressing back the trunk against the perineum, or by the introduction of the fingers into the anterior axilla. In such cases an extensive episiotomy is very useful. At times, too, the blunt hook is a very considerable help in the extraction; it is best placed in the posterior axilla, but in certain circumstances it may be placed also in the anterior. Naturally, there is a risk—when the hook is not placed directly in the axilla but lies on the humerus—of fracture or separation of the epiphysis of the bone. Other authors recommend bringing down the posterior arm or both arms, so that the circumference of the shoulders is diminished and at the same time a hold is given for extraction. Spiegelberg objects to this 'loosening' of the arms in living children from the fear of injuries to the joints and bones. According to him, when the amount of room is sufficient for the manœuvre of 'loosening' the arms, the extraction may be carried out by the means already described. Of considerable importance in these attempts at extraction is marked bearing down on the part of the patient, and pressure from above exerted by another person (Kristeller). When all means of delivery fail, the child will be killed—gradually suffocated from the imperfect expansion of the lungs or pressure on the cord. In such circumstances the best method to overcome the dystocia is to carry out

unilateral or bilateral division of the clavicles[1] (Cleidotomy) (v. Herff, Phänomenoff). In this way the transverse measurement of the shoulder is very considerably diminished.

In some cases, certainly, even these methods fail, and then evisceration must be practised. Lichtenstein records a case where cleidotomy, after removal of the head, failed. The shoulders were then pushed back, and version and extraction carried out after evisceration.

In the Göttingen Women's Clinic, no giant children in the true sense of the word (over 5000 grm.) have been born. If the limit of weight is set somewhat lower—about 4500 grm.—then there were, among 4200 births, 40 children with a weight of over 4500 grm.; and of these, 30 were boys and 10 girls. In one case, perforation was necessary; four times the forceps—once on account of impending asphyxia of the child, and once on account of fever on the part of the mother; once version was necessary; and in one case freeing of the arms and head in a footling presentation. On two occasions atonic post-partum hæmorrhage occurred. In 38 of the cases the presentation was a cephalic one; once it was a footling, and once a transverse. In a large number of the cases, tears of the vagina and perineum were observed, and in two there was a complete tear into the rectum. In several of the cases, subinvolution was noticed during the puerperium.

Of partial gigantism (see also p. 248) indications may occur *in utero*. The extremities and the head are most frequently affected. In some cases the condition is one of unilateral gigantism.[2] In other cases some portions of the soft parts of the body or of the extremities, the trunk, and the face may be affected in such a way that marked changes occur which are termed elephantiasis. In this condition, the connective tissue, the fat, the blood-vessels, and the lymphatics all take part. (On lymphangioma, and the difficulties in delivery due to it, see p. 116.) Such formations when completely circumscribed must be considered as tumours, and, according to the portion of the tissue chiefly affected, called lymphangiomata, fibromata, or angiomata. Ballantyne pictures a case of congenital elephantiasis affecting the whole of the right lower extremity (see also Cusson, Guinon, Lindner, and others).

[1] Division of the clavicles was first definitely recommended by Spencer as a means of overcoming difficulties in dealing with certain cases of impaction of the trunk of the fœtus. In these cases he advised division of the clavicles to reduce the width of the child's shoulders.

[2] Unilateral gigantism, or hemi-hypertrophy, and partial gigantism may be distinguished. The reader must remember that a number of cases of partial gigantism —of the extremities, for example—have been described which are not due to elephantiasis, nor to the presence of circumscribed tumours. All grades of enlargement may occur from general gigantism, through unilateral and partial gigantism, to simple hypertrophy of the individual cells of a part.

Literature

Ziegler, *loc. cit.*, p. 15; Kaufmann, *loc. cit.*, p. 24; Kleinhans, *v. Winckel, loc. cit.*, p. 34; Küstner, *Muller, loc. cit.*, p. 15; Spiegelberg and Wiener, *loc. cit.*, p. 15; Olshausen and Veit, *loc. cit.*, p. 44; Runge, *loc. cit.*, p. 44; Ahlfeld, *loc. cit.*, pp. 24 and 34; Thoyer-Rozat and Schwab, '*Riesenwuchs*,' *Geb. Ges. Paris* (March 19, 1903), *ref. Zentralbl. f. Gynäk.* (1903), p. 1307; Ballantyne, 'The Problem of the Post-mature Infant,' *Journ. of Obst. and Gyn. Brit. Empire* (Dec. 1902), vol. ii.; Fuchs, '*Riesenwuchs und Partus serotinus*,' *Münchner med. Wochenschr.* (1903), Nos. 33 and 34; v. Herff, '*Kleidotomie*,' *Arch. f. Gynäk.*, vol. liii., p. 542; v. Winckel, '*Dauer der menschlichen Schwangerschaft*,' *Samml. klin. Vortr.* (Volkmann), Nos. 292 and 293; Eltze, '*Geburten von Kindern über 4000 grammes*,' *Diss. München* (1903); Freslug, '*Partus serotinus und Riesenwuchs*,' *Diss. Groningen* (1904); Moisnard, *Schwangerschaft und Geburt bei übermässig grossem Kinde*,' *Diss. Paris* (1903); Lütjens, *Diss. Greifswald* 1903 (*Kasuistik*); Jakoby, '*Riesenwuchs*,' *Arch. f. Gynäk.*, vol. lxxiv., pt. 3; Guéniot and Pierra, '*Atonische Nachblutungen bei Riesenwuchs*,' *L'obstétrique. Jahrg.*, vol. ix., p. 5; Starcke, '*Riesenwuchs und Dauer der Graviditat*.' *Arch. f. Gynäk.*, v[illegible] lxxiv., pt. 3; Grube, '*Übertragung und Riesenwuchs*,' *Geb. Ges. Hamburg* (April 18, 1906), *ref. Zentralbl. f. Gynäk.* (1906), p. 773; Rieck, *Ebenda*; Schubert, '*Riesenwuchs*.' *Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxiii., p. 4; Vaccari, '*Riesenwuchs*,' *Becken und Dauer der Schwangerschaft*, '*Monatsschr. f. Geburtsh. u. Gynäk.*, vol. xxiv., p. 1; Ballantyne, *loc. cit.*, p. 15, p. 302; Cusson,' *Kongenitale Elephantiasis*,' *Diss. Paris* (1905); Guinon, '*Kongenitale Elephantiasis*,' *Geb. Ges. Paris* (March 31, 1907), *ref. Zentralbl. f. Gynäk.* (1907), p. 1013; Lin[illegible]r, '*Kongenitale Elephantiasis*,' *Geb. Ges. Wien* (April 22, 1902), *ref. Zentralbl. f. Gynäk.* (1903), p. 51; Cyrille Panaiotoff, '*Ursache des Riesenwuchses*,' *Diss. Montpellier* (1907); Maclean, *Brit. Med. Journ.* (Feb. 23, 1907); Lichtenstein, *Geb. Ges. Leipzig* (Feb. 17, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 677; Waller, *Trans. Obst. Soc., London*, vol. i., p. 309 (1860); Varnier, *Obstétrique Journalière*, p. 185 (1900); Beach, A. P., *Med. Record*, vol. xv., p. 271 (1879); Ettingshans, *Samm. klin. Vortr.*, No. 358; Prochownik, *Zentralbl. f. Gynäk.* (1889), No. 33; Spencer, H., *British Med. Journal* (April 13, 1895), p. 808.

CHAPTER XVI

Congenital Dropsy—Emphysematous Putrefaction

For difficulties in delivery caused by marked over-distension of the bladder, and of the ureters, cystic kidneys, tumours of the liver, aortic aneurysm, tumours of the testis in the abdominal cavity, and hydrometra see the corresponding chapters. For fœtus *in fœtu*, see p. 327.

Congenital Ascites

Congenital ascites is more often the cause of dystocia than was formerly thought.[1] The literature on these cases is a very extensive one. Pure ascites is seldom observed: it is found most commonly with general fœtal dropsy, and with dropsy of the mother. Very frequently an hydrothorax is present at the same time. Ascites is found, further, with hydrammios; less commonly, however, with hydrammios in uniovular twins. The most frequent cause of this condition is no doubt syphilis—and especially syphilitic gummata of the liver—leading to severe congestion in the portal circulation. As further causes, are described—severe defects in the circulatory system, heart failure, absence of the ductus venosus Arantii (Paltauf), compression of the large vessels by tumours in the abdomen or by an over-distended bladder, fœtal chronic peritonitis, and hypoplasia of the urinary apparatus (Opitz). In macerated fœtuses, too, at times, large quantities of free fluid are found in the abdominal cavity. The causes of fœtal ascites are therefore often various. (See also Hohl.)

[1] Fordyce has collected sixty-three cases of this condition, and among these there were eight instances of syphilis and nineteen of hydramnios. Syphilis of the mother was present in three of twenty-four cases recorded by J. Y. Simpson. Ballantyne has collected seventeen cases in which distension of the bladder was associated with fœtal ascites. As he points out, it is an interesting fact that in comparatively few of the cases has any disease of the liver been present; but in nearly half of the cases which have been subjected to an autopsy, acute or chronic disease of the peritoneum has been found. In a case recorded by Kleinhans there was present atresia of both ureters, atrophy of both kidneys, over-distension of the bladder, a congenital umbilical hernia, and atresia of the œsophagus.

In a case of Ballantyne's, the fluid had a specific gravity of 1007; an alkaline reaction, contained albumin and globulin, and a distinct trace of oxyhæmoglobin.

The obstetrical importance of fœtal ascites lies in the fact that, at times, very marked difficulty in the delivery of the child may arise from the over-distension of the abdomen — difficulties which, eventually, may necessitate artificial aid. The majority of such fœtuses are, however, born spontaneously: for, on the one hand, they are frequently premature, and, on the other hand, the abdomen, being full of fluid, is very compressible and capable of adapting itself. Even marked cases do not lead to such bad results as are often observed in cases of hydrocephalus: for example—Küstner, in a review of the literature, has not been able to find a case in which rupture of the uterus occurred from over-distension of the abdomen.[1] Hohl has collected thirty-three cases, of which certainly only five were delivered without assistance. However, they were nearly all severe cases: in five cases delivery was effected by traction on the head or the shoulders which were already born; in one, by traction on the feet; in nine, by opening the abdomen; and in two with the forceps. In eleven cases, several operations were necessary. Hohl also describes several cases collected from the old literature, in which the midwife or the doctor had torn off the head or an arm.[2] In two cases also of twins, ascites was recorded;

FIG. 47.—FŒTAL ASCITES AND ANASARCA. Anterior aspect. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] In four cases out of sixty-three, the mothers died as a result of the prolonged labour and the operative interference required (Fordyce).

[2] In a case recorded by Eden, both lower limbs separated at the hip-joint during attempts to deliver, and the external genital organs were also destroyed by traction with the craniotomy forceps. After perforation of the abdomen, however, the child was easily extracted.

and on several occasions Hohl found that hydramnios was present. The most common presentation in cases of fœtal ascites is a cephalic one; but footling presentations are also observed more frequently than normally. Küstner found, in his review of the literature, about half as many footling presentations as head presentations. Transverse presentations infrequently occur. In a case observed by us the presentation was a brow; while Hohl also records a face presentation. In several cases during labour, weak uterine pains were present as the result of the marked distension of the abdomen or of the uterus. In marked degrees of fœtal ascites, the labour usually takes place in the following way: namely the head or the breech is born easily, and then the further delivery of the child is arrested. Less commonly, the marked over-distension of the abdomen interferes even with the delivery of the head from the pelvis. In such cases the use of the forceps will deliver the head, and then the delivery is definitely arrested. An examination with the half or the whole hand, under an anæsthetic, is then necessary. If this is not done and forced traction is made, the neck may be broken, or even the whole head torn off. Too short a cord, dicephalic or other double monsters, spina bifida, sac-like appendages or tumours, and locked twins must all be thought of. If the head is torn off, version and extraction should be practised: otherwise, version—except in transverse presentations and uncommon indications—would scarcely come into consideration. Küstner is opposed to version

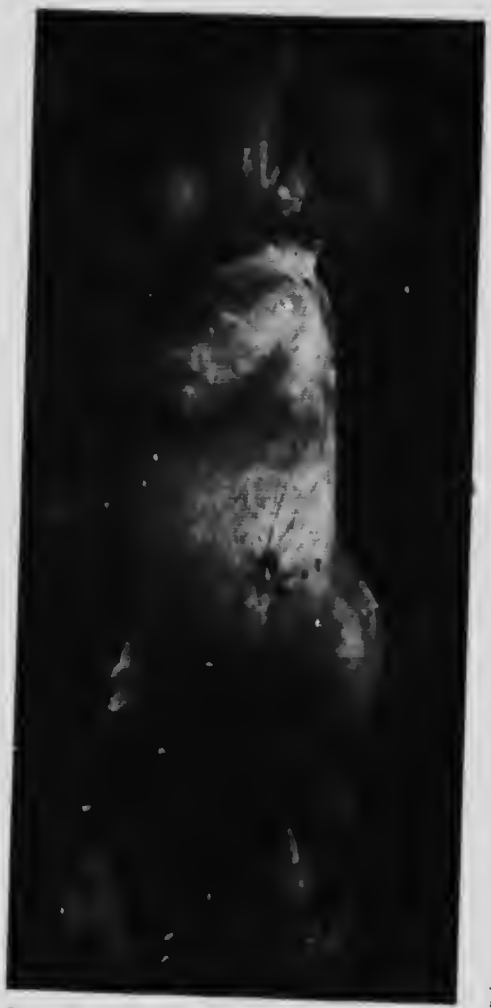


FIG. 48.—FŒTAL ASCITES AND ANASARCA. Lateral view, showing the site of perforation. (Specimen from the collection of the Göttingen Women's Clinic.)

when the head is torn off, and prefers to carry out extraction of the trunk with the cranioclast. As a general rule, in a cephalic presentation, an attempt should always be made first by careful traction on the shoulders, as the children are generally small, and the fluid in the abdomen can readily adapt itself to pressure. In well-marked ascites the abdominal walls — especially in dead, macerated fœtuses are at times so stretched, thin, and lacerable, that they give way to marked pressure with the fingers and allow the fluid to escape. Most commonly a spontaneous discharge of this kind occurs through an umbilical hernia (case of Kleinhans). In one of our cases (see p. 288), in extraction by the pelvis, the ascitic fluid perforated into the rectum at the level of the pouch of Douglas and escaped in this way. Extraction may be rendered easier by attempting to rotate the abdomen backwards into the hollow of the sacrum or to the side. If a careful extraction by the shoulders is impossible, then a diminution in the size of the abdomen is necessary. Puncture should be carried out: this method always being employed before mutilating operations—such as perforation or evisceration —are performed; as such children—especially in cases of fœtal peritonitis—may be delivered alive.[1] Very great difficulty may be encountered when the ascites is associated with tumours in the abdominal cavity, or with over-distension of the bladder. In ascites, with an over-distended bladder, it may be necessary to puncture first the abdomen and then the bladder.[2] In pelvic presentations, too, a careful attempt at extraction is to be practised; and if this fail, the methods described are to be employed. Many authors (P. Frank, Aubenas, and others) have successfully punctured the scrotum in pelvic presentations, on the assumption that the processus vaginalis peritonei in these cases commonly remains open. Lastly, it must be pointed out that well-marked fœtal ascites may hinder the proper expansion of the lungs as a result of the high position of the diaphragm (Extra-uterine acquired asphyxia).

In the Göttingen Women's Clinic, we have had under observation two cases of marked ascites with difficulty in delivery. Both cases occurred in the same woman. General œdema, was also present, but marked ascites was the most conspicuous condition. In both cases syphilitic disease of the liver could be recognised as the cause. The first labour occurred on July 27, 1907; the woman had already had

[1] Fordyce mentions two cases: in one the child lived for nearly a month, and in the second it recovered after the abdomen had been aspirated and 500 grm. of fluid withdrawn.

[2] In a case recorded by Stevens, after delivery of the head with forceps, it was found necessary to open the thorax, and perforate the bulging diaphragm with scissors. Three and a half pints of clear straw-coloured fluid at once escaped, and the further delivery of the child took place easily.

several abortions. On examination on July 2, marked œdema of both legs was present; the heart and lungs were sound, but there was a good deal of shortness of breath from the over-distension of the abdomen, and the high position of the diaphragm. The circumference of the abdomen measured 119 cm. The urine was normal. During labour there was inertia of the uterus, and, on account of weakness of the fœtal heart-sounds, an attempt was made at extraction—the child being in a footling presentation. Great difficulty was experienced. Suddenly, when the breech was visible, a large quantity (some two litres) of yellow fluid escaped from the anus of the child while marked traction was being made downwards; after which, further delivery took place easily. The child was dead, and the abdomen had now collapsed. There was general œdema present, and a discharge from the anus of the fluid described continued for a long time. The placenta was markedly œdematous, and weighed 2200 grm. Severe post-partum hæmorrhage occurred, which was arrested by an injection of cornutine. The weight of the child—a female—without the ascites was 2800 grm.; its length, 47 cm.

The second labour occurred on June 24, 1908. The patient, on account of hæmorrhage, caused by premature separation of the placenta, was placed in the inverted position. Examination showed a brow presentation. During the first stage there was inertia uteri, and towards evening marked hæmorrhage. For this reason—and, also, on account of the brow presentation and the failure of the fœtal heart-sounds—internal version (both feet being brought down) was performed without difficulty. Both feet were œdematous, and the tissues very friable. Further extraction failed on account of the marked ascites. As the child was already dead, the ascitic fluid (to the amount of three to four litres) was evacuated after perforation of the abdomen. Extraction then readily followed. The placenta was again very œdematous and weighed 2360 grm. There was marked atonic post-partum hæmorrhage, which ceased on the administration of cornutine.

Literature

Küstner, *Müller, loc. cit.*, p. 15; Ahlfeld, *loc. cit.*, p. 24; Olshausen and Veit, *loc. cit.*, p. 44; Spiegelberg and Wiener, *loc. cit.*, p. 15; Hohl, *loc. cit.*, p. 15; Bochénski, *Gynäk. Ges. Lemberg* (May 31, 1906), *ref. Zentralbl. f. Gynäk.* (1907), p. 369; Unger, '*Fötale Peritonitis*,' *Monatsschr. f. Geburtsch. u. Gynäk.*, vol. xxix., p. 5; Fordyce, W., *Teratologia*, vol. i., pp. 61, 143 (1894); Kleinhans, *v. Winckel, loc. cit.*, p. 34; vol. ii., pt. 3, p. 1671 (1905); Ballantyne, *loc. cit.*, pp. 15, 355–362 (1902); Eden, T. W., *Trans. Obstet. Soc. London*, vol. xlv., p. 45 (1904); Stevens, T. G., *Trans. Obstet. Soc. London*, vol. xxxvii., p. 5 (1896).

Hydrothorax

Congenital hydrothorax occurs very rarely indeed, and but few references to it can be found in the literature. It is frequently combined with ascites and other dropsical conditions; for example—in severe disturbances of the circulatory system, heart failure, and macerated fœtuses. Besides the difficulties in delivery which such a pleural effusion may produce, this fœtal anomaly has, further, an obstetrical interest, as the children affected with it may suffer after birth from marked asphyxia as a result of compression of the lungs (Asphyxia acquired after birth: see case recorded by Spiegelberg).[1] The dystocia occurs after the birth of the head in cephalic presentations, or of the breech in pelvic presentations; less commonly, before the birth of the head (Hohl's case). In such cases, forceps must be resorted to, and after the delivery of the head the true cause of the difficulty can be ascertained. The diagnosis is not, however, always easy. If the thorax is not born with the help of the various manœuvres described on p. 281, opening it by puncture or perforation must be considered. In pelvic presentations the abdomen must first be opened and then the diaphragm perforated. By this procedure it will be possible to diagnose the hydrothorax by the recognition of fluctuation through the diaphragm. Cases, however, of true hydrothorax are very uncommon. Spiegelberg's case, cited above, belongs to this class. Hohl has also collected the cases from the old literature. The majority are cases of combined accumulations of fluid in the abdomen and in the thorax (cases of Severinus and Carus). Hohl describes three cases of true hydrothorax: one of his own, and two from the literature (Gottel's case and one described without the name of the author). In one case, the head, engaged in the pelvic inlet, descended somewhat lower, and then remained stationary in spite of excellent pains. It was finally delivered by a difficult forceps extraction. The delivery of the trunk was associated with great difficulty, but succeeded at length with the help of one finger and Smellie's hook introduced into the axillæ.

In the dead child, delivered in this way, an enormous hydrothorax was discovered. In the case observed by Hohl himself, on account of presentation of the right arm, the membranes being intact, version was performed. The hydrothorax was recognised on the

[1] In this case labour occurred prematurely at the thirty-fifth week, and the child was born spontaneously, twenty minutes after the rupture of the membranes. It presented some anasarca, and a greatly distended abdomen and thorax. The autopsy revealed considerable pleural effusion into both pleural cavities; only two aortic valves were present, and the membranous part of the interventricular septum was absent.

introduction of the hand on account of the obstruction which ensued. The diaphragm was opened with Smellie's scissors, through the abdomen under the costal arches, and a large quantity of fluid evacuated, after which the child was born easily and spontaneously.

LITERATURE

Hohl, *loc. cit.*, p. 15; Küstner, *Müller, loc. cit.*, p. 15; Kleinhans, *v. Winckel, loc. cit.*, p. 34; Spiegelberg and Wiener, *loc. cit.*, p. 15; Ballantyne, *The Diseases and Deformities of the Fœtus*, vol. i.; L. Seitz, *v. Winckel, loc. cit.*, p. 15; Rührlen, *Schmidts Jahrbuch*, vol. xciv., p. 198.

Œdema of the Skin and General Dropsy

This occurs either as a pure anasarca, general congenital œdema,[1] or, more commonly, combined with congenital collections of fluid in the body cavities. In these cases it consists either of a pure fluid transudation, or, less commonly, of a watery gelatinous infiltration[2] of the subcutaneous tissue. Sometimes a marked hyperplastic condition of the skin and subcutaneous tissues is also present; while not infrequently thickenings of the skin are present, of the nature of elephantiasis: as, for example, in the remarkable case recorded by Steinwirker, elephantiasis congenita cystica[3] (see illustration in Olshausen and Veit). In association with fœtal dropsy, œdema and dropsical

[1] Clifford White has recorded an interesting case of general œdema of the fœtus occurring in University College Hospital. The mother, thirty-three years of age, had had ten pregnancies. The eldest child was alive and well; the next two children died soon after birth; and the remaining six pregnancies ended either in the birth of premature dead children or in miscarriages. The child presented by the face, and was delivered with forceps. There was delay in the birth of the shoulders, necessitating the introduction of the finger into the anterior axilla. Extensive œdema of the tissues was found to be present, and the skin of the neck was torn by the finger in several places, giving exit to a considerable quantity of serous fluid. The body was then easily delivered. The child's heart beat for some minutes, and it made several attempts at respiration. The mother made a good recovery. Wasserman's reaction was negative in the mother, and the spirochæta pallidum could not be demonstrated in the liver or in any of the fluids of the fœtus. The child, a female, weighed 6 lb., and showed well-marked œdema, ascites, and hydrothorax. The viscera, blood-vessels, thoracic duct, and umbilical cord were normal. Microscopic sections of the tissues showed well-marked œdema of the skin, subcutaneous tissue, and muscles. The placenta weighed 3 lb. 9 oz., and showed slight œdema only.

[2] This, Ballantyne suggests, may be due to the undeveloped or mucoid state of the subcutaneous tissues when attacked by the dropsy.

[3] In this condition the subcutaneous tissue is greatly thickened, and cysts are found in it containing serous fluid. Meckel's case is figured by Ballantyne. He regarded it at first as an acephalus, as it appeared to consist solely of a trunk with limbs bearing a fleshy spongy mass instead of a head; but on making a section through it he was surprised to find underneath a well-formed fœtal face.

conditions are occasionally also observed in the mother, as well as dropsy of the placenta and hydramnios. Whether the dropsy of the mother occurs primarily and that of the fœtus secondarily, or vice versa, cannot always be determined with certainty; but both are possible. Hereditary syphilis, anomalies of the cardiac ostia (closure of the foramen ovale, permeability of the ductus arteriosus), diseased conditions of the mother's blood (hydræmia, leukæmia), disturbances in the circulation through the umbilical veins are given further as causes of this condition. In cases of chondrodystrophia and in neardiaci, general dropsy is also frequently seen.[1] (For the old literature up to 1849, see Hohl.)

The fœtuses are almost always incapable of living, and frequently die during pregnancy or during delivery. With regard to difficulties in delivery in such cases, there are a large number of instances in the literature where such difficulties were encountered. Cases of dystocia are seen, least commonly of all, in pure anasarca (cases of P. Ruge and Fuhr: see Kleinhans). In the case recorded by Ruge, the extraction of the child as a footling presentation gave rise to great difficulty, as the œdematous and infiltrated skin produced a stiffening of the fœtus similar to the stiffening of the muscles met with in cases of rigor mortis, and this proved a great hindrance to the delivery of the child. In Fuhr's case the difficulty was overcome by the fact that the œdematous fluid trickled away from numerous tears in the skin which occurred during the extraction. The difficulty in delivery, or in the performance of the various obstetrical manœuvres, does not only depend on the general enlargement of the child, but also on the fact that in many cases marked friability and fragility of the tissues, as the result of the œdematous infiltration, is also present. In the case of ascites and universal œdema—already mentioned (p. 288)—during

[1] The general character of these cases of universal dropsy of the fœtus seems to show that they are in reality of the cachetic or anæmic variety of œdema—that is, due to the presence of some toxic substances in the tissues. These are probably of the nature of waste products, and they may act in one of three ways: either by stimulating the secretory activity of the endothelium, or by injuring its vitality and so increasing its permeability, or by setting up an osmotic flow of water into the tissues. It may well be that the primary condition is a failure in the excretory functions of the placenta, and that this leads to an accumulation of waste products in the tissues of the fœtus. The placental change may be due to some maternal disease. On the other hand, the œdema may be due to changes in the fœtal blood, and this has been suggested as a cause. Whether these changes are merely of an anæmic character, or are brought along by the presence of certain poisonous substances in the blood remains to be proved. The œdema in these cases is comparable to that seen in the adult, as the result of such general diseases as diabetes or cancer.

