A MANUAL

OF

DISEASES OF THE NERVOUS SYSTEM

BY

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DISEASES OF THE NERVES AND SPINAL CORD

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this tissue has undergone the atrophy which ultimately supervenes. Thus we may have two classes of cases belonging to this type, one with enlargement of muscles, few or many, the other with only wasting of muscles, the two corresponding in position and in the general course and relations of the affection.

In another form there is never an interstitial growth of fat, and, probably, no considerable increase of fibrous tissue; simple atrophy of the fibres is its pathological characteristic, while primary shrinking of the muscles is its external manifestation. The shoulder muscles are prone to suffer most, and in some cases there is a peculiar affection of the face. These cases are also more variable in the time of life at which the symptoms begin. But these varieties, as we shall see, are connected by cases which to some extent combine the various features. Such combined forms forbid the separation of the types, which, nevertheless, frequently maintain so distinct a course in many members of a family, even through several generations that we are compelled to recognize their distinctness, although we cannot divide them altogether.

The essential element in the disease has been spoken of as a “qualitative” defect, which entails an imperfect development, manifested sooner or later by the defective vitality of the proper elements of the tissue. But there is reason to believe that the defect is not always merely qualitative. In some cases the defect in certain muscles is so absolute at so early a period, as to make it almost certain that these muscles or parts of muscles are congenitally absent, and that the germinal defect is thus quantitative as well as qualitative. The muscles in which this apparent failure is observed vary in the several forms, and will be mentioned in the account of these.

The peculiar form of atrophy, which differs from the others in the early affection of the muscles supplied by the peripheral nerve, is separately described, and its features are not included in these remarks.

PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS (LIPOMATOUS MUSCULAR ATROPHY, &c.).

The affection thus designated usually manifests itself during the later developmental period of childhood, and clearly depends on a morbid developmental tendency, which is often present in several members of the same family. It is characterised by a progressive change in the size, and diminution in the power, of many muscles. The apparent cause of the change has been already mentioned.

Isolated cases, which can now be recognised as examples of this disease, were recorded in England in 1830 (Sir Charles Bell) and in 1847 (Partridge), and in Italy in 1838. A series of cases was described by Meryon in 1852, and Oppenheim in 1855, but enlarge-
ment of the muscles was not conspicuous in these. Several remarkable examples had already come under the notice of Duchenne, who was busy exploring the field of muscular paralysis with the aid of 'localised electricity.' He recognised its novel features, and published an account of it in 1861, under the name by which it has since been generally known.

**Etiology.**—Our knowledge of the causes of the disease is limited to a few general facts. Males furnish the majority of the cases; they suffer at least four, and perhaps seven times as frequently as females. In the latter, moreover, the malady is slighter in degree, later in development, and less frequently causes death.

The disease occurs rather less commonly in isolated cases than in family groups. The number in a family has varied from two to eight. As many as eight brothers suffered and died in the family described by Meryon, while all the daughters escaped. In a family known to me, four sons have suffered and none of the daughters; in another instance two daughters are affected and no sons. Thus there may be a tendency in a family to the affection of one sex, and not the other; but, on the other hand, children of both sexes may suffer in the same family. In many instances in which several members of one generation are affected, no antecedent cases can be traced in the family; the malady, while congenital, is not hereditary. In other families antecedent cases can be traced, and these are invariably on the mother's side. The disease is thus transmitted by women who are not themselves its subjects. In a case in which four brothers suffered, the mother's brother and sister were likewise affected. Again, a brother and sister were affected, one daughter of a second sister, and three daughters of a third sister. In another instance, a boy suffered, and his sister, unaffected, had two sons diseased and a daughter free, of whose children two sons were the subjects of the malady. Thus the congenital tendency is exclusively due to the maternal element in the embryo. This is also shown by a fact many times observed, that the children of the same women, by different husbands, suffer in the same way.

