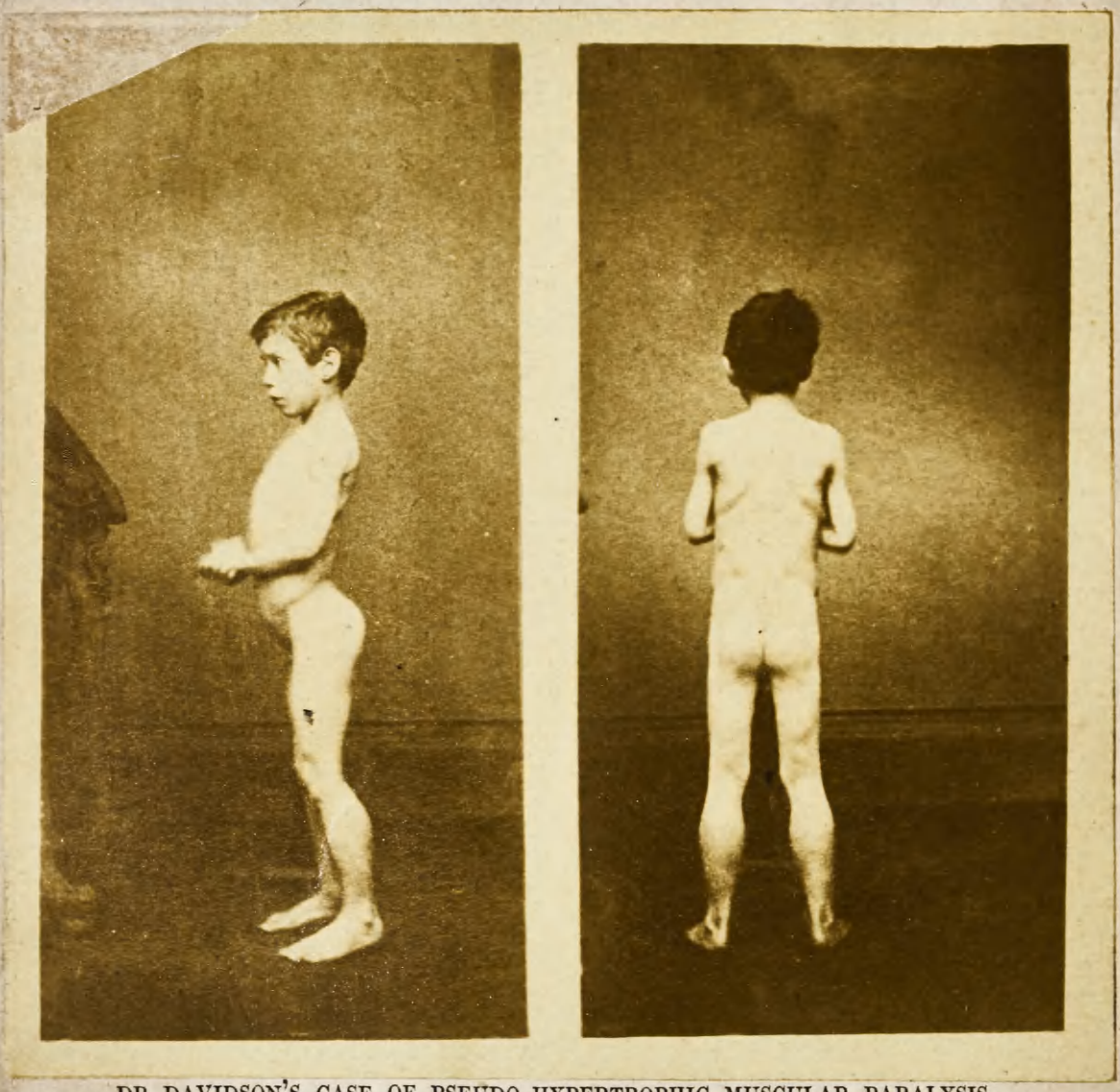

DR DAVIDSON'S CASE OF PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

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Original Articles.

I.—ON PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

(With Photographs.)

