On the pathology of pseudo-hypertrophic muscular paralysis : with remarks on a so-called degeneration of the nervous system / by Geo. S. Middleton.

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THESIS FOR THE DEGREE OF M.D.

ON THE PATHOLOGY

OF

PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS,

WITH REMARKS ON A SO-CALLED DEGENERATION OF THE NERVOUS SYSTEM.

BY

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ON THE PATHOLOGY OF PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS, WITH REMARKS ON A SO-CALLED DEGENERATION OF THE NERVOUS SYSTEM.

OPPORTUNITIES for making examinations of the tissues in cases of pseudo-hypertrophic muscular paralysis have been so infrequent, that I very readily undertook the investigation of the parts in two cases which died in Glasgow within the last two years, and were subjected to *post-mortem* examination. I was the more inclined to do so as the chief interest centres in the condition of the nervous system, and I had previously devoted a considerable amount of time to the microscopic examination of the brain and the spinal cord in cases in which these were affected.

The cases to which I refer occurred in the practice of Dr. Robertson at the Town's Hospital, and *post-mortem* examinations were made, the one by Dr. Newman and the other by Dr. Steven. To Dr. Robertson I am indebted for the clinical history of these cases, which will be found in the Appendix. Dr. Newman handed over to me the parts which he had removed for microscopic examination, and Dr. Steven also kindly supplied me with portions of the cord and muscle in his case, which had, however, been previously examined both by himself and by Dr. Coats, the latter having very briefly described in his *Manual of Pathology* the appearances met with. I have made a very detailed and minute examination of over 200 microscopic sections from these cases, and the results are recorded in the following pages.

Report by Dr. Newman of the Post-mortem Examination of Case I.

"*External appearance.*—The body is greatly emaciated and almost perfectly free from adipose tissue, and is also very anæmic. There is great atrophy of the muscles, particularly those of the extremities.

"*Muscular system.*—Examination of the muscular system throughout the body gives the following results. The superficial muscles of neck are almost normal, with the exception of the sterno-mastoid, which is pale in colour, but does not present marked evidence of the changes which are so observable in some of the other muscles. The deep muscles at the back of neck and scaleni muscles are pale in colour, but otherwise present the characteristic appearances of healthy muscle. The superficial muscles of the back present very much the same appearances as the deep muscles of the neck, while the deep muscles of the back are very pale in colour, and in only some parts can any trace of muscular structure be seen. The intercostal muscles are not much affected. The deltoid, the flexor sublimis digitorum, and the ext. indicis are found to be greatly changed, so that it is with difficulty that any trace of fibrillation can be made out. The glutei maximus, medius, and minimus on both sides of the body are so much altered that there is some doubt as to their identity, so much so that the question is raised whether the tissue examined is muscular or adipose; those muscles are very pale in colour, soft, but not oily in appearance. In the quadriceps ext. and gastrocnemius this change is not so far advanced as in the gluteal muscles, but still more marked than in other muscles of the body.

"*Nervous system.*—The brachial plexus and the ulnar nerve do not present anything remarkable in their appearance, and the sciatic also appears to be normal.

"*Brain.*—The dura mater is firmly adherent to the calvarium, but is easily removed from the base of skull. Brain otherwise normal.

"*Spinal cord* does not present any change to the naked eye.

"The *heart* is very small and the left ventricle is firmly contracted. There is no increase in subpericardial fat; the myocardium is very slightly paler than normal, and on careful examination with the naked eye no evidence of fatty or other changes can be detected; all the valves are strictly normal.

"The *large blood-vessels* are normal.

"*Lungs.*—Both apices and posterior parts are firmly adherent

by old fibrous adhesions; both apices are firmly consolidated, and on section present the characteristic appearance of chronic catarrhal pneumonia followed by caseation. In the right lung, about an inch and a half from apex, there is a cavity the size of a walnut, containing soft caseous material. The lower lobes of both lungs appear to be normal. There are a few miliary tubercles distributed throughout the right lung.

"*Liver.*—Practically normal.

"*Spleen.*—Normal.

"*Kidneys.*—Are slightly enlarged, capsules easily separated, and on section, beyond slight hyperæmia, there is nothing to be made out."

All the parts kept for microscopic examination were preserved in alcohol, without the use of any other hardening agent. In preparing sections, the parts were soaked in water to remove the spirit, they were then put into a solution of sugar for several hours, and afterwards transferred to a solution of gum for some hours more. I found that the longer they were kept in gum the more easily they were cut. All the sections were made with Cathcart's * ether freezing microtome. When cut they were floated off the knife into boiled water, and allowed to remain sufficiently long to remove all trace of the sugar and gum. Sections of muscle were examined unstained in glycerine, in glycerine and acetic acid, and in acetate of potass solution; stained with logwood, both in acetate of potass and, clarified, in Canada balsam; and stained with methylaniline, in acetate of potass. Sections of the nervous system were examined unstained in glycerine and acetic acid; stained with methylaniline, in acetate of potass, and, when clarified, in Canada balsam; stained with logwood, carmine (Beale's solution), or picrocarmine, in Canada balsam.

Muscles.—The muscles examined were those of the neck and the deep muscles of the back, the deltoids, the flexors and the extensors of the right forearm, the gastrocnemii, the left gluteus maximus, and the tibialis anticus: the muscular tissue of the heart and of the bladder was also examined.

The voluntary muscles presented the changes which have been described by all previous observers. These may be considered under four divisions.

* *Edin. Clin. and Path. Journal*, 9th Feb., 1884.—I have found Cathcart's microtome an exceedingly satisfactory instrument. Without it I could not easily have accomplished the work which this examination has entailed, and which has all been done without trouble in my own house.

1. Variation in thickness of the muscular fibres.
2. Hyaline or colloid degeneration, or coagulation necrosis, of the muscular fibres.
3. Increase of connective tissue.
4. Fatty infiltration.

In all the muscles examined the fibres were found to differ greatly in diameter; and, in addition, many of them presented marked irregularity of thickness. Both of these conditions were much greater than in normal muscle, and were most pronounced in the muscles which had undergone least fatty infiltration; but the smallest fibres seen were in muscles which were almost totally replaced by fat. Thus, in the deep muscles of the neck, fibres were seen of normal size, about $\frac{1}{400}$ inch in diameter, while most were much smaller, many being only $\frac{1}{1000}$ inch; in the gluteus maximus, on the other hand, where scarcely any muscular fibres remained, these did not measure more than $\frac{1}{2000}$ inch in diameter. Irregularity in size was very apparent in some of the fibres, especially in those which had undergone hyaline degeneration.