In the majority of these cases there is no evidence of the presence of either maternal or paternal syphilis, and in very few instances has fœtal syphilis been demonstrated as present.

the extraction of the child—which was by no means difficult—the skin of the leg peeled off like a cuff. Such observations have been made on many occasions. Whole extremities and the head have been torn off in forceps extraction—and this, too, even when traction has been applied with certainly by no means excessive force. It is evident that definite rules of treatment cannot be given for every case. As the possibility of dropsical children remaining alive is highly problematical, the majority of obstetricians, including Spiegelberg, recommend that delivery should be terminated not by manual manipulation but by the use of instruments. Even then, tearing off of the head may still occur: for example, in a forceps delivery. In the most marked forms of dropsy, nothing except embryotomy is available, which in these circumstances can hardly be considered as a set obstetrical operation. Very often the presence of such dropsical children may be suspected. The œdema may be recognised—especially when some part of the child prolapses—by the depression resulting from the pressure of the finger. Further, in cases of marked œdema of the mother, one should always be prepared for the presence of œdema of the fœtus. (For the cases of ascites and general œdema which have been under observation in the Göttingen Women's Clinic, see p. 288.)

LITERATURE

Hohl, *loc. cit.*, p. 15; Kleinhans, v. Winckel, *loc. cit.*, p. 34; Spiegelberg and Wiener, *loc. cit.*, p. 15; Ballantyne, *loc. cit.*, p. 15; fig. 32, p. 23[illegible] Ahlfeld and Veit, *loc. cit.*, p. 44; Jarho, *Diss. Marburg* (1902); Kreisch, *Muncher med. Wochenschr.* (1901), p. 1387; Neelson, *Berliner klin. Wochenschr.* (1852), p. 30; Betschler, *Klin. Beitr. zur Gynäk.* (Breslau, 1862), p. 200; Steinwirker, *Diss. Halle* (1872); Keiller, *Edinb. Med. and Surg. Journ.*, April 1855; Lindner, *Geb. Gesellsch. Wien* (April 22, 1902), *ref. Zentralbl. f. Gynäk.* (1903), p. 51; Seuleeq, *Geb. Gesellsch. Paris* (March 11, 1907), *ref. Zentralbl. f. Gynäk.* (1907), p. 1013; Meckel, *Arch. f. Anat. u. Phys.*, p. 149 (1828); Clifford White, *Proc. Roy. Soc. Med. London* (1912), vol. v.

Congenital Myxœdema

Congenital myxœdema (Myxidiocy), if we may judge from the sparse literature relating to it, is very rare. In this disease, due to a defect of the thyroid gland, the skin and the subcutaneous tissue has a swollen, œdematous, or myxœdematous, character. Whether the condition is due to the presence of mucin,[1] or a mucin containing œdematous fluid, or only to an œdematous infiltration, is not quite certain (see Ewald). Ewald considers, as it appears to me, together with sporadic cretinism, cases of so-called congenital myxœdema with total

[1] Any excess of mucin there may be in the subcutaneous tissues is to be explained by the fact that there is an overgrowth of the connective tissue, and that young connective tissue is rich in ground substance and, therefore, in mucin.

congenital aplasia of the thyroid (Athyreosis, thyreoaplasia), in contrast to endemic cretinism, which, he maintains, very seldom occurs as a congenital condition. Kaufmann does not think the designation, 'sporadic cretinism,' a very good one. These cases of so-called sporadic cretinism may present later[1] the condition of dwarfism, idiocy, myxœdematous skin, swollen lips, and thick tongue. As a result of the changes in the skin in congenital myxœdema, and the increase in the size of the child produced thereby, its delivery may be rendered difficult. Weirich has recently recorded two cases of congenital myxœdema; in both cases the labour was long, and one case required the application of forceps. Sarabia also reports two cases of congenital myxœdema. A review of the literature, however, leads one to the conclusion, that many conditions not belonging thereto have been included under this heading.

LITERATURE

Kaufmann, *loc. cit.*, p. 24; Kleintans, *v. Winckel, loc. cit.*, p. 34; Ballantyne, *loc. cit.*, p. 15; Ewald, *'Die Erkrankungen der Schilddrüse,' Nothnagel, loc. cit.*, p. 53, vol. xviii.; Weirich, *Diss. Jena* (1901); Sarabia, *Verhandlg. d. Span. gynak. Gesellsch., ref. Zentralbl. f. Gynak.* (1904), p. 1174; Charrière, *Diss. Paris* (1907).

Emphysematous Putrefaction

Emphysematous enlargement of the fœtus is the result of decomposition of its tissues. When the bag of membranes is ruptured and organisms find their way into the uterus (introduced by the examining fingers, by instruments, or by the passage of germs from the vagina or the cervix), the contents of the uterus undergo putrefaction, especially if the temperature of the mother is raised. This decomposition occurs most rapidly with macerated children; less quickly with recently dead children. It is therefore found, primarily, in cases of difficult and protracted labour—especially with a contracted pelvis; for in these cases, as is well known, the membranes often rupture prematurely, and numerous attempts at delivery have been made.

The organisms causing the decomposition are the *Bacillus coli*,[2] *Bacillus ærogenes capsulatus*, and anærobic bacilli (Krönig and Lindenthal). As a result of this decomposition of the fœtus, gas develops in the uterus, giving rise to the condition termed tympania uteri or physometra. The presence of gas may be recognised by percussion

[1] In the majority of cases it is not until the child is one or two years old that the cretinoid condition is recognised.

[2] The *B. coli*, in all probability, is able to produce gas only in the tissues of diabetics. Emphysema of the fœtus, physometra, and emphysema of the uterine wall are in practically all cases produced by the *B. ærogenes capsulatus*.

over the highest part of the uterus. Following on these putrefactive processes the skin of the fœtus peels off in blisters and patches, the umbilical cord acquires a greenish yellow colour, and, finally, the whole fœtus becomes discoloured and smells, as does also the liquor amnii. Later, a formation of gas may also occur in the fœtal tissues, especially in the subcutaneous tissue and in the cavities of the body (Emphysematous putrefaction).

It is very often possible in these cases to recognise crepitation due to the accumulation of gases. In well-marked cases, the whole body appears to be blown out, the skin is tightly stretched and crepitates under the slightest pressure, the limbs are swollen, the trunk distended, and when punctures or incisions are made into the soft parts these collapse[1] (Spiegelberg). Such a marked condition of emphysematous putrefaction is seldom observed at the present day, because of the better practice of asepsis, and the better training of midwives and doctors, as well as the fact that a doctor is more commonly present at cases of labour. In former years, when these conditions did not obtain, it was seen fairly often (see Hohl). Difficulty in delivery of the child from these changes not uncommonly ensues, and, even at the present time, such cases do occur. The difficulty may certainly readily occur when the labour comes to a standstill, as is so frequently observed in cases of contracted pelves, from secondary uterine inertia, weak pains resulting from tympania uteri, and tetanus uteri. In these cases, however, the delivery of such fœtuses as rapidly as possible is indicated by the threatened, or existing, septic infection of the mother. In any manipulations that may be necessary, the tissues will be found to be rotten and friable. In the treatment of dystocia from emphysematous distension it is best to puncture the thorax or the abdomen, whenever necessary, to let out the gas. Above all things, all manipulations should be avoided which may lead to tearing off of fœtal parts, as in that case the attendant is almost always compelled to introduce the hand into the infected genital tract, and so is very likely to promote the spread of the infection.[2] Spiegelberg recommends 'that the fœtus should be seized with the craniuoclast, and that then the gas should be let out by perforation of the thorax or abdomen.' Still more difficult to answer is the

[1] Although in a certain number of cases of so-called gas sepsis, the presence of the *B. œrogenes capsulatus* is no doubt a post-mortem phenomenon, yet a number of cases have been recorded in which it was proved to be present in the tissues ante-mortem.

[2] The *B. œrogenos capsulatus* may gain access to the uterus in one or two ways. The first and most important is by the examining finger or instruments; in the second place, it may occasionally be present in the circulating blood. As it is always present in human fæces, it is very likely to be introduced into the uterus when the external genitalia are insufficiently cleansed.

question, What is the best method of procedure when the mother's soft parts are more or less undilated? Dilatation with a Bossi's dilator, or similar instrument, is best avoided in such cases, as by their use, as is well known, tears of the cervix frequently occur, which may very readily become infected, and so serve as new doors of entrance for the dreaded general infection. This danger also exists in vaginal Cæsarean section which has, however, been often employed to effect the delivery of the child or as a preliminary to total extirpation of the uterus. All these operations have a very doubtful prognosis. Other obstetricians, therefore, employ a more waiting policy—repeated douching of the uterus with a Fritsch's catheter, and the copious application of iodoform to the uterine cavity.

Hohl discusses this question very fully. According to his investigations, putrefaction of the fœtus and the dystocia that possibly results was known to Celsus, Smellie, Mauriceau, Röderer, and others. Dugès suggests that putrefaction converts the fœtus into a sort of tampon, so that it does not undergo the usual movements of the mechanism of labour, but fills up and closes the birth canal. His view is so far right that tetanus of the uterus is very often associated with a putrid fœtus. Hohl gives a complete review of the cases in which difficulty in delivery occurred.

Literature

Spiegelberg and Wiener, *loc. cit.*, p. 15; Wächter, *Diss. Munchen* (1875); Hohl, *loc. cit.*, p. 15; Seitz, '*Die Veränderungen von Fötus und Plazenta nach dem Tode der Frucht*,' v. *Winckel, loc. cit.*, p. 15; vol. ii., pt. 2, p. 276; Runge, *loc. cit.*, p. 24.; Welch and Nuttall, *Johns Hopkins Hospital Bull.* (1892), vol. iii., p. 81; Dobbin, *Johns Hopkins Hospital Bull.* (1897), vol. viii., p. 24; Chiari, *Prag. med. Wochenschr.* (1893), vol. xviii., p. 1; Halban, *Monatssch. fur Geb. u. Gyn.* (1900), vol. xi., p. 90.

CHAPTER XVII

The more Important Congenital Diseases and Malformations of the Skin

Here we must consider first the amnion (see p. 11). The so-called Simonart's ligaments may be torn either *in utero* or during birth, and may then remain as more or less long threads attached to the skin. These amniotic bands[1] may produce other changes in the skin—such as constrictions, which may extend down to the bones; scars in the skin, and spontaneous amputations with recent or old changes. In other cases the amniotic adhesions form long cylindrical processes between the superficial surface of the fœtus and the amnion. When these processes or bands tear away from the surface of the body, an area devoid of skin remains, forming a wound (Ahlfeld). Such circular defects of the skin[2]—which, according to their age, may be cicatrised over, or still raw—are found especially in the neighbourhood of the occiput. The skin on the peripheral side of the constricting groove is either œdematous or atrophied.

These anomalies have an obstetrical interest, inasmuch as the bands, if not torn through, may give rise to difficulties in delivery. Thus a midwife said that she heard the tearing of such a band during the delivery of the child as a 'crack.' A case is also on record where the midwife had to cut the amniotic bands after the birth of the child, so as to completely separate it from the mother (see Küstner).

Calcification of the superficial layers of the skin has also been observed—generally combined with calcification of the membranes—so that difficulty in delivery may arise (so-called union of the fœtus with the uterus).

Of congenital syphilitic affections of the skin I will just mention syphilitic pemphigus and the maculo-papular syphilide.

[1] In cases of malformations, the fact that no traces of any amniotic adhesions can be found does not prove that they were not originally present, as no doubt they may entirely disappear during the course of a pregnancy.

[2] Specimen No. 801, Univ. Coll. Hosp. Med. School Museum—that of an anencephalic fœtus—shows several such scar-like areas in the skin on the dorsum of the right foot. The most marked is situate just in front of the outer malleolus. They are no doubt the scars left by old amniotic adhesions.

Congenital disappearance of the pigment (congenital leucopathia, leucodermia) may further be mentioned: this is either of a universal—albinism (albinos)—or only partial nature. The albinos are completely pigmentless, the skin is clear white and transparently rosy. The functions of the skin are unaffected. The hairs are yellowish white like silk,[1] and the iris and choroid are free from pigment.[2] Albinism is usually found in several members of the same family.

In albinismus partialis, scattered pigmentless portions of the skin are found, consisting of white spots, which generally are irregularly outlined.[3]

In contrast to these are the cases of congenital hypertrophy of pigment, and a circumscribed increase of pigment.[4] In these cases other portions of the skin—the papillæ and the horny layer—may undergo hypertrophy. The pigmented flat areas are distinguished as flat nævi;[5] the others, as wart-like nævi[6] (see Lesser).

Congenital and permanent dilatations of the blood-vessels of the skin occur as telangiectases[7] and angiomata; they are called, as a rule, nævi vasculosi (Port-wine mark). They may vary very much in size. As is well known, popular belief attributes them to some maternal impression (see also Kaufmann). The congenital angiomata at times are so numerous that they may involve the whole of an extremity (Lesser). As such angiomata, from the possibility of their being injured, may constitute a great danger for small children, their early surgical treatment is indicated.

Congenital dilatations of the lymph-vessels (lymphangiectasis and lymphangiomata) are not uncommonly observed in the hand. They are in some cases so extensive that they may involve the whole of an extremity, or cover a large part of the body. In the case mentioned of Malcolm McLean (p. 117), so large a lymphangioma of one arm was present that it led to difficulty in delivery.[8]

Elephantiasis congenita belongs also to this chapter (see the section relating thereto). Quite recently, I saw a child with a lymphangioma

[1] In a case recorded by Folker the hair was red.

[2] The pupil, therefore, appears red and the iris pink; and, as the retina has no protection from light, photobia is always present.

[3] Partial albinism is, of course, more noticeable in coloured races. In some tropical countries it is said to be endemic. Marey records the case of two negroes who had six children, of whom three were black and three white.

[4] Nævus pilus.

[5] Nævus pigmentosus.

[6] Nævus venosus.

[7] The term 'telangiectasis' is usually employed for a condition which is not congenital, and which is due rather to an enlargement of pre-existing vessels than to the formation of new ones.

[8] Moncorvo, of Rio Janeiro, has recorded a series of cases in which the disease developed *in utero*, but had increased after birth. He found no filaria, but attributed the solid œdema to *Streptococci*.

in the right axilla the size of a fist which had given rise to no difficulty in delivery.

A very rare congenital disease of the skin is ichthyosis congenita (Fish-scale disease), known by the pathologists as hyperkeratosis universalis congenita. Kaufmann describes it as follows: 'The skin appears covered with polygonal in many places disk-like horny plates, which are separated from one another by fissures and clefts crossing in various directions—especially surrounding the joints and are composed of thick layers of horny epithelium enclosing lanugo; the horny layers are also present in the widened hair follicles; the thickened (6 to 10 mm.) epithelial cells burst apart from one another—hence the resemblance of the skin to that of a half-roasted sucking-pig or the designation 'Harlequin Foetus'; the fingers and toes are stunted as a result of the shortness of the stiffened skin; the rigid mouth remains open; the eyelids and the lips are defective; the eyes are covered by the conjunctivæ in a condition of ectropion; the skin, covered with its horny plates,[1] becomes directly continuous with the mucous membrane of the alveolar processes; and the feet are in a position of club foot' (see Ballantyne and Ziegler).

These children are usually born one to two months prematurely; but full-term children may also be born affected with ichthyosis. They die as a rule after a few hours;[2] or, at latest, on the day after birth (for a detailed account of the anatomy of the condition, see Jarisch). Death results in part from the marked alteration in the skin; in part from the great difficulty in feeding, due to the condition of the mouth. The etiology of this rare condition is still very doubtful. Hereditary transmission is not recognisable,[3] although cases have been recorded in which several children of the same mother have suffered from ichthyosis. It is probable that the disease begins in the third to fourth month of pregnancy. After birth, the temperature sinks very rapidly. In many cases of ichthyosis the liquor amnii has been deficient. It is not certain whether these two conditions stand in any causal relationship to one another, Ahlfeld is inclined to regard the oligohydramnios as secondary. Treatment, as has been said, is useless.[4]

[1] Unna maintains that this condition is quite different from ordinary ichthyosis, and says that all the histological phenomena are attributable to one condition: namely—a firmer connection of the epithelium, mainly limited to the surface. The skin is too small for the body—hence the fissures and the ectropia and contractions.

[2] In the less severe cases, life may be prolonged for some little time; thus Crocker records the case of a child with ichthyosis congenita which lived three months, although its vitality was very low all through its life.

[3] This statement is too absolute: the disease is hereditary in some cases, but by no means in all. Kaposi records a case in which all the five sons of an ichthyotic mother were ichthyotic, while her three daughters were free from the disease.

[4] Thyroid extract, in small doses, has been found of considerable value in some cases.

Baths must mainly be employed, which no doubt do the children some good. For less extensive cases, Ahlfeld recommends rubbing with ointments and fats (cod-liver oil).

The nails very seldom exhibit any congenital affections. By anonychia is understood the partial or complete absence of a nail. Sometimes this condition is observed in several members of the same family. It may affect all the fingers and toes or only one; the bed of the nail, however, and the skin surrounding the nail, may be well formed.

Of congenital affections of the hair the most important is alopecia congenita, which may be either partial or total. The disease may remain stationary permanently; or, after a longer or shorter time, may disappear. Anomalies of the teeth and nails are sometimes observed at the same time. In many cases the influence of heredity can be recognised.[1] In congenital partial alopecia larger or smaller areas of the body—which in later life may only increase very little in size—are devoid of hair. At the same time, the sebaceous glands may be imperfect or undeveloped. Many authors regard an anomalous condition of the amnion as the cause of alopecia congenita.

Of anomalies in the colour of the hair, poliothrix circumscripta has been observed—that is, a partial congenital want of pigment, in which white tufts of hair, without any accompanying decolorisation of the corresponding portions of the skin, may be transmitted through many generations.

Dermoid tumours are very frequently observed in the skin. They occur as the result of cutting off of portions of the branchial clefts during embryonic life, or by inclusion of the skin or epithelial structures in the depth of the corium, or in the subcutaneous tissues (Kaufmann). They are found most frequently where, during embryonic life, infoldings of the epithelium, or fœtal folds or clefts, are met with clothed with epithelium. Microscopically their wall exhibits the structure of the skin.

Lastly, I must mention briefly the changes in the skin which are found in cases of maceration (vesicles with exposed red portions of the underlying skin), mummification (dry, leather-like skin in premature children, reddish from the want of fat), in congenital icterus (intense yellow colour—at times combined with hæmorrhages); in congenital heart disease, white pneumonia, malformations of the lungs (blue or cyanotic); in congenital infectious diseases (measles, scarlet fever, and smallpox); in lithopædion (calcification of the superficial parts of the skin); in contracted pelves (pressure-marks and gangrene); in operative termination of labour (contusions of the skin or tearing of the soft parts); in a caput succedaneum (serous infiltration of

[1] Nicolle and Halipré record that in one family there were thirty-six individuals in six generations with defective hair and nails.

the scalp, and superficial bleeding and excoriation); in a cephalhæmatoma (distension of the skin by blood present between the pericostenm and the bone). I have already mentioned adhesions of the skin in various parts of the body.

Literature

Jarisch, '*Hautkrankheiten*,' *Nothnagel, loc. cit.*, p. 53; vol. xxiv., p. 1; Kaufmann, *loc. cit.*, p. 24; Küstner, *Müller, loc. cit.*, p. 15; Lesser, *Hautkrankheiten*; Ahlfeld, *loc. cit.*, p. 24; Ballantyne, *loc. cit.*, p. 15; Lenglet, *Diss. Paris*, 1902 ('*Übersicht über angeborene Hautkrankheiten*'); Döring, *Diss. Erlangen*, 1901 ('*Angeborene Haarlosigkeit*'); Böhler, *Diss. Freiburg*, 1901 ('*Ichthyosis cong.*'); Raelot, *Diss. Paris*, 1902 ('*Alopecia cong.*'); Macé '*Ichthyosis*,' *Geb. Gesellsch. Paris* (Jan. 17, 1907), p. 916; Singer '*Alopecia cong., Übersicht*,' *Diss. Erlangen* (1906); Caripuy '*Part. Alopec. cong.*,' *Geb. Gesellsch. Paris* (March 25, 1907), p. 944; Folker, *Lancet*, May 31, 1879; Marey, *Amer. Journ. Med. Sci.* (1839); Moncorvo, *Annales de Derm.* (1893), vol. iv., p. 233; Crocker, *Diseases of the Skin* (1905), vol. i., p. 516; Unna, *Histopathology*, p. 1377; Nicolle and Hudipré, *Annales de Derm et de Syph.* (1895), vol. vi.

CHAPTER XVIII

Double Malformations—Double Monsters

A detailed description of this interesting condition, with an exhaustive bibliography, is given by Schwalbe, Ahlfeld, Strassmann, Marchand; also by Küstner, Ziegler, G. Veit, and others.

A definition of what is included under the term, 'Double monsters' (as Schwalbe rightly points out) is not easy, and, indeed, a sharp distinction between a double malformation and a single malformation is impossible[1] (see Parasitic double monsters and teratomata). For this reason Schwalbe contents himself with a morphological definition, which is not based upon the mode of origin. According to this, a double monster is defined as one which shows at least some partial doubling of the axis of the body. By such a definition, it is possible to separate the double monsters from cases of the duplication of single limbs. The fœtal bodies which are united with one another in double monsters are either completely or incompletely developed — Duplicitas completa and Duplicitas incompleta. The first group is by far the most frequent. To the second group, whether the cephalic or pelvic extremity is affected, belong the teratomata, in which the condition is that of a completely stunted parasite (Ziegler).

In cases of duplicitas completa the united bodies may be equally or unequally developed. In unequally developed twins, the well-developed twin is termed an autosite; the imperfectly developed twin, a parasite or a bigerminal teratoma. As no heart develops in the parasite, its nutrition must be carried on by the well-developed twin.

Triple formations and triple monsters are very rare. As Strassmann points out, a complete triple formation[2] is excessively rare, as the most common condition is rather that of the union of a complete double monster with an incomplete double monster.

In many malformations—for instance, in dicephalus and diprosopus—

[1] This difficulty will be realised when the reader remembers that such opposite conditions as a sacral parasite or a double penis, and pygopagous twins—such as 'Rosa' and 'Josefa'—all belong to the class of double monsters.

[2] In the extraordinary case reported by Reina and Galvagni, the monster had three heads, three upper limbs, and two united trunks with two lower limbs. A single set of male genitals was present.

there is the appearance of a single body; but the X-rays show that complete or partial doubling of the spinal column is present.

To give a classification of double monsters, satisfying all demands, is not easy. Many writers adopted formerly—and, indeed, some even at the present time do—the well-known scheme of Förster. He describes double monsters as follows:—

1. Monstra duplicia katadidyma—Duplicitas anterior (superior);
2. Monstra duplicia anadidyma—Duplicitas posterior (inferior);
3. Monstra duplicia anakatadidyma—Duplicitas parallela.[1]

A second group, Duplicitas cruciata, is no longer recognised. According to Strassmann, the terms 'Monstra anadidyma' and 'Monstra katadidyma' may give rise to misunderstanding; so they in his opinion they are better given up. Schwalbe, indeed, asserts that this classification has produced 'a great deal of confusion' in the literature. The reader must remember that the terms used in the classifications of different authors have often very different meanings.

Taruffi divides double monsters into three groups:—

1. The double monster is due to the union of two bodies in the region of the epigastrium and thorax;
2. The fœtuses are united together by the head;
3. The union is in the region of the pelvis.[2]

On his system, Ballantyne bases his classification.[3]

[1] (1) A monster divided above, but single towards the podalic pole; (2) a monster divided below, but single towards the cephalic pole; (3) a monster separate above and below, but united in the middle.

[2] (1) *Thoracopagus*: united by the epigastric region, and more or less of the thorax; (2) *Syncephalus*: united by the heads, more or less, with trunks separate or fused; (3) *Dicephalus*: united by the pelves, with heads and often necks and part of thorax separate.

[3] Ballantyne's classification is as follows:—

POLYSOMATOUS TERATA (*Two or more individuals involved*)

1. Twins, entirely separate, but in a single chorion (Monochorionic).
2. Twins, united only by the vessels of their umbilical cords (Allantoido-angiopagous).
   (*a*) Paracephalic. (*b*) Acephalic. (*c*) Amorphous.
3. Twins, united more or less completely (Double Monsters).
   (*a*) Symmetrically united.
      (1) Syncephalic. (2) Dicephalic. (3) Thoracopagus.
   (*b*) Asymmetrically united.
      (1) Cephalo-parasitic.[4] (4) Thoraco-parasitic. (6) Lecano-parasitic.
      (2) Prosopo-parasitic. (5) Gastro-parasitic. (7) Melomelic.[5]
      (3) Trachelo-parasitic.
4. Triplets, quadruplets, quintuplets.
      (1) Entirely separate. (2) United.

[4] These groups (Nos. 1 to 6) include the various forms of parasitic double monsters attached to the different regions of the body—from the head to the pelvis.