By A. DAVIDSON, M.A., M.D., *Physician to the Liverpool Northern Hospital; late Assistant Physician to the Children's Infirmary; Lecturer on Pathology at Liverpool School of Medicine.*

PSEUDO-HYPERTROPHIC Muscular Paralysis is a form of muscular weakness, found almost solely in boys, in which some or all of the affected muscles, instead of being atrophied as in Wasting Palsy (Progressive Muscular Atrophy), become on the contrary enlarged and hard, and so give rise to the very puzzling appearances by which the affection is characterised. The disease is an uncommon one, but its strange characters make it very interesting. As no description of it is, so far as I know, to be found in any English publication, with the exception of a short note by Dr Lockhart Clarke, in the 19th volume of the Pathological Transactions, I purpose in this paper to give some account of the disease; first, relating the cases that have come under my own observation, and then making some remarks on the nature and progress of the malady, these remarks being founded partly on the more extended observations of Duchenne, who first described it.

CASE I.—The first case that came under my notice was that of the boy whose photograph accompanies this paper. He was brought to the Liverpool Children's Infirmary in the autumn of 1871 for advice, on account of the difficulty he had in walking, and his frequent falling as he moved along. When he was stripped for the purpose of examination, it was found, as may be seen in the photograph, that the curve of the spine in the lumbar region was greatly increased, the belly jutting forward and the shoulders being thrown back. This unnatural curve disappeared when he sat or lay down. It was seen also that the muscles of the calf of each leg were much larger and harder than natural, and this was the case also, to a less degree, with the posterior muscles of the thighs, the glutei, lumbar spinal muscles, and the external oblique muscles of the abdomen. The upper half of the body contrasted strongly with the lower, being somewhat emaciated, the upper arms especially being thin and wasted. He stood with his legs apart, and the heels slightly drawn up from the ground. The latter point was more evident when he walked, and indeed resembled a slight talipes equinus. In walking, he had a most peculiar gait,—a waddle,—as if he needed to balance the body first on one leg and then on the other. If he attempted to go quickly, he fell. He could readily stoop so as to touch the floor, but it was with excessive difficulty that he could again raise himself erect, requiring great muscular straining, and having to assist the movement by means of the hands placed on the knees. When sitting, however, he could recover himself from the bent position with comparative ease. The electro muscular contractility of the lower limbs seemed to be diminished, but was not absent. The cutaneous sensibility was normal.

The following is the history given by his mother. He is nine years of age. He was delicate from birth. He did not walk till he was two years old, and always walked feebly. When about five years old, he had two attacks of convulsions without any evident cause. After that time the difficulty of walking increased, and it was then observed that his belly projected. During the last two years he has

lived away from home and out of his mother's observation; and since his return she has found him much worse than before. He never walks any length without frequently falling; and the slightest blow will knock him over. Of late his arms also have shown signs of weakness. His appetite and general health are good. Though his mother states that he is quite intelligent, yet it is evident to me that his mental power is considerably below the average of boys of his age.

This case is an excellent example of Duchenne's paralysis, and was shown by me to the Medical Society of Liverpool last October. He remained under my care in the Children's Infirmary for some weeks, and was treated by the application of faradization regularly during that time, but without the slightest benefit. Since then he has become perceptibly worse, though he is still able to walk about.

CASE II.—A boy, aged 13, came under the care of Dr Stephens, my colleague, at the Children's Infirmary, who has given me leave to record his case. This boy exhibited the same symptoms as the first, in a less degree. He had the same lumbo-sacral curvature, the peculiar gait in walking, the enlargement of the calves and hamstring muscles, and the inability to raise himself from the stooping posture when he stood or knelt. If, however, he stooped while sitting, he could raise his body again with perfect ease. In his case there was little or no enlargement of the lumbar spinal muscles, or of the glutei. The muscles of his arms were thin and weak. He was in good health and quite intelligent.

His mother's account of him was that he was two years old before he walked, and that for some time he walked weakly; but afterwards well enough. When he was about eight years old he began to show difficulty in walking, and three years ago the present symptoms began, the frequent falling, the jutting out of the stomach, &c. During a residence of several months in the Children's Infirmary some improvement occurred in his symptoms, under the electrical treatment and strychnine.

In this boy's case the disease seems to be hereditary, for

his mother's brother, a watchmaker, is also affected with muscular paralysis. In his case the attack occurred when he was 16 years old, at first merely giving rise to weakness and awkwardness in walking, but leading ultimately to total inability to support himself on his legs. He is now 31 years of age. There are no signs of spinal paralysis. The muscles of his legs are feeble, but not much wasted, and he still uses them and the muscles of his back to propel the chair to which he is confined. The deltoid and the muscles of the upper arm have little power, and are much wasted, but he is able to use his hands freely in following his occupation. It was impossible to make a more full examination of this man's case, or to ascertain any facts as to the earlier symptoms, but I was satisfied that the disease was the same as that of the boy his nephew; though unlike the other recorded cases, it had begun at a much later period of life, and had terminated its progress spontaneously.