The condition variously designated as hyaline or colloid degeneration, or coagulation necrosis,* was met with in all the muscles, but chiefly in those in which fat was least abundant. Numerous fibres were affected, presenting a vitreous-like appearance when viewed by transmitted light with a low power; the hyaline portions were broken up into clumps of considerable diameter, with an almost vitreous-like fracture. Only a few of these presented any trace of transverse striation, but many of the patches were the continuation of fibres which showed fairly well marked striation, and some of these were bifurcated. In other cases the remaining portion of the fibre presented only a granular appearance. Some fibres also presented this granular appearance without the hyaline condition. In sections stained with methylaniline, the hyaline masses had a reddish tint, suggestive of, but not so rosy or bright in colour as, that taken on by amyloid material. In many cases fibres so affected were broken across, the fractured ends being connected only by the sarcolemma. It is also noteworthy that this change was most marked in those muscles which otherwise presented least departure from the normal. Can it be that this degeneration is the first stage in the disappearance of the muscular fibres? The same degeneration has been described by other observers, but not by all who have written on this subject.

* For an illustration of this, see Coats' *Manual of Pathology*, p. 106.

Increase of connective tissue between the muscular fibres, and between the various fasciculi, was very marked in all the muscles, but was most pronounced in those muscles which presented least of the fatty change. The fibres of this connective tissue for the most part lay parallel to the fibres of the muscle. In a few sections it looked as if degenerated muscular fibres were directly continuous with these connective tissue fibres, but this cannot be stated with certainty. In this tissue were numerous nuclei, many of them very large, and corresponding to the description given by Clarke and Gowers in their case. The walls of the vessels were considerably thickened and surrounded by connective tissue, but the increase of connective tissue did not bear any distinct relation to the distribution of the vessels.

Infiltration with fat was the most prominent feature in all the muscles. This reached its maximum in the gluteus, and in the deep muscles of the back (erectores spinæ), was very extensive in the gastrocnemii, the tibialis anticus, the deep muscles of the neck, and the deltoids, less marked in the flexors of the forearm, and least marked in the extensors of the forearm, and in the superficial muscles of the neck. The fat cells were large, like those of a lipoma, and, where muscular tissue still remained, they were arranged in rows parallel to the muscular fibres, and enclosed in tubes of connective tissue, so that a single row was frequently seen isolated from the other tissues. The cells nowhere contained fat crystals. In the gluteus maximus very few muscular fibres were seen, many of them granular, and all of very small size, as already noted.

In the heart there was none of this infiltration, nor were any of the fibres affected with hyaline degeneration. Indeed, the only thing noted was increase of the connective tissue, and this was not very pronounced.

The bladder was hypertrophied, measuring, when hardened in spirit, almost half an inch in thickness. Here also there was an increase of connective as well as of muscular tissue, but there was no infiltration with fat.

Nervous System.—The parts of the nervous system examined were the cord at various levels throughout its whole length, the medulla oblongata, the pons, the corpora striata, the ascending frontal convolution on both sides, portions of the brachial plexus, of the ulnar and the sciatic nerves, and of the lumbar sympathetic ganglia.

In every section one's attention was at once drawn to the pre-

sence in enormous numbers of small whitish glancing bodies, scattered everywhere throughout the tissues, but much more numerous in the white substance than in the grey. These bodies varied in size, the largest measured being $\frac{1}{100}$ inch in diameter, and the smallest $\frac{1}{2000}$ inch. The smaller ones were rounded or oval in form, but the larger ones more irregular in outline, looking as if they were formed by the junction of several smaller ones. In some places they were so closely set as to present the appearance of a string of glancing beads. In unstained sections most of them were clearly demarcated from the surrounding tissue, but round many of them the nerve-tissue was darker than elsewhere, and into this deeper coloured tissue they shaded rather indefinitely. Under a low power they appeared semi-opaque and homogeneous, but under a high power (Hartnack's eye-piece No. 3 and objective No. 7) they were less glancing and more granular-looking. Their distribution was quite indefinite, and bore no relation to the blood-vessels of the tissues, and when examined in longitudinal sections of the cord they were seen to extend longitudinally quite as far as transversely. In such longitudinal sections it was evident that they pushed aside and compressed the fibrous structures of the cord. The smaller of these bodies might have been taken for corpora amylacea, though none of them presented concentric striation. It was impossible, however, to make this mistake with regard to the larger ones, which corresponded more nearly with what have been described as colloid bodies. In order to make out their nature, various staining agents were employed. Methylaniline rendered them somewhat reddish, but this colour was discharged soon after the sections were mounted in acetate of potass. Iodine stained them of a pale yellow colour, and the addition of sulphuric acid did not make any change. They, therefore, cannot consist of amyloid material. Osmic acid rendered them brown; a black colour was developed only in some of them, chiefly those which have been described as surrounded by a darker tissue than normal. With carmine and picrocarmine they were unstained, but they took on the colour of logwood. In sections coloured with logwood, deeply stained irregular patches were seen in great numbers, their centres being less stained than their peripheral portions. These patches looked almost like collections of round cells closely aggregated, but on more minute examination they were found to consist of glancing bodies surrounded by irregularly condensed and puckered fibrous tissue. Sections dehydrated in

alcohol, and clarified with turpentine, showed these bodies less lustrous, and more opaque and granular. Ether had no effect on sections dehydrated with alcohol, save to render the whole section, including these bodies, more transparent.

From the fact that I had seen similar bodies previously in sections of nervous tissue, I concluded, as I had done before, that they were merely an accident of the mode of hardening, an opinion strengthened by their irregular and wide distribution. I could find no description of similar appearances in any of the text-books of Pathology or in works on the Diseases of the Nervous System which I consulted; but, on commencing a perusal of the recorded pathological observations on pseudo-hypertrophic paralysis, I found Mr. Kesteven described a similar appearance, looking on it as an example of a lesion described by Dr. Batty Tuke and Dr. (now Professor) Rutherford.* On referring to their paper, I found their description corresponded closely with that above given. In a later paper,† while they unfortunately termed the lesion "miliary *sclerosis*," they guarded "the reader against the assumption that this term indicates that this is a form of gray degeneration (the sclerosis of French writers). We adopt the term because it simply indicates rounded hardened patches." In order to determine whether I was dealing with the same bodies, I treated sections with fuming nitric acid and fuming sulphuric acid. These had the effect of determining a change fully described and figured by Drs. Tuke and Rutherford as occurring in their case. The glancing bodies lost their lustre and their homogeneous or granular appearance, and were converted into masses varying in aspect, some looking like clusters of grapes arranged round a centre, others like rosettes made of tubes rounded or blunted at the ends, and very similar to tubes of myeline which I had often observed at the margins of unstained sections of brain-tissue when pressure had been applied to the cover-glass, and which reminded me of tubes of macaroni. When treated with turpentine this appearance was lost, and sections under the low power looked as if they were riddled with holes. On examination under a high power, these spaces were found to contain a very fine reticulum, the neuroglia, with here and there an axial nerve cylinder. This appearance of loss of tissue was still more pronounced in

* "On a New Lesion observed in the Brain of an Insane Person." *Edin. Med. Jour.*, Sept., 1868. P. 204.