[5] This group (No. 7) includes the cases in which an accessory limb, or part of one, is inserted upon another normal limb.

Ahlfeld, who genetically attributes all the various forms to the occurrence of fissures, gives the following classification :—

1. Total Fission.
   (*a*) Equally developed forms (Homodegous twins—Thoracopagus, Craniopagus).
   (*b*) Unequally developed forms (Fœtus papyraceus—Acardiacus, Epignathus, Sacral teratomata, Inclusio fœtalis, Transplantatio fœtalis).
2. Partial Fission.
   (*a*) Fission beginning at the cephalic extremity (Duplicitas anterior—Dicephalus, Dicephalus, Ischiopagus, Pygopagus).
   (*b*) Fission beginning at the pelvic extremity (Duplicitas posterior—Dipygus, Janiceps).
3. Multiple Fission.
   (*a*) Overgrowth of the whole body (Giguntism).

Marchand, in his excellent account of this question in Eulenburg's 'Real-encyclopaedia,' gives the following system :—

1. Duplicitas symmetros.
   The bodies of originally double embryonic rudiments of equal value are united with one another, and are either equally developed (equal), or one of the two has not developed to the same extent as the other (unequal)—Double monsters, Conjoined twins.
   (A) Duplicitas completa.
      (*a*) The union is confined to the lower end of the body—Monstra duplicia cum conjunctione inferiore.
      (*b*) The union is limited to the middle of the body or proceeds upwards—Monstra duplicia cum conjunctione media.
      (*c*) The union is limited to the upper end of the body or proceeds downwards—Monstra duplicia cum conjunctione superiore.
   (B) Duplicitas incompleta.
      The doubling affects not the whole germ, but only a portion of it. The formation may be symmetrical or asymmetrical.
      (*a*) The doubling is limited to the lower end of the body—Duplicitas incompleta inferior.
      (*b*) The doubling is limited to the upper end of the body—Duplicitas incompleta superior.
      (*c*) Triplets and multiple formations.
2. Duplicitas asymmetros.
   The two bodies are developed from two originally unequal

asymmetrical rudiments, of which the one (always imperfectly developed) is more or less enclosed by the other and nourished by it—True parasitic double monsters.

(*a*) Teratoid tumours.

In his comprehensive work, Schwalbe gives a classification which is based on a purely morphological point of view:—

1. Free double formations, Twins either completely separated from one another, or only united by their placentæ.
   (*a*) Equally developed embryonic rudiments—Gemini monochorii æquales (uniovular twins). According to Schwalbe, the uniovular twins which are only united by the placenta are also double formations.
   (*b*) Unequally developed embryonic rudiments—Gemini monochorii inæquales (acardii).
2. United double formations.
   (*a*) With equally symmetrically developed individual parts—Duplicitas symmetros (to this division belong the double symmetrical and single symmetrical forms).
   (*b*) With unequally—that is, asymmetrically—developed individual parts—Duplicitas asymmetros (parasites).

The old literature is to be found in Hohl.

For Hohl's remarks on the obstetrical importance of double malformations, see pp. 332 and 334: as well as for G. Veit's classification of these malformations from the point of view of the progress of labour.

All double malformations develop from a single ovum.[1] For this reason they have always a single placenta, and a single chorion, and are always of the same sex. If, as Schwalbe maintains, a difference in the sex is possible, then such an occurrence must be considered as an example of true hermaphroditism. Schwalbe has observed a case of pseudo-hermaphroditism in an acardius.[2] As a general rule the female sex predominates very markedly in double monsters.[3]

The question as to the special genesis of double monsters is one of great interest. Among the earlier writers the idea prevailed that these malformations occurred in this way: namely, that two

[1] Whatever view may be taken of the exact mode of origin of double monsters this, at any rate, is an established fact.

[2] This case is not entirely satisfactory, since the sexual glands were not examined microscopically.

[3] Of 883 cases collected by Taroffi, 540 were of the female sex, 315 of the male sex, and in twenty-eight the sex was not determined. Förster calculates the proportion of females to males as about three to one.

embryos, originating from a single ovum, approached one another *in utero*, came into contact, and finally became united with one another (see Ahlfeld and Schwalbe). Another curious impression was that such monsters were the result of an improper quality of the seed. It is still a question—as it was formerly—whether these malformations arise from the union or the division of the embryonic rudiments. According to the 'fission theory' (Ahlfeld, Förster, Lenckart, Joh. Müller, and others), they develop in this way: namely, that an originally single embryonic rudiment later divides. According to the 'fusion theory' (Geoffroy St. Hilaire, Panum, Dareste, B. S. Schultze, and others), two originally separate embryonic rudiments are present, which, however, in their later development become united with one another to a greater or lesser extent. In Schwalbe's opinion, such a difference—namely, union or division in the sense of two opposite exclusive theories—cannot be maintained. In order to come to a decision upon this question, as he points out, there are three means available: Double formations may be investigated at very early periods of fœtal life; the experimental method may be pursued; or an attempt may be made at the reconstruction of the complete monster, so as to define the latest period at which it can have arisen. (For further information, see Schwalbe's detailed and lucid remarks; only his conclusions can be given here.) The period of development, during which these double formations may occur, lies between the time of the formation of the first segmentation groove and the completion of the blastocyst stage; for the separate double monsters a very early, and for the asymmetrical monsters a somewhat later, period of development may be admitted. As regards[1] the question of their formal genesis, we can, according to Schwalbe, only say that a division of the egg-substance must be assumed to occur. Beyond this general fact, the various forms must each be considered separately as regards their exact mode of origin.

Recently, P. Strassmann has examined the question very closely. The period of the development of double formations is, according to his conclusions, in the very earliest stages of segmentation. As regards the causation of double monsters and also of uniovular twins, he

[1] The fact that double monsters possess a single amnion proves that they must be formed at a very early period of development: namely, at a time earlier than the age of the youngest described human ovum—the Teacher-Bryce ovum, estimated to be about thirteen to fourteen days old, in which the amnion is already fully formed, although the embryo is still in the trilaminar stage.

There is no evidence that the single amnion in these cases is derived from the union of two originally separate amnions. Even if the origin of the double monster takes place at a later period than this, it must still occur before the completion of the blastocyst stage, since each individual may contain organs formed from all the three layers of the blastocyst.

considers that the entrance of many spermatozoa[1] will not account for it, any more than a division of the segmentation globule will account for the development of uniovular twins. The occurrence of ova with two nuclei is also very doubtful.[2]

Strassmann says further: 'In all probability the two embryonic rudiments are not completely separated originally, but remain united to one another by an area of germ material, and, according to their mode of development and the possibility of their further union with

[1] The view that polyspermia may lead to the formation of double monsters appears to be contradicted by the observations of Hertwig, Sobotta, and others. By treating the ova of some animals with certain chemical substances it is possible to bring about their penetration by several spermatozoa at the same time. This appears, as a general rule, to exercise a most deleterious effect upon the ovum, and to result in the production of many abnormal forms of cell division, but not in anything resembling the formation of a double monster. Bovert points out that in such a case the development does not proceed further than the blastocyst stage. When two ova are allowed to fuse, and to be fertilised by a single spermatozoon, then a giant formation ensues. He lays down the rule that whenever multipolar cleavage figures appear in a cell the result is always pathological.

There are, however, a number of authorities (including Schultze, Dareste, Duval and Ballantyne) who believe that polyspermia is probably an important factor in the production of double monsters.

[2] Ova containing two germinal vesicles can undoubtedly occur in the human subject, and the observations of Franqué, Stöckel, and v. Schumacher have proved this. In the event, however, of such an ovum becoming fertilised by two spermatozoa, then four separate polar bodies must be discharged, and two separate blastomeres be formed, leading to the development of two areæ germinativæ and two chorions—a condition never seen in true uniovular twins. There is no evidence to show that the single chorion of uniovular twins ever results from the fusion of two originally separate chorions.

With regard to the exact period at which the developing ovum acquires the power of forming a double monster there is a good deal of difference of opinion. Some writers hold that this tendency is present in the normal ovum or spermatozoon; others, that it is the result of an abnormal fertilisation; while others, again, think that it is present for the first time when segmentation of the ovum begins. All the available evidence seems to show that the last is the most probable mode of origin; namely, that the conditions which lead to the development of a double monster first make their appearance after the normal fertilisation of a normal ovum. We may, then, accept the view of Spemann that double monsters are formed by a division of the substance of the ovum, and that this may occur at any period of development, from the appearance of the first segmentation groove up to the stage of the complete formation of the blastocyst.

In favour of this view may be mentioned the extreme similarity which exists between uniovular twins and double monsters, and the fact that it is possible in various ways to bring about the formation of double monsters in the normally fertilised eggs of animals at different stages of their development.

Double monsters undoubtedly owe their origin to the presence of two embryonic rudiments on one blastocyst. Their external form and mode of union must depend upon the original position and the convergence or divergence of the axes of the two resulting primitive streaks.

one another, they form equal, unequal, or parasitic double monsters. The possibility of their developing separately, or partly united, depends—leaving out of account a portion of germ plasm lying latent in the embryo and capable of separate development—upon the relation of the axes of the embryonic rudiments, and their proximity to one another at their dorsal, ventral, lateral, or cranial portions. Even a position of the embryonic areas, originally close together, may be altered at a very early period into a different situation, which will ultimately determine the mode and variety of their union.'

For incomplete cases of doubling, this author is also inclined to invoke a double embryonic rudiment, except that the 'doubling' only appears at a later stage and less completely. Other authors regard the doubling as the result of division of a single embryonic area—especially in the case of anterior doubling.[1]

Our knowledge of the ultimate causes of double monsters is still of a very hypothetical nature (see Ziegler and Schwalbe's remarks on the experimental results). From clinical and experimental conclusions, traumata of various kinds are probably of most importance. Whether physical, chemical, and osmotic conditions—which certainly play a part in experiments on animals—can also have an influence on man, is more doubtful.

As regards the union of double monsters, a junction of similar parts always occurs (skull with skull, pelvis with pelvis, liver with liver, brain with brain, lens with lens, sclerotic with sclerotic). Geoffroy St. Hilaire has termed this the 'Loi d'affinité de soi pour soi.' According to Strassmann, the use of the term 'coalescence' or 'union' may lead to confusion, as it is usually employed for the union of portions of the body in the act of healing a wound, or the formation of adhesions; while double monsters are always united by a more intimate connection. For this reason, he thinks, the condition is better termed an 'incomplete separation,' or, in a sense, a 'flowing together' of germs originally double. He shows this very clearly in cases of double arm (Symbrachius). The hypothesis of Geoffroy St. Hilaire (mentioned above) is rejected by Ahlfeld, as he assumes that Hilaire meant thereby a power of attraction of similar organs for one another, which, as Ahlfeld rightly points out, does not exist. Schwalbe, therefore, correctly says that this statement is often misunderstood, and that it is rather to be regarded as the expression of a matter of experience, if the union of double monsters occurs as is

[1] Marchand, for instance, regards an incomplete anterior doubling as the result of dichotomy of an originally single rudiment.

explained above. The rule has,[1] further—although rarely—some exceptions (see Schwalbe).

With regard to the size of double monsters—especially of the symmetrical variety—that of the two individual parts of the malformation is almost always below the normal size of a healthy fœtus of the same age, as is indeed often the case in uniovular twins. This fact is of great importance as regards the prognosis and the course of labour, as we shall learn more fully in considering the obstetrical importance of these malformations. However, not infrequently it happens that both individual parts attain the size of a normal fœtus. Thus Schwalbe found in a case of ileo-xiphopagus, the length of each of the fœtuses to be over 55·5 cm.—a measurement which is much above the normal. In a case, too, of sternopagus (see p. 343), each individual had almost the normal length and weight.

The umbilicus may be single or double—monomphalous or diomphalous double monsters. A single umbilicus is found, as a rule, when the thorax and abdomen are united; and when it is present, the lower half of the body is single. A double umbilicus, on the other hand, is found in fœtuses united by the backs, pelves, or heads. With a single navel, there is one cord; with a double navel, two cords. Even with a double navel the cord, however, may be—at least, partially—common to both: a dichotomously divided cord. Relatively frequently, a double cord has a single insertion into the placenta—especially in pygopagus. The single cord, with a single navel, usually contains four arteries and four veins; but the vessels may be united with one another.[2]

A question of great interest is how far these united beings are dependent upon one another mentally and physically. I will limit myself to definite facts only, and must refer the reader for further details to the very interesting observations of Schwalbe and Strassmann. The double monsters which have survived have in several instances been carefully investigated from this point of view, and it has been found that the question must be answered in different ways, depending upon whether the double monster is composed of two equally or unequally developed bodies. With equal development of the two halves, the fœtuses may be considered as two separate individuals—as, for example, in the case of the Siamese twins—and are quite independent of one another both physically and mentally. The case (more fully described on p. 310) of the pygopagi, 'Rosa' and 'Josefa,'

[1] This law, in the case of double formations, means that the arrangement of the two primitive streaks, and the embryos resulting therefrom, is such that the two bodies cannot do otherwise than come into contact by similar portions of each; and if union takes place, such union will affect, therefore, similar organs in each.

[2] There is often only one umbilical artery—a condition which Schatz regards as the least-developed form of a hemiacardius. The amnion may be entirely absent.

demonstrates the same thing. This case proves also that the independence of the two is not complete. Strassmann rightly points out that the independence of the functions of the two individuals, united in a double monster, depends primarily upon the degree of union of their nervous systems, and, secondarily, upon that of their circulatory systems. Naturally, the conditions are quite otherwise in the parasitic double monsters. Here, as is shown by the nomenclature, the parasite is primarily dependent upon the autosite for its nutrition. As regards signs of life, there appears to be at times some slight independence of both fœtuses. Schwalbe, for example, examined a case of epigastrius parasiticus, in which the parasite was removed by operation, and it showed no independent movements. One knee of the parasite was inflamed and suppurating,[1] and when this was touched the autosite cried and made movements of protecting itself, so that some union of the nervous systems of the two individuals must be assumed. In the very low forms of parasitic double formation—the teratomata—naturally, there can be no question of the presence of definite physiological signs of life.

Complete observations on the psychical and bodily relations in double monsters have been carried out recently by Hemeberg and Stelzner. They had the opportunity of examining carefully the pygopagi, 'Rosa' and 'Josefa,' the so-called Bohemian sisters.[2] I must refer the reader to the original work. I can only touch here on some interesting points. Heredity was not present in this case, for the mother had borne four other healthy children. The delivery occurred at the normal end of pregnancy, and took place very easily—in five minutes—in such a way that the head of Rosa was born first, then the two pairs of lower limbs, and finally the trunk and head of Josefa.[3] Two placentæ and two umbilical cords were alleged to be present. For the first month the children were fed on the bottle; then the mother's milk became established, and the children were suckled by her for two years. In one year they learned to walk, in two years to speak. As school-children, they were capable of such mobility that they could climb plum-trees. In their fourth year they both suffered from a somewhat obscure infectious disease. In their twelfth year, Rosa suffered from diphtheria with high fever and delirium, but Josefa remained unaffected. During an attack of chorea—from which Josefa suffered alone—the menses ceased in both; but since their fourteenth year

[1] In this case the temperature of the autosite was markedly raised.

[2] For two interesting photographs of the Bohemian sisters the reader should consult Gould and Pyle, *Anomalies and Curiosities of Medicine* (1897), pp. 180, 181.

[3] This is the usual mode of delivery in pygopagous monsters; and the fact that labour as a rule takes place easily is probably the reason why this form of monster so often survives.

menstruation had been regular in both of them. In other double monsters menstruation has usually occurred at different times, as has also defæcation and micturition. The pains associated with the menses were experienced sometimes by Rosa, sometimes by Josefa. Later, Rosa suffered from an attack of intestinal catarrh, accompanied by abdominal pain, loss of appetite, and diarrhœa. Between the attacks of diarrhœa, formed stools were passed from the healthy intestine of Josefa. Their physical condition was as follows: A common anterior and posterior surface could be distinguished without difficulty, and, when they stood up, the anterior surface was turned to the observer; Rosa (144 cm. high) was placed on the right, Josefa (142 cm. high), was on the left. Together they weighed 85 km. (287 lb.). The right leg of Rosa and the left of Josefa were anterior and internal, and the left leg of Rosa and the right of Josefa posterior and external. If the twins were regarded from behind, the spinal columns appeared to bend towards one another at an angle of about forty-five degrees. The middle portion of the thoracic vertebræ of the spinal column of Josefa showed a marked lateral curvature towards the right, while that of Rosa pursued a straight course. The spinous processes could be palpated downwards as far as the spinous process of the seventh or eighth thoracic vertebra. Just about this level the spinal column begins to assume a horizontal direction and blends into a strong bridge, the middle of which is situated some 92 cm. (when the twins are standing) above the level of the floor. The connecting bridge of tissue is about 23 cm. long, its thickness—that is to say, the circumference of that portion which would have to be divided if one wished to separate the twins—measures 94 cm. Palpation of the dorsal surface of the point of union does not permit of any certain decision being arrived at as to the configuration of the bones. Only a single—apparently normally formed—coccyx can be felt through the skin. Regarded from behind, the two posterior and external thighs (the right of Rosa and the left of Josefa) form buttocks of apparently normal configuration, on which the twins sit and lie without inconvenience. They use, too, the ordinary closet. Both pelves appear to have a normal inclination, although the outer half of each is placed at a lower level than the inner half. The examination of the genital organs showed the following condition: If the two median limbs are raised up, with the twins in the dorsal position, in the space between the anterior median and the posterior external pair of limbs is a vulva some 14 cm. in length. If the vulva is opened up, it appears as an isosceles triangle bounded by three large labia majora. The anterior border of this is formed by a single labium about 14 cm. long and covered with hair; while the two lateral labia, which meet posteriorly, are about 10 cm. in length. Besides these, there are two small labia minora, 4 cm. long, which pass forwards to the clitoris.

In addition to a double urethra, double vulvar orifice, double vagina, and completely separated uteri, there is a single perineum, anus, and rectum.

With regard to the movements of the body, the twins are able to use their feet in progression in different ways. Most commonly, progress takes place so that the right or the left feet are put forward together.[1] In this way they can move tolerably quickly, and mount stairs. Auscultation and percussion gave normal results, and situs inversus was not present, as is so often the case in thoracopagi. The colour of the iris and of the hair is the same in both. The sensibility is normal, and only a small zone of common sensibility is present in the region of the union. The sensibility of the anus, the introitus vaginæ, urethral orifice, and of the clitoris is in common. Both twins feel the desire to go to stool at the same time, and also the passage of the fæces and of the urine. When one of the twins drinks more than the other, she has a more frequent desire to pass urine. The frequency of the pulse-rate is always different. Going to sleep and waking up do not occur at the the same time. Their appetite varies, as does their liking for definite articles of food. Differences of opinion often occur and quarrels result, which in former years not infrequently degenerated into a bout of fisticuffs. The fact, communicated to them by other persons, that the death of one must be followed by that of the other, has caused them much mental distress. Concordance of dreams and transference of thoughts is not present.[2]

The two authors give, further, a résumé of the literature of Pygopagi.

[1] Not infrequently, however, one of the sisters walked forwards, dragging the other after her walking backwards.

[2] On April 15, 1910, the sisters Rosa and Josefa Blazek were admitted to the surgical clinic of the hospital at Prague. On their admission it was found that the abdomen of Rosa was considerably enlarged, while that of Josefa showed no change. The possibility of pregnancy was strenuously denied; but on April 17, Rosa was delivered of a healthy male child, which survived. On the twelfth day of the puerperium, Professor Pitha made an examination, and found the following condition: The perineum was somewhat torn, and the septum—which originally separated the two vaginæ almost in the whole of their extent—had been extensively torn away from its attachments. The vagina of Josefa still presented the characters of that of a nullipara, while that of Rosa those of the vagina of a woman shortly after her confinement. The uterus of Rosa was enlarged and corresponded to the size of the uterus at this period of the puerperium; that of Josefa was normal in size, anteverted and flexed. It was impossible to determine the exact measurements of the pelves, partly because of the shortness of the vaginæ and partly because of the resistance of the abdominal walls. The breasts of both were enlarged and both were secreting milk.

The labour appears to have taken place rapidly and easily; but as no medical man was present the mode of delivery and presentation of the child is uncertain.

During the first days of the puerperium the temperature and pulse-rate of the two sisters was different—Rosa showing some slight elevation of temperature. During the whole of the pregnancy of Rosa, and up to eight weeks before her confinment, Josefa continued to menstruate regularly.

For a discussion of the civil rights of double monsters, see Chapter XXII. Other questions, which in earlier days were of considerable importance, no longer possess any interest. In 1770, for example, Werther (quoted by Hennebürg and Stelzner) discussed at great length whether at the Day of Resurrection double monsters would appear separated or united, and whether there existed a monstrosity of the soul as well as of the body.

Some general pathological and physiological questions are of interest. In disease associated with fever, the twin affected may have its temperature raised by several degrees—three degrees or more. This was observed, for example, in the case of the celebrated Hindu xiphopagus, 'Doodica' and 'Radica,' who were separated by Doyen in 1902. Doodica suffered from tuberculous peritonitis with high fever, while the temperature of her sister remained normal. The same condition occurred in the Brazilian xiphopagus, 'Rosalina' and 'Maria.' Rosalina had an attack of influenza with a temperature of 104·2°, while the other child remained quite unaffected.

These cases do not support the assumption that the occurrence of fever is to be attributed to changes in the blood. Experiments with iodide of potassium, methylene blue, and sodium salicylate have shown that the interchange of fluids between the two bodies is regular and rapid, and a substance introduced into one twin quickly appears in the urine of the other. Hennebürg and Stelzner have shown also that when the mouth and nose of one of pygopagus twins is closed, restlessness and attempts to get free occur in it after some twenty seconds; while the second twin continues to have a calm unaltered respiration. Death in cases of double monsters occurs in each at the same time or shortly after one another; an interval of some hours has, however, been observed. In all probability death ensues from the transmission of poisonous bodies, shortly before death, from the dying to the sound twin.

As regards the viability of double monsters it is clear that life is impossible in severe malformations of the brain, or of the internal organs—especially the thoracic viscera. Such children die *in utero* or soon after birth. This result is often promoted by the presence of other severe malformations—such as hemicephalus, hydrocephalus, and umbilical herniæ. According to Schwalbe, in the symmetrical double monsters, the best prognosis is in those in whom the two individual members are united by a similar part of the body of the smallest possible size—as in xiphopagi and pygopagi. In asymmetrical double monsters, the prognosis for the autosite depends on the size and the position of the parasite (see epignathus and sacral parasites). It is clear, then, that it is mainly cases of sternopagus, ischiopagus, dicephalus, xiphopagus, and pygopagus which may survive and even

attain an advanced age. The great majority of these unfortunate individuals gain an existence—precarious, but certainly profitable—from their condition, in show-booths, or frequent the Universities and centres of population, and exhibit themselves to the students, and at medical societies.

If the two individuals possess extensive portions of the body in common, it is impossible for them to survive. To this class belong, for instance, the cephalo-thoracopagi (see p. 319). In some cases, the separation of the individuals has been attempted—in part, at least—always with the same unfortunate result (with one exception, the case of König). In the majority of the cases, any attempt at separation of the individuals is not desired—certainly, not by their relations—as if this be done the means of their making a livelihood will be lost at the same time. Still, in some cases, separation is indicated on account of an incurable disease in one of the two. An absolute indication for separation is furnished by the death of one individual, which will certainly very quickly affect the still sound individual. Schwalbe appropriately compares these indications with those for Cæsarean section in the dead or dying mother. In former years the separation was attempted by the application of a ligature, tightened from day to day. At the present day surgical methods of separation are alone to be considered. The conditions are most favourable in those xiphopagi in whom there is present only a bridge of skin and cartilage. Equally good is the prognosis in those cases in which only a thin bridge of liver substance common to both is present, which can be easily divided with a Paquelin's cautery. In cases of sternopagi there is usually present such a degree of union of the important organs (heart and intestine) that any operation for their separation is not to be thought of. So far as I can gather from the literature, the separation of symmetrical double monsters has been attempted on five occasions (cases of König,[1] Böhm,[2] Chapot-Prévost,[3]

[1] In König's case two Swiss sisters, born in 1689, and united belly to belly, were separated by means of a ligature drawn tighter from day to day, and the operation afterwards completed by a knife. Both children are said to have lived—the only recorded case in which such a result followed. The connecting band of tissue consisted, however, only of skin and cartilage.

[2] In 1866, Böhm related a case of a xiphopagus in which the connecting bridge of skin containing some cartilage was divided and the wound sutured. One of the children, feeble at the time of the operation, died three days afterwards; the other survived and was alive and well at the age of five years.

[3] How difficult any such operation may be is illustrated by the case of 'Rosalina' and 'Maria'—the thoracopagus operated upon by Chapot-Prévost in May 1900. The cavities of the pleura, pericardium, and peritoneum all communicated, and during their separation the pleura of Maria was opened so that a pneumothorax developed, into which blood escaped from a wounded vessel. Maria died on the eighth day after the operation, but Rosalina survived.

Doyen,[1] and a case from Switzerland[2] with the name of the surgeon not given (see Strassmann and Schwalbe).

The Chief Varieties of Double Monsters

A. United Equally Developed Twins—Symmetrical Double Monsters

1. *Duplicitas anterior.* That is, anterior doubling, with union of the posterior parts of the body (see Ziegler).