CASE III.—A boy, aged 12 years, first seen by me in the end of 1871. His mother's statement was that he had always been weak in his back; that he did not walk till he was 2½ years old, and even afterwards never walked strongly. If he fell, he could not get up again. He grew up in the same state, walking feebly and in a peculiar manner, but otherwise healthy. He never had convulsions. When he was between 10 and 11 years old, he lost the power of supporting himself on his legs at all, and lay helpless. He was then admitted into the Northern Hospital, where he remained six months, and was treated by faradization without benefit. I understand that while he was in hospital he lay in bed helpless, able to move his legs, but unable to stand or walk, and not even to raise himself easily. But his legs appeared plump and well nourished, and the lumbar spinal muscles were so much enlarged as to resemble tumours. His present condition is as follows:—He is very pale and emaciated. He lies helpless on his side in bed, with the knees drawn up, and the toes pointing downwards. The only power of movement he has in his legs is that of slightly flexing the toes. Notwithstanding this feebleness, the muscles of the

calf, the ham, and the buttocks remain moderately large and hard, and though too feeble to move the limb, yet have an appreciable power of resistance. The anterior muscles of the leg are quite inert. The muscles of the back are swollen and hard, especially in the lumbar region, and so also is the latissimus dorsi. In the arm, the flexor muscles of the fore-arm are much swollen, and the triceps to a less degree; the others are wasted. He is able to use his hands so as to play with toys, and by means of his arms and back can assist in raising himself; but he cannot feed himself. He speaks and eats well enough. He has frequent cramps in the muscles at night. He is growing gradually weaker, and suffers now from chronic bronchitis.

These three cases, then, are examples of the disease which Duchenne was the first to discover, and which he has named Muscular Paralysis with apparent hypertrophy, or Paralysis with hardening of the muscles. The disease has also been described by Eulenberg, Heller, and other German writers under the name of Lipomatosis musculorum luxurians. Cases have also been recorded by Mr Adams, Dr Hillier, and Dr Down, in the 19th and 21st volumes of the Pathological Transactions of London. The most complete account of the malady is to be obtained from a series of papers by Duchenne in the Archives Générales de Médicine for 1868.

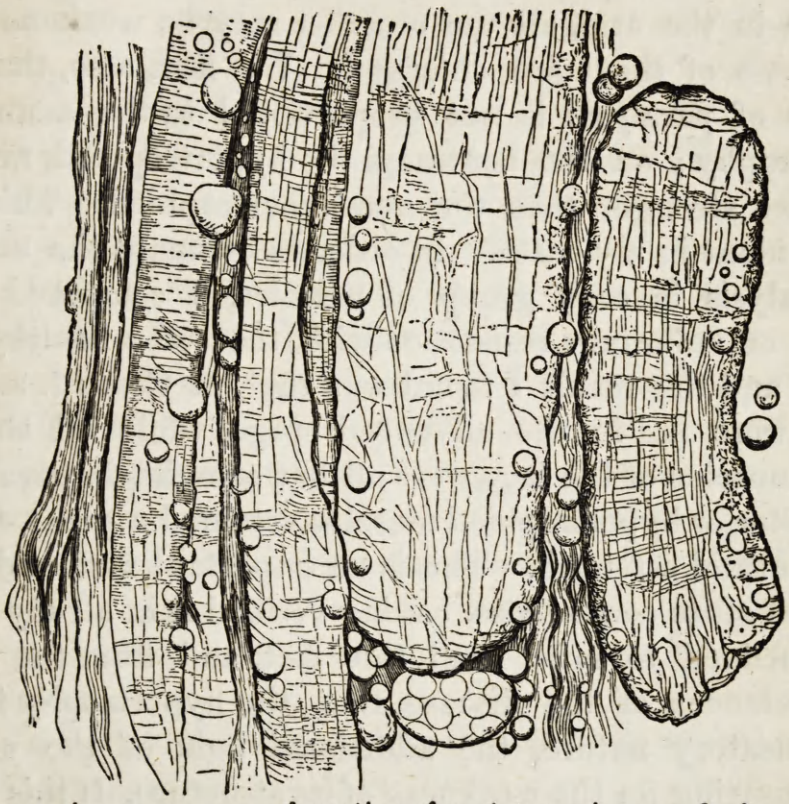
By far the greater number of cases have been observed in boys. Duchenne records one case in a girl, and one or two cases have happened in adults.