† "On the Morbid Appearances met with in the Brains of Thirty Insane Persons." *Edin. Med. Jour.*, October, 1869. P. 289.

sections from which the acid was removed by soaking in water, and which were then stained with logwood, dehydrated, clarified, and mounted in the usual way. What were formerly deeply-stained patches were now clear spaces, contrasting strongly with the surrounding tissue. When stained with carmine this condition was not so apparent. Sections treated with nitric acid, and then washed and stained with osmic acid, showed none of those clear spaces, but darkened patches as in other sections not so treated. Strong ammonia had no effect on these bodies. These, therefore, have all the characters of the so-called "miliary sclerosis" of Tuke and Rutherford, and must be the condition described under that name.

In further elucidation of the nature of these bodies, I examined all the microscopic specimens of nervous tissue in my possession, amounting to some hundreds of sections from over thirty different cases. In this large series of observations I found similar glancing bodies in a great many sections, but only in one or two cases as numerous as in this one. They were present only in those instances in which the parts had been primarily or wholly hardened in spirit, never where the parts had been first fixed by hardening in chromic acid or the bichromates and finally preserved in spirit. In the cases described by Drs. Tuke and Rutherford, and by Mr. Kesteven, the tissues had *always been immersed for some time in alcohol* before being finally hardened in chromic acid. Believing that the condition was due to the action of alcohol, I had alternate portions of a normal cord hardened in alcohol and in chromic acid; but, as I was in haste to get the work finished, they were in these fluids for only a week. On making sections I found that while the tissue hardened in chromic acid presented none of these bodies, that in alcohol was freely strewn with them; indeed, they were quite as numerous as in the case under investigation. As it so happened, the fact that the cord had been in alcohol for so short a time was an advantage; for in the sections numerous drops of myeline were seen everywhere, especially abundant in the white matter, and some of the glancing bodies presented the appearance already described as being brought about by treating sections with nitric acid. These latter looked just as if they were formed of drops of myeline run together. The conclusion, therefore, to which I have come is, that these bodies are simply the result of hardening in alcohol, and that they are formed by the action of the alcohol on the myeline of the nerve-tubules.

In this view I believe I am confirmed by a description of the lesions found in a case of paraplegia by Lockhart Clarke.* He does not state how the parts were hardened, but he describes and figures (as a lesion) bodies very similar to those found in this case. "Around the spinal canal, and in the transverse commissure, a large number of corpora amylacea were also observed. In the surrounding white columns, especially the posterior and the lateral, many of the nerve-fibres had evidently suffered from either partial destruction or complete disintegration of their white substance. At the lowest roots of the same pair of nerves—the fourth cervical—the principal lesion was in the neighbourhood of the posterior commissure —in which were observed two or three small spots of transparent granular disintegration—and in the anterior decussating commissure, the front of which, at the bottom of the anterior median fissure, was partially replaced by a heap of fat globules; but many of these bodies were changed from the perfectly spherical form by compression against each other, and in this shape bore a strong resemblance to *aggregated particles of the white substance of nerves.* In other regions of the cord similar particles were found in different parts of the grey substance, collected into groups, or arranged in strings like beads, and appearing like nerve-fibres composed of series of particles of their own white substance."

Drs. Tuke and Rutherford did not consider their lesion the same as Clarke's; nor did they lose sight of the possibility of their "miliary sclerosis" being the result of the agents used in hardening, but they negatived this idea because they had "subjected healthy brains to precisely the same treatment, and other observers have done the same, and no such appearances have been produced. Further, in the case described, this lesion was found only in the white matter of the atrophied half of the cerebellum, although the half that was not atrophied was treated and examined in exactly the same way." † I cannot reconcile their statement with my own observations, nor can I say whether they are still of the same opinion.

I do not profess an intimate acquaintance with all that has been written on the pathological appearances met with in lesions of the nervous system, but, holding the view that this particular appearance is merely the result of the action of alcohol on the tissues, and must therefore be of frequent occur-

* "Pathological Investigations in a Case of Paraplegia." *Brit. and For. Med.-Chir. Rev.* 1864. Vol. xxxiii, p. 486.

† *Edin. Med. Jour.*, Sept., 1868. P. 210.

rence, I think it is strange that, in the course of my reading, I have seen descriptions of similar appearances only by Kesteven, Tuke and Rutherford, and Clarke. Possibly its infrequent mention is to be attributed to the facts that others, like myself at an earlier period, have attributed it to its true cause, and thought it unnecessary to notice it, and that, in recent years, workers in nervous pathology have discarded alcohol as a hardening agent.

But while alcohol has been justly condemned, I have nowhere met with a description of the reasons. Hence I have been induced to go into detail in regard to the appearances found in this case. Besides leading to confusion by the formation of these glancing bodies, alcohol destroys the fine structure of nervous tissues. In transverse sections of the cord, for instance, the tubular contour of the nerve-fibres in the white substance is almost entirely lost, the white columns presenting, both stained and unstained, the appearance of a fibrous network, with the axial cylinders dotted over it. In sections hardened in chromic acid or the bichromates, on the other hand, the tubular contour is beautifully preserved.

Other objections to the use of alcohol are that cavities are formed in the tissues, and crystals deposited. In every case in which the nervous tissue had been hardened in alcohol, holes were very much more numerous in the sections than in cases hardened in chromic acid. Many of these are due to blood-vessels, the section of the vessel having dropped out of its cavity; but a much greater number of them are due partly to the removal of the myeline to form glancing bodies, and partly to irregular contraction of the tissues. I therefore am very chary of accepting a hole in a section of nervous tissue as evidence of softening and disintegration. In various of the hitherto recorded cases of pseudo-hypertrophic paralysis such holes have been described, as they have been also in other diseases of the nervous system. In the case under investigation they were exceedingly numerous, particularly in the brain. They were frequently also constant in position; *e.g.*, in almost every one of a dozen sections of the medulla a hole was seen just external to the nucleus of the hypo-glossal nerve; but while this constancy is so far in favour of its being a lesion, I am of opinion that it is the result simply of an elongated spindle-shaped cavity formed as above described. The same explanation applies to the cavities or empty spaces frequently found around the ganglion cells and the vessels. In addition to such holes, which have regular margins, there were, of course,

numerous irregular tears seen, the result simply of the manipulation of the sections.