Indirect hereditary tendencies, such as are indicated by the occurrence of diseases of the nervous system, can be traced so rarely that it is doubtful if they have any influence. Neither the age of parents, nor their intemperance, appears influential, and their consanguinity becomes effective only when raised in energy by repetition. Thus in a family known to me, the intermarriages during five genera-

* Nevertheless, Meryon's cases ('Med.Chir. Trans.' 1859) certainly belonged to this variety. Conclusive proof of the affection is afforded by cases in collateral families which have come under the writer's observation in near and distant branches of the same family. Other evidence of the fact is described in a 'Clinical Lecture on Pseudo-hypertrophic Paralysis' (London, Churchill, 1879). The chief literature is referred to in that lecture; the numerous papers that have appeared since have added chiefly to our knowledge of the characters of the allied forms of idiopathic atrophy, &c., and the relations of these to the pseudo-hypertrophic variety, and are referred to on a later page.

**Pseudo-hypertrophic Muscular Paralysis.**

The disease always manifests itself during the period of development, sometimes in the early stage of growth, at the close of infancy, often only during mid-childhood, rarely not until growth is nearly ended. In a third of the cases, the first symptoms are noted when the child first attempts to walk, which is usually a little later than in healthy children; very rarely indeed, the child has never walked. In about another third the child seems well until it is four, five, or six years old, and then impairment of power attracts attention. In three quarters of the cases, the disease manifests itself before the tenth year. Rarely, the patient is conscious of no symptoms until after puberty, at the age of eighteen or twenty, but in such apparently late onset, there has been enlargement of muscles long before power becomes impaired, and the disease began much earlier than it seemed to do. One patient, for instance, in whom weakness was only noticed when she was twenty, had been often "chaffed," when a young girl at school, on account of her "tea-kettle calves." Neither social state nor general constitutional condition seems to influence the occurrence of the disease, but its manifestation has sometimes been apparently accelerated by influences that disturb the general health; it has been first noticed, for instance, during convalescence from some general illness.

**Symptoms.**—Impairment of power usually attracts attention before any change is observed in the size of the muscles, or if these are noticed to be large, it is with feelings of parental pride rather than with suspicion, in spite of the fact that the children often walk clumsily, fall with ease, and rise with difficulty. The act of going upstairs is especially difficult to them; the child has to take hold of the banisters and pull himself up.

The muscles may at first present nothing unusual, especially in slight cases, or if the child is fat, as is frequently the case. But at the age of five or six years an unusual enlargement of certain muscles is usually conspicuous, especially when there is a contrast between these and other muscles which are small. If enlargement is almost universal, it is usually great and conspicuous. The enlarged muscles usually at a time become still larger in comparison with the others, but afterwards they cease to increase and ultimately become smaller, first relatively and then absolutely. This change occurs earlier in some muscles than in others, and it may give place to a condition of distinct atrophy.

Among muscles that are most frequently large, those of the calf take the first place. They sometimes attain a remarkable size. I have measured a calf 14 inches in circumference in a boy of twelve. The muscles in front of the lower leg are less frequently enlarged, but sometimes project beyond the edge of the tibia. The extensors of the
enlarged, the former more frequently than the latter, but occasionally only in one part. Both these muscles are sometimes wasted. The forearm muscles suffer in only a small minority of the cases, and the intrinsic muscles of the hand usually escape altogether.

This escape of the intrinsic muscles of the hand affords a very marked contrast to spinal muscular atrophy, in which they suffer early; but

knee are often big; occasionally the rectus or vastus internus is alone increased in size (the rectus in Fig. 147), and the other parts may be normal or small; less frequently all parts are small. The flexors of the knee commonly escape. The glutei are frequently conspicuously large; the flexors of the hip are, of course, inaccessible to observation, but they are usually feeble, and no doubt diseased; there is generally enlargement of the lumbar muscles, and disease of a peculiar and important character in those of the shoulder.

