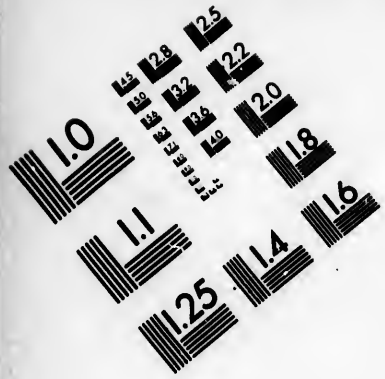

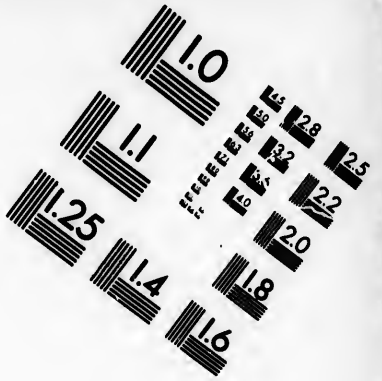

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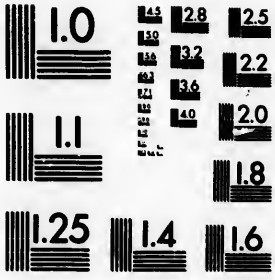

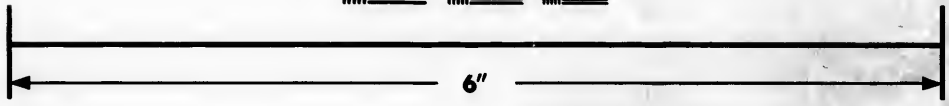

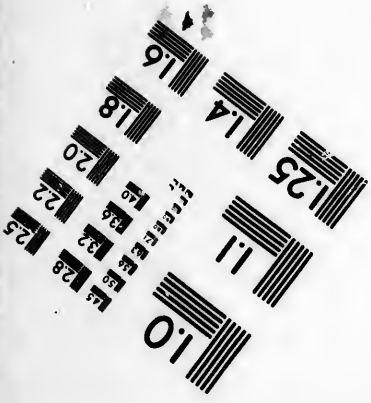

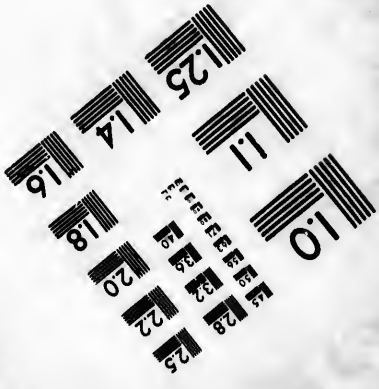

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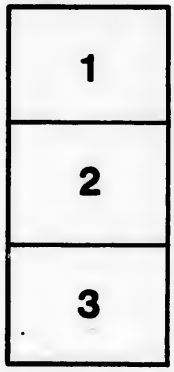

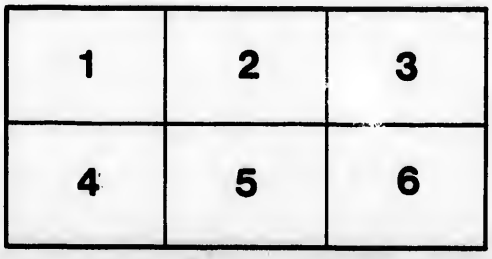

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Stewart J.

ABSTRACT OF A CLINICAL LECTURE ON A CASE OF THE "JUVENILE FORM" OF PROGRESSIVE MUSCULAR ATROPHY (ERB'S "DYSTROPHIA MUSCULARIS PROGESSIVA").

BY JAMES STEWART, M.D.

Professor of Materia Medica and Therapeutics, McGill University, Montreal; Physician to the Montreal Dispensary; Director of the University Dispensary for Diseases of the Nervous System.

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GENTLEMEN,—The patient whom I exhibit today, through the kindness of my friend, Dr. Wilkins, presents in a very marked degree all the essential features of a disease which has only recently been described. The case is one of what Erb calls the "Juvenile Form" of progressive muscular atrophy.