Crystals were much more abundant in the sections of the brain than in the sections of the cord. They are common in sections hardened in alcohol, and present various forms, many being only semi-crystalline. The most common form is a feathery one, which in sections of the convolutions mounted in glycerine frequently present an appearance suggestive of the flowering on a window pane in frosty weather. They are often so abundant as to obscure the structure of the tissue. With transmitted light they are dark, but in sections stained, clarified, and mounted in balsam, they are quite transparent. Besides these forms rhomboidal crystals, frequently in clusters, were often seen much more crystalline in appearance than the former. In order to remove the possibility of these being due to the sugar and gum in which the tissues had been soaked, the sections as cut were floated off into boiled water, and allowed to remain in it for hours. Besides, they were seen in sections cut by the hand, and never exposed to either sugar or gum. Nor were they due to the staining agents, which, especially logwood, frequently deposit crystals, with the appearance of which I am perfectly familiar; for they were seen quite as well developed in unstained sections as above described. I cannot say definitely what these crystalline substances are, but they are soluble in ether, and insoluble in fuming nitric acid; probably they are unusual forms of cholesterin, or of some fatty crystal.

For the reasons above stated, therefore, I am opposed to a treatment of the cord and the brain for pathological investigations which is often recommended—viz., a preliminary immersion of the parts in alcohol for 24 hours or more, preparatory to hardening in Müller's fluid, &c. The structures of nervous tissue should be first fixed in their natural position by the use of chromic acid or the bichromates before they are preserved in spirit. The objections to these hardening agents are no doubt considerable, for they render the tissue both very brittle and difficult to stain with carmine (which is by far the best staining agent for nervous tissue). I have no experience to lead me to say which of them is the best fluid, but I think that with proper attention the parts may be examined before they become brittle, and removed to spirit, which will then do no harm. As to staining, I have found that sections hardened in the bichromate of ammonium give very good results when dehydrated in alcohol, and clarified with

turpentine, without any staining. In any case, it is better to have the natural appearance of the parts retained than to have beautifully cut and stained sections which do not show the structures in their proper condition.

I now proceed to the description of the various portions of the nervous system examined.

Spinal Cord.—Throughout the whole length of the cord the central canal was of normal size and well marked, except in the lumbar region, where its calibre was obliterated, its epithelial lining being in close apposition all round. This condition, however, is by no means uncommon in this situation, and cannot be looked upon as abnormal. Numerous holes were seen in both grey and white matter, besides spaces around vessels and ganglion cells; for the reasons above stated I do not look upon these as of pathological significance. The vessels in the pia mater were distended with blood, as also were some of those in the sections; but there was nowhere any exudation of leucocytes around the vessels or elsewhere. The walls of the vessels were not abnormally thick.

In the *upper cervical* region, the ganglion cells of the anterior cornua were well developed, both as to number, size, clearness of outline, and length of processes: none of them showed pigmentation. The largest measured about $\frac{1}{330}$ inch in greatest diameter, and the smallest rather less than $\frac{1}{1000}$ inch, the majority being between these sizes. They were gathered into groups quite in a normal manner. The cells of the internal group were much smaller than those of the other groups, throughout the whole length of the cord; but this in my experience is the normal condition, and these cells did not present any appearance of granular degeneration or atrophy. A more striking fact was that in many sections, not only of the cervical region, but lower down, the cells of this inner group were not of the same size on the two sides, the largest on the one side measuring $\frac{1}{1000}$ inch, while on the other, cells of double that size were seen. As the ganglion cells are very irregular in form, and of considerable length, I think that this may be explained by the section of the cells on the smaller side representing not a section through their largest diameter, but through one of their smaller diameters. However that may be, I am not inclined to look upon the condition as abnormal. In the postero-lateral group, the cells were somewhat smaller than in the anterior groups, and the posterior vesicular column was indistinctly represented by a few scattered cells, well developed. There was no evidence of

sclerosis anywhere. In a few of the sections the anterior commissure was traversed by fibres extending from the fibrous tissue in the anterior median fissure; but this condition is described as normal by Lockhart Clarke.*

In the *cervical enlargement*, sections coloured with picrocarmine presented, scattered over the white substance, patches stained of a deeper hue than the rest, resembling somewhat the appearance observed in sclerosis of the lateral columns. When examined by the high power, the depth of staining was seen to be due to the meshes of the reticulum being thickened at various points. This thickening was very local, a small central nucleus with radiations being frequently observed; possibly these were enlarged Deiter's cells. Otherwise the sections were much as in the upper cervical region. The ganglion cells were well developed in every way.

Throughout the *dorsal* region the amount of grey matter was very small, the broadest diameter of the anterior cornua being only $\frac{1}{6}$ or $\frac{1}{8}$ of the same diameter of the white matter. The ganglion cells were much fewer in number than in the cervical and lumbar regions, and smaller in size, the largest often not measuring more than $\frac{1}{1000}$ inch in its long diameter. The best developed groups were the antero-lateral and the posterior vesicular column. Occasionally the one side showed better development than the other.

In the *lumbar* region the ganglion cells were abundant, large, and well developed, those of the internal group being as usual smaller than the others, which measured from $\frac{1}{500}$ inch to $\frac{1}{1000}$ inch in diameter. No difference could be detected on the two sides, and there was no evidence of sclerosis anywhere. Here also the anterior commissure was occasionally traversed by fibres running from the anterior median fissure.

Throughout the cord, sections stained with methylaniline showed some of the ganglion cells of a reddish tint, disappearing after being mounted for some time in acetate of potass.

The *medulla oblongata*, the *pons*, and the *corpora striata* presented nothing to note save glancing bodies, frequent holes, and numerous crystals.

In the *ascending frontal convolutions* similar appearances were presented, and in addition, along the surface of the grey matter there were here and there very small patches more deeply stained than the rest of the grey tissue. These patches were found to contain more connective tissue than the other parts.

* "Researches into the Structure of the Spinal Cord."—*Phil. Trans. of the Royal Society.* 1851. Vol. cxli, p. 614.

The portions of the *brachial plexus*, *ulnar*, and *sciatic* nerves, and of the *sympathetic ganglion* examined did not reveal anything abnormal. In the nerves, glancing particles, very much smaller in size than those occurring in the cord and brain, but evidently of the same nature, were seen dotted over the nerve-fibres. The tubular character of the nerves was lost, just as in the other tissues.

The notes of the *post-mortem* examination which was made by Dr. Steven will be found in the Appendix, under Case II. In this instance the parts given me for examination were portions of the spinal cord at various levels, and a portion of the erector spinæ muscle. These were hardened from the first in a four per cent solution of bichromate of ammonium, and afterwards preserved in spirit. The sections were made and mounted in the same way as in the preceding case.