Of all the muscles of the body, next to those of the calf, no one is enlarged more frequently or in greater relative degree than the infraspinatus. It often stands out so conspicuously that its edge is apt to be mistaken for that of the scapula (Figs. 148 and 149). The infraspinatus is sometimes also prominent, but its condition is usually concealed by the trapezius, which is little involved. The deltoide is also frequently large; the serratus rarely. The pectoralis is never enlarged, but, on the other hand, its lower half is wasted or absent (see p. 518) in a large proportion of the cases, and with this the latissimus dorsi, which has the same action in depressing the raised arm (see p. 26). The teres major may share the wasting of the latissimus.

The other muscles of the arm suffer in diminishing degree and frequency from above downwards. The triceps and biceps are sometimes

* E.g. Scheff, 'New York Neurol. Soc.,' Oct. 2nd, 1888; Baumler, 'Sudwest. Neurol. Versamml.,' Freiburg, 1888. I have once met with wasting of the extensors of both phalanges of one thumb, and enlargement of the abd. indicis has been observed (Taylor, Clin. Soc., April 24th, 1881), and fatty growth in the thenar muscles (Burger, 'Arch. f. Psyeh,' Bd. xiv).
intermediate between this and the third type (see also Westphal, 'Charité Annales,' 1887, xii, p. 447), but in the patient shown in Fig. 152 there was considerable enlargement of the masseters. The tongue has been increased in size in a few instances. The other muscles supplied by the cranial nerves always escape.

The diseased muscles are weak, but the impairment of power is to some extent irrespective of the change in size. The muscles that are abnormally small are generally weaker than those that are abnormally large; and in the latter the weakness increases with the wasting. In the legs, the greatest weakness is in muscles that are inaccessible to examination—the flexors of the lips; next in order of weakness come

the extensors of the knee, and the extensors of the hip. The muscles below the knee usually retain considerable power for a long time, and the extensors of the ankle fail before the flexors. In the upper limbs the depressors of the arm are usually alone weakened during the early period of the disease, but subsequently the shoulder muscles suffer, then the triceps and the biceps, while the muscles that move the hand commonly retain good power to the last.

The distribution of weakness in the legs causes certain peculiar defects of movement which are very characteristic, and some are even all but pathognomonic of the disease. The difficulty in going upstairs is especially due to the weakness of the extensors of the knee and hip. The defect of the extensors of the hip causes the gait to have a peculiar oscillating character, in which the body is so inclined as to bring the centre of gravity over each foot, on which the patient successively throws his weight, because the weak glutes medius cannot counteract the inclination towards the leg that is off the ground, unless the balance is exact. The greatest defect, however, is in the power of rising from the floor, and the most characteristic peculiarity is the mode in which this is achieved, if it is still possible, and no objects are near by which the patient can aid himself. He commonly has not sufficient power to extend the knees when the weight of the trunk is on the upper extremity of the femur, which is then a lever in which the power, applied between the fulcrum and the weight, acts at least advantage. He therefore places his hands on his knees, as in Fig. 150; and his arms thus bring much of the weight of the upper part of the trunk on the femur close to the fulcrum, between this and the power, which can then act at greater advantage. Moreover, the mere weight of the head, which is in front of the arms, tends to aid the extension.

This, indeed, may effect the extension of the knee without the aid of the extensor muscles, as anyone may ascertain by observing the mobility of the patella in this attitude. When the knees are extended, the power of the extensors of the hip may be sufficient to raise the body into the upright position, or the patient may aid them by an upward push with the hand as he takes it off. E, however, these extensors are weak, the hands are often moved higher and higher up the thighs, grasping alternately, and thus pushing up the trunk. To get thus the requisite support, the knees must not be quite extended; and if their extensors have no power, the device cannot be employed, and the patient is altogether unable to rise. In many cases, especially when extension of the hip is easy, the patient achieves the extension of the knees in another way; he puts the hands on the ground, stretches out the legs behind him far apart, and then, the chief weight of the trunk resting on the hands, by keeping the toes on the ground and pushing the body backwards, he manages to get the knees extended, until the trunk is supported by the hands and feet, all placed as widely apart as possible (Fig. 151, 2). Next the hands are moved alternately along the ground backwards, so as to bring a larger portion of the weight of the trunk over the legs. Then one hand is placed upon the knee (Fig. 151, 3), and a push with this, and with the other hand on the ground, is sufficient to enable the extensors of the hip to bring the trunk into the upright position.