The patient is a male, aged 21 years. His occupation, up to the time he was compelled to cease working from his present trouble, was that of a farm laborer. He complains of weakness of his back and legs. He first noticed this weakness three and one-half years ago. At that time he experienced difficulty in dragging his body after his feet when getting into a carriage or in ascending a stair. He could, at this time, raise his feet without difficulty, but to move his body, he found it necessary to use his hands to drag himself along. About two years ago he first noticed that he was very apt to fall, and on at-

tempting to rise from the horizontal position he found it necessary to use his hand to drag himself up. He never suffered from any serious illness. He attributes his present trouble to a fall which he received three and one half years ago. On careful enquiry, however, he acknowledges that for a long time previous to this accident, he disliked ascending a stair, because he found it both difficult and tiresome. Two and one half years ago he received a second injury; on this occasion a weight fell on his head, rendering him insensible for half an hour, and giddy and stupid for several days. His parents are dead, but cause of death is unknown. He has a brother living and in good health. Had no sisters. As far as he knows there has been no similar trouble to his in any of his relations.

Present Condition.—You will notice the peculiar gait which he assumes when he walks across the floor: 1st, he walks with his feet far apart; 2nd, he walks on the front part of his feet, the heels being raised from the floor, and 3rd, the gait is of a more or less waddling character. Nothing abnormal can be found in connection with the circulatory, respiratory, digestive or genito-urinary systems. There are no symptoms of any affection of the brain or cranial nerves.

On stripping the patient the marked difference in the size of certain muscular groups is at once noticeable. In the upper extremities, the contrast between the well developed muscles of the forearms and the atrophied ones of the upper arms is

very striking. The circumference of the thickest part of the upper arms is an inch less than the fore arms. A still greater disproportion exists between the muscles of the thighs and those of the legs, the circumference of either calf being an inch greater than the circumference of either thigh at the thickest part. The following muscles of the upper part of the body are in a state of more or less complete atrophy: The pectoralis major and minor, of each side, are considerably atrophied, especially the costo-sternal portion of the former. The lower half of each trapezius has almost entirely disappeared. There is scarcely a trace left of the rhomboids. The latissimus dorsi of each side is very much atrophied, as is also the whole group of the spinal extensors. The biceps of each arm is greatly wasted, and what there is left of it is in a state of active contraction, preventing the full extension of the arms. The brachialis anticus of each arm is also in a state of advanced atrophy; the triceps is only slightly affected. The coraco-brachialis, the supra and infra-spinati, as well as the deltoids, are normal. None of the muscular groups in the fore-arms or hands have suffered.

In the lower extremities the following muscular groups are in a state of more or less complete atrophy: The glutei of both sides, and the ilio-psoas. The quadriceps of each thigh is more extensively atrophied than any other group in the lower extremities. The peronei of the right side are considerably atrophied, while those of the left side

have escaped. The calf muscles are hypertrophied. When the patient is in the erect posture there is marked lordosis. All the atrophied muscles are firm. They are not the seat of any fibrillary twitchings. The patient is quite unable to raise himself from the horizontal to the erect position, even with the aid of his hands. He, however, can accomplish this by getting a support to his chin, and thus using the muscles of the neck to drag his body upwards. The patellar reflex is absent. The plantar reflex is exaggerated, while the cremaster and abdominal are normal on the right side and absent on the left. The epigastric reflex is present, but the scapular is absent. The atrophied muscles do not respond to the faradic current. They are *not*, however, the seat of the degeneration reaction. Sensibility is normal. There is no interference in the vesical or rectal reflexes.