The *muscular tissue* presented a most marked fatty infiltration, with some increase of connective tissue, and great loss of muscular fibres with diminution in the size of those remaining. None of these showed any appearance of hyaline degeneration.

Spinal Cord.—In this case no glancing bodies were seen, and holes were much less frequent in the sections than in the preceding case. With methylaniline some of the vessels and of the ganglion cells were coloured of a somewhat rosy or pinkish hue, but none were distinctly amyloid; the colour was soon discharged in acetate of potass. In all the various regions of the cord the white matter showed very clearly the tubular character of the nerve-fibres. The tubules varied very much in size, the largest ones being, of course, at the periphery of the cord. It was doubtful whether some of those were not abnormally large, as they were found to measure as much as $\frac{1}{2500}$ inch in diameter, the common size being about $\frac{1}{10000}$ inch. All, however, presented the axis cylinder quite distinct, and not enlarged.

In the *cervical enlargement* the ganglion cells were abundant and well developed, save in the internal group, which, as usual, contained cells much smaller than those of the other groups, but not presenting any appearance of atrophy. The majority of the cells in the other groups measured about $\frac{1}{500}$ or $\frac{1}{600}$ inch in diameter.

In the *dorsal* cord the cells were smaller, few of them reaching in size $\frac{1}{500}$ inch. The vessels were distended with blood, but no hæmorrhages, and no exudations of leucocytes were seen.

In the *lumbar* cord the conditions were very similar to those noted above; but there was more of the appearance of deficiency of the cells of the one side as compared with the other. The ganglion cells varied in size from $\frac{1}{1000}$ inch to about $\frac{1}{350}$ inch. One very minute hæmorrhage was seen in the anterior commissure, pushing the fibres of the commissure before it in a knee-like bend.

Illustrations of the cervical and lumbar enlargements in this case will be found in Coats' *Manual of Pathology* (p. 434), and in the text he gives expression to the opinion that the cells in the anterior and internal groups are probably deficient. As I have already stated, I believe the cells of the internal group are normally smaller than the cells of the other groups, and that the condition noted in this and in the other case is, therefore, not abnormal. With this view I believe Dr. Coats is now inclined to agree.

In reviewing the literature of this subject I have found recorded seventeen *post-mortem* examinations, to the original reports of eleven of which I have found access. In seven of these the examination of the spinal cord and nervous tissues gave negative results—viz., in the cases recorded by Meryon,* Eulenburg-Cohnheim,† Charcot‡ (Duchenne's case), Brieger,§ Schultze,‖ and Ross.¶ In Brieger's case there was congestion of the vessels of the white substance, with collections of leucocytes in the perivascular canals, in the brain and basal ganglia, but not in the spinal cord. Similar collections of leucocytes have been described by Bramwell and others, but I regard them as an accidental complication and not essential to the disease. In Brieger's case they are sufficiently explained by the presence of tubercular meningitis. Their significance was indicated by me in a previous paper;** such exudations are generally met with in cases which have been characterised by cerebral excitement. In Schultze's case it is stated that

* *Med. Chir. Trans.* 1852. Vol. xxxv, p. 75. *On Paralysis.* 1864. P. 202.

† *Verhandlungen der Berliner Med. Gesellschaft.* 1866. P. 191. (Not seen).

‡ "Note sur l'état anatomique des muscles et de la moelle épinière dans un cas paralysie pseudo-hypertrophique."—*Arch. de Physiol.* 1872. P 228. (Not seen.)

§ *Deutsches Arch. f. Klin. Med.* 1878. xxii, p. 200.

‖ *Virch. Arch.* 1879. Vol. lxxv, p. 475.

¶ *Diseases of the Nervous System.* Second Edition. Vol. i, p. 1017.

** "On Vascular Lesions in Hydrophobia and in other Diseases characterised by Cerebral Excitement."—*Journal of Anat. and Phys.* 1880. Vol. xv, p. 88.

the connective tissue in the ulnar nerves was perhaps in excess. It is, I find, extremely difficult to say what amount of connective tissue is normal to the peripheral nerves, so that I am inclined to believe that, in his case, the nerves, like the other parts, were quite normal.

The case recorded by Barth * is generally looked upon as having been amyotrophic lateral sclerosis, rather than pseudo-hypertrophic muscular paralysis. I did not, however, see his record of it.

The next recorded case that I find is that of Kesteven, † who describes dilatation of the perivascular canals, and numerous circumscribed spots of granular degeneration. He figures very distinctly the glancing bodies described above, and the conditions met with in his case are to be ascribed to the action of the alcohol in which the tissues were preserved.

The most important case hitherto recorded is that by Clarke and Gowers, ‡ who describe extensive lesions mainly of the grey matter. The form of degeneration they describe is one with which I am not familiar. The tissues were preserved in chromic acid, but had they been exposed even for a short time to the action of alcohol, I should have been inclined to attribute the appearances found to the action of that agent, especially as it is distinctly stated that globules of myeline were accumulated in various situations. No inference from the facts is drawn in their conjoint paper, but more recently Gowers § has indicated his belief that the lesion had probably little connection with the primary disease, being rather a secondary result of long-standing paralysis and frequent attacks of pulmonary trouble.

Bäg || describes an abnormally transparent condition in the neighbourhood of the posterior cornua, and an increase of the interstitial tissue with rarefaction of the nerve-fibres in the whole length of the cord, similar in extent and form to that found in degeneration secondary to disease of the brain. I had access only to the extract of his paper in Virchow's

* "Beiträge zur Kenntniss der Atrophia musculorum lipomatosa."—*Arch. der Heilk.* 1871. Bd. xii. P. 121.

† "The Microscopical Anatomy of the Brain and Spinal Cord in a case of Imbecility, associated with Duchenne's Paralysis."—*Journal of Mental Science.* 1871. Vol. xvi, p. 563.

‡ *Med.-Chir. Trans.* 1874. Vol. lvii, p. 247.

§ "Pseudo-Hypertrophic Muscular Paralysis." A Clinical Lecture. 1879. P. 42.

|| "Tre Tilfalde af Pseudohypertrophia Muscularis."—*Hospital Tidende.* 2 R., Bd. 4, p. 441. Virch. *Jahresbericht der Medicin.* 1877. Pt. II, p. 133.

Jahresbericht, and cannot say whether he substantiates his statement with illustrations. It seems to me that he has quite probably mistaken for sclerosis a condition similar to that described in some of the cervical sections in my first case. When I first examined my own sections I was inclined to the belief that I was dealing with sclerosis, but subsequent experience of more perfectly prepared and coloured sections led me to alter that conclusion.