What is the nature of the malady, and how are the peculiar symptoms which accompany it to be explained? The parents appear generally to think that the child is suffering from "spine disease," that is disease of the vertebral column, and I have found that medical men have sometimes been led, by an imperfect examination of the case, to express a similar opinion. But if we examine the vertebral column we find nothing abnormal except the unnatural curve which exists at its lower part, and this curve disappears whenever the patient sits or lies down. Again, the disease does not appear to depend on any affection of the

cerebro-spinal axis. It has none of the characters of such affections: and, moreover, in the only case (one of Eulenberg's) where a post-mortem examination has been recorded, Cohnheim, who made the examination, could find no change in the microscopic characters of the spinal cord. It may be that further investigations may discover that the nervous system is primarily involved; but for the present we must look on the disease as one of the muscular structure—a muscular degeneration and consequent paralysis—affecting at first only the lower limbs, but by degrees spreading upwards and attacking the trunk and arms also. Something of the nature of this degeneration may be learned even during life, by removing a small portion from one of the affected muscles for microscopic examination. Duchenne employs for this purpose an ingenious instrument which he has invented, and calls *emporte-pièce histologique*. I have succeeded without difficulty in removing pieces of the muscle for examination in the following way:—After freezing the skin with the spray-producer, I have made an incision into the muscle with a tenotomy knife, and by introducing Lister's probe-pointed forceps into the wound, I have been able to seize a piece of muscle and tear it away. The accompanying drawing of the microscopic appearances in this disease is from a piece of the gastrocnemius obtained in this way from Case II. The appearance of the ultimate muscular fibres is altered, the transverse striæ being very faintly marked, in some altogether absent. No true fatty degeneration exists, but outside the sarcolemma are seen an abundance of fat granules and globules of all sizes, and a large quantity of fibrous tissue, sometimes in wavy bundles, at other times forming loose meshes. Some of the ultimate muscular fibres are diminished in breadth. In some places the appearance of the minute structure might suggest that the muscular fibrillæ had been converted into the fibrous tissue. Duchenne's description of the microscopic appearances closely corresponds with the above. From him, too, we learn that these characters are not peculiar to the enlarged muscles such as the gastrocnemii, but are present

also in the other weakened muscles which are not enlarged.*

We conclude, then, that this degeneration of the muscles is the essential part of the disease, and that it leads first to weakening, and ultimately to complete paralysis of these organs.



Microscopic appearances of portion of gastrocnemius muscle from case II. Magnified 600 diameters. Fully described in text.

We have next to consider in what relation the enlargement of the muscles stands to the disease. This apparent hypertrophy, associated as it is with, at least, partial paralysis, is the most striking of all the symptoms. Duchenne has given the disease its name from this very character—Pseudo-hypertrophic Muscular Paralysis. We have already seen that it is not present in all the degenerated muscles,

* Eulenberg and Cohnheim's account of the appearance of the muscles in the case they examined after death differs somewhat from this; but in their case (a boy aged 13) the disease was much more advanced. They found all the striated muscles, except the heart and diaphragm, greatly altered. Those of the legs had a fatty appearance, and in some parts could scarcely be distinguished from the subcutaneous adipose tissue. Under the microscope the muscles appeared to be infiltrated with adipose tissue, but the structure of their ultimate elements was not altered, except that the volume of the fibres was diminished.

but only in a limited number. In the lower limbs, for example, which are first attacked, all the muscles are affected with the paralysis, but usually only the gastrocnemii, the hamstring muscles, and ultimately the glutei and lumbar spinal muscles exhibit the hypertrophy. The anterior muscles of the leg remain of normal size or are diminished. So also in the arm, all the muscles may be weakened, but only some of them are enlarged. We find, also, that the degree of paralysis is not proportional to the amount of hypertrophy; on the contrary, the hypertrophied muscles are much stronger than the non-hypertrophied. This fact was evident in my cases. In order to estimate the amount of paralysis in each group of muscles, I compared their power of voluntary resistance with that of a healthy person; * and the result I found was that, in Cases I. and II., which were not in the advanced stage, while all the anterior muscles of the legs were all but completely paralysed, the posterior hypertrophied muscles retained a great deal of their natural strength. This is an important fact, and I am satisfied of its correctness, having verified it in all my cases. I am inclined to infer from this fact, and from the other circumstances of the disease, that the hypertrophy is of a compensatory nature, the additional bulk of the muscle compensating for the weakness of its structure. If this be so, why is it that in the legs the posterior muscles alone usually possess this compensatory hypertrophy? † It is because they have the greatest amount of work to do. For, though all the muscles of the lower limbs act in keeping the body erect and in walking, yet, by far the greatest amount of

* The examination of the electro-contractility of the muscles in this disease has given such varied results, that I have taken no note of it in these remarks.

