The Primary Muscular Dystrophies.

A thesis presented by

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The Primary Muscular Dystrophies

The interesting group of diseases which are included under the name of the Primary Muscular Dystrophies, were until recent years looked upon as diseases of the Nervous System, due to changes in the Spinal Cord. During the last few years, however, a great deal of attention has been directed to their study, & careful investigations by many competent observers have resulted in their now being considered to be Primary affections of the muscles themselves. They are marked by certain well defined characters which distinguish them from those diseases in which the muscular system is more or less extensively affected secondarily to some lesion of the Nervous System, central, or peripheral.

These distinctive characters are:—

(i) Changes in the muscular system, which do not correspond with any known nerve distribution; the muscles or groups of muscles ~~affected~~ affected undergoing slow but very sure & progressive loss of power.

(ii) Sensation is not impaired.

(iii) The electrical reaction of the diseased muscles is not changed qualitatively, but only quantitatively. Their electrical irritability is

much reduced to both the galvanic & faradic currents, but there is never any reaction of degeneration.

(iv) The Sphincters are not affected, nor are the laryngeal, pharyngeal & diaphragmatic muscles.

(v) A very marked tendency for the disease to attack several members of the same family & to pass from one generation to another. They are essentially congenital in character, though merely potential at the time of birth. It is by no means uncommon especially in the Pseudo-hypertrophic variety to find cases without any previous family history so that they are congenital rather than hereditary.

Having thus distinguished them as a group from other groups of diseases, considerable difficulty is experienced in dividing the group into its various classes. Certain types have been described, but many, if not the majority, of the cases do not conform to any one of the types, but are intermediate in character, uniting the symptoms of various types in differing proportions. In whatever way we may subdivide the group, it cannot be too strongly urged that all the types described are probably simply

varieties of a single condition which has been well named by Erb, Dystrophia Muscularis Progressiva. Probably the subdivision of the group suggested by Erb is the best that has so far been placed before us, from a scientific point of view. It is as follows.

Dystrophia Muscularis Progressiva.

I Cases which occur in childhood.

II Cases which occur in youth.

I(a) Hypertrophic forms { (i) Pseudo-hypertrophy (ii) Real hypertrophy

I(b) Atrophic forms { (iii) Primarily affecting the face (iv) not affecting the face.

II is the type usually known as Erb's Juvenile Type.

Before proceeding to a further consideration of the various types, it will be well to give a brief sketch of the more important steps in the historical evolution of the group. As long ago as 1830 Sir Chas. Bell, & in 1847 Partridge, described cases of what is now recognised as Pseudo-hypertrophic Muscular Paralysis. Meryon,* in 1852, seems to have been the first who carefully studied this disease & as the result of Post-mortem examination he came to the conclusion that it was a disease primarily of the Muscular System & unassociated with any lesion of

* Med-Chirurg Transactions vol XXXV p. 73.

the cord, or other part of the Nervous System.
In 1864 the same observer* published the result of another carefully made autopsy which completely confirmed the views which he had announced twelve years before, & suggested that the degenerative changes in the muscles were possibly due to defective nutrition. In 1861 Duchenne† described fully & carefully cases of the Pseudo-hypertrophic type, but failed to recognise till 1868 their essentially muscular origin. In 1855 the same observer‡ described a condition of atrophic paralysis, now recognised as the Facio-Scapulo-Humeral type of Landouzy & Déjérine, which he classed with Progressive Muscular Atrophy of adults, naming it "Atrophie Musculaire Progressive de l'enfance". He assumed that, like the Adult Progressive Muscular Atrophy, this had its origin in changes in the Spinal Cord. In 1885 Landouzy & Déjérine published‡ the results of careful investigation of a number of cases which were, as above stated, identical with Duchenne's Progressive Muscular Atrophy in Infants and

† "De l'Électrisation Localisé" 3rd edit. pp. 595 etc.
‡ "De l'Électrisation Localisé" 3rd edit. pp. 578 etc.
‡ Revue de Médicine Feb-April 1885
* Charcot "On Paralysis" p. 210

proved as the outcome of Post Mortem examination that his conclusions as to their Spinal origin were incorrect & that, on the other hand, they were not dependent upon, or associated with, any demonstrable lesion of the Nervous System, but were apparently primarily muscular affections. Leyden*, in 1875, wrote a paper in which he proposed to separate the hereditary forms of Muscular Atrophy from those of the Duchenne type & pointed out the strong points of resemblance between these hereditary atrophic forms & the Pseudo-hypertrophic type. Other important contributions to our knowledge before 1891 were the able monograph by Gowers (1879) on Pseudo-Hypertrophic Muscular Paralysis, in which he brought especially to view the fact that it is exceedingly common to meet with cases which combine in various proportions the Hypertrophic & Atrophic varieties & a paper by Erb+ published in 1882, in which he first described cases of what is now known as Erb's Juvenile Type of Muscular Dystrophy. In 1891 Erb published a treatise[a] (a translation of which has been published by the New Sydenham

*Klinik der Rückenmarkskrankheiten II p. 525.

+Handbuch der Electrotherapie p. 389.