Brigidi * found in his case in the sympathetic ganglia dilatation of the veins, atrophy and pigmentation of the nerve-cells, and sclerosis. No other observer who has mentioned the examination of these ganglia has described a similar condition. In the extract of the case in the *Record* there is no mention of the cord and other nervous centres.

Ross † in his first case observed atrophy of the ganglion cells in different groups, at different levels of the cord. Notwithstanding this, however, after having had the opportunity of examining a second case with negative results, he has come to the conclusion that pseudo-hypertrophic paralysis cannot be attributed to disease of the nervous system.

Drummond ‡ met with a case presenting a bulging of the lumbar enlargement, evidently congenital, and disintegration of the lateral grey network of fibres, somewhat similar to that described by Clarke and Gowers. The parts were hardened in spirit and Müller's fluid. The illustrations from his case do not seem to me clearly to indicate a lesion.

Bramwell § has recorded, with numerous illustrations, a somewhat similar case, with a bulging in the cervical region. Besides this bulging, attributed to a congenital malformation, he describes collections of leucocytes round the vessels, and lacerations of the grey matter. I have already indicated my opinion of such conditions, but neither in Bramwell's work, nor in the only clinical report of his case as yet apparently published ‖ is there any note of the patient's condition and symptoms at the time of death. The sections of cord shown would, apart from the bulging, pass for normal. He states that some of the nerve cells appeared to be hypertrophied. I know of no standard size for comparison.

Pekelharing ¶ gives a detailed description of lesions which

* *Imparziale*, 28th Feb., 1878. *Lon. Med. Rec.*, October, 1878.

† *Diseases of the Nervous System*, 1st Ed., 1881. P. 207.

‡ *Lancet*, 1881. Vol. ii, p. 660. *Diseases of the Brain and Spinal Cord*, 1883. P. 335.

§ *Diseases of the Spinal Cord*, 1882. P. 201.

‖ Dr. Milner Moore, *Lancet*, 19th June, 1880.

¶ Ein Fall von Rückenmarkserkrankung bei Pseudomuskelhypertrophie. *Virch. Arch.* 1882. Vol. lxxxix, p. 228.

he believes he has met with in a case of this nature. Among these are dilatation and irregularity of the central canal in the cervical and dorsal regions, with obliteration of it in the lumbar region, also an accumulation of nuclei round it in its whole length. Then there are enlarged vascular canals on either side of the central canal; also a distinct atrophy of the anterior and median groups of ganglion cells, besides a condition similar to Clarke and Gowers' disintegration. In my experience the size of the central canal is exceedingly variable; it is usually surrounded by nuclei, and on either side there is generally a large vessel which, in the process of hardening, frequently has shrunk away from the surrounding tissue. These conditions I therefore look on as normal, and the illustrations accompanying his paper show the ganglion cells to be perfectly normal in number and size. With this conclusion I am glad to find that Schultze * agrees, as he does also with the view that Drummond's and Bramwell's plates represent essentially normal tissue.

Finally, in Pekelharing's paper there is found a synopsis of a case described by Gœtz,† who observed congestion of the vessels of the cord, and throughout its whole length a gelatinous proliferation of the connective tissue, with atrophy of the nerve-fibres of the white substance, and a peculiarly sharp separation of the various cell groups, but no atrophy of any of them. Pekelharing believes, however, that Gœtz's illustrations of his case support his own views by showing atrophy of the anterior and internal groups in the lumbar region. I have been unable to get the original report of this case, but the extract leads me to the opinion that it is a case the structures of which had been altered by hardening in alcohol.

My own observations and an examination of the cases hitherto recorded, lead me therefore to the conclusion that no alteration of pathological significance has been met with in the nervous system. The lesions said to have been observed are either accidental conditions, or not characteristic of this affection. There is even less indication of disease in the nerves and cord than one would expect to be the secondary result of the pronounced muscular degeneration. The conclusion to which we are driven is that the disease is primarily one of the muscular tissue. That it is of a congenital nature is indicated by the disease being one of early life, and by its more or less hereditary character; but I cannot discover any basis for speculation as to the immediate seat of this inherent vice.

* *Virch. Arch.* 1882. Vol. xc, p. 208.

† *Aerztliches Intelligenzblatt.* München. 1879. P. 419.

The malformations met with in two instances in the cord are interesting as indicating a tendency to congenital malformation. The cause of death in almost all instances, so far as I have seen, has been some affection of the lungs, generally of a phthisical nature. This may be accounted for to some extent by impaired respiration due to diminished muscular power.

Note.—Since the above was in type my attention has been drawn to an extract from a monograph on "Muscular Pseudo-hypertrophy," by Dr. Gradenigo,* of Padua, who views the disease as one of a myopathic nature, and objects to the term paralysis being applied to it. The description given of the changes found by him in the muscles corresponds generally with that of the appearances in my own cases; but he seems to have found dichotomous division of the muscular fibres much more commonly than I have, and regards it as perhaps characteristic of this affection. In the extract there is no mention of the condition of the nervous system, but it is stated that the pathogenesis of the disease, at first myopathic, is later on neuro-myopathic. It is evident, therefore, that he agrees with the view that the case in favour of an affection of the nervous system has not been made out.

* *Contribuzione alla Patogenesi della Pseudo-ipertrofia muscolare.* Milano: Fratelli Rechiedei. 1883. *London Med. Record*, 16th June, 1884.

APPENDIX OF CASES.

Reports by Dr. Robertson.

Case I.—"James S., age 16, admitted into Town's Hospital 8th April, 1882. Father of drunken habits; died from disease of kidney. Mother living; said to be of sober habits. Has a younger brother ill like himself, and another brother and two sisters, who are well.

"Patient is fairly intelligent; has cleft palate (posterior); no evidence of constitutional syphilis. Present illness began several years ago—cannot tell how many. He first felt his back sore when he rose from bed; afterwards, in running, the right leg would suddenly give way; at the same time calves of both legs got big; people looking at his big legs wondered that he should be so weak in walking. About 3½ years ago arms began to get weak, this being years after the legs.

"*Present Condition.*—Special senses and mind correct. *Lower extremities:* both feet in associated position of talipes equinus and varus; hamstrings contracted; legs moderately wasted; thigh muscles greatly so; intrinsic muscles of feet a little wasted. He can flex both legs on thighs beyond their contraction, but contractile power of the left thigh muscles weaker than that of the right ones. When legs are fully flexed, the extensor power of muscles in front of thigh is noticed to be very weak; he can flex and extend thighs on pelvis better; can flex and extend toes—all these movements are feebler than normal. Anterior abdominal muscles also weak, but proportionately less so than the thighs. He can expand the chest fairly well, and the intercostals are observed to act. Chest is ricketty — chicken-breasted — attributed to fall when an infant. When lying on back he cannot rise to erect posture, but he can turn from the back to the sides, or from the sides to the back; can flex and extend the head and upper part of the back pretty well. There is no appreciable change in the appearance of the back—neither fuller nor more wasted than normal.