Pygopagus

In this very rare malformation, the twins are united in the pelvic region (sacrum and coccyx). The two bodies are usually placed side by side. According to Strassmann, there are two spinal columns; but nearly always only one sacrum and one coccyx. The genital organs and the termination of the intestines have usually each a common opening. The spinal cords are united below, and the aortæ and venæ cavæ generally communicate. As has been already mentioned, many malformations of this kind are capable of surviving.[3]

Ischiopagus

The fœtuses are united to one another in the region of the pelvis in such a manner that the axes of the bodies form an acute or obtuse angle. The sacra are opposed to one another; there is only one umbilicus; the cavity of the pelvis is common to both; and the anus, the

[1] In February 1902, Doyen operated upon the celebrated xiphopagus, 'Radica' and 'Doodica.' The operation was undertaken because Doodica was suffering from tuberculous peritonitis. The conjoined ensiform cartilages were easily separated with a bistoury. The peritoneal cavities were opened, the connecting bridge of liver exposed and crushed with a special pair of compression forceps. The bridge of tissue on either side was ligatured with catgut and then carefully divided. Six days later, Doodica died suddenly from the rupture into the peritoneal cavity of a tuberculous abscess situated in the pelvis. Radica rapidly gained in weight after the operation, and her general health much improved.

[2] In 1881, Blaudet and Bugnion attempted to separate the conjoined sisters 'Marie' and 'Adele,' born in Switzerland on June 26. The operation was undertaken on October 9; but Adele died six hours afterwards, and Marie, on the next day, of peritonitis.

[3] A pygopagus, no doubt, results from the union at their posterior extremities of two embryonic rudiments situated on one blastocyst. At the site of the union, the parts involved are to some extent stunted in their further development; while the other portions of the embryonic rudiments develop normally. The yolk-sac, originally single, ultimately becomes divided.

In considering the feasibility of operating upon such cases it must be remembered that probably the two spinal canals and peritoneal cavities would be opened, and that there are usually large anastomosing vessels between the two circulatory systems.

end of the intestine, and the genitalia are single or double. The lower extremities are present, twofold or fourfold. According to Strassmann, these relatively frequent malformations seldom live, and no such monster has attained the age of one year.[1] Schönbek published, a short time ago, a very interesting case of Ischiopagus parasiticus with dystocia. The difficulty arose from the fact that the extremities arising from the trunk at a right angle become extended above the pelvic brim. The mother suffered from eclampsia.[2]

Dicephalus

In this case there are two separate heads, and the doubling may also affect the neck (Dicephalus dianchenos, in contrast to dicephalus monanchenos), the thorax, and the trunk. The division of the spinal column is best shown by means of an X-ray photograph (see Fig. 49).[3]

In these malformations, union of the internal organs often occurs. Dicephalus tetrapus, D. tripus, D. dipus, D. tetrabrachius, D. tribrachius,

[1] The 'Jones Twins' lived, apparently, for one year and eight months. Their weight at birth was twelve pounds, and their length twenty-two inches. Lying on their mother's back they could both be nursed at the same time. They were born in Indiana, June 24, 1889, and died on February 19 and 20, 1891 (Huff).

[2] The mode of conjunction in a case of ischiopagus will be realised best if the reader assumes that the two fœtuses have each been divided by a sagittal incision extending from the anus to the umbilicus and including the symphysis pubis, and that then the bodies have become fused in such a way that the right and left halves of the divided and separated tissues of the one fœtus have become united to the left and right halves of the tissues of the second fœtus. As a result of this, the right half of the symphysis of one will be joined to the left half of the symphysis of the other, and the two sacra will be placed opposite to one another. If the incision involves the anus and the umbilicus, then the anus and umbilicus should be common to both—and this is usually so; and if we imagine the incision extending back as far as the sacrum, then the sacra would be united at their lower extremities —a not uncommon condition in these monsters.

The genital organs are composed of a half from each fœtus, and, as a result of the manner in which the bodies are united, the lower limbs are situated at right angles to the long axes of the trunks, which are usually placed in the same straight line. It is interesting to note that cases of ischiopagus are very rare indeed among the lower animals, and it is doubtful if any true example of this condition has been met with. It is obvious that no question of the separation of the two halves of such a monster could arise.

[3] Fig. 49 shows very well what is usually noted in these cases: namely, that the division of the bodies is much more marked than might be assumed from the external appearances.

Cases of anterior doubling are extremely common in fish embryos; and experiments have shown that such malformations can be produced by trauma. In the majority of cases the condition is brought about by the fusion of two separate embryonic areas; but in a certain number it may, undoubtedly, be due to the division of an originally single embryonic rudiment, and this may occur possibly even at a latter period of development than the blastocyst stage.

and D. dibrachius may be distinguished (see Fig. 50). In certain conditions, dicephali are capable of living (see Ahlfeld).[1]

Diprosopus

In this case there is union or doubling in the cephalic region without complete separation of the heads, only the face being more or less doubled. The cavity of the skull is single or double. The malformation is one which approaches very nearly to that of dicephalus. The various forms of Diprosopus triophthalmus, D. tetrophthalmus, D. diophthalmus, D. tetrotus, D. triotus, D. diotus, D. distomus, D. monostomus, D. tribrachius, and D. dibrachius may be distinguished. In many cases an encephalus or hydrocephalus have been observed at the same time (see Figs. 51, 52). Such fœtuses, on account of the extreme malformations of the brain and pharyngeal cavity, are not capable of living (Ahlfeld).

Fig. 49.—Radiogram of Dicephalus Dibrachius Dipus.

In rare cases, in the least degree of duplicitas anterior, merely a doubling of the jaw, mouth, and nose is present (Ziegler).[2]

[1] The best-known example of this variety of monster was 'Ritta' and 'Christina,' born in Sardinia on March 13, 1829. These twins, of female sex, had four upper and two lower limbs (Dicephalus tetrabrachius dipus). The vertebral columns, separate above, were joined below by a rudimentary os innominatum, and there was also a union of the two manubria. Other celebrated instances were the 'Scottish Brothers,' who lived for twenty-eight years; the 'Tocchi Brothers,' and 'Marie and Rosa Dronin.'

[2] All gradations may be met with between this class and the preceding one: namely, from the variety mentioned here of diprosopus diophthalmus to that in which the upper half of the body is entirely doubled.

2. *Duplicitas posterior.* The whole trunk is doubled, and the twins are united by the cranial portion of the head.

Craniopagus

According to the place of union, the condition is called craniopagus parietalis (for illustration, see Ziegler), craniopagus frontalis (Küstner), craniopagus occipitalis (Schwalbe). The brains are usually separate from one another; but in cases of extensive union, coalescence of parts of the brain may also occur.[1] As a rule, the fœtal bodies are so placed that the angle made by the two fœtuses —in craniopagus occipitalis, for example—is generally, or very often, an acute one. The union does not always affect the corresponding parts (bones of the skull); so that here there is an exception to the above-mentioned 'Law' of Geoffroy St. Hilaire. In many cases it can also be seen that the abdomen of the one child is turned to one side, and that of the other child to the other side. The length of life of a craniopagus is generally short, but it has lasted as long as ten years.

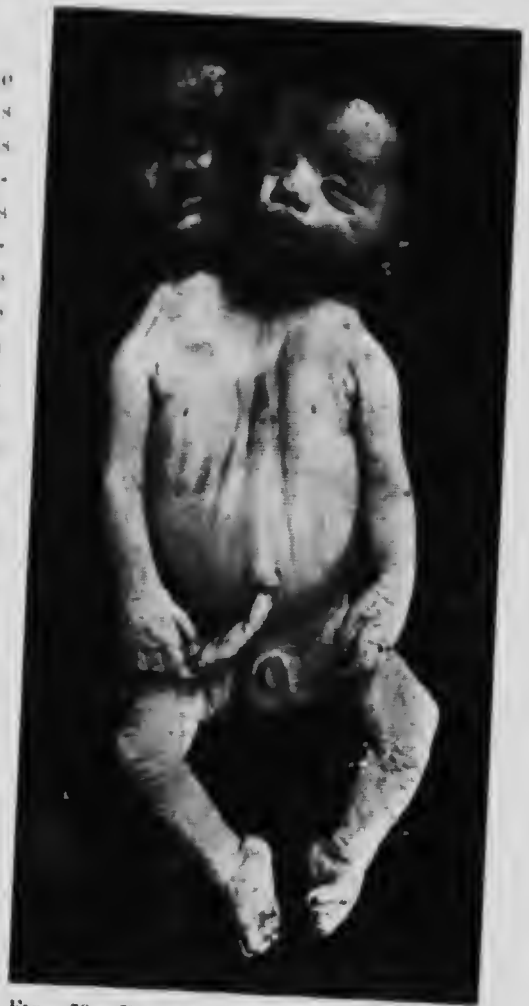


FIG. 50.—DICEPHALUS DIBRACHIUS DIPUS. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] The union is usually quite a superficial one, affecting only the bones of the skull. All these forms of monster are excessively rare.

The explanation of the formation of these monsters is no doubt to be found in the union at their cephalic extremities of two originally separate embryonic rudiments. We may conclude that this takes place at a comparatively late period of development, from the fact that the cephalic pole is developed subsequently to the complete formation of the primitive streak.

Any operative interference can [illegible] be of evail when, as is usually the case, the brains are separate.[1]

The parasitic form of this malformation is termed epencranius; but it is also known as craniopagus asitieus.[2] Cases of cephalopagus also occur in which while one individual is well developed, only portions of the other are present

.—Dicephalus with Hemicephalus. (Specimen from the collection of the Göttingen Women's Clinic.)

[1] In the historical case described by Munster, two female children, born in 1495 and joined by the foreheads, lived to be ten years old. After the death of one, an attempt was made to separate the other; but the skull was opened and the meninges injured at the operation, and she soon succumbed.

[2] The most celebrated case of this kind is a specimen, No. 166, in the Teratological series in the Museum of the Royal College of Surgeons England. It consists of the skull of a Bengalee child with a second imperfect skull attached to the anterior fontanelle, which is widely extended. The base of the second skull is much contracted, all the bones of the face are much smaller than those of the supporting skull. The faces are turned almost in opposite directions, so that the left frontal bones of one skull articulate by suture with the right parietals of the other, and vice versa. The child, a male, died at the age of four years from the bite of a cobra. The brains were distinct, but the dura mater of each adhered to that of the other at the point of contact. The movements of the features of the upper half were purely reflex; the movements of its eyes did not correspond with those of the child, and the eyelids were usually open, even during sleep.

generally, the head and trunk, ... ly the head (see Strassmann,[1] and the remarks of Ahlfeld).

Cephalothoracopagus, Syncephalus, Janus, and Janiceps

... these cases there is present a union of the forehead, face, and s... m, as well as of the internal organs of the neck, thorax (less commonly, there are two separate thoracic cavities), and, at times, also of the abdomen.[2] The large brain is usually single, the medulla oblongata, the cerebellum, the pons and corpora q... l... onita are double (for the creations of the bones of the skull, of the brain, and of the internal organs, see the instructive illustrations of Schwalbe). The union is such that the one half belongs to the one twin; the other half to the second twin. Two main types may be distinguished—the rat... cephalus (Janus) symmetros (according

FIG. 52. DIPROSOPUS DISTOM... TE... TH... THALMUS DIOTUS, WITH HY... CEPHALO... AND HARE-LIP. (Specime... col... on of the Göttingen Wome... Clinic.)

[1] Strassmann records the case of a craniopagus of the fifth month, to the head of which a portion of a ...ood fœtus—consisting of the ...ead, a part of the trunk, and the right arm—was attached, so that its occiput looked towards the forehead of the first. He explains the condition in the following way: Originally there was present a double monster united by the head. One of the two fœtuses was weaker than the other, and its cord correspondingly thinner. During its growth, the first twin, by its movements, brought about the separation of the cord of the second, causing its death and finally a rotation of the point of attachment.

[2] There is usually ... single œsophagus, stomach, and intestine as far as the attachment of the omphalo-mesenteric duct: beyond this, the intestin... is double. All the other thoracic and abdominal organs are double in a typical case.

to Schwalbe, better termed Cephalothoracopagus dysymmetros), and the much more frequent Janus (Janiceps) asymmetros. In the first form there are present two well-formed faces—an anterior

FIG. 53.—CEPHALOTHORACOPAGUS ASYMMETROS. (The opposite side of the fœtus shown in Plate VI.)

and a posterior; the two fœtuses being in contact with one another—breast to breast, face to face. Each fœtus has its own separate occiput.[1] In the second asymmetrical form (better termed, according

[1] To understand the mode of union in a cephalothoracopagus, the reader may assume two symmetrical fœtuses divided by a sagittal incision, from the umbilicus upwards, extending back to just in front of the spine, and to the sella turcica in the skull; and the two fœtuses then becoming fused in such a way that the right half of the divided portion of the body of the first becomes joined to the left half of the divided portion of the body of the second, and vice versâ. In this way a cephalo-

PLATE VI

CEPHALOTHORACOPAGUS ASYMMETROS

(Specimen No. [illegible] University College Hospital Medical School Museum Obstet. Sect.
From a case under the care of the late Dr. G. K. Batten.)

to Schwalbe, Cephalothoracopagus monosymmetros or C. asymmetros), there is only one well-developed face. Not infrequently in this malformation a condition of cyclopia (see Ziegler), synotia, and closure of

Fig. 54.—Radiogram of a Cephalothoracopagus Asymmetros.

the oral cavity is present in the second imperfect face. Röntgen rays readily show the development of the face from the right and

thoracopagus disymmetros, with two posterior surfaces and two anterior surfaces, will result; the two symmetrical faces formed by the union of the left and the right halves respectively of the faces of the original fœtuses. If we further assume that the right and left halves which go to form one of the anterior surfaces are not so well developed as those of the other anterior surface, then we shall have the condition known as Cephalothoracopagus monosymmetros or C. asymmetros, in which the second face may be almost entirely undeveloped, and the second anterior surface of the upper part of the body only imperfectly represented.

It is an interesting fact that in some cases of cephalothoracopagus the craniopharyngeal canal, in which originally the stalk of the stomadæal portion of the pituitary body lies, remains open.

left halves of the two fœtuses (Strassmann).[1] Such malformations are generally born dead, or die soon after birth. In rare cases, a Janus parasiticus has been described (see Ahlfeld).[2]

Dipygus.

In this very rare malformation there is a doubling of the lower extremities and of the lower part of the body. The upper portions are generally single, rarely showing an incomplete fissuring. Dipygus dibrachius and dipygus tetrabrachius may be distinguished. The doubling of the spinal cord may commence at varying levels. These malformations have been often confused with examples of prosopo-thoracopagus (Schwalbe).[3] Dipygus parasiticus is more frequently met with; in this case the autosite may remain alive.[4]

3. *Duplicitas parallela.* In this condition there is a doubling of the whole trunk, or a doubling of the anterior or posterior extremities of the body, so that they are placed parallel to one another. The union and fusion of the fœtuses is in the region of the thorax and, in many cases, also in that of the abdomen.

[1] If, as we may assume, the formation of this form of monster is due to the fusion of two originally separate embryonic rudiments, then the union must take place at a time before the two cardiac tubes unite to form the primitive heart. Instead of these two tubes fusing with one another—as they normally should—in this malformation we must suppose that they fuse with the corresponding tubes of the other embryonic plate.

[2] Specimen No. 838, Univ. Coll. Hosp. Med. School Museum—a female monster with fused heads and chests, and two symmetrical faces—is a good example of a cephalothoracopagus disymmetros; and specimen No. 839 (see Plate VI and Figs. 53 and 54) is a good example of the asymmetrical variety. Specimens Nos. 844, 845 and 846 are also examples of cephalothoracopagus asymmetros in the pig of varying degrees—the last showing on the side of the less-developed face synotia, cyclopia, and a well-marked proboscis formation.

[3] This variety of malformation occurs not infrequently in fishes, and has been produced experimentally in them by Kopsch. By means of the electric current, he has succeeded in producing a division of the posterior part of the body in the young embryo of the trout, twenty-four hours before the appearance of the head. Each of the two separated halves developed a medullary tube, and a number of myotomes and lateral plates. It is obvious, then, that in the formation of a dipygus we must consider not only the possibility of the partial fusion of two embryonic rudiments but also—so far as experimental results go—the division of an originally single embryonic rudiment.

[4] The parasitic forms of Duplicitas posterior are much commoner than the symmetrically developed forms; whereas in the case of Duplicitas anterior just the opposite condition obtains.

Thoracopagus

This is the most frequent of the symmetrical double monsters occurring in Man.[1]

The bodies are united only by the thorax, and are placed

FIG. 55.—THORACOPAGUS TRIBRACHIUS TRIPUS. The third common lower extremity is stunted and ankylosed. (Specimen from the collection of the Göttingen Women's Clinic.)

parallel to one another. According to the position and extent of the union, cases of sternopagus may be distinguished in which the bodies are united together mainly by the

[1] In 110 cases of double monsters, recorded by Taruffi, seventy-one were instances of thoracopagus.

sternum.[1] In a thoracopagus, the thoracic cavity, as a rule, is common to both fœtuses; as is also the diaphragm. The hearts may be separated, and are then often malformed; or may be united. Fusions,[2] too, are very frequent in the intestine—as in the case of a sternopagus occurring in the Göttingen Women's Clinic, and described by v. Oynhausen. It is generally very difficult to draw a sharp line of distinction between cases of thoracopagus and sternopagus.[3]

In the case of a xiphopagus, there is most commonly only a band-like union in the region of the xiphoid process of the sternum. The bridge consists of skin, cartilage, vessels, and (rarely) also a portion of liver. To this group belong the well-known Siamese twins. (For a full history of their life, as well as the results of the autopsy, see Ahlfeld and Schwalbe.) Xiphopagi, and less commonly sternopagi, are, judging by these examples, capable of living; and separation by surgical measures has, in such forms, the greatest chance of success. (For an account of the separation of the sternopagus, 'Maria' and 'Rosalina,' by Chapot-Prévost, and for the operation carried out by Doyen, in the case of the xiphopagus, 'Radica' and 'Doodica,' see note 1, p. 314.) According to the number of the upper and lower extremities present Thoracopagus tetrabrachius (see Plate VII), T. tribrachius, T. dibrachius, T. tetrapus, T. tripus, and T. dipus may be distinguished.

Prosopo-thoracopagus

In these cases, besides the thorax and abdomen, the head and neck are also united.[4] This variety of malformation is very rare in Man (see

[1] In the true thoracopagus two sterna are present—one on either side; each made up of the right and left halves respectively of the sterna of the two fœtuses. In the sternopagus, in which usually only the lower halves of the sterna are united, the portion making up the right half of the conjoined portion of the sternum belongs to one fœtus, and that the left half belongs to the other fœtus.

[2] When the hearts are double, each heart belongs to one of the fœtuses, and they are not made up—as in a cephalothoracopagus—by the separate union of the two primitive heart-tubes of the two fœtuses with one another. When double, they are, however, often joined by a band of tissue.

The lungs are separate, but the two diaphragms are united—as are also the livers, in some cases. The intestinal tracts are often single over an area extending from the end of the duodenum to the site of origin of the omphalo-mesenteric duct.

[3] If the account given above of the development of the double heart of a thoracopagus is correct, then we may assume that the union of the two embryonic rudiments to form such a malformation with two hearts takes place at a later period of development than when only a single heart is present; and, further, we may conclude that the union of the two embryonic rudiments to form a thoracopagus takes place at an earlier period of development than when a sternopagus is formed, and this in its turn at an earlier period than when a xiphopagus results.

[4] This form of monster forms a type intermediate between the thoracopagus and the cephalo-thoracopagus. If we assume a union of the two embryonic rudiments at an earlier period than in the case of a thoracopagus, and a partial fusion of the two heads and necks, then we have the type of a so-called prosopo-thoracopagus.

PLATE VII.

THORACOPAGUS TETRABRACHIUS TETRAPUS.

(Specimen from the collection of the Göttingen Women's Clinic.)

Schwalbe). (For Thoracopagus parasiticus, Epigastrius, Inclusio fœtalis, see p. 326.)

Rachipagus

This is a malformation which, according to Schwalbe, has only once been observed in Man, but which has been often described in the sheep. In it the thoracic and lumbar portions of the spinal column are united.

B. United Unequally Developed Twins—Asymmetrical Double Monsters

All these malformations may be observed in cases of Duplicitas anterior, Duplicitas posterior, and Duplicitas parallela. (For the meaning of the terms 'autosite' and 'parasite,' see p. 301.) Neither the terminology nor the classification of these malformations is very settled. I will only briefly mention the most important.

Dipygus parasiticus—Heteradelphus [1]

This is a malformation which might be included under the heading Duplicitas parallela. According to Schwalbe, however, infra-umbilical tumours of this kind belong in part to the class Duplicitas posterior. According to the place of insertion of the parasite on the thorax or on the epigastrium, the malformation is distinguished as thoracopagus parasiticus (Thoracomelos) or thoracopagus inæqualis; or as epigastrius or omphalopagus (when attached in the region of the umbilicus) (see Fig. 56). A union of the autosite with the parasite may also occur in the region of the breast or of the abdomen. The one twin is usually well developed, the other presents the appearance (as Strassmann correctly describes it) as if its head were sunk in the abdominal or thoracic cavity of the autosite. This burying may also take place in such a way that only the more or less well-developed anterior half [2] of the body appears to be attached to the autosite (see Schwalbe). The following useful classification of these asymmetrical malformations of the ventral surface of the abdomen is given by Schwalbe:—

(a) *Supra-umbilical Attachment of the Parasite*

1. All the principal parts of its body can be recognised in the parasite corresponding to the Hemiacardius (Acardius anceps, see p. 355):

[1] The term used by Isidore Geoffroy St. Hilaire for this condition.

[2] The cases recorded by Wirtersohn and Hesse, and figured by Schwalbe, are good examples of this kind. In each of these the parasite is so well developed that it is only the nature of the attachment between the two which demonstrates that we have to do with a true parasite and not with a case of duplicitas anterior.

Duplicitas asymmetros ventralis supraumbilicalis, cum Hemiacardio parasitico.[1]

2. Only portions of the anterior half of the body can be recognised in the parasite: Duplicitas asymmetros supraumbilicalis, cum Acardio parasitico acormo.

3. Only portions of the posterior half of the body can be recognised in the parasite: Duplicitas asymmetros supraumbilicalis, cum Acardio parasitico acephalo.[2]

4. The parasite consists of merely an unformed mass: Duplicitas asymmetros supraumbilicalis, cum Acardio parasitico amorpho.

(*b*) *Infra-umbilical Attachment of the Parasite*

In this case the same subdivisions may be recognised as in (*a*).

To this class of malformation[3] belongs the case of the Indian, 'Laloo,'[4] described by Virchow (Strassmann). In the parasite, the upper half of whose body was to some degree inverted into the autosite, the arms was absent. There were present a stunted penis and scrotum, two long arms with stunted hands, and two feet malformed in part—the toes of which moved either spontaneously or at the will of the autosite. From time to time the parasite passed a few drops of urine. At the time of the appearance of Strassmann's monograph, in 1905, Laloo was thirty-two years old. To this class belongs also the Genoese, 'Colloredo' (see Ahlfeld). Colloredo,[5] who was born in Genoa, in 1617, exhibited himself in Europe during many years for money. A xiphopagic parasite of some degree of development was attached at the ensiform process; it consisted of a head, thorax, left upper extremity, and two club hands with three fingers. Whether a heart was present or not was doubtful; movements of respiration were visible, the eyes were closed, the mouth open, and, at times, there was dribbling of saliva. The parasite took no nourishment.

Lastly, I must mention an interesting case, observed and fully described by Schwalbe. It was that of an epigastrius which was born

[1] Or Thoracopagus parasiticus, or Xiphopagus parasiticus.

[2] Or Epigastrius.

[3] Namely, Thoracopagus parasiticus or Xiphopagus parasiticus.

[4] He was born in Oudh, India, and was the second of four children. The stunted trunk of the parasite was attached to the lower right side of the sternum of the autosite by a bony pedicle, and, lower down, by a fleshy pedicle. Perspiration and elevation of temperature appeared to occur simultaneously in the autosite and the parasite.

[5] In 1642, he was said to have been again examined when in Scotland, and is accredited with being married and the father of several children—all normally developed (Gould).

spontaneously in a cephalic presentation. The well-developed autosite vomited after each meal and did not thrive. On account of suppuration of one knee the epigastric parasite was removed by operation. It consisted of the lower half of the trunk, two lower extremities, well-formed male genitalia, lumbar spinal column, sacrum and coccyx, and rudiments of the upper extremities. At the operation it was found that both peritoneal cavities were widely united, and that the abdominal organs of the parasite lay between the right and left lobes of the liver of the autosite, compressing the duodenum and displacing the stomach. The abdominal organs of the autosite and of the parasite were not connected, so that the operation was a simple one. The autosite, however, died from collapse, two hours afterwards. In the parasite, there were malformations in the intestinal tract and in the urinary system. Menge published, a short time ago, a case of an apparent epignathus removed successfully from a child twelve weeks old.[1]

FIG. 56.—THORACOPAGUS PARASITICUS. (Specimen from the collection of the Göttingen Institute of Anatomy.)