The shortening and contraction of certain muscles lead to another group of symptoms—distortions due to permanent alteration in the position of joints. Some of these are produced, as are distortions in other forms of muscular weakness, by shortening of the less affected opponents of the weaker muscles. Thus the knee joints become fixed by the contraction of the flexors, and the elbow by the contraction of
the biceps when the triceps has lost all power. These contractions only occur late, and are usually facilitated by the habitual flexion of the knee and elbow joints. But the deformity at the ankle-joint, which results from contraction of the calf muscles, commences earlier, before their opponents are weak, and is the result of shortening of the muscles. As a consequence of it, the patient cannot get the heels well upon the ground, and the foot cannot be flexed passively beyond a right angle. The gradual increase of the contraction results in considerable "taillies equinus," and as power lessens, the patient is unable to walk less, and the consequent loss of the extension involved in the act permits a rapid increase in the contraction. The feet, as Fig. 152 shows, soon assume a posture of extreme extension, the dorsum being in a line with the front of the leg, or forming with it a convex curve. A subluxation of the ankle-joint takes place, and the articular surface of the astragalus, its anterior extremity, and that of the os calcis, form three prominences under the skin. When this reversal of the ankle occurs, the tibialis anticus can no longer act as a flexor.

Another deformity, which is due chiefly to muscular weakness, is curvature of the spine. An antero-posterior curve, with the concavity backwards, is an early symptom of the disease, and it may become extreme, the upper part of the trunk being carried so far back that a vertical line from the scapula falls an inch or more behind the nuchal. It is due, not to the weakness of the trunk muscles, but to that of the extensors of the hip, in consequence of which the pelvis is inclined forwards, carrying with it the lower lumbar vertebrae; hence the upper part of the trunk has to be held far back to keep the centre of gravity of the body over the feet. The proof of this mechanism is that when the patient sits, and the pelvis is supported on the ischial tuberosities, the lordosis disappears. It is, indeed, replaced by an opposite curve, in which the back becomes convex, clearly due to the weakness of its extensor muscles. This curve may become very great, as in the case shown in Fig. 163. The weakness of the spinal muscles also permits the occurrence of lateral curvature (Fig. 152), influenced, in its direction, by the habitual posture and the preponderance of weakness on one side or the other.

Fig. 152.—Late stage of pseudo-hypertrophic paralysis; a boy fourteen years old, with muscular contraction and wasting, and lateral curvature of the spine.

PSEUDO-HYPERTROPHIC MUSCULAR PARALYSIS.

The electric irritability of the muscles is only altered when distinct weakness or wasting has set in, when it is lowered alike to faradism and voltaism. There is never any trace of degenerative reaction.

The ingo-jerk may be at first normal, but as the extensors of the knee become feeble, it is always lessened and gradually disappears. It is never excessive, and in all advanced cases it is lost. Sensation is unaffected, and so also are the sphincters in the vast majority of cases. Very rarely there has been, towards the end, a slight difficulty in the retention or expulsion of urine, to be regarded, perhaps, rather as a complication than as an effect of the disease. All other functions of the nervous system are commonly normal, including those of the sympathetic. The mental development of the subjects of this disease is generally beyond that of other children of the same age, doubtless on account of the indirect influence of a malady which withdraws them from active amusements; mental defect is a rare complication.