"*Upper Extremities.*—The forearms are small, but do not give the impression of much wasting; a little wasting in the

metacarpal spaces, and on the webs between the thumb and forefinger (state of thenar eminences not noted); upper arms (biceps, &c., and triceps) distinctly wasted; neither deltoid much wasted in appearance, but the left is smaller than the right; both anterior and posterior axillary folds are greatly wasted. He can flex and extend both wrists and all the fingers of both hands; can flex and extend left forearm but feebly; cannot flex right forearm on arm; while attempting flexion of latter, the forearm involuntarily pronates; can extend this arm; can withdraw right arm from side, but cannot bring the right elbow close in to the side again. He can raise the shoulders by levator angulæ scapulæ; and none of the actions of the scapula are lost, though they are rather weak. He registers 4 lbs. on dynamometer with right hand, and 7 lbs. with left (I register 56 lbs.). He can voluntarily pronate and supinate the forearm, but the supination, particularly of the right arm, is not very complete.

"Face and tongue muscles seem normal; swallows fairly well, notwithstanding palatal deficiency. Sensation normal in all forms. Reflex responses scarcely appreciable in legs; better in belly; cremasteric feeble but distinct. Response in legs to interrupted current weak; effect of continuous current could not be clearly ascertained; pain so great in skin, even with 15 cells of Leclanché's battery. Feels heat by hot sponge quite well.

"*Bowels and Bladder* correct. Puberty distinctly reached since trouble began.

"*5th January, 1883.*—Legs much more wasted; contraction much greater—legs upon thighs, and thighs on abdomen; can move the feet only very feebly—cannot flex or extend them more than half an inch; so with the legs on thighs, but if the thighs are withdrawn from the pelvis somewhat forcibly, he can bring them back on abdomen readily; no apparent change in the state of arms, or hands, nor of back nor head; can move head in all directions—he cannot raise his body when seated in the erect posture, should he bend forwards. He can swallow well, and there is no wasting of tongue. Intercostals act fairly well. Sensation nowhere impaired. General health weaker. Bronchitis and some phthisical indications."

The *post-mortem* examination was made by Dr. Newman on 11th January, 1883. The report is incorporated in the paper.

Case II.—"Walter W., aged 13½, admitted into Town's Hospital 21st March, 1882. Patient's mother is living; father is dead. Mother all her life has been subject to fits apparently

hysterical. Two of her children died from inflammation of brain during teething; another son is bow-legged, he being the only other child alive besides Walter. The latter, our patient, was never stout, and did not walk till he was three years old; afterwards he was able to run about till he was ten. He then first complained of sore feet in walking, and was subject to sweating of the head. Soon afterwards he found it difficult to rise from the floor. At that time his mother had not thought his legs to be either thinner or stouter than they should be. He was able to walk fairly well till two years ago. His general health at that time, and previously, was not very good, appetite being indifferent. His arms began to get weak about a year after the legs, and for nearly two years he has been unable to take his food himself. Sometimes he had a difficulty in passing water, but he never wet the bed. Sense of feeling in no part of his body has been impaired. He has had no pain anywhere except in his feet after walking, but he often complained of headache.

"The following note was taken of his condition on the 8th April, 18 days after admission, there being little change, except increasing general weakness in the interval:—Upper extremities—He can flex and extend both wrists, though feebly, and can abduct and adduct fingers and thumbs, as well as flex and extend them; cannot flex left forearm, but can extend it feebly by stages. He cannot pronate the left arm fully, but can supinate it better. The right arm is in much the same condition as the left, but rather worse. Though the forearms are small, they have not a wasted appearance, nor is there distinct wasting of the hands, except a little of the thenar eminences. He can very slightly flex or extend the forearms upon the arms, and he has even less power of movement at the shoulder joints; but he has a little power in raising the shoulders, especially the right one, and in drawing them back to the spine. When the arms are put out from the side, he can bring them in again slowly, and by stages. The scapulæ do not stand out prominently. There is great wasting of the deltoid muscles, and of the anterior and posterior axillary folds. Though he has a little power in the hands, they are so feeble that he is quite unable even to move in the least the index on the dial of the dynamometer. In contrast, a healthy boy, age 10, turns the index to 25 lbs. The full current (Faradic) of a Weiss' battery has no effect on the flexors, but a little on the extensors, particularly of the left forearm.

"*Lower extremities.*—He can flex and extend toes, but has

scarcely any power of movement at the ankles. He has a little, but not much, power of flexion and extension at the knee and hip joints. He cannot stand, nor put his feet on the floor, nor can he sit without support; upon the whole, the left leg is worse than the right. The left calf measures $8\frac{3}{4}$ inches in circumference, the right $8\frac{1}{2}$; in a healthy boy, age 11, they measure each about $9\frac{1}{2}$ inches. The bellies of the calf muscles stand out as small, soft, bulbar-like masses. The left leg is slightly contracted at the knee, and the foot is in the position of talipes equinus. The thighs, though small, have not a wasted appearance.

"When the head is bent down on the chest, he can manage to raise it by an effort, but if it fall back he cannot bring it up should he at the time be leaning back, though if sitting quite erect he can manage to raise it with difficulty. There is no incurvation of the back, and the abdominal muscles have considerable power, and are fairly resistant to the finger when he is sitting up. He requires to be lifted into and out of bed. He seems to have full power in opening and shutting the mouth, and also of moving the tongue and eyes in all directions. The intercostal muscles are weak, though some action is observable.

"*Reflexes.*—The plantar reflex in both feet is distinct; right cremasteric is obtained; abdominal and epigastric are observable; no deep reflex.

"*Sensation, &c.*—No impairment of general sensibility; distinguishes impressions of heat and cold perfectly. Special senses are correct, and is fairly intelligent.

"Heart normal, but feeble in action; urine correct; some bronchitis, and probably diffused tubercle.

"The day after the foregoing note was taken patient was shown to the Pathological Society of Glasgow. His general enfeeblement afterwards rapidly progressed, and he died on the 18th of the same month, the lung symptoms being the most prominent before death.