Those cases where only supernumerary extremities, or a rudimentary thorax without extremities, are present, and also those formations, known as teratomata, in which rudimentary extremities, or other parts of the body of a fœtus, are enclosed, belong to this class of malformation. Such teratomata may be situated under the skin of the abdomen or the thorax,

[1] Schwalbe mentions a case (which he regards as unique), met with by Kermauner, in which an umbilical cord was attached at one extremity to an acardius acephalus, and at its other extremity inserted into the epigastrium of a macerated three-months' fœtus.

or within the walls of the thorax or the abdomen. They are known as inclusio fœtalis subcutanea, or as inclusio mediastinalis, when they are enclosed in the anterior or posterior mediastinum (see Ahlfeld). The terms inclusio abdominalis or engastrius are used when the tumours are placed in the abdomen. Such fœtal rudiments lie either free in the abdominal cavity, between the organs, or are contained in a cyst. Some cases have been described where the autosite has recognised spontaneous movements in the abdomen (Strassmann, case of Highmore). In the majority of cases, the fœtuses are already macerated or mummified. In many instances such tumours, after a longer or shorter time, suppurate, and the abscess then bursts into the general peritoneal cavity, or into some of the hollow viscera such as the bladder or intestine. In considering the diagnosis of obscure abdominal tumours these teratomata must always be thought of. The old literature, as Ahlfeld rightly remarks, contains under these abdominal inclusions various other conditions not belonging thereto: namely, simple dermoid cysts and tubal pregnancies. Very frequently the teratomata are either retro-peritoneal, or situated between or behind the mesentery.

Similar parasitic formations are also found at the posterior extremity of the body in the coccygeal and perineal region. Thus there may be, for example, an increase in the number of the lower extremities which are attached to the anterior or posterior surface of the lower part of the trunk (see Ziegler). Such malformations are termed polymelia or pygomelus.[1] They belong to the groups, ischiopagus, pygopagus, or dipygus; hence they are termed ischiopagus parasiticus, pygopagus parasiticus, or dipygus parasiticus. The parasite alone is termed epipygus. Lastly, teratomata also occur in the region of the coccyx, which are enclosed by the skin of the autosite, and in which rudiments of fœtal organs and portions of the skeleton are found. (For further details, see under Coccygeal tumours, p. 251.)

Epignathus

In the cephalic region, too, parasitic double formations are observed. The parasite most frequently springs from the base of the skull, and protrudes as a shapeless mass from the cavity of the mouth (see Fig. 57). Definite parts of the body are rarely recognisable; but it more or less deforms the mouth, and on this ground its removal must be

[1] An interesting case of this kind is that of Anna Marie Przesonyt, recorded by Pitha. At birth she was normally developed; but soon after, a lump appeared on the sacrum, which in her third year burst, giving exit to a considerable quantity of fluid and accompanied by the appearance of an accessory lower limb. This was successfully removed by Pitha, and appeared to consist of two limbs fused together. The femur articulated with the trunk by a ball-and-socket joint.

attempted. The mass (in Fig. 57) consists, in its upper part, of skin; in its lower part the covering of the tumour passes over into the mucous membrane of the mouth. These tumours may also contain connective tissue, cartilage, bones, teeth, portions of brain, rudiments of intestine, or muscle and other organs. Rarely well-developed extremities are found. Such malformations are termed Epignathus,[1] Prosopagus parasiticus, Sphenopagus, or Pinnopagus.

FIG. 57.—EPIGNATHUS. (Specimen from the collection of the Göttingen Institute of Anatomy.)

With regard to the origin of the epignathi, the hypothesis put forward by Ahlfeld namely, that by an epignathus is understood an acardiacus amorphus, which is united with the oral cavity, generally with the hard gums, of its twin brother—is even at the present time accepted by many authors. According to Schwalbe, this hypothesis can no longer be considered as correct, as it does not take into account the results of more recent embryological investigations—especially as regards the amnion and the pedicle of the allantois in Man. Much more probably an epignathus arises from one of the segmentation cells, or of a portion of the germ-plasm, which in the earliest stages of the development of the autosite is separated off (Marchand-Bonnet's hypothesis: see Schwalbe). A very interesting case of epignathus has been described by Rosenow, and termed by him Epiglossus; it was confined to

[1] Like the true coccygeal tumours the epignathi may be divided into four classes: (1) The umbilical cord of a second fœtus is attached to the gums of a fœtus or in their neighbourhood; (2) more or less well-developed portions of the body of a second fœtus are attached within the oral cavity of a fœtus; (3) a tumour having the structure of a teratoma; or (4) a tumour, having a structure resembling that of a dermoid, is found growing from the oral cavity.

the tongue, and had lead to macroglossia (see under Macroglossia, p. 100).[1]

Cases in which such parasitic malformations are included within the cavity of the skull are termed Enencuitis. In very rare cases such inclusions have been seen in cases of Hygroma colli. (For Craniopagus parasiticus, see p. 318.)

Triple Monsters

Triple monsters (Trigemini conjuncti) and multiple monsters are extremely rare. They occur in the same way as double monsters, and are more frequent in the Animal Kingdom than in Man. Double inclusions in the abdominal and cranial cavities, on the other hand, are found relatively frequently (see Strassmann, Schwalbe, and Ahlfeld). Hohl gives the older literature (see Triecephalus).

[1] There are many points of resemblance between cases of epignathus—the Endoprosopus amorphus of Taruffi—and true coccygeal tumours; and this refers not only to their structure, but also to their probable mode of origin. In considering true coccygeal tumours, the fact that they occur in close anatomical relationship to the neurenteric canal was pointed out, and also the view held by some writers that they originate from this structure and the tissues surrounding it. The intimate relation that an epignathus bears to the buccal element of the pituitary body and the cranio-pharyngeal canal has not failed to attract attention, and has lead to the suggestion that these tumours may be derived from this structure. In many of the cases, however, they are not actually attached to this region, but only in its neighbourhood. Ahlfeld's hypothesis—that in an epignathus an Avarditacus amorphus is drawn into the buccal cavity of a healthy twin, and there acquires a secondary attachment—appears to be negatived by the fact that they are both contained within the same amnion. The view that these tumours are due to the separation of a portion of the germ-plasm at an early stage of development, and that this possesses the potentiality of developing into a more or less well-developed embryo, or only into a teratoma, is widely held at the present day. If this view is taken, then the relationship of these tumours to the base of the skull and hard palate is explained by the attachment of such a portion of germ-plasm to the infolded ectoderm of the stomodæum, the apex of which forms the buccal portion of the pituitary body. If we accept a similar suggestion of the separation of a portion of the germ plasm for the origin of the true coccygeal tumours, may not the connection of such a tumour with the neurenteric canal be a purely accidental one, and due to the passage of the separated cell or mass of cells into the canal and its further development in this position?

In a similar way, cases of encranius and of inclusio abdominalis would be explained by the development of the separated portion of germ-plasm within the medullary groove or the primitive entodermal cavity. It is further conceivable that such a separated portion of germ-plasm, or one of the early segmentation spheres, may be situated in any part of the embryonic cell mass of the normally developing embryo, and in this way the problem of the occurrence of teratomata in different organs of the body would receive a probable solution.

CHAPTER XIX

The Obstetrical Importance of Double Monsters

Double monsters—especially the [illegible] doubling of the upper or lower half of the [illegible] obstetrical interest. It is evident that [illegible] usually associated with such conditions, [illegible] monster through the pelvis very difficult [illegible] circumstances, even impossible. If the diagnosis [illegible] by the medical attendant or midwife, rupture of the uterus [illegible] to occur, in unfavourable conditions; and this happens [illegible] frequently as a result of the operative measures adopted by the [illegible] attendant, if the labour does not proceed normally, or marked complications are present. An unfavourable termination, indeed, is often to some extent prevented by the fact that the fœtuses—as in twin pregnancies—are generally small[1] from shortening of the gestation period, as a result of over-distension of the uterus, or from an insufficient amount of nutrition. The relative frequency of pelvic presentations, and the additional fact that a large number of these monsters die *in utero*, and are therefore very compressible and capable of being moulded, is also an advantage. On the other hand, a considerable number of cases occur where these factors cannot be reckoned on: in which the fœtuses are of the average size, are not dead, and do not present in a favourable position (see, for instance, the case described by v. Oynhausen from the Göttingen Clinic). Further, from the rarity of such malformations, no one obstetrician can have any large experience of this subject. Such labours take place usually as a surprise, and their course depends rather on the obstetrical dexterity, and not so much on the obstetrical knowledge, of the practitioner concerned.

The physical signs which such malformations exhibit are not in the least characteristic. Very frequently hydramnios is noted in the history. My remarks are concerned more especially with symmetrical

[1] Tur, who has made a special study of the size of double monsters, finds that in symmetrical double monsters, occurring naturally, each half usually attains the size of a normal fœtus; while in the case of artificially produced monsters this does not hold good.

malformations, in which all those symptoms may be recognised that are present in cases of normal twin pregnancy: namely, œdema of the lower extremities and of the labia, unwieldiness of the patient, dyspnœa, palpitations of the heart, and excessive movements of the child.

According to some authors, such malformations are commoner in multiparæ than in primiparæ.[1] As Hohl points out, the abdomen in some cases has been described as projecting anteriorly, the loins and iliac regions appearing empty. Very little indeed can be done in the way of diagnosing such conditions during pregnancy. In a certain number (some 25 to 35 per cent.) of the cases, the pregnancy does not reach its normal termination. The presentation is most commonly a cephalic one[2] (see the observations of Hohl).

With regard to the course of the labour, it should be noted that this not uncommonly takes place spontaneously. According to Hohl's statistics, of 119 cases, delivery occurred naturally in seventy-three; of thirty-one cases collected by Playfair, delivery was natural in twelve (Spiegelberg). In two of the cases quoted by Hohl, the forceps was necessary; once the blunt hook, three times version, eight times version and extraction, once extraction of the head and arm, once bringing down the arm, once perforation of the chest and abdomen, twelve times extraction only, once embryotomy, twice Cæsarean section, and fourteen times several operations combined (forceps, perforation, and version). Among the cases in which artificial aid was required, four of the mothers died; twenty-nine did well; and in eleven instances the result is not given. Among those in whom the delivery terminated naturally, forty-seven did well; while in twenty-seven cases the result is not stated. Of the children delivered artificially, twenty-seven were still-born and five were born alive. Of those delivered spontaneously twenty-two were born dead, twenty-nine alive, and in twenty-two the result is not stated. From these statistics it is clear that the prognosis is better for the mother than one would *a priori* have supposed.[3]

During labour, the diagnosis is almost always first made when the expulsion of the child is arrested, and, in cephalic presentations, after the birth of the head. In many cases from an external examination

[1] Of twenty-one cases recorded in the *Trans. of the Obstet. Soc. London* (in which the information is given), in seventeen cases the mothers were multiparæ and four primiparæ.

[2] Of forty-six cases collected from the *Trans. of the Obstet. Soc. London*, including twenty-four from Playfair's paper and twenty-two other cases, in twenty-seven there was a cephalic presentation, and in fifteen a breech or footling presentation.

[3] Of forty-nine cases recorded in the *Trans. of the Obstet. Soc. London*, in twenty-three labour occurred spontaneously, in nine it was ended by the application of the forceps, and in seventeen by traction on the feet or podalic version. Of fifty-three cases in all, published in these *Transactions*, in only one case, apparently, did the mother die—a case recorded by Playfair, which occurred in 1724.

a diagnosis of twin pregnancy has been made. According to G. Veit (whom we have to thank for the best description of double monsters, from an obstetrical point of view), such a malformation may be excluded with certainty when two separate amniotic sacs can be recognised, or when a portion of a child lying uncovered can be felt near another part still covered by the membranes. If two dissimilar poles of the body present during delivery—for example, a head with the breech—a xiphopagus must be first thought of, as it is only in this malformation, on account of the thin cord-like bond of union, that such a rotation of the bodies on their axis is possible. In the same way, when similar poles of the child present during birth, the possibility of a double malformation must be borne in mind.

Further than this, if the first child, in an assumed twin pregnancy, presents as a transverse presentation, a double monster should be thought of (according to G. Veit), since a transverse presentation of the first of twins is very uncommon.[1] Küstner gives the following very important rule: 'If, in a certain twin pregnancy, the attendant is compelled for any reason to introduce his hand into the uterus, the opportunity should be seized to examine the bodies for a possible bond of union.' This author further points out that in cases of double monsters the most marked absence of any set method of procedure has been observed in the operative measures adopted: as, for example, the removal of any limb already born, forcible attempts at delivery with forceps in cases of diprosopus—as bad a treatment as the application of forceps in a well-marked hydrocephalus—and forced attempts at extraction by the pelvis in doubling of the lower extremities. As on all occasions when dystocia occurs, so especially in this class of case, the golden rule holds good: 'If the labour is arrested, the half or whole hand should be introduced, under an anæsthetic, and the cause determined.' This procedure will enable the extent of the union and its situation to be recognised. As excessive haste is quite unnecessary in cases of double monsters, so a hasty mode of treatment is also to be avoided. It is precisely when some practitioner employs forcible extraction in such a situation—as, for example, the arrest of the labour after the birth of the head—that accidents not infrequently occur. It is best for the mother and the child when the development of the two parts of the double body is as complete as possible, and when the union between them is as slight as possible (see Strassmann).

As has already been briefly mentioned, the delivery of double monsters in a relatively large number of cases is accomplished by the natural forces. As a result of this, it is often possible to observe in such cases a definite mechanism of labour. The recognition of this fact is very

[1] According to Strassmann, this occurs in only 2·5 per cent. of all cases of twin pregnancy.

important in the treatment of such conditions. As the prognosis for the child (according to the observations published) is very unfavourable, no attention need be paid to the life of the child. According to the statistics collected by Conradi, of eighty-one double monsters born, five lived for forty days to eight months; two lived to their second year, and one survived to a later age; all the others died either during or soon after birth. Strassmann mentions the romance of Holtei ('The Vagabonds'), in which in a droll fashion the joy of some poor artistes is depicted when a double monster is born to them, and their grief when, after a short span of life, their possible source of income is again taken from them (see also Saltarino who, in an interesting book, has depicted the life of such a monster).

To G. Veit belongs the merit of having brought all the double monsters under a definite system with regard to the course of delivery. He divides them into three groups depending upon the possibility of the occurrence of definite mechanism of labour:—

Group I.—This includes incomplete doublings of the upper or lower pole of the body (Duplicitas anterior and Duplicitas posterior). The union is here a very intimate one, and the doubling not very extensive. To this class belong cases of diprosopus, dipygus, and cephalo-thoracopagus.

Group II.—This includes those fœtuses which are united to one another by the upper or lower poles of the body (Duplicitas posterior and Duplicitas anterior). In these cases the union is less intimate, and the doubling very complete. The bodies may spontaneously assume, during birth, a position in one and the same straight line, or may be brought into such a position. This includes cases of craniopagus, pygopagus, and ischiopagus.

Group III.—To this class belong, according to Veit, marked forms of Duplicitas anterior and Duplicitas posterior: that is to say, fœtuses which are united to one another by the trunk—thoracopagus and dicephalus.

Group I

Mechanism of Labour in cases of Diprosopus, Dipygus, and Cephalothoracopagus

In this class, a mechanical difficulty occurs during delivery from the size of the doubled parts. In these cases, conditions are present similar to those seen in excessive enlargement of some part of the child from other causes (hydrocephalus, spina bifida, hernia cerebri, tumours of the coccyx). In a case of diprosopus, on making an internal examination, an excessive number of sutures and bones may be felt; while in a case of hydrocephalus (for which this malformation most commonly

is mistaken), the wide sutures and fontanelles, and bones of the thinness of paper, may be distinguished. A combination of diprosopus and hydrocephalus has, however, been observed (Fig. 52).

In a case of dipygus, when the lower pole of the body presents, three or four lower extremities may be found on palpation, but only a single body above.

When the conjoined parts are not too large, the pelvis of ample size, the pains good, and the presentation favourable, such labours can occur spontaneously—especially when the fœtus is premature; often, however, with a prolonged second stage. In the case of a diprosopus or a cephalo-thoracopagus, the condition is more favourable when the monster is born in a pelvic presentation as the enlarged head passes more readily in an after-coming than in a fore-coming position. If the labour comes to a standstill and the natural forces prove insufficient, then artificial aid must be given. In some cases where the circumference of the head is not too large the forceps may be employed; but, on the whole—as in cases of hydrocephalus—it is better to avoid the forceps. In the majority of cases, as early as possible—namely, as soon as the necessary conditions are present—it is best to perform perforation, followed up by cephalotripsy or cranioclasm. Naturally with dead children only perforation is usually required.[1] Prophylactic version, even although the after-coming head may pass better through the pelvis, is best avoided. In cases of cephalothoracopagus the passage of the head through the pelvis may be attended with difficulty, as the broad shoulders with the four arms are too large. In this case the conditions are similar to those obtaining with giant children and cases of hydrothorax (see p. 289). In such circumstances perforation, cleidotomy, and evisceration may be necessary. In many case of cephalo-thoracopagus, hemicephalus is also present, which increases the difficulty both of the diagnosis and the treatment—especially as hydramnios is so often associated with hemicephalus (see p. 39).

The birth of a dipygus in a cephalic presentation offers little difficulty, provided that, if only three extremities are present, one is not attached obliquely or ankylosed. In a case of dipygus with a pelvic presentation, the doubling of the breech may cause some slight difficulty.

[1] Of seven cases belonging to this group, recorded in the *Trans. of the Obstet. Soc. London*, in four (two presenting by the head and two by the feet) labour terminated spontaneously. In one, a cephalic presentation delivery was effected with the forceps; and in the two recorded by Playfair there appears to have been some difficulty in delivery. In the first of these the forceps failed, and the large head was perforated. An arm was then brought down by the aid of the blunt hook and amputated. This gave room for the introduction of the hand, and the feet were brought down and the two bodies delivered.

In the second case the shoulder presented, and version was performed; the body of the second child followed the head.

It is best in these circumstances to bring down all the lower extremities; or, in a transverse or oblique presentation, to perform version (bringing down the legs), as in this way the circumference of the breech is markedly reduced. In dipygus tripus (according to most authorities), the united lower extremity should be brought down, as it may become placed transversely across the pelvic inlet, and so render further delivery difficult or impossible.

Group II

Mechanism of Labour in cases of Craniopagus, Pygopagus, and Ischiopagus

As Veit points out, delivery in these cases occurs most readily when the united fœtal bodies, at the very commencement of labour, are situated in one straight line or can be so manipulated that they pass through the birth canal one after the other; and this statement gives a clue to the treatment. With craniopagus, in cephalic presentations, for the reasons stated, podalic version should always be performed whether the case is one of Craniopagus frontalis, C. parietalis, or C. occipitalis. In cases, too, where the union of the two fœtuses is by the forehead or the occiput, so that the two fœtal axes are placed at an angle to one another, it is important to carry out the extraction of the second fœtus first. When in a case of craniopagus both fœtuses present by the feet, if they are of small size spontaneous delivery, resulting from a possible change of position, may be waited for; although, as Küstner points out, in this case it is even more important that similar parts should not pass through the pelvis at the same time. In the case described by Hochstatter (Küstner), both fœtuses presented by the feet; the extraction of the four arms caused great difficulty, but the birth of the heads followed very quickly one after the other. If in a case of craniopagus the double heads present, version by the feet is indicated, as this in any case is not likely to offer great difficulty.[1]

In pygopagus and ischiopagus, the condition is most favourable when the free end of the body, or the head, presents. Care must, however, be taken that the lower extremities do not become extended along the body of the unborn child. In the case described by Schönbeck (Ischiopagus parasiticus, see p. 315), an obstruction to delivery arose, as the lower extremities, attached to the trunk at an angle, caught on the pelvic brim. In this case, the weight of the monster (without the brain) was only 2300 gr. If the arms become extended, they must be brought

[1] In a case of craniopagus frontalis recorded by Tweedie, a hand and foot presented, and the monster was delivered by traction on the feet of the first child. The two heads followed one another, the feet of the second child being born last. The mother was a secundipara, and the monster, born prematurely at the seventh month, lived five minutes.

down before the entrance of the shoulders into the pelvis.[1] If, in cases of ischiopagus, and pygopagus, the united bodies present, the best plan is (as in dipygus) to bring down all the four lower extremities. Then the fœtuses, if they are small or of average size, may be born spontaneously. B. S. Schultze described, at the second Gynæcological Congress at Halle, a case of ischiopagus in which the fœtuses with their united lower extremities passed through the pelvic canal at the same time, and were born alive; the head of the posterior fœtus being delivered with the neck of the anterior one.[2]

Group III.

Mechanism of Labour in Cases of Dicephalus, Thoracopagus, and Sternopagus

It is difficult in this group to consider all the many varieties of monster from the same standpoint as regards their treatment. As Küstner points out, in the birth of such monsters there is only one common characteristic: namely, when the head of one fœtus is born, the possibility and probability is that the portion of the fœtus still movable in the uterus—either as a whole, or, when it is united, as in a dicephalus, as a part—will become fixed above the pelvic brim, and so render the further progress of the delivery impossible. The treatment for the different forms belonging to this group is a varying one. If, for instance, a certain amount of mobility exists between the head which is born and the upper part of the body (as in cases of thoracopagus and dicephalus tetrabrachius), then it may be possible to bring the part lying transversely above the pelvis into a favourable position. In this case spontaneous delivery or version by the feet may be possible. In other cases, however (as in dicephalus dibrachius), the head which is born may be so closely attached to the portion of the body lying transversely above the pelvis that any such movement is quite impossible.

In dicephalus, a pelvic presentation is a favourable one; as then the

[1] The usual mechanism of labour in cases of pygopagus is for the first twin to be born in a cephalic presentation, followed by the second twin presenting by the feet. This mechanism occurred in two of the three cases collected by Playfair; one of these was that of the Hungarian Sisters. The Bohemian Sisters, 'Rosa' and 'Josefa,' were born in the same way; while in the case of 'Millie-Christine,' the presentation was first by the stomach and afterwards by the breech. The difficulty that may arise in such a case is well illustrated by Marchand's case, in which the medical attendant at first attempted to deliver by traction on two of the legs and only succeeded finally when all four were brought down.

[2] In a case of ischiopagus described by Sternberg, a diagnosis of twins was made. The first head was delivered with great difficulty with the forceps. As the body did not follow, the arms were brought down; a third arm was then recognised. An examination with the whole hand showed that the second head was arrested above the pubes. The further description of the labour is not very clear; but, apparently, the second child was born in a position of condiplicatio corpore, the head pressed into the abdomen. The first child made a few attempts at breathing.

two heads very frequently pass through the birth canal one after the other. It is in this case advisable to draw the trunk, when expelled, up over the abdomen of the mother so as to bring the posterior head into the hollow of the sacrum, the second head then slips up anteriorly above the pelvic brim and remains behind. This mechanism is often favoured in this particular malformation by the fact that the children are not infrequently below the average degree of development. In cases of dicephalus with cephalic presentations the following possibilities may occur during birth: With a large pelvis and a small head, after the birth of the first head, the second may follow or be drawn with the forceps into and through the pelvis; the trunk then follows, or is born spontaneously. On the other hand, the second head may remain lying above the brim, and, in spite of good pains and a large pelvis, may not enter the pelvic cavity (Küstner). In these conditions version may be prevented by the presence of the short stretched band of union, and even an internal examination may be very difficult. At times, however, the second head may be felt externally. As the shoulders cannot enter the pelvis, the head which is partly born remains impacted in the vulva. The application of the forceps to this head is naturally useless, and embryotomy must be considered. In such cases it is best to practise decapitation, and afterwards podalic version; or, what is less favourable, cephalic version. In certain cases, with a small child and large pelvis, the delivery of the second fœtus may be brought about by the occurrence of spontaneous evolution (traction being made on the arm); although this is not without danger: namely, the risk of the occurrence of an impacted shoulder presentation and rupture of the uterus, as the lower uterine segment in such cases is almost always very much stretched. If the circumference of the shoulders is very large, then unilateral or bilateral cleidotomy must be practised. In cases of dicephalus with several arms and with doubling of the upper half of the body, the degree of mobility is much greater; so that after the birth of the head podalic version can be performed. In cases of dicephalus with multiple lower extremities and breech presentations, all the lower extremities should be brought down. The same is the case when a third lower extremity, united and frequently ankylosed, is present. In a few cases dicephalic fœtuses have survived.[1]

[1] Of twelve cases of dicephalus recorded in the *Trans. of the Obstet. Soc. London*, seven in Playfair's paper, and five since that date, in two cases the first head, after its birth, was decapitated and version performed; in two cases, the forceps was applied to the first head, and the second head delivered in the same way; in three cases, after the birth of the first head, the body was born by a process of spontaneous evolution; while of the remaining five cases, in four the breech presented, and in one the head presented and version was performed—all five cases being delivered by traction on the lower limbs. In these last cases, when care was taken to cause the two heads to enter the pelvis, one after the other, they passed through without difficulty.

In thoracopagus and xiphopagus the conditions are similar, but more favourable than those in dicephalus tetrabrachius. Küstner points out that even when there is a broad band of union of the xiphopagus and even when portions of the liver lie in it, the mobility of the two individuals is so marked that in the majority of cases thoracopagi and xiphopagi can be born without much difficulty. According to the observations of Playfair, of eighteen thoracopagi, eight were born spontaneously; in five cases, version and extraction were necessary; and in four cases instrumental assistance. One woman died undelivered.[1]

When the union between the fœtuses is broad and short, the prognosis is not so good as it might appear, from the above remarks; and especially when the parts common to the two individuals are well developed—as in the case of thoracopagus observed in the Göttingen Clinic, where the union was a very firm one and there was a common thoracic cavity, a common sternum, and closely united ribs.

If in a case of cephalic presentation one head has entered or escaped from the pelvis, then delivery—either occurring spontaneously or with slight help—is possible; as the head of the second child may pass through the pelvis with the thorax of the first, or pressed into its abdomen. In xiphopagi the degree of mobility, due to the presence of a band-like union between them, is often so marked that they may be born rotated upon one another, as were the Siamese twins.[2]

Schönfeld[3] carried out the separation of xiphopagi *in utero* with a knife wrapped round with linen. The fœtuses were born dead. The presentation of both fœtuses by the feet is the most favourable position for the progress of the delivery, and this fact may be made use of in the treatment of the condition. When the diagnosis is made sufficiently early—as, indeed, is very seldom the case—version should be performed, and all the feet brought down. Even when the head of the first fœtus is already born in a cephalic presentation, and the second head remains above the pelvic brim, bringing down the lower extremities of both fœtuses should always be attempted first; all the feet being drawn down together, and the second head delivered last of all.