You will at once notice the striking difference there is in the patient before you, and the one* whose case we enquired into last week, and whom most of you have seen. When comparing these two cases, it is at once observable that we have to do with dissimilar clinical pictures, although they are both frequently described as one and the same disease. The following are the marked points of difference between them: 1st, they differ as to the localization of the atrophy. In the patient affected

* The patient referred to is a man, aged 37, who has the ordinary spinal variety of progressive muscular atrophy. The wasting commenced three years ago in the small muscles of the left hand.

with the spinal variety of the disease, the atrophy commenced in the small muscles of the hand, in the interossei, thenar and hypo-thenar groups. The wasting is confined to these small muscles. In this patient the atrophy affects the trunk muscles principally, while the hand muscles are perfectly free from any form of wasting. They differ also as to the condition of the affected muscles. In the spinal case they are soft and flabby, while in our patient here they are firm, hard, and have a knotty feeling. In the man previously seen, the atrophied muscles are the seat of fibrillary twitching, while the muscles in this boy's case are free from these fibrillary movements. Another marked difference is that in the case of the spinal form there is neither true nor false hypertrophy of the muscles, while there is here, especially in the calf. Other points of difference are the ages at which they make their appearance. The spinal form is essentially a disease of advanced adult life, while the juvenile form is seldom or never seen after the twentieth year. They are both slowly progressive diseases; the juvenile is, however, much slower than the spinal variety. In the latter the periods of intermission are comparatively short and seldom, while in the former they are long and frequent. They differ also as to the complications that may arise during their course. Last week, when we were examining the patient affected with the spinal form, I pointed out to you that there was marked trembling of his tongue when he protruded it This is sufficient evidence that there is commenc-

ing bulbar paralysis in his case, and is the beginning of a series of symptoms that will before very long lead to a fatal ending. In the patient before you no such complication exists. In all the cases of the juvenile form of progressive muscular atrophy described up to the present, no such complication has existed. Secondary sclerosis of the pyramidal columns is not infrequent as a result of the changes that take place in the spinal form. It does not occur in the juvenile form. When we come to discuss the pathology of the disease, it will then be clear to you why these complications are so frequently present in the one case and never present in the other. Another marked point of difference between these two forms of atrophy is the fact that one is much more amenable to treatment than the other, the juvenile form being much more likely to have a favorable ending than the spinal.

They differ also in their pathology. In speaking last week of the appearances found post mortem in the spinal variety of the disease, I mentioned that the essential change was a slowly progressive obliteration of the multipolar cells in the anterior horns of grey matter of the spinal cord. The local muscular changes were simple atrophy of the muscular fibres. There is no increase of connective tissue, no deposition of fat, and no hypertrophy of the muscular fibres. Now in the juvenile form the changes are wholly seated in the muscles. The multipolar cells of the anterior horns of grey matter remain free, as do also the peripheral nerves.

The muscular changes consists in atrophy of the muscular fibres, with here and there fibres which have undergone hypertrophy. In advanced cases hyperplasia of the connective tissue is very marked, and lying between the connective tissue fibres is seen only a small quantity of muscular fibres in an advanced state of atrophy, which, however, still retain their transverse striation. The most important change is the hyperplasia of the interstitial connective tissue, and next to this is the deposition of a more or less quantity of fat. It is probable that the increase in the muscular fibres is the first phase of the morbid change, and that the later appearing connective tissue hyperplasia gives rise to atrophy of the muscular fibres. These changes, as we will presently discuss, are essentially those found in cases of pseudo-hypertrophic muscular paralysis, and the so-called hereditary form of progressive muscular paralysis. This hereditary form of muscular atrophy has been described by Friedreich and others, but it is essentially the same disease as we are now considering. When the disease is hereditary and sets in about puberty, the muscles affected are those of the upper arms and trunk, while if it sets in during childhood the atrophy is principally confined to the muscles of the lower extremities.

The disease commonly called pseudo-hypertrophic muscular paralysis, differs but little, if at all, from the disease with which the patient before you is affected. Clinically, the only difference appears to be, that in the pseudo-hypertrophic paraly-