"*P. M.—42 hours after death.*—The spinal cord to the naked eye seemed healthy. The gastrocnemii, gluteal and deltoid muscles more particularly appeared to have been converted into fat, even the striated character being scarcely traceable at many parts. The other muscles of the extremities and trunk were similarly changed, though generally in a less degree, especially the trunk muscles. Heart weighed 3 oz.; wall of left ventricle and other muscular structures appeared to be quite healthy; kidneys, liver, and spleen normal in aspect; some pleuritic adhesions.

"Specimens of the spinal cord, nerves, and muscles were removed for microscopical examination."

The *post-mortem* examination was made by Dr. J. L. Steven, but he has no record of it. The above notes were made by Dr. Robertson.

The two following cases were first seen in the Royal Infirmary, where they were under Dr. Perry's care, and afterwards at Motherwell, in May, 1884, by the kindness of Dr. P. C. Smith. Their mother, a fairly intelligent woman for her position (wife of a coal miner), gave the family history as follows:—

Children of the family—

1. James, died of scarlet fever: a healthy boy.
2. John, æt. 13, healthy: at work in pit.
3. Thomas, æt. 11: affected.
4. Francis, æt. 9: affected.
5. William, died of infantile diarrhœa.
6. Michael, æt. 5: healthy.
7. A girl, æt. 4,
8. A girl, æt. 14 months, } both healthy.

Mother's family.—Consisted of six sisters and five brothers. The sisters are all married, and all have large families, none affected with any paralytic lesion save the two cases under notice. Two of the brothers died young (4½ and 7) of bronchitis; another died in early infancy; none of these presented any indication of this disease. Two brothers married (one since killed) and had families; all well.

Father's family.—Nine brothers; and two sisters who died of fever. The only history of anything like nervous disease in any of the brothers or their families is that of a girl, æt. 26, who had epileptic fits after an injury to the head, and is now in an asylum.

Case III.—Thomas M., æt. 11 years. In this boy's present condition it would be impossible to diagnose the case as one of pseudo-hypertrophic muscular paralysis, for there is no hypertrophy, and he is so helpless that none of the phenomena characteristic of the disease can be elicited. He is sometimes lifted out of bed into a chair, but he is utterly unable to assist himself. Even when supported on both sides he is quite unable to stand erect or to place his feet to the ground; in that position his legs are flexed on the thighs, and the thighs on the abdomen. There is very little evidence of talipes equinus. He can sit on a chair, doing so with his legs bent under him, and kept somewhat though not very widely apart. Neither when seated nor when standing is

there any lordosis; on the contrary, there is well marked curvature of the spine with the convexity backwards.

The boy is emaciated, but the emaciation is general, and does not strike one as specially affecting the upper limbs, as in his brother's case. The pectoral muscles are much less atrophied than in Francis, as are also the muscles of the neck, scapulæ, &c. The muscles of the spine and the intercostals partake of the general emaciation. There is no apparent hypertrophy of the calves or of the thighs. The thenar and hypo-thenar eminences are emaciated, but not like the condition in progressive muscular atrophy. There is no claw-like deformity of the hands. Measurement of limbs:—

	Right.	Left.
Calf, ...	$9\frac{1}{4}$ inches.	$9\frac{1}{4}$ inches.
Thigh, ...	$11\frac{3}{4}$ „	$11\frac{1}{4}$ „
Forearm, . . .	$6\frac{3}{8}$ „	$6\frac{1}{2}$ „
Upper arm round deltoid,	$6\frac{1}{4}$ „	$6\frac{3}{8}$ „

No fibrillation of the muscles; no mottling of the skin; no gross impairment of sensation. The knee-jerk is absolutely lost; plantar reflex very weak; cremasteric reflex not examined. There is no ankle clonus. The mental condition is quite normal.

When about 13 months old, and just trying to walk, he had in succession hooping-cough, bronchitis, and diphtheria, which kept him in bed for about 14 months. It was only when $2\frac{1}{2}$ years old, therefore, that he really began to walk, which he did quite suddenly. When about 3 years old he had measles. He never had any convulsive attack. The weakness in his limbs was first noticed when he was between 4 and 5 years of age; his gait was observed to be rocking, and he had a tendency to fall all of a heap. His mother cannot say that she ever observed any increased muscular development in his limbs, which is still so marked in his brother; but she says he had the same difficulty in rising from the ground, in going up stairs, or in climbing on a chair. This, and Dr. Robertson's second case, seem to indicate that in some of these cases there is no stage of apparent hypertrophy. The almost total helplessness has come on only since March; he was then able to walk a little; he can now scarcely turn himself in bed without assistance.

Case IV.—Francis M., æt. 9. Up to between four and five years of age he was a healthy boy. He had measles when he was about 14 months old; he never had convulsions; and has had no other illness. He was somewhat slow in learning to speak; but he never evinced any mental inaptitude. The first symptom noted was "rocking" gait, which was attributed

to his stoutness. The waddling was so marked that his mother was advised by some of her neighbours to chastise the boy to cure him of what was looked upon as a bad habit. It was only after this had lasted for a considerable time that the weakness of the limbs was observed.

He is now typically the subject of pseudo-hypertrophic paralysis, exhibiting the characteristic phenomena in walking, rising from the ground, and in ascending a stair. He can climb on a chair only with assistance. He is still able to attend school. The muscles of the upper limbs are much atrophied; the deltoid and triceps are excessively small, and there is almost no pectoral fold; the trapezius and other muscles about the clavicles and scapulæ are much emaciated. The muscles of the forearms are not so markedly wasted. The thenar and hypothenar eminences are small. The spinal muscles are firm and full. There is well marked lordosis. When lying on one side he can abduct and adduct the leg with difficulty, moving it only a little way. The thighs and calves are relatively very large, and are quite firm to the touch. He stands with his feet several inches apart, and prefers having something to lean upon. There is no tendency to talipes equinus: indeed, he is flat-footed. There is no *griffe des orteils.*

Measurement of the limbs:—

	Right.	Left.
Calf, ...	10¼ inches.	10 inches.
Thigh, ...	12 „	11½ „
Forearm, . . .	6½ „	6½ „
Upper arm round deltoid,	6 „	6¼ „

No fibrillation: no mottling of skin: no loss of sensation: temperature not tested, but legs are said to become cold very easily. Knee-jerk absolutely lost: plantar reflex weak: cremasteric not made out as the testes are not fully descended. No ankle clonus. Mental condition quite good.

Since the above note was made this boy has unfortunately met with a fracture of the middle of the shaft of the left femur. It was induced by a very trivial cause, falling down *two* steps of a stair. It is not known exactly how he fell. It will be interesting to note whether union takes place readily and properly, as I am not aware of any observations on the occurrence of such an accident in any case of pseudo-hypertrophic muscular paralysis. If the account of the accident be correct, it would almost seem as if there had been some degeneration of the bony tissue in this case.

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