If the heads fail to enter the pelvis and version is no longer possible,

[1] This is not quite accurate; Playfair's actual words are: 'One died undelivered; eight were terminated by the natural powers—in three of which the feet and in three the head presented; in two the presentation was doubtful; six were delivered by turning or traction on the lower extremities; four were delivered instrumentally.'

[2] Accurate details of the mode of delivery are not forthcoming; but the mother averred that the head of one was born with the feet of the other.

[3] Forceps were applied to the head of the first fœtus on account of protracted labour; and, as after the delivery of the head the body did not follow, the whole hand was introduced into the uterus and the site of the union felt. After the division of the band, which contained cartilage, the extraction of the first child was completed, and the second child was turned and delivered.

then, after a cautious attempt with forceps has failed, perforation is indicated. During extraction by the lower limbs (following the recommendation of Spiegelberg), in order to obtain room the trunk should be brought as far as possible into one of the oblique diameters of the pelvis; as in this way, possibly, the tendency for the head to remain above the promontory of the sacrum, or above the anterior pelvic wall, may be avoided. In order to bring the posterior head first into the hollow of the sacrum it is necessary, as in cases of dicephalus, to draw the delivered trunk well up over the abdomen of the mother so that the other head may move up anteriorly above the pelvic inlet and remain behind. In some cases it may be necessary to remove the anterior half of the double body so as to draw down the posterior head in the manner described. In such a case the forceps also may be necessary. With large foetuses in pelvic presentations, during the extraction, the evisceration of one child may have to be considered. The last possibility is that one child is born in a cephalic or pelvic presentation and the other lies transversely above the pelvic brim; in this case, with the first in a pelvic presentation, version of the second child must be carried out. When the first child presents by the head and the second transversely, there are three ways which may be adopted of dealing with the condition. In the first method, the feet of the child already born in part are drawn down, as well as those of the foetus lying transversely, and then the extraction is carried out in the manner described. As Strassmann points out, after version, the feet of the undelivered foetus can be more readily reached than those of the half-born foetus. In the second method, the feet of the first half-born child are drawn down, and then by means of the bond of union between the two bodies the second is converted into a cephalic or breech presentation. This method can only be employed when the uniting band is broad and firm, as there is the possibility of tearing the bond of union.

Lastly, the following method may be adopted: The feet of the child lying transversely are drawn down by the head already born, the two trunks are delivered together, and, finally, the head of the second child. If no one of these three ways leads to any result then recourse must be had to evisceration. The decapitation of the head already delivered is useless, as it in no way renders the delivery easier.

It should only be considered when the head is born and no farther progress takes place; in this case, by decapitation, more room is afforded for the evisceration. If the one head is born and the second has engaged in the pelvis, and the delivery comes to a standstill, an attempt can then be made to deliver the engaged head with the forceps. If extraction with the forceps fails, then perforation must be carried out. A similar procedure must be adopted when the fore-coming head

will not escape; first an attempt to deliver with forceps, then perforation. After the delivery of the perforated head, or the delivery of the head with the forceps, an attempt can be made to draw down the feet.[1]

Labours with triple formations are very seldom observed. The spontaneous delivery of such fœtuses is naturally only possible when they are not at full term. Thus in Fucello's case, the monster (a tricephalus) was very small, but lived two days. It cried and sucked, (so it is alleged) with all three mouths. In another case (recorded by Reina), of tricephalus tribrachius dipus (see Küstner), dystocia occurred. After three days of labour and fruitless attempts with forceps, Reina perforated the fore-coming head, and discovered behind this another head; so he amputated the delivered head. Even then the forceps failed to extract the second head, and perforation was necessary; and after the amputation of this head another third head had to be amputated: only then did the extraction of the child finally succeed. I believe, however, that Küstner and Strassmann are right when they doubt the correctness of this treatment.

Labour in parasitic double formations usually proceeds spontaneously. It is true that epigmathi, in certain circumstances, may develop into large tumours; but, during birth, they are very movable and compressible, and not infrequently it is possible to carry out puncture or removal of the tumour. As such tumours pass through the pelvis better with the after-coming head, in cephalic presentations version should be practised, as in this way a good hand-hold for extraction is afforded.

On the other hand, in true fœtal inclusions, dystocia occurs more frequently (case of Schaumann; see Küstner, Buhl, and Schönfeld). In such difficult cases the practitioner must proceed, with regard to diagnosis and treatment, in accordance with the numerous rules given. As therapeutic measures, perforation, puncture, and evisceration must be considered. Thus in the case of inclusio fœtalis,[2] recorded by Schaumann, puncture of the markedly distended abdomen was necessary. (For a case of dystocia due to an ischiopagus parasiticus, see the case of Schönbeck, p. 315.)

If, in conclusion, I give a brief résumé of my observations (see

[1] to eight cases recorded in the *Trans. of the Obstet. Soc. London*, since 1867 (the date of the publication of Playfair's paper), in four cases delivery was effected by traction on all four lower limbs, the heads being delivered last, one after the other. In three cases the head of the first twin was born followed by its body and lower limbs, and then the lower limbs of the second twin followed by its body, and, finally, its head. This was the mechanism of labour in the case of the Bohemian twins, 'Rosa' and 'Josefa.'

[2] In this case (figured by Ahlfeld in his *Atlas*, Plate viii, Fig. 7), there was a large cyst in the abdomen of the autosite, containing a malformed fœtus, which consisted mainly of two lower separate extremities and one fused upper extremity with nine fingers.

also Hohl), in all cases of double monsters the advice must first be given not to proceed too hurriedly, as haste is generally not necessary. Only in this way can a want of method in the treatment, which has been drawn attention to, be avoided. In the great majority of cases, success can be obtained without mutilating operations. Attention should be paid to the normal mechanism described as often occurring in the various forms. As delivery, judging by experience, occurs best with a pelvic presentation of both fœtuses, an endeavour should be made whenever possible to bring down all the feet. In all cases everything must be done in the interests of the mother, and for this reason the labour should be allowed to proceed as carefully as possible. These fœtuses are only very seldom capable of living; and this is so because other malformations are frequently present. On these grounds almost all obstetricians unanimously reject cæsarean section, even when the fœtus is alive. Spiegelberg rightly says: 'One dare not, for the problematical preservation of the life of the child, place in jeopardy the life of the mother.' For the same reason Strassmann rejects cæsarean section, symphysiotomy, and, indeed, cutting operations of any kind. Küstner does not express himself so strongly, but says: 'Cæsarean section should only be considered in the highest degrees of double monsters, when a complete examination of the surface of the body has shown that the capability of survival is certain.' He continues, however: 'So long as such a diagnostic examination of the surface of the body is possible, other simpler obstetrical operations, such as version, will be possible. If cæsarean section is chosen, it must be certain that it is the least dangerous operation for the mother in the existing conditions.' (See also Hohl, on the question whether cæsarean section is permissible in monsters or not.) In principle, the classical cæsarean section is (according to our view) to be rejected, as apart from the want of viability on the part of the child the case is not usually an aseptic one (the membranes are long ruptured, and repeated examinations have been made). This is not the case with extra-peritoneal cæsarean section,[1] especially recommended by Sellheim, which in certain cases (large fœtuses, and broad and firm union) may occasionally be indicated—possibly, in conjunction with mutilating operations or separation of the fœtuses.

Intra-uterine separation of the fœtuses—as, for example, was undertaken in the above-mentioned case of Schönfeld—can only be considered

[1] Although, when first introduced, extra-peritoneal cæsarean section was thought to be especially suitable for cases in which the uterus was possibly infected, yet experience has shown that it is better not performed in such cases, and that its indications, as regards the question of the presence of infection of the uterus, are almost the same as those of the classical operation.

correct treatment in the rarest possible cases, as the fœtuses will necessarily be killed; and, by such an operation carried out in the dark, a severe injury may, in certain circumstances, be inflicted upon the mother. It may be permissible, however, if it is possible to draw down the uniting band, and if podalic version can still be carried out. (See also Hohl on the intra-uterine separation of united twins.)

The recommendations of Ahlfeld, on the treatment of cases of double monsters, are very important from the practitioner's point of view. For 'doubtful' cases, he strongly recommends *morcellement*—the mutilation, step by step, and removal of portions of the body—as this procedure is the safest for the mother in the end, even when it does not at once procure a more hopeful outlook.

Ahlfeld concludes his remarks on this subject with these words: 'Accordingly, the practitioner should, in the case of the protracted delivery of a double monster, reduce in size that part which lies in the pelvis and is most easily reached, partly by the emptying of any of the cavities of the body which may be accessible, partly by the removal of some of the limbs, and partly by decapitation.' His recommendations present a marked contrast to those given above. The practitioner, however—whose knowledge of this difficult subject is usually somewhat defective—will do better, in the interests of the mother, by a careful application of the short rules for treatment cited above than by blind attempts to effect delivery by force; it is evident that he can more readily remember them than the detailed instructions laid down by various writers—which should, however, be carried out by preference if he is familiar with them.

In the Göttingen Women's Clinic, a case of thoracopagus tetrabrachius tetrapus came under observation a short time ago. The case has been fully described by v. Oythausen. The patient was a working woman, thirty-eight years of age, who had had seven living children. She stated, on her admission to the Clinic, that she had had labour pains since early in the morning. The abdomen was pendulous and markedly distended, measuring some 124 cm. in circumference. External examination through the excessively stretched abdominal walls was very difficult; the fœtal heart-sounds, however, were clearly heard below the umbilicus. After the rupture of the membranes, the head was born, with a few strong pains; the rest of the child's body did not follow, however; nor did it with the application of traction from below with forceps, or pressure from above. The child made a few gasping attempts at breathing. Chloroform narcosis was induced. On abdominal examination, the body of a second child could be felt lying in an oblique position, the head in the right iliac fossa. On internal examination with the whole hand a broad bridge of union between the two bodies could be felt fixed behind the symphysis. The

diagnosis of a double monster was now made. An attempt was first made to bring the body of the second child into a longitudinal position, so as, if possible, to extract the two bodies without separating them. A foot of the second child was drawn down and a sling put round it, but the attempt at version failed. The thoracic cavity of the fore-lying child was now perforated, and, lastly, the head and a portion of the chest removed with Siebold's scissors. A renewed attempt to perform podalic version on the second child now easily succeeded. The resulting extraction, however, presented great difficulty, as the torso of the embryotomised child was fixed in the right iliac fossa. The lower part of the body of the second child which had been brought down was therefore removed, and it was now possible without difficulty to extract by the feet of the first child the remaining portion, consisting of the lower half of the body of child number one and the upper half of the body of child number two. The placenta followed at once.

During the fruitless attempts at version, severe uterine bleeding took place; and severe symptoms of collapse appeared, which were not relieved by large doses of camphor. The uterus, in spite of the administration of cornutine, contracted very imperfectly. Examination showed, high up on the right side, a tear in the cervix which extended out into the broad ligament, but apparently did not communicate with the peritoneal cavity. The uterus, the tear, and the vagina were plugged, and a tight abdominal binder was applied. The woman remained in a state of marked collapse, and saline infusion and large doses of camphor only improved the symptoms temporarily. There was no external post-partum hæmorrhage. With signs of increasing exhaustion and dyspnœa, death occurred four hours after delivery.

The autopsy confirmed the diagnosis of an incomplete rupture of the cervix. The occurrence of this was probably not due to the version, but rather to the injury inflicted by the jagged stump of the body. There had been no further hæmorrhage, but the uterus presented the appearance of a large relaxed sac. The unfortunate result is to be attributed partly to the size of the child, partly to the very broad and firm bond of union. Treatment according to the rules already given was not possible.

The post-mortem examination of the fœtuses gave the following result:—

The length of the thoracopagus was 45 cm.; the fœtuses were of the male sex, of equal length and, externally, equally developed; the weight of the two together amounting to 4010 grm. (8·75 lb.). These figures do not actually represent the true weight, as during the various operations the greater part of the blood escaped. The occipito-frontal circumference of each of the slightly moulded, heads of

the two fœtuses measured 35 cm.; the weight of the placenta was 750 grm.; its size, 24 cm. by 28 cm. The monster had four upper and four lower extremities; in one fœtus a club foot was present, and the arm of the other was contracted at the elbow-joint. The two fœtuses, besides their length and weight, showed all the other signs of maturity. The lanugo had disappeared from the skin with the exception of the region of the shoulders and of the sacrum. The hair of the head was about 1 cm. in length, and the nails were horny and reached the ends of the phalanges. The development of the subcutaneous fat corresponded to that of a full-term fœtus. While the descent of the testes had not occurred in either of the fœtuses, yet this is not certain evidence against their maturity.

The fœtuses were united in such a way that the two lower thoracic apertures were turned towards one another, and the bond of union was at the sternum. The union extended downwards as far as the common navel; but the two fœtuses were not placed facing one another in the median line, but more ventro-laterally.

After the removal of the soft parts over the thorax, it was evident that there was a sternum common to the two, with a curve concave upwards, which in its whole length shows no sign of any division. To this sternum the ribs, strongly curved on their posterior surfaces, and the rib cartilages of each fœtus were attached. The cartilages of the last true ribs were united with one another. In the angle formed by the last ribs on the anterior and posterior surfaces there was a cartilaginous projection, which resembled the two ensiform processes. The conclusion that these processes were of this nature was the more probable as the one on the anterior surface, from its mode of projection, appeared to belong to fœtus number two, and that on the posterior surface to fœtus number one. Each fœtus had two clavicles situated in the normal position. In order to render the thoracic viscera visible, the whole of the anterior chest-wall was removed. This disclosed a common pericardium, and in it—to external appearances, at any rate—a common heart, measuring $3\frac{1}{2}$ cm. in height and $5\frac{1}{2}$ cm. in breadth. It lay with the origin of its vessels directed upwards and backwards, and its apex—or rather, border—directed forwards and downwards. On the anterior surface there was placed, running in a sagittal direction, a shallow groove which divided the organ externally and unequally into two main portions, in such a way that the larger part appeared to belong to the second fœtus. The examination of the interior of the heart confirmed these conclusions, as will be shown later.

Each fœtus possessed two auricles, which communicated with one another by a wide open foramen ovale. Further, the right auricle of one fœtus communicated with the right auricle of the other,

so that one could speak of a single cavity. The two left auricles were divided from one another by a septum. There were in all four auricles.

The position of the external division, by the groove mentioned above, corresponded to its internal division, into two hearts; and while the second fœtus had two ventricles, not communicating with one another, the first fœtus had only one, which was in open communication with the left ventricle of the second fœtus. In the single ventricle of the first fœtus there opened, close to one another, the valvular opening from the right and the left auricle. From the left ventricle of the second and from the single ventricle of the first, the aorta took origin; and from the left auricles, the two pulmonary arteries. These vessels, in the second fœtus, were united by a wide ductus Botalli; in the first fœtus, by a very narrow one. Into the right ventricle opened a common vena cava inferior; while the superior venæ cavæ passed separately up to their entrance into the common right auricle.

Each fœtus had two lungs, which in their situation and in their relation to the blood-vessels showed nothing abnormal. The lungs of the fœtus whose head was born spontaneously, contained air; those of the second fœtus were atelectatic. Situs inversus was not present in either fœtus, and the structures in the neck and the other organs of the thorax were normal.

The arrangement of the abdominal organs was as follows: There was present a common diaphragm and a common abdominal cavity; situs inversus of the abdominal organs was not present in either fœtus. A liver was placed in each hypochondrium, and these organs were united with one another by a broad band of liver-tissue. There were two gall-bladders, and the vessels of the liver and the gall-ducts were present in either fœtus. Between them lay a common duodenum, about 3 cm. in length, to the middle of which, with a triangular enlargement, was attached a common jejunum, some 50 cm. long. At the end, the jejunum was dilated in an ampulla-like manner, and passed into two separate ilea which, after a course of about 25 cm. opened on either side into the cæcum and large intestine. On both sides a vermiform process, about 3 cm. long, was present. No separation of the large intestine into its various divisions and flexures was present. On the posterior surface of the common jejunum, at the ampulla-like enlargement, a Meckel's diverticulum originated, the blind end of which formed a filamentous union with the mesentery of the first fœtus.

The two excretory ducts of the two equally-developed pancreatic glands united into a very short common duct which, in its turn, opened into the duodenum rather on the side of the first fœtus. All the other

abdominal organs were present separately in each fœtus, and were normally developed.

The vessels of the umbilical cord showed a remarkable arrangement. A microscopic section through the cord showed two arteries and two veins. From the navel, one artery passed to the two pelves and gave off from its sides two branches which united with the two hypogastric arteries of each fœtus. The two umbilical veins passed from the umbilicus to the two livers. The two urachi, after a short course, were obliterated on both sides close to the bladder, and in the umbilical cord no trace of them could be found microscopically. The single insertion of the umbilical cord into the placenta was not quite central, the vitelline duct and sac were not to be found.

To sum up: The condition is one of a mature monomphalous autositic thoracopagus tetrabrachius, living up to the time of birth. The heart forms the only exception to this description. As regards this, the first fœtus is maldeveloped as compared with its brother, as it has only one ventricle from which it is able to send mixed blood only, both into its greater and lesser circulations. The viability of a double monster depends mainly, as has been pointed out, on the conditions of its respiratory and circulatory systems. In the foregoing case the question whether the complicated structure of the heart would have allowed a sufficient amount of oxygen containing blood to pass into the extensive circulatory system can be negatived with certainty.

The remaining deformities—the club foot and the contraction of the arm at the elbow-joint, which in the ventro-lateral position of the fœtus could be recognised as one of the two internal arms—are to be regarded as accidental, and may be explained without doubt as due to want of room in the uterus.

The nature and mode of union of the fœtuses is, on the whole, that common to cases of thoracopagus—as we see from the descriptions of Virchow, Veit, Marchand, Ahlfeld, Dönitz, Kortüm, Völker, Kamann, and others. For example, the common sternum, common thoracic and abdominal cavities, and common navel; further, one diaphragm and pericardium. The heart consists generally of two single organs, more or less united with one another; in this respect the foregoing case is of interest, as it presents a transition to one common heart. Divided externally only by a slight groove, internally it showed a common right auricle, and altogether three ventricles. According to Ahlfeld, it is the parasite which usually has no heart; but as the participation of the two fœtuses in the defect of development is otherwise equal, one can hardly speak of the case as one of parasitic double malformation. The common course of the jejunum is found in the majority of the described thoracopagi, and, further, the origin of Meckel's diverticulum from the end of the common portion of intestine, is not an exceptional

condition. Marchand found that the common course of the small intestine reached only to the insertion of the ductus omphalo-mesenteriens. The processes, too, described above as ensiform processes, are also described by Marchand. A bridge of union between the livers is nearly always found: often, indeed, in double malformations, with less extensive union—as, for example, in xiphopagi.

The assertion of some authors (Schultze and Förster) that in monomphalic double malformations the fœtus placed on the right always shows situs inversus has been denied by Perls, Tarnffi, and others (quoted by Marchand). Eichwald's statement also, that the liver at least of one of the fœtuses shows situs inversus, has been shown to be untenable by the more recent researches of Martinotti and Lochte, and the foregoing case also serves to refute this view.[1]

In the pathological collection of the Göttingen Women's Clinic there are the following specimens of double monsters, sent to the Clinic by medical men:—

Catalogue No. 129.—*Dicephalus dibrachius dipus.* A female, of about 7 to 8 lb. in weight. The monster showed an hydrocephalus of medium size together with a marked microcephalus and fissure of the spinal column. It was born, according to the history, as a breech presentation, after the feet were brought down; but the extraction was very difficult, and only succeeded after spontaneous rupture of the hydrocephalus.

Cat. No. 153.—*Dicephalus dibrachius dipus.* A male, weighing about 6 lb. The heads are said to have been born one after the other. The monster lived some hours.

Cat. No 164.—*Diprosopus hydrocephalus distomus, tetraphthalmus dibrachius diotus.* Weight, about 6 lb. Of female sex. Tolerably marked hydrocephalus. The median eyes lay in a single cavity. Each fœtus showed an exactly corresponding unilateral hare-lip. The monster was delivered by perforation.

Cat. No. 112.—*Sternopagus tetrabrachius tetrapus.* Of female sex: about 8 lb. in weight. A well-marked union in the thoracic region, extending up to the neck. There are four arteries and one vein in the umbilical cord. During birth, the condition was discovered after the decapitation of one fœtus, but otherwise there is nothing said of the course of the labour.

Cat. No. 207.—*Thoracopagus tetrabrachius tripus.* The mother was a primipara. The monster, a female, was about 4 to 5 lb. in weight. Possessed four well-developed arms and three legs, one of which, the posterior, was shortened and crippled (contractures and web-like formation). The anus was absent. It was born spontaneously with

[1] As Ballantyne has pointed out, situs inversus is only an occasional complication of double monsters, and even in separate twins it is a rare occurrence.

good pains. According to the description of the practitioner in attendance, the larger head was born first with its face looking towards the symphysis pubis, the hands following almost immediately, then the second smaller head was born with the face turned towards the perineum; [illegible] arms raise, too, the hands followed almost at once. The umbilical cord was torn at the time of birth.

LITERATURE

Schwalbe, *loc. cit.*, p. 15, pt. 2 (1907); Ziegler, *loc. cit.*, p. 15; Olshausen and Veit, *loc. cit.*, p. 44; Köstner, *Müller, loc. cit.*, p. 15; Spiegelberg and Wagner, *loc. cit.*, p. 15; Strassmann, *v. Winckel, loc. cit.*, p. 24, vol. ii., pt. 3, p. 1726; Kleinhans, *v. Winckel, loc. cit.*, p. 34; vol. ii., pt. 3, p. 1364; Rohl, *loc. cit.*, p. 15; Ahlfeld, *loc. cit.*, pp. 24 and 34; Runge, *loc. cit.*, p. 24; Henneberg and Stelzner, *Berliner klin. Wochenschr.* (1903), Nos. 35 and 36; G. Veit, *Volkmanns Sammt. klin. Vortr.*, Nos. 164 and 165 (1879); Sabarino, *Abnormitaten* (1900); Froriep, *Neue Notizen*, vols. iii. and viii.; v. Oynhausen, *Diss. Göttingen*, *Arch. f. Gynäk.*, vol. lxxxvi., p. 1; Tieber, '*Dicephalus tribrachius*,' *Prager med. Wochenschr.* (1902), No. 28; Zwinjarzki, *Diprosopus syncephalus, hemicephalus, triophthalmus, thoracopagus, tetrabrachius, ref. Zentralbl. f. Gynäk.* (1903), p. 1316; Kamann, '*Zwei Thorakopagen*,' *Arch. f. Gynäk.*, vol. lxviii., pt. 3; Köstner, *Verein. Breslauer Frauenärzte* (M[illegible]5, 1903), *Thoracopagus*; Stanze, '*Epignathus*,' *Diss. München* (1902); Rosenow, '*Epiglossus*,' *Diss. Kiel* (1901); Leopold, '*Dicephalus dibrachius*,' *Arch. f. Gynäk.*, vol. lxxii.; Nickles, '*Dikephalus*,' *Diss. Breslau* (1903); Diepgen, '*Zwei Thorakopagen*,' *Diss. Freiburg* (1902); Guérin-Valmale, and Lagnière, '*Xiphodym*' *Gib. Gesellsch.* (Paris, December 17, 1903), *ref. Zentralbl. f. Gynäk.* (1904), p. 1088; Strassmann, '*Über Zwillinge u. Doppelmissb.*' *Geb. Gesellsch.* (Berlin, October 28, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 146, with report of discussion (November 28, 1904); Jolly, '*Geburt und Trennung von Xiphopagen*,' *Festschr. f. Olshausen* (Stuttgart, 1905); Weinberg, '*Diprosopus hemicranius cum rachischisi totali anencephalus et amyelus*' (*Württemberg med. Korrespondenzbl.*, 1905), *ref. Zentralb. f. Gynäk.* (1905), p. 1523; Förster, '*Die Entstehung der Doppelbildungen*,' *Verhandl. d. phys. med. Gesellsch. in Würzburg*, vol. xxxvii., No. 6; Scharfenak, *Geb. Gesellsch. in Leipzig* (October 27, 1904), *ref. Zentralbl. f. Gynäk.* (1905), p. 174; Ostreil, *Thoracopagus, ref. Zentralbl. f. Gynäk.* (1905), p. 192; Bokelmann, '*Dikephalus mit Anencephalus*,' *Geb. Gesellsch.* (Berlin, October 27, 1905), *ref. Zentralbl. f. Gynäk.* (1906), pp. 33 and 59; W. Freund, *Entstehung der Embryome, Naturforscher, Vers.* (Stuttgart, November 16 to 22, 1906), *ref. Zentralbl. f. Gynäk.* (1906), p. 1214; R. Meyer, '*Embryonale Gewebseinschlüsse, u. s. w.*,' *Ergebnisse d. allg. Path. u. path. Anat. d. Menschen u. d. Tiere*, vol. ix., pt. 2 (1905), pp. 517 and 705; Nádasy, '*Syncephalus triotus thoracopagus tetrabrachius et tetrapus*,' *Gynäk. Gesellsch.* (Budapest, May 29, 1906), *ref. Zentralbl. f. Gynäk.* (1907), p. 1342; Schönbek, '*Ischiopagus parasiticus mit Geburtsstörung*,' *Zentralbl. f. Gynäk.* (1908), p. 707; Menge, *Fränk. Gesellsch. f. Geburtsh. u. Gynäk.* (February 3, 1907), *ref. Zentralbl. f. Gynäk.* (1908), p. 265; Sellheim, '*Extraperitonealer Kaiserschnitt*,' *Hegars Beitr. z. Geburtsh. u. Gynäk.*, vol. xiv., No. 1 (1909); Henke, '*Craniopagus frontalis*,' *Monatsschr. f. Geburtsh. u. Gynäk.* vol., xxix., pt. 5, p. 665; Reina and Galvagni, *Atti dell' Acad. Gioena Catania* (1834), quoted by St. Hilaire, *Histoire des Anomalies* (1836), vol. iii., p. 341; Ballantyne, *loc. cit.*, p. 44 and p. 235; Tweedie, *Lancet* (August 26, 1876); Marchand, '*Missbild.*,' *Real-Encyclopädie der gesamten Heilkunde* (1897); Playfair, *Trans. Obstet. Soc. London* (1867), vol. viii., p. 300; Hertwig, *Handbuch der Entwicklungslehre*; Schumacher and Schwarz, *Anat. Anzeig.* (1900), vol. xviii.; Stöckel, *Arch. f. mikros. Anat.*, vol. lii.; Spemann, *Archiv f. Entwicklungsmech.*

vol. xvi., p. 533; Koenig, *Miscellanea curiosa sive Ephemeridum*, Annus viii.; Leop. Carol (1869), Observ. 145; Quoted by Londonin, '*Les Monstres doubles operable*,' *Revue Scient.* (Paris, 1892), vol. iii., p. 73; Böhm, *Virchow Archiv* (1866), vol. xvi., p. 152; Blaudet and Bugnion, *Revue de la Suisse Rom.* (1882), vol. ii., p. 121; Chapot and Prévost, *Gaz des Hopitaux* (1901), vol. lxxiv., p. 329; Doyen, *Verhandlung der Chir. Congress* (Berlin, 1902); Huff, *Amer. Journ. Obstet.* (1890), vol. xxii., p. 923.