sis, we have lipomatosis, while in the juvenile form of muscular atrophy, hypertrophy is not necessarily present, and if present it is true and not false. If this is the only difference it is quite plain that it would be better to describe the juvenile form of muscular atrophy as being sometimes attended with a true and sometimes with a false hypertrophy of the muscles, rather than describe two separate diseases. Pathologically there is no difference between them. They are both myopathic and *not* neuropathic disorders. All the recent autopsies in cases of pseudo-hypertrophic muscular paralysis agree in the particular that no changes in any portion of the spinal cord are present, the changes found being confined to the muscles and differing in no way (except in a great degree of lipomatosis) from those described as being present in cases of the juvenile form of muscular atrophy. Changes have been described as being found in cases of the pseudo-hypertrophic paralysis in the ganglion cells of the anterior horns, but this was some years ago, and before the much improved methods of the histological examination of nervous tissue were known. Seeing that in a number of recent cases examined by such competent observers as Recklinghausen, Schultze, and Ross, where improved methods were made use of, it follows that little or no value can be attached to the alleged changes found by the observers of even a few years ago.

Erb is a firm believer in the essential identity of these two diseases. Speaking of the juvenile form

of muscular atrophy he says * "there is a particular form of disease of the muscles which consists partly in hypertrophy with subsequent atrophy of the muscular fibres, partly in hyperplasia of the insterstitial connective tissue with more or less lipomatosis. Whether the changes in the muscular fibres or in the connective tissue is the primary event, or whether they are simultaneous appearances has not yet been definitely settled. There are no changes in either the peripheral or central nervous system. It is a very chronic and slowly progressive trouble. Clinically the disease is characterized by affecting in the upper part of the body, the pectoral, the trapezii, latississmi dorsi and other shoulder muscles, the muscles of the upper arm, while those of the forearm and hand escape. In the lower part of the body the muscles that suffer are those of the abdomen and the extensors of the back, the muscles of the thigh, calves, and the peroneal group. Cases of this disease in the past have been mostly described as ordinary cases of progressive muscular atrophy, a few as pseudo-hypertrophic muscular paralysis and hereditary muscular atrophy. If the disease appears in the earliest childhood, and if there is no lipomatosis it is what has been called hereditary muscular atrophy. If there is a high degree of lipomatosis, especially of the lower extremities it is what has been called

* Erb: Ueber d. juvenile form d. progressiven Muskelatrophie u. ihre Beziehungen zur sogen. Pseudo-hypertrophie d. Muskeln-Deutsches Arch. f. Klin. Med. xxxiv. 5 u. 6 p. 467.

pseudo-hypertrophic muscular paralysis. These three, hitherto separately named affections, are in reality one and the same disease. It is quite a distinct disease from the spinal form of progressive muscular atrophy." It follows therefore, according to Erb, that there are two distinct forms of progessive muscular atrophy—a neuropathic form and a myopathic form. In the patient whose case we examined into last week, we had a good example of the neuropathic or spinal form. The patient before you now is a good example of the myopathic form. For the former or neuropathic form of the disease Erb proposes the name "*Amyotrophia Spinalis Progressiva*," while for the latter or myopathic variety of the disease he suggests the name "*Dystrophia Muscularis Progressiva.*"

TREATMENT.—Before this patient came under the care of Dr. Wilkins, the atrophy had made such progress, that it was hopeless to expect benefit from any form of treatment. Where the disease is however seen early, there is fair grounds for hoping that in a small number of cases, arrest of it or even recovery may follow well directed treatment. As already mentioned, this form of muscular atrophy is more amenable to treatment than the spinal variety. There are very good grounds for believing that both forms would not be so fatal if more systematic and scientific attempts were made in their treatment. Physicians, as a rule, when they diagnose a case of muscular atrophy, pronounce it both "interesting" and "incurable." Seldom is even the attempt made to prevent the further progress

of the degeneration. In the present state of the therapeutics of this subject, it is not possible in the very great majority of cases to prevent the progress of the disease. The few cases that have yielded to treatment are a sufficient proof that in the near future we will be much better able to combat this degenerative process. I would strongly advise you in all cases of progressive muscular atrophy, but especially in that form of the disease under consideration, to make persistent efforts to cure. The only therapeutic means of any promise is electricity, especially galvanism. The galvanization of the atrophic muscular groups should be performed very gently, otherwise the process may be quickened in place of retarded. It should be continued until it is quite clear that it is useless. Should it be of no effect, faradization of the affected muscles, or even general faradization should be resorted to.