† Duchenne and Lockhart Clarke refer to a remarkable case under the care of Dr Bergeron, in which nearly all the muscles of the body were enlarged. The appearance of this boy is described by L. Clarke—"Every visible muscle of his body, except the pectorals, was enormously developed; his head even appeared swollen, and the temporal muscles stood out like convex shells. Yet when the poor boy attempted to walk, he laboured to get *along*, presenting the most grotesque appearance; and when laid on the ground, he was wholly unable to rise by his own efforts." Duchenne describes him as an infant of the Michael Angelo type. This case may appear to contradict the theory of compensatory hypertrophy, but more details of the case would be needed to enable one to judge of its significance.

muscular effort is exerted by the posterior muscles. In walking, little muscular force is required of the anterior muscles in moving the leg forward. In standing, the principal effort required of them is in fixing the leg at the knee; and it is just here that their weakness in this disease is shown. For it is in order to make up for the inability of these anterior muscles to fix the knee-joint sufficiently for standing, that the belly is projected forward, and the increased lumbo-sacral curve formed. From the same inability to fix the knee arises the marked difficulty these patients have in raising the body after stooping; for this difficulty disappears in a great measure in the sitting posture. The raising of the heel from the ground, or equinism, as Duchenne calls it, which is one of the characters of this disease, is explained by this excessive action of the muscles of the calf over those of the front of the leg; and the peculiar mode of walking seems to arise partly from the depression of the toes, requiring the foot to be raised higher from the ground, and partly from the necessity for balancing the body more exactly on the legs, alternately, so as to make up for the weakness of the muscles which ought to support it on them.

The progress of this disease has been divided by Duchenne into three stages.

1. The first stage is characterised by weakness of the muscles of the lower limbs; showing itself usually either at the time when children should begin to walk, or some years after they have walked. The consequent difficulty they have in keeping the body erect gives rise to those peculiarities in the attitude, and in the mode of progression, which we have seen exist in the disease. This stage lasts several months, or even a year or more; and if the malady be recognised at this time, and subjected to the proper treatment, the patient may recover; but if the case has passed for any length of time into the second stage, then, according to Duchenne's experience, it will proceed steadily to a fatal termination. If the relative of the boy mentioned above (Case II.) suffered, as I believe

he did, from Duchenne's paralysis, then he is an example of spontaneous cessation of the disease in the second stage.

2. The second stage is marked by occurrence of the characteristic hypertrophy of certain of the affected muscles. By this time the muscular weakness, which at first only affected the lower half of the body, may have extended to the upper half also. (Cases I. and II. are examples of this stage.) This stage may last for years, even till an advanced period of youth; but it ultimately passes into—

3. The third and last stage. The paralysis now increases in severity and extent, as was seen in Case III. Duchenne's description is as follows:—"The young patients can no longer stand upright; they always remain in the recumbent posture, without any power to change the position in which they may be placed, and the arms, if not hitherto affected, soon lose all their movements. With this aggravation of the paralysis, the hypertrophied muscles may sometimes be seen to melt away, as it were, and then all the limbs and the trunk become atrophied *en masse*. Although in this stage the patients are reduced to great weakness, they may nevertheless live for a considerable time: they are usually cut off by some intercurrent disease."

The prognosis in this disease is most unfavourable. All the cases that came under Duchenne's notice in the second stage resisted treatment, and either advanced to the third stage, or terminated fatally. Two cases, however, which he recognised in the first stage of the disease, before hypertrophy had occurred to any great extent, recovered under treatment by faradization.

The above cases and remarks will, I hope, be sufficient to indicate the general characters of the disease. There are many other details which might have been discussed, but for which I would refer the reader to Duchenne's own paper. The disease is at present very imperfectly understood; and it is only by much more extended observation of its origin and progress—and especially by more complete investigation of its anatomy after death—that we can expect to arrive at an improved knowledge of its pathology and treatment.