CHAPTER XX

Acardiacus—Monsters without a Heart

Acardiacus is a rare malformation: many experienced obstetricians have never met with it. It is found almost always in twins separated from one another, and situated within a common chorion, and therefore in homologous uniovular twins with a common placenta. The one twin, as a rule, is well developed; the other is malformed in the manner immediately to be described. The name acardiacus is not strictly correct, as not uncommonly slight rudiments of the heart are found. On the other hand, however, many of the so-called parasites are entirely without a heart. A functionating heart is certainly never present.[1]

In the great majority of cases, the well-developed foetus is born first,[2] and only exceptionally the acardiacus. In such circumstances, the possibility of a second foetus being present must always be thought of. Here, however, I may remind the reader that a condition of acardia in a single foetus, as a result of 'intra-uterine decapitation,' cannot always be excluded (see the very interesting case recorded by Landau, under anomalies of the amnion, p. 11). Acardiacus occurs as a result of an extensive anastomosis of the circulatory systems of the two foetuses. The blood-pressure predominates in the circulatory system of one of the twins in such a way that the heart of this, the stronger twin, carries on the circulation in both; and the blood-stream in the other weaker twin is reversed, so that arterial blood flows centripetally, to

[1] All the acardiaci have this characteristic feature: they possess a circulation in common with that of their co-twin, and the motive force of this circulation is furnished by the heart of the latter. For this reason they are often termed placental parasites; but, as Ballantyne points out, this is not strictly correct, for one of them is really a parasite upon the other by means of the vessels in the placenta. Taruffi terms them 'disomata omfalo-angiopaghi'; but Ballantyne prefers the term 'allantoido-angiopagous' twins, as etymologically more correct.

[2] Of eight cases recorded in the *Trans. of the Obstet. Soc. London* (in which the details are given), in six cases the normal child was born first, and in two cases the acardiacus.

the acardiacus, through its umbilical arteries.[1] As a result, a more or less complete atrophy of the heart, the lungs, and the trunk of the malformed twin occurs, and it is nourished by the normally developed one. Very frequently, marked congestion occurs in the umbilical veins of the acardiacus, owing to insufficiency of the venous outflow, with resulting hypertrophy and œdematous infiltration of the subcutaneous tissues. In this way may be explained the marked disturbances in delivery (to be described later), which may occur with these malformations. The acardiaci—in whom the absence of a heart is universal—with the above-mentioned qualifications, are divided as follows: —

I. Acardiacus acephalus Holo-acardius acephalus[2]

This is the most frequent variety.[3] In it the head (and usually the arms), thorax, heart, large vessels, lungs, and the liver (in rare cases, present in a rudimentary form) are absent: while the lower abdominal organs, the pelvis, and the lower extremities are usually well developed. Portions, however, of the bowels and the whole of the pancreas, spleen, kidneys, and bladder may be absent. The lower extremities, as has been mentioned, are very frequently œdematous. The bones of the head, the cervical spinal column, and the bones of the upper extremities are usually entirely absent, as well as the brain and the upper part of the spinal cord with the nerves proceeding from it.

[1] The arrangement of the circulatory system in an acardiacus acephalus is very well shown in a specimen described by Keith. In this case, the single umbilical artery, on entering the abdomen, gave off three branches, which supplied the alimentary canal and the diaphragm, and ended in a median dorsal vessel—the aorta (?). Opposite the fifth cervical segment, the aorta gave off a vessel representing the vertebral artery, and two others representing the right and left carotid arteries. The aorta had no connection with the heart: the latter was a small three-chambered organ, consisting of an auricle, a ventricle, and a third chamber—possibly, representing the bulbus arteriosus. A vessel was given off from this chamber and passed to the base of the occipital region, where it ended. With this doubtful exception, the heart was entirely a venous structure, and had no connection with the arterial system; and, as far as it was functional, can only have assisted in forcing the blood along to the placenta. In the diaphragm, three veins united to form the umbilical vein: the first probably representing the cardinal veins, the second the right jugular vein, and the third coming from the alimentary tract. The main features of the circulation were, that the heart—if functional at all—drove the blood along the veins to the placenta, and that it had lost or never had any connection with the dorsal aorta, through absence or obliteration of the arteries of the visceral arches.

[2] Schatz divides the acardiaci into two main groups: the holo-acardiaci and the hemi-acardiaci. In the first group, the heart is entirely absent; in the second group, it is present in a more or less rudimentary state.

[3] Torulli was able to collect 108 cases of acardiaci acephali from the literature, and Förster states that acardiaci formed 18 per cent. of his collected cases.

PLATE VIII.

ACARDIUS ACEPHALUS

(Specimen from the collection of the Göttingen Women's Clinic.)

Some authors distinguish as acardius paracephalus, those forms of this malformation which show a rudimentary formation of the head. They belong, however, rather to the acardius acormus (see p. 455). According to the disposition of the extremities, the following subdivisions may be distinguished: Acardius acephalus syncnus, A. monopus, A. dipus,[1] A. apus, A. monobrachius, A. dibrachius, A. abrachius (see illustration of an acardius monopus in Küstner).

Bkart de la Faille has described a case of epignathus with which two acephali were united by the umbilical cord (see Schatz).

II. Acardiacus amorphus—Holo-acardius amorphus

This has the form, most commonly, of a round solid tumour—often the size of a fist—covered with skin, or of a formless mass into which is inserted an umbilical cord—frequently with a velamentous insertion and with only one umbilical artery (as is the case in the majority of acardiaci).[2] The condition is due to the fact that development has remained stationary at a very early stage. In rare cases, the extremities are represented by small projections. On a section of this formless (often hairy) lump—even macroscopically, more certainly microscopically—besides connective tissue generally, rudiments of organs can be recognised (vertebræ, muscle, intestine, cystic spaces, etc.). Schwalbe has described a case of acardius amorphus in which, besides a completely developed femur (the size of that of a fœtus of eight to nine months of age), definite rudiments of one of the bones of the leg, of the pelvis, of the spinal column, and of the

[1] Specimen No. 813, Univ. Coll. Hosp. Med. School Museum, is a good example of an acardiacus acephalus dipus dibrachius. At the upper part of the anterior surface of the trunk is an irregular shallow groove, in which there are two small cystic bodies surrounded by a few hairs. The right lower extremity terminates in a single digit; the left consists of a small heart-shaped prominence with a nipple representing the foot. The right upper extremity is represented by a rudimentary hand with three digits; the left, by a rudimentary hand with two digits. The co-twin was a living female,[3] and there was a single placenta and single amnion. A radiogram shows only a faint shadow of dorsal and lumbar vertebræ with a suggestion of a pelvis.

[2] As Geoffroy St. Hilaire says: 'There are neither abdominal nor thoracic viscera, nor any intestinal canal: only a cavity, surrounded by cellular and fatty tissue, with possibly a serous membrane and some vessels. The mass is no more than a purse of skin, of which it would be difficult to determine the nature, were it not for the umbilical cord, to the end of which it appears to be suspended.'

[3] Of seventeen cases recorded in the *Trans. of the Obstet. Soc. London*, ten were of female sex and seven of male sex.

brain were found.[1] The formless acardiaci amorphi may also be distinguished as anideus (εἶδος = 'form'), in contrast to the better-developed mylacephalus (μύλη = 'mola or mass'; κεφαλή = 'head'), which exhibits a somewhat more highly developed, more human form.

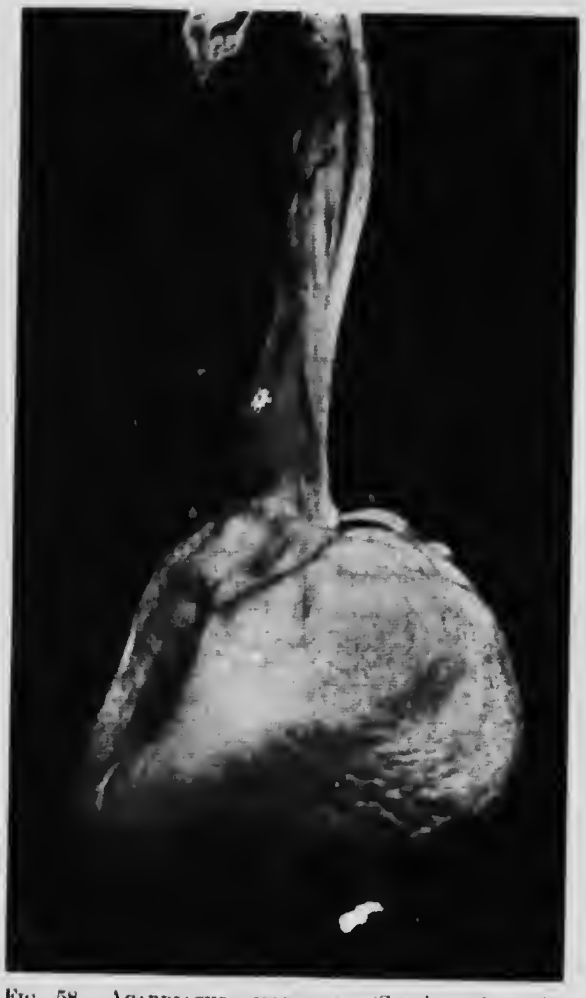


FIG. 58.—ACARDIACUS AMORPHUS. (Specimen from the collection of the Göttingen Institute of Anatomy.)

than the amorphus. The acardiacus amorphus is rarer than the acardiacus acephalus.

[1] In a specimen described by Dr. Herman, the mass formed an oval elastic body, about 6 cm. by 4·25 cm. by 2·5 cm., covered by skin, portions of which were covered by dark-brown hair. The interior consisted of œdematous connective tissue with fat; there was no trace of intestine, nor of any solid organ. A mass of bone and cartilage lay almost in the middle, bearing the characters of one of the cervical vertebræ. A thick cord, apparently the spinal cord, ran through the vertebra, and terminated in a tubercle on the surface.

III. Acardiacus acormus[1]—Pseudo-acormus[2]—Holo-acardius acormus

This variety is very rare, and has been observed in only a few instances (see an illustration in Schwalbe). The condition is one of a monster in which only the head is developed, while the remainder of the body is very rudimentary. The umbilical cord is inserted into the region of the neck.

IV. Acardiacus anceps—Hemi-acardius

This form is, according to Ahlfeld, the best developed of all; for the head and upper arms are present in a rudimentary state. Frequently, also, there is a rudimentary heart, rudimentary lungs, larynx, and diaphragm. This malformation forms a transition from the true acardiaci to the normally developed twin fœtus.[3]

As regards the mode of origin of acardiaci, the views held at the present day are very divergent. The original view—which, as we shall see later, has been revived in a modern form—was that the future acardiacus was imperfectly developed in its very earliest stages, so that the formation of the heart remained in abeyance. The defects, therefore, present in this malformation are to be regarded as primary, and not as a result of the imperfect circulation (Dareste and Panum; see Schatz, 'The Acardiaci and their Relations'). Claudius was the first to recognise the inversion of the blood-stream as the most important

[1] Κορμός = 'trunk.'

[2] Barkow gives the main characters of the acardiacus acormus as follows: The head is the best developed part; but even this is often much malformed. Rudiments of the vertebræ and ribs are generally present; some of the muscles are represented, but their anatomical arrangement is much altered; the sense organs are usually present, and the digestive tract, in part, but situated outside the abdomen; the brain, medulla, and nerves are, in part, present; the heart is entirely absent, and the pseudo-acormus is nourished by a branch of the umbilical artery of its co-twin. According to Schwalbe, only four certain cases have been described.

[3] Ahlfeld suggests that the presence of trabeculæ carneæ in the heart of such a fœtus seems to show that it has functionated for some time. Schatz has described a case in which the blood of the normal twin passed through the umbilical arteries into the aorta, from this to the heart of the abnormal twin, and by the inferior vena cava and the umbilical veins, back to the placenta again.

Schwalbe gives the following useful classification:—

CHORIO-ANGIOPAGHI.—One twin with a rudimentary or not completely functionating heart, or with complete absence of the heart. In this individual there is reversal of the circulation (*Acardii*).

1. Heart rudimentary (*Hemi-acardii*).
2. Heart absent (*Holo-acardii*).
   - (*a*) The defect is chiefly localised in a definite part of the body.
     - Upper half of the body wanting (*Holo-acardius acephalus*).
     - Lower half of the body wanting (*Holo-acardius acormus*).
   - (*b*) The defect is not localised in any definite part of the body.
     - General defect, so that a more or less formless mass results (*Holo-acardius amorphus*).

factor in the development of the acardii.[1] His hypothesis, however, that the heart of the sound twin gradually brings to a standstill the circulation of the other, is certainly false.[2] It is, further, erroneous, as he refers the time of the formation of the malformation to a period in development at which the heart is already formed. The heart, according to his view, undergoes secondary atrophy from imperfect nutrition produced by thrombosis of the coronary arteries. Ahlfeld has shown, however, that the development of this malformation must be referred to a very early period of development. He explains it in the following way (see his 'Textbook of Midwifery'): 'At the end of the first week of development, the connective tissue of the allantois grows out with considerable rapidity, from the insertion of the abdominal stalk to the inner surface of the chorion, and sends connective-tissue stems, containing fœtal vessels, into the chorionii villi. If two fœtuses are situated within the one chorion, the two allantoides distribute themselves almost equally over the area available, and homologous twins are formed, which share the placenta in equal or almost equal parts. But if the allantois of the one precedes in its development—in even a small degree—that of the other, then the first appropriates the placental area completely, or almost completely, for itself. In its development, the second allantois no longer finds any unappropriated part of the chorion, so that it has to unite with the allantois of the first; or it may unite with a small area of the chorionii villi, but its major portion is inserted into the other allantois. The second fœtus, with its imperfectly situated area of nutrition, will infallibly be stunted in its growth; and, indeed, not infrequently dies if the blood in the allantoic vessels of the first fœtus does not find its way into the body of the second fœtus, whether this occurs through the united allantoic vessels which are in communication with one another, or whether the reversal of the circulation in the allantois occurs in the existing vessels of the second fœtus. Thus the blood in the vessels of the first fœtus[3] passes into the body of the second

[1] Hempel was, however, really the first (in 1850, as Schwalbe shows) to recognise the reversal of the circulation, and to point out that the blood flowed to the acardiacus through the umbilical arteries and from it through the umbilical veins.

[2] He, however, recognised the two main grounds on which later hypotheses have been founded: namely, that the failure of the heart and other organs is secondary, and that the acardiaci are derived from normal twins.

[3] If the allantois of the first fœtus entirely surrounds that of the second, the allantoic vessels of the two will lie in intimate contact, and union between them will readily occur. The ultimate result will be that the first fœtus will possess a well-formed placenta, while the second will not have any placenta at all, and its umbilical cord will be inserted into that of the first. If, further, the amnion of the second fœtus remains small, then its vessels will pass to the cord of the first fœtus along the membranes—that is to say, they will have a velamentous insertion. If the vessels of the second fœtus unite with a small area of the placenta, as well as with the allantois of the first, then the ultimate result will depend upon which of these two circulations is the stronger, as in the end this one will prevail and the other will atrophy and disappear.

fœtus, and the nutrition of the latter fœtus is at this stage either carried out by two allantoic circulations proceeding simultaneously, or the imperfectly nourished circulation of the second fœtus ceases, and it is entirely nourished by the first fœtus. The heart of the second fœtus, then, only develops so long as a special circulation persists; if this ceases, it dwindles; and later, either fails altogether or remains in a rudimentary form. In the same way, the organs situated round the heart are not developed—the lungs, diaphragm, liver, etc.; while those organs are well developed which lie at the point of entrance of the allantoic vessels into the body of the second fœtus and in the pelvic cavity; and so the pelvis, the genitalia, the kidneys, intestine, and the lower extremities are relatively well developed in the majority of the cases.' According to Ahlfeld's view, an acardiacus is to be regarded, therefore, as an allantoic or placental parasite (Gemellus placento-parasiticus). I have entered into a detailed description of Ahlfeld's hypothesis because it has found many supporters.

Schatz, who has discussed this question in numerous publications—based, mainly, on the ground of very painstaking observations—regards this view as incorrect, and numerous authors have expressed their agreement with him. According to Schatz, the conception of Ahlfeld—that the overpowering of one heart takes place in the extensive capillary anastomosis of the two allantoides—is physically and physiologically unthinkable.[1] He follows this idea out further, and comes to an essentially different conclusion, which is founded on the theory, put forward by him, of a third common circulation, which plays a part in the ætiology of hydramnios. Without entering here into the discussion, the most important points of Schatz's theory must be mentioned. This third circulation is formed, in uniovular twins, at the area of contact of the two fœtal circulations in the common placenta. In this area there are a number of chorionic villi—a zone of anastomosis—common to both twins. Here the changes are enacted which produce the reversal of the blood-stream of the passively participating twin, so that its circulation becomes an off-shoot of the active twin (see Ahlfeld). Schatz's own hypothesis may be stated in the following terms: The first postulate necessary for the development of an acardiacus is some obstruction in the venous flow of the first twin (A)—namely, in the fœtus itself or in the umbilical cord. The arrested blood finds a way out through the venous channels to the second twin (B). As a result of this,

[1] Schatz recognises three varities of acardii:—

(*a*) Acardii, in which the heart has not developed at all.

(*b*) Acardii, in which the condition first appears after the heart has been formed and commenced to functionate, and in which it arises from certain changes in the vitelline circulation—at times associated with primary or secondary heart failure; so that an allantoic or placental circulation is never formed.

(*c*) Acardii, in which the condition occurs after the formation of the allantoic or placental circulation, and is produced by some change in it.

A contains a lessened quantity of blood, while the circulatory system of B is over-filled. The heart of B drives, from its surplus, blood through the arterial anastomosis into the arterial system of A with its weak heart, and brings this heart more or less quickly to a standstill. The acardii (I cannot enter here any further into a consideration of the related macro-cardii, micro-cardii, hemi-cardii, hetero-morphous micro-cardii and macro-cardii) develop according to Schatz :—

1. Occasionally through a primary failure of the heart associated with an arterial and venous anastomosis in the placenta.

2. The great majority, however, arise from a secondary failure of the heart, produced through and by some fault of the circulatory system ; most commonly, through some obstruction to the circulation in the venous outflow from the placenta to the heart of the affected acardiacus. This obstruction is generally situated at the umbilicus, or in the veins of the umbilical cord. Schatz gives a complete table of the acardiaci and their relations.[1]

Marchand, and other recent writers, consider that the acardiaci originate through some primary defect. According to this view, the pre-disposition to the formation of an acardiacus is already present before there can be any question of an anastomosis of the vessels. According to Marchand, Ahlfeld's hypothesis no longer has any value at the present day, as it does not take into account the advances in the field of embryology, and especially as a free allantois does not exist in the human embryo. Marchand's theory attributes the cause to an unequal division of the segmentation cells of the two embryonic rudiments which results in an unequal division of the yolk substance and a retardation in the development of the vascular layer of one embryo.[2]

According to Schwalbe, the question as to the genesis of acardiaci cannot be answered in a uniform manner. For each form—indeed, for

[1] The last place in which such an obstruction to the circulation can arise is in the placenta—and here it may be due to thrombosis or hæmorrhage. The time at which (during pregnancy) an acardius may be formed is, according to Schatz, very variable. He thinks that the hemi-acardii may originate at any period of pregnancy ; while the holo-acardii acormi probably originate at a time when both the vitelline and allantoic circulations are in existence. He does not consider that the second late twin would, as Ahlfeld holds, be in any way prejudiced as regards its nutrition ; such a prejudicial effect could only be produced by some hindrance to the circulation.

[2] As Marchand says : 'If one embryonic rudiment is smaller than the other, the development of its mesoderm, its abdominal stalk, and its amnion will certainly be interfered with. If we imagine a primarily maldeveloped embryonic rudiment, with an imperfect attachment to the chorion or to the abdominal stalk of its co-twin, then a more or less imperfectly formed fœtus, without an independent placenta, will result ; although the more perfectly the smaller embryo is formed the greater the share it will have in the placenta. A certain result of such a mode of development is the formation of a well-marked anastomosis between the vessels of the two.'

each separate form—a special investigation must be undertaken. Generally speaking, only this can be said—namely, that while acardiaci can arise through primary defects of development, yet secondary degenerations may also play a rôle. In all well-marked defects, the fact that they occur at a very early period of development must be accepted.

Schwalbe, indeed, attributes very great importance to the theory of primary defect, for he says: 'For the holo-acardii, and a large number of the hemi-acardii, a primary defect or a very early partial destruction of one germ must be invoked.'

Bauermeister, among gynæcologists, has worked at this question in recent times. He comes to the conclusion that the natural system of acardiaci and their relations, constructed by Schatz, is founded on unproved premises. According to Bauermeister, a primary malformation—a primary acardia—must be assumed for the majority of acardiaci.[1]

V. Winckel advocates an entirely different theory, and Benda and others agree with him. According to this view, these malformations occur from amniotic constrictions. The aplasia of the heart follows as a result of the diminished work to be performed for the upper half of the body. While this explanation cannot be a general one, yet the possible development of acardii in this manner must—at least, in my opinion—be admitted.

Acardiaci have also been observed in cases of triplets, and the cases have been collected by Br. Wolff. According to these statistics, eleven triplet and four quadruplet births are described in which one or two acardii were born as well as the normal fœtuses.

Acardiaci may, in certain conditions, produce severe obstruction to delivery. Hohl writes especially fully on this subject, with a review of the old literature. Acardii may, according to him, occur both in primiparæ and multiparæ.[2] The mothers of these fœtuses are nearly always very fertile women. According to I. F. Meckel, the pregnancy usually reaches its normal end, and the twin born with the acardiacus is quite mature. Of the twenty-two cases collected by Hohl, in eleven cases the time of the delivery is given; of these, in three cases,

[1] The majority of recent writers look upon acardiacs as due to imperfect or arrested segmentation; and, while this might possibly be due to the abnormalities which are present in the circulation, it is more likely that the abnormal conditions of the circulation are the result rather than the cause of such imperfect development. If this view is the correct one, then the error in development must occur at a very early period—before, indeed, the earliest stages of the development of the circulatory system. Experiments on the ova of animals have shown that such formations can be produced artificially; and all the evidence available at present seems to show that acardiaci are the result of the faulty and imperfect development of one half of a twin blastoderm.

[2] Of seven cases recorded in the *Trans. of the Obstet. Soc. London* (in which the information is given), three of the mothers were multiparæ and four primiparæ.

the pregnancy reached its normal termination, and in eight cases it was interrupted prematurely: once in the sixth month, four times in the seventh month, once in the eighth and the ninth months, and once no definite time is given. According to the statistics of Tiedemanns, fourteen of the fœtuses were born at full term and thirty-three prematurely. A footling presentation is the most frequent in acardiaci. In eight of Hohl's cases the presentation was noted, and in all it was of this variety.[1] The well-developed twin is almost always born first, as has been pointed out, and the acardiacus one-half to twelve hours later. In one case, the acardiacus amorphus was expelled only after three days (Hohl). Hohl mentions the following important physical signs as a help in the diagnosis of the acardii during delivery:—

1. The varying and peculiar rudiments of the head or its complete absence.
2. The imperfectly formed or entirely absent upper extremities.
3. The upper conical or rounded end of the trunk, more or less distant from the umbilicus, with or without ribs.
4. The preceding birth of a sound child.
5. The absence of movements after the birth of the first child.
6. The presentation by the feet which are usually abnormal in form.
7. The imperfectly developed genital organs.

Kleinhans rightly adds:—

8. The failure of the fœtal heart, which sign No. 4 naturally pre-supposes.

The signs Nos. 4, 6, and 7 are only of slight value, judging by the recorded cases.