The rate of progress of the disease and its duration vary much. After some years, or between ten and fourteen, the power of standing becomes lost in consequence of the increasing weakness and the contraction of the calf-muscles. When the patient ceases to walk, the muscular disease makes more rapid progress; deformities become greater, and the patient may become almost helpless, except in the hands, and yet live on for several years. Death is sometimes due to some intercurrent malady, as an acute specific disease, but generally the lessened respiratory power causes some chest affection to develop, or one that should be trifling to become grave. Life is thus ended by acute pneumonia or bronchitis, or by chronic lung disease,—a form of VOL. I.
pneumonic phthisis or broncho-pneumonia, which develops gradually, with little febrile disturbance. There is never sufficient paralysis of the respiratory muscles to cause death directly. The duration of the stage of helplessness depends very much on the care which the patient can obtain.

In the cases in which muscular power remains good until after puberty, the progress of the disease is generally slow. The patient may reach the age of thirty before power is much impaired. It is possible that, in some cases, the disease never attains a considerable degree. More frequently, however, after slight symptoms have lasted for some years, a rapid increase occurs, and very few patients reach the age of forty. The course of the disease is slower in girls than in boys, and females furnish a relatively large proportion of the late cases.

Varieties.—The chief varieties of the disease depend on the age at which it commences, and on the condition of the muscles, whether they are large or small. In rare cases a single muscle may be large, and the rest small, as in Fig. 154, in which only the vasti were increased in size. Or everywhere and from the first the muscles are smaller than normal, and they progressively waste. Such cases are not uncommon; in the first group of cases described (by Meryon), enlargement of muscles was inconspicuous. Many cases in which all muscles are small belong properly to the form considered in the next section. The cases described by Meryon must be regarded as examples of the pseudo-hypertrophic form (see p. 505), but they present many points of resemblance to the "simple atrophy" described further on.

Complications.—Congenital mental weakness, due apparently to defective development of the brain, sometimes complicates pseudo-hypertrophic paralysis. In rare cases there have been indications of some other morbid condition of the central nervous system, such as epilepsy. It is uncertain in what light the slight occasional affection of the bladder is to be regarded, whether as an invasion of the vesical muscles or as a central complication. Vigorous has recorded a case in which the symptoms of pseudo-hypertrophic paralysis were combined with the peculiar rigidity of Thomas's disease. Of course the subjects of the disease are liable, like other children, to various affections of the nervous system; I have seen both chorea and polio-myelitis as merely accidental complications.

Pathological Anatomy.—It is rare, at the time of death, for any muscles to be actually larger than natural, and most of those that are affected are usually below the normal size. They are pale and yellowish in colour, and often, to the naked eye, resemble perfectly masses of adipose tissue. The resemblance is not merely one of aspect. As seen under the microscope, it may be difficult for the observer to realise that he is not looking at a fatty tumour. Nothing may be at first visible but fat-cells, precisely like those of adipose tissue. Among the cells, however, are tracts of nucleated fibrous tissue, and a closer examination of these shows that the tracts contain also muscular fibres (Fig. 155), most of them much narrower than normal. They are also irregular in width; a broad fibre, for instance (as in the figure), suddenly becoming narrow.

The fibres for the most part preserve their transverse striation, but where they are narrowed this may have in part disappeared, either by granular degeneration, or, more commonly, by a simple fading of the strata. In the narrowed fibres the striæ are sometimes further apart than normal. In other parts broad fibres may be seen, normal or nearly normal in aspect (Fig. 156) coursing among the fat-cells, and accompanied by a smaller amount of fibrous tissue. Fibres occasionally present fatty degeneration, a longitudinal striation or fissuring, vitreous ("waxy") degeneration, or vacuolation, but these are rare. Some empty sarcolemma sheaths may be seen where the narrowing of the fibres is greatest. In muscles that still preserve some red tint, the amount of fat is less, and there is often a relatively larger amount of interstitial fibrous tissue. Very rarely, in some part of a muscle, there has been only wasting of the fibres, without the interstitial change, present elsewhere (Singer, in the triceps).