Further, the general œdema of such fœtuses—especially of the lower extremities—so frequently present, must be noted as of importance in the diagnosis; the pressure of the finger forming a pit-like depression. In one of the cases collected by Hohl, in which the trunk terminated at the dorsal vertebræ, the enlarged liver could be felt.

The diagnosis may be extremely difficult when the acardiacus, as rarely happens, does not present as a pelvic presentation. In such cases, fœtal ascites, an enormously distended bladder, tumours situated on the buttocks or on the surface of the body, and sac-like appendages must be thought of. In a case recorded by Raether, the condition was diagnosed as a submucous myoma. During pregnancy, diagnostic signs pathognomonic of these malformations can hardly be present. In some cases, hydramnios is met with. The amniotic sacs are

[1] Of twelve cases recorded in the *Trans. of the Obstet. Soc. London*, four presented by the pelvic extremity, two by the head or upper part of trunk, and in six cases the presentation is not mentioned.

generally of varying size, and the acardiacus has, as a rule, the lesser quantity of liquor amnii (Ahlfeld). Whether in reality (as Kleinhans suggests) the recognition of only one fœtal heart is a sufficient hint for the diagnosis of an acardiacus during pregnancy seems to me doubtful.

In the fourteen cases collected by Hohl, delivery occurred seven times spontaneously, and in seven cases had to be ended artificially: twice by the application of forceps, and five times by the extraction of the acardiacus acephalus. The mother died in two cases: in one, of sepsis, and in another of convulsions. In six cases, the extraction was very difficult from the marked swelling and 'voluminous rotundity' of the trunk and the thickness of the abdominal walls.[1]

In other cases, version and extraction were necessary, and also evisceration; and in further cases, procedures which hardly deserve the name of operations (Pasquali, and others). When the headless extremity of the monster presents in the upper aperture of the pelvis, the forceps is not a suitable instrument for the extraction of the child (as Hohl rightly points out). It readily slips off, if the ends of the blades do not overlap, and, if they do, locking of the forceps is impossible. Different obstetricians, who have attempted to deliver with the forceps, have been compelled on these grounds to perform version and extraction. It is best to undertake these manœuvres first of all, if the headless extremity of the fœtus presents. As the fœtal parts, from the œdema present, are very readily torn during extraction, the cephalotribe must often be applied. In footling presentations (which are very common) extraction should be carried out when the necessary indications are present. This is usually easy, although many cases have been recorded in which, partly from general anasarca and partly from ascites, severe obstruction to delivery has occurred. C. Mayer met with such a case (quoted by Hohl), the delivery of which he describes as the most difficult operation—lasting at least two hours—which he had experienced in his midwifery practice. He delivered the first well-developed child with the forceps. On making a further examination, he found a right foot presenting, and, as extraction was indicated, he tried to carry it out by traction on the foot. As there was some evidence of obstruction, the second foot, lying against the abdominal wall, was drawn down, and, during this manœuvre, the abdomen was found to be of enormous circumference. In this, Mayer found a confirmation of his

[1] Of seven cases recorded in the *Trans. of the Obstet. Soc. London*, in five cases delivery occurred spontaneously; in one case, slight traction had to be made; and in the last case (recorded by Schofield, in 1879) great difficulty was experienced. Traction on the legs resulted in their tearing off, and it was found impossible to extract the breech (which now presented) with the blunt hook: finally, after a very prolonged labour, the markedly œdematous body of the acardiacus was expelled naturally. The mother made a good recovery.

conclusion—drawn from the œdematous condition of the first foot—that the child was dropsical. As a renewed attempt had no result, the perforator was introduced into the abdominal cavity, but only a little fluid escaped. The blunt hook, passed through the opening into the abdomen of the child and pressed against the pubes, was used to reinforce the pull on the feet. As this procedure also led to no result, Mayer introduced the whole left hand into the uterus, and found the trunk, without head or arms, of enormous circumference. Under guidance of the left hand, the perforator was pushed up to the upper part of the trunk, introduced into it, and a considerable quantity of fluid evacuated, and then with the hook and the hand the extraction was completed.

Walther has described a similar case. He was called to a case of hydramnios, with uniovular twins and premature labour at the eighth month. After the birth of the first twin, an excessively large acardiacus gave rise to very severe obstruction to delivery. The practitioner had, by various useless attempts at delivery, torn off the lower extremities. The lower uterine segment was dangerously thinned. Opening the child's abdomen and partial evisceration, undertaken by Walther, led to no result. In the same way, the introduction of a blunt hook into the spinal column, and the rotation of the upper half of the body, failed to effect delivery. Cephalotripsy, with the simultaneous rotation of the upper half of the body on its long axis (a procedure which Walther strongly recommends for such obstructed labours) was, finally, successful—as in Raether's case (see p. 363).

Nacke also met with a very difficult case (described in conjunction with Benda). After the expulsion of the first premature twin, fœtal heart-sounds could not be heard. On internal examination, a soft mass could be felt—the upper end of the fœtus—which was thought to be a breech, by the practitioner. Nacke, who was called in consultation, was able to make the diagnosis of an acardiacus. The thighs were so œdematous and voluminous that they were as large as the trunk. On account of the danger of rupture of the uterus (which was closely applied to the child), Nacke did not dare to bring down the feet. The blunt hook, therefore, was inserted into the fore-coming part of the trunk, and pulled on with gradually increasing force. It tore out, however, several times, and an attempt to open the abdomen failed. At last the monster was successfully delivered with the blunt hook inserted higher and higher up into the body, after the attempts at delivery had lasted for an hour and a half. The anatomical examination of the specimen (Benda) showed the presence of the rudiment of a heart, and marked malformations of the feet. Benda proposed to call the malformation Hemitherium posterius,[1] and accepted as a cause of its formation

[1] A case of hemi-acardius or acardiacus paracephalus.

v. Winckel's theory of the development of acardii through the action of amniotic bands.

Further interesting cases of dystocia with acardiaci have been described by Pasquali, Raether, Albert, and others. Pasquali practised podalic version in a case where the upper end of the trunk presented during birth. During the extraction, a leg tore off at the knee. Delivery was effected at last by the introduction of a blunt hook into the anus, and traction thereon.

Raether diagnosed, in his case, a submucous fibroid: the foetus presented by the upper end of the trunk, so that he could not reach the feet. He attempted, therefore, the removal of the supposed fibroid. A correct diagnosis was first made after the delivery of the trunk.

Lastly, Albert has also described a case of dystocia with acardiacus. Evisceration was necessary: in spite of which, the feet were torn off during the succeeding extraction.

In cases of acardiacus with pelvic presentations and dystocia, the pelvis which is generally well developed—especially the region of the symphysis—gives a good hold for the insertion of the blunt hook, and extraction can almost always be carried out in this way.

If the case is one of acardiacus amorphus, or acardiacus acormus, the same procedure should be adopted as in a case of the removal of a separated head remaining impacted in the birth canal. The use of the combined handgrip, the craniochast, and the forceps must be considered.

In the pathological collection of the Göttingen Women's Clinic there is a good specimen of an acardiacus amorphus (Cat. No. 134), which was presented by a practitioner living in the neighbourhood of the clinic (see Fig. 58). It is an acardiacus amorphus, with velamentous insertion of the cord, which contains one artery and one vein. The specimen came from a multipara—a woman twenty-six years old, who had had four children—and was expelled with a well-developed six-months' living female child, which died on the fourth day after birth. The other children were healthy.

Literature

Schwalbe, *loc. cit.*, p. 15; Strassmann, *v. Winckel, loc. cit.*, p. 15; vol. ii., pt. 3, p. 1726; Ahlfeld, *loc. cit.*, pp. 24 and 34; Hohl, *loc. cit.*, p. 15; Schatz, *Die Acardii und ihre Verwandten* (1898); Olshausen, *loc. cit.*, p. 44, and *Arch. f. Gynäk.*, vol. liii., p. 144; Schatz, *Sitzungsberichte d. naturf. Gesellsch.* (Nov. 1900), No. 5; Spee, *'Referat über Acardiacus und dessen Genese,' Schwalbes Jahresbericht* (1898), p. 227; Elben, *Diss. Berlin* (1821); Claudius, *Die Entwicklung der herzlosen Missgeburten* (1859); Breus, *Wiener med. Jahrb.* (1882); Spiegelberg and Wiener, *loc. cit.*, p. 15; Ziegler, *loc. cit.*, p. 15; C. Mayer, *Verh. d. Gesellsch. f. Geburtsh.* (1846), vol. i., p. 128; Walther, *Zeitschr. f. prakt. Ärzte* (1903), No. 1; Küstner, *Müller, loc. cit.*, p. 15; Runge, *loc. cit.*, p. 15; Kleinhans, *v. Winckel, loc. cit.*, p. 15, vol. ii., pt. 3, p. 1031; Br. Wolff, *Arch. f. Gynäk.*, vol. lix.; Pasquali and Bompiani, *ref. Zentralbl.*

f. Gynäk. (1886), p. 350; Ruether, *Geb. Gesellsch. Hamburg*; Bretschneider, '*Drillinge und Acardiacus*,' *Geb. Gesellsch.* (Leipzig, Feb. 16, 1903), *ref. Zentralbl. f. Gynäk.* (1903), p. 608; Bucura, '*Acardiacus*,' *Wiener Geb. Gesellsch.* (Feb. 9, 1904), *ref. Zentralbl. f. Gynäk.* (1904), p. 1411; Hochheimer, '*Acardiacus*,' *Diss. München* (1904); Bauereisen, '*Acardiacus*,' *Arch. f. Gynäk.*, vol. lxxvii., pt. 3; Benda and Nacke, *Zentralbl. f. Gynäk.* (1907), p. 408; Hunziker, '*Acardiacus amorphus, injiziert mit Quecksilber*,' *Beitr. z. Geburtsh. u. Gynäk.*, vol. iii., pt. 3; Schubert, '*Acardiacus*,' *Gynäk. Gesellsch.* (Breslau, Jan. 21, 1908), *Mon. f. Geburtsh. u. Gynäk.*, vol. xxviii., pt. 3; Kehrer, '*Acardiacus mit Lebergeschw.*,' *Arch. f. Gynäk.*, vol. lxxxv., pts. 1 and 2; R. Meyer, '*Amorphus*,' *Geb. Gesellsch.* (Berlin, April 3, 1908), *ref. Zentralbl. f. Gynäk.* (1908), p. 1453; Taruffi, *loc. cit.*, p. 44; vol. ii., p. 248; vol. viii., p. 440; Ballantyne, *loc. cit.*, p. 44; Keith, *Trans. of the Obstet. Soc. London* (1901), vol. xlii., p. 99; Spencer and Lawrence, *loc. cit.*, p. 98; Herman, *Trans. of the Obstet. Soc. London* (1893), vol. xxxiv., p. 2.

CHAPTER XXI

Malformations of Extra-uterine Fœtuses

It is obvious that fœtuses developed outside the uterine cavity may be malformed in various ways. The literature on this subject is not very extensive, and we have to thank v. Winckel chiefly for a comprehensive study of the matter. Some observations are also to be found in the monograph of Werth on Extra-uterine Gestation. As regards the frequency of these malformed fœtuses there are very divergent opinions.[1] Some authors regard them as very rare; others, as not so rare; and others, again, including Winckel, as comparatively common—occurring in at least 50 per cent. of the cases. Thirteen cases met with by v. Winckel all showed, without exception, definite malformations.[2] Most of these consist of deformities and other disfigurements as the result of external causes. According to v. Winckel's investigations, the head is primarily affected. In this are found, mainly, changes of form, displacements, flattening, compression, slight depressions, and fractures of the bones; and also hydrocephalus, hydromeningocele, or hemicephalus. Of eighty-seven cases collected, alterations in the form of the skull were found in 66 per cent. and hydrocephalus in 10·4 per cent.

V. Winckel considers hydromeningocele, encephalocele, and spina bifida—which, according to him, are various stages of the same affection

[1] Among the cases of extra-uterine gestation operated upon in the later months of pregnancy (collected by Sittner), of 122 children seventeen were malformed, and in five cases the malformation probably played a part in causing the death of the fœtus. Of seventy of the children, thirty-seven were less developed, fourteen were of the normal size, and nineteen were larger than intra-uterine fœtuses of the same period of development. Of the 122 children, fifty-nine died within the first four weeks, and sixty-three (or more than half the number) survived for a longer period than this. Of the sixty-three who lived, eighteen were lost sight of, fourteen died within the first year, eight in the second year, and the remaining twenty-three were alive and well, at varying periods, after the operation.

[2] The majority of extra-uterine fœtuses which reach full term, or die in the latter months of pregnancy, are less well developed—both as regards their length and weight—than intra-uterine fœtuses of the same period of development. The largest extra-uterine fœtus described is one delivered by Hamilton by laparotomy, which weighed 5 km. (11 lb.).

as all due to the same cause; namely, the result of marked and long continued compression of the internal and external jugular veins, and compression of the thorax—which, as the pressure of the chin against the thorax becomes more marked, must always be increasing in amount. According to this view, these diseases are to be regarded as the results of congestion.[1]

Next to the head, the breech and the extremities are most commonly affected. The most frequent condition is pes varus. The frequently associated pes valgus is generally a result of the pes varus (see v. Winckel). Incomplete and complete fractures may also be observed in the lower extremities as well as in the upper.

In the region of the thorax and abdomen, kyphosis and scoliosis,[2] umbilical hernia with eventration (Werth), spina bifida, and hypospadias have been described. In 5 per cent. of the cases collected by v. Winckel, amniotic constricting bands were present.

According to this author—besides the limitation of room—a small amount of liquor amnii, changes in the placenta, and transitory contractions of the embryonic fetal sac must be included among the etiological factors. Werth cannot admit the contractions of the sac as a cause, but thinks that the other factors are sufficient explanation.[3]

The fact must be mentioned that the well-marked deformities, especially of the head, which have been met with in some cases after a short period of extra-uterine life, may completely disappear.[4]

With regard to the part played in the etiology by amniotic

[1] Of v. Winckel's cases, the head was affected in 75 per cent., the pelvis and lower extremities in 50 per cent., the upper extremities in 40 per cent., the thorax, abdomen, and genitalia in 3 to 4 per cent.

[2] Specimen No. 7118, Univ. Coll. Hosp. Med. School Museum, is a six months' male fœtus, which was removed alive from a case of secondary abdominal pregnancy—probably following tubal abortion. It measures thirteen inches in length; the left side of the lower jaw and face are compressed; the legs are bowed, and the feet are in a condition of pes varus; the toes of the right foot are twisted; and there is marked lateral curvature of the spine.

[3] Werth is inclined to believe that some of the cases, in which the wall of the sac contained a great deal of muscle tissue, were in reality cases either of interstitial gestation or of pregnancy in the undeveloped horn of a unicornate uterus.

[4] In a case recorded by Taylor, the child weighed seven pounds, and was fully developed. There was a marked depression over the anterior portion of the right parietal bone, and the tissues over the larynx and anterior part of the neck were also depressed, there was talipes calcaneo-valgus of the right foot, and genu recurvatum of the right knee. Four months after delivery, the shape of the head is said to have undergone considerable improvement; but the child died suddenly of convulsions, some five months after its birth; so that probably some injury to the brain was also present.

constricting bands, Werth[1] does not agree with the suggestion put forward by v. Winckel that mechanical causes are of chief importance. He believes rather that disturbances of nutrition play an important part, and that the ovum, embedded in the tube, suffers from the unfavourable conditions of the circulation, from the hæmorrhages, and the deeply spreading zone of necrosis of the tissues at the periphery of the embryonic implantation cavity.

Literature

V. Winckel, *Über die Missbildungen von ektopisch entwickelten Früchten und deren Ursachen*, *loc. cit.*, p. 15 (1902); Werth, *Die Extrauterinschwangerschaft*, v. *Winckel*, *loc. cit.*, p. 15, vol. ii., pt. 2, p. 783; Taylor, *Trans. of the Obst. Soc. London* (1891), vol. xxxiii. p. 115; Sittner, *Archiv. für Gynäk.* (1898), vol. lxxxiv. p. 1.

[1] Werth argues that, in the majority of cases, the fœtus, in the early months of an extra-uterine gestation, is as well developed as an intra-uterine fœtus of the same age, and thinks that the factors which interfere with its further normal development mainly come into play in the later months of pregnancy. Further observations on the exact anatomical condition of young fœtuses from extra-uterine gestation sacs are necessary to prove this point.

CHAPTER XXII

The Legal Rights of Monsters

Very little information on this question is to be found in the text-books and manuals devoted to medico-legal medicine, and surprisingly little elsewhere.

In olden times, the fate of all malformations was a sad one.[1] The Romans were allowed to kill a monster after it had been inspected by five neighbours; and even this formality was not necessary, according to the Laws of the Twelve Tables.[2] According to Hohl, the natives of Peru kill all malformed children. Whether these observations—now some sixty years old—still hold good at the present day, I am unable to say. According to the laws of later times, it was not allowable to kill malformed living children. In accordance with this view, one school of obstetricians maintained that the malformed child should be safe-guarded as far as possible, even *in utero*; and that, therefore, perforation and such mutilating operations should be avoided. For this reason they recommended Cæsarean section, when necessary, so as to preserve the life of the malformed child. Happily, this error of obstetrical practice was not allowed to remain long unrectified.

In the earlier laws, a distinction was made between Monstra and Portenta. As Monstra, were distinguished those beings which were born of a woman, but had no human head. They were held not to be human; legal rights were completely denied to them, and they were regarded as not competent. In distinction to these, those beings were termed Portenta whose appearance was human, but who were deformed in any way; these were allowed complete legal rights. To this class a large number of all malformations belongs—namely, fœtuses with maldeveloped sexual organs, and a larger or smaller number of

[1] As Ballantyne points out, the birth of a monster during the Early and Middle Ages was a Divine warning—it meant that a deity must be propitiated; and no doubt this was one of the reasons why—at any rate, in Europe—such monstrosities were almost invariably killed.

[2] The Twelve Tables of the Roman Law, set up under the presidency of Appius Claudius, 303 B.C.

limbs. In the Common Right, which existed in Germany before the introduction of the Civil Law Book (based on the Corpus juris Justinian),[1] the prevailing view was, that the question as to whether a malformation should be given legal rights or not, depended upon whether it did or did not possess a human form.[2] If it had not human form, then it was a monster, and had no legal rights. Even after the establishment of the Common Book of Laws, the question, from the scientific aspect, was not settled. The Civil Book of Laws contains only the definition (see sect. 1) · 'The legal right of man begins with the completion of birth.' While a few writers (Cosack, 'Text-book of the Civil Laws,' vol. i., p. 60; Kuhlenbeck, 'From the Pandects to the Civil Book of Laws,' vol. i., p. 80) assume that one cannot speak of a man, when a living fœtus is born which is so undeveloped that it has not the form of a human being, and that therefore no legal rights can be ascribed to it; yet the prevailing opinion (see Planck, 'Commentary on the Civil Book of Laws,' vol. i., p. 58; Gareis, 'Commentary on the Civil Book of Laws,' p. 6; Staudinger, 'Commentary on the Civil Book of Laws,' vol. i., Annot. to sect. 1) is based on the standpoint that the Civil Book of Laws does not recognise the possibility of the birth of a monster. Everything which is born of a woman is rather regarded as human, and therefore has legal rights, both as regards the penal and the civil laws. The mother, therefore, who kills her monstrous child, is punished for murder (sect. 211, 'Book of Civil Laws').[3] If the mother die during the birth, she will be succeeded by

[1] The Justinian Code of Roman Law, A.D. 528.

[2] The English law is thus stated by Blackstone: 'A monster which hath not the shape of mankind, but in any part evidently bears the resemblance of the Brute Creation, hath no inheritable blood and cannot be heir to any land, although it be brought forth in marriage; but although it hath deformity in any part of its body, yet if it hath human shape it may be an heir.' This, he adds, is a very ancient rule in the law of England; and he observes that the Roman law agrees with our own in excluding such births from succession, yet accounts them, however, children, in some respects, where the parents (or, at least, the father) could reap any advantage thereby, esteeming them the misfortune rather than the fault of that parent. But our law will not admit a birth of this kind to be such an issue as shall entitle the husband to be tenant by the courtesy, because it is not capable of inheriting.

In the case of a man marrying a woman having possession of an estate of inheritance, and by her having issue born alive and capable of inheriting her estate, the man, on the death of his wife, holds her lands for life as a tenant by the 'courtesy' of England.

[3] The law knows no such principle as 'Euthanasia.' As Mr. Justice Hawkins laid down: 'It is equally criminal to accelerate by one hour the death of a person as to cause it.' No degree of monstrosity or unshapelinces sanctions the destruction of life born of woman—either by medical attendant or friend. The question of monstrosity does not arise, therefore, in criminal cases (as in trials for infanticide, or abortion), but only in civil actions (Tidy). In a French case, for child murder, the prisoner was acquitted because the child was acephalous!—but no such decision is recorded in our English Courts.

the monster, even if it is not capable of surviving[1] and dies within a short time. The monster is in this case succeeded by his relations (father, sisters, etc.). The legal situation is, of course, a different one if the monster is born dead. In this case the occurrence of the birth is legally of no importance as the dead fœtus has no legal protection.[2]

As regards malformations of the genital organs (Hermaphroditism)[3] the earlier laws recognised so-called Neuters—that is, persons of no sex (Prussian General State Laws of 1794). According to the 'Book of Civil Laws,' they are reckoned as of the predominating sex. Thus anyone who claims a certain sex must in the private right prove that he belongs to it. In private right, the distinction of sex—and therefore the question whether any person is masculine or feminine, apart from family rights (validity of an engagement or marriage)—is

[1] It must be remembered, that in these cases, the meaning of 'born alive' is different from the meaning of the same expression when used respecting infanticide. In questions of tenancy by the courtesy, it has been decided that any kind of motion of the child—such as a twitching and tremulous motion of the lips—is sufficient evidence of live birth. As regards infanticide, proof of a conclusive separate existence of the child is demanded, before live birth is admitted.

[2] In all cases where questions of law are involved, it will be best for the medical man not to attempt to give an answer to the question as to what is implied by the phrase 'not possessing the shape of mankind,' but to content himself with a full description of the deformity and to leave to the Court the responsibility of deciding the question. It is fairly certain that none of the human monsters which have survived their birth for any length of time could be denied to have had human shape.

[3] The determination of the true sex may be a matter of considerable difficulty; and, as we have pointed out in considering hermaphroditism—in cases in which it is of great importance that the question should be decided one way or the other—an exploratory incision, to determine with certainty the nature of the sexual glands, might be justifiable. The French law allows a child to be registered as of indeterminate sex; but the English law does not recognise such persons, and all children must be registered either as male or female. In cases where no definite diagnosis has been made, they are entered as boys at Somerset House. It is said that, on an average, some three persons a year apply to the Registrar-General to have their sex altered. Legally, persons cannot choose which sex they will be, but it must be fixed for them by medical opinion.

In the case of a monster, if it had been shown that the individual was possessed 'of the shape of mankind,' and was therefore capable of inheriting—in the case of a title or entailed inheritance of land—the sex would have to be determined as well as the capability to inherit. In the case of an entailed estate, if only one child was born—and that an hermaphrodite—it would be necessary to decide 'the sex that doth prevail'; for, 'according to the sex that doth prevail, so will it succeed.' If two children were born, and the second was a male, then, if in the first (the hermaphrodite) the male peculiarities prevailed over the female, he would succeed; but if the female peculiarities prevailed over the male, then the second child, being a male, would succeed. It is obvious that in such a case an exploratory incision might be of the greatest importance. In the case of the marriage of such persons, the only question the law takes into account is that of the ability to consummate the marriage.

of no great importance. It is otherwise, however, with respect to public rights. For example, as regards penal rights, in such questions as the following: rape (only possible on a female), seduction of a girl under sixteen years of age; and, as regards civil rights, the question of a vote and liability to military service.

The legal position of the so-called Siamese twins is exceedingly doubtful. They have legal rights in any case, and it is to be assumed that each one has them separately. The one can therefore inherit rights on the ground of succession, which the other cannot. If one were bodily injured, while the integrity of the other was not involved, the demand of the first would be sufficient for criminal proceedings ('Book of Civil Laws,' sects. 223, 232). The following question might be very interesting: How would the judge decide whether one of the twins had been guilty of a crime, or whether one of them had committed a murder, intentionally?[1]

In proof of the fact that judicial questions are not quite of an idle nature, Henneberg and Stelzner report that on a railway journey undertaken by the pygopagi 'Rosa' and 'Josefa,' their manager took only one ticket for the two. The railway authorities required two. The manager took the matter into Court, but lost his case, and was compelled to pay the costs—amounting to about 1600 francs.

As the two twins have an active interchange of the fluids of their bodies, the vaccination of one would no doubt suffice.[2] We may suppose, however, that Baptism would have to be administered to both.

[1] The original Siamese twins, 'Eng' and 'Chang,' were undoubtedly two independent individuals, both mentally and physically. Their original complete independence of thought became, by long habit and experience, less apparent, and they developed a mutual interdependence, which was one of the most striking features of their characters. There can be no doubt that any action performed by one of them would have had to be considered as the result of entirely independent mental processes, in which the volition of the second need not have been in the least degree involved, except in so far as it might be necessary for the physical accomplishment of such an action.

[2] The Siamese twins suffered from smallpox, measles, and ague at the same time, and the paroxysms of ague occurred synchronously in the two. No doubt vaccination of the one would have protected them both, although iodide of potassium administered to one could not be detected in the urine of the other.

INDEX

F

G

H

I

J

K

L

M

N

O

P

R

THE END

PRINTED BY
SPOTTISWOODE AND CO. LTD., COLCHESTER
LONDON AND ETON