[a] "Dystrophia Musculoris Progressiva" Leipzig 1891

Society vol. 148. (1894)] in which he methodised the knowledge at that time at our disposal, which, as the previous notes show, was little more than a collection of more or less disconnected types, & proved + that they were all simply varieties of a single condition, classifying them as before stated.

Having given this historical sketch of the more important points in the evolution of the group of the Primary Muscular Dystrophies, I propose, before entering on a general description of the various types, to describe certain cases which have come under my immediate notice during a period of office as Resident Medical Officer to the London Temperance Hospital.

The first patients were three brothers aged 13, 10, & 5 years respectively, each of whom exhibits a different stage of the Pseudo-hypertrophic type.

Family History after most searching enquiry no trace could be found of any muscular or nervous disease in any member of the three previous generations. Their father, a previously healthy man, died at the age of 35

+ New Sydenham Society Translation pp. 248 et seq.

of "blood poisoning". Their mother appears to be a healthy woman of the working class. There are no other brothers or sisters. The income of the household seems to have been of a very limited & somewhat precarious character & consequently the children had never been very well nourished probably.

Personal History All of them had had measles though not very severely & the second boy had also had whooping cough, but no clear connection could be established between these illnesses & the apparent time of onset of their present maladies.

State on admission None of them had any pain at all. In all the temperature was slightly subnormal.

Circulatory System Percussion revealed considerable enlargement of the area of cardiac dulness in the two elder boys, the apex beat being at the seventh rib in the nipple line in the eldest & at the sixth rib in the nipple line in the second. On auscultation no cardiac murmurs were heard but the cardiac action in the cases of the two elder boys showed some amount of irregularity. The pulse rates were 96, 76 & 88 respectively & the pulse

showed no change in tension or volume from the normal.

Respiratory System Percussion & Auscultation revealed nothing abnormal. Respirations were 24, 24, & 28 per minute respectively. Neither of them had any cough.

Alimentary System nothing unusual was found except persistent constipation in the second boy & very bad teeth in the youngest.

Nervous System Sensation & the special senses were unchanged. They all slept well. The boys were all up to the average in intelligence, the eldest rather above it, his school master speaking very highly of him as "one of my most promising pupils."

Urinary System The urine in all was acid, of normal Specific Gravity & contained neither deposit nor Albumin. A faint trace of sugar was found ~~in~~ on applying the yeast test in each case.

Locomotory System In this system occur all the characteristic phenomena of the disease & consequently each case must be described separately.

S. J. aged 5 years, his mother states, began to walk when about eighteen months of age.

Recently she has noticed that he has not been able to walk so far as he used to do without fatigue, & that he drags his feet a good deal. He exhibits the disease in an early stage. At present the muscular system is not visibly much affected in any part except the muscles of the Calf, which are enlarged & hard & to a lesser degree the Infra-spinatus, Deltoid & Rectus Femoris which are similarly affected. There is a marked tendency to Talipes Equinus. When asked to rise from the floor he goes through a series of ~~manoeuvres~~ manoeuvres which is very characteristic of this disease. He first turns over on to his face, flexes his knees so as to rise on to his hands & knees, then extends his knees so as to rise on to his hands & feet. Taking his feet as a fixed point, he now gradually moves his hands nearer to them until he is able to place one of his hands on the corresponding knee & then the other. It is then necessary to extend his hip in order to assume the erect attitude & this he does by gradually climbing with his hands up his thighs, till, by suddenly throwing back his shoulders, he is able to shift the Centre of Gravity further back & so extend his hip.

When erect he stands with his legs widely separated in order to provide himself with as broad a basis as possible & it is observed that he has a well marked lateral curvature of the spine. He also experiences great difficulty in going up stairs & overcomes it - some what as follows. Supporting himself on the balusters & fixing his body with the arm thus occupied, he swings one leg laterally on to the stair above gradually bringing it to a position in front of him. He now rests his free hand on his knee & partly pulls his body forward by means of the balusters, partly jerks it forward thus extending the leg & completing the step. This process has to be repeated for each succeeding step. The difficulty experienced in rising from the floor & in going up stairs is mainly due to weakness of the extensors of the hip & knee which results in difficulty in raising the foot from the ground to clear even small obstacles & hence even in its earliest stages this disease is characterised by a singular proneness to falling. The movements of his arms are good in all directions. He walks with a characteristic oscillating gait. The knee jerks are slightly diminished

owing to the commencing affection of the Rectus Femoris. The affected muscles react fairly well to both Galvanism & Faradism, but those of the right calf are less responsive than those of the left.

A. T. aged 10 years, his mother states, began to walk when two years old. About three years ago she first noticed that his feet were beginning to "turn in" & that he began to be "shaky on his legs." The early stages of the disease corresponded closely with those described as existing in his younger brother. About eighteen months ago he ~~legs~~ began to get much worse & at the end of six months had entirely lost all power in his legs. She thinks that since that time he has been stationary.

Examination of the muscular system shows that the Muscles of the head & neck are apparently unaffected. The muscles of the Shoulder show the following changes. The Infraspinatus & Deltoid are largely atrophied. The Latissimus Dorsi & Pectoralis Major especially in its lower part are much atrophied. He can raise his arm to a right angle & can touch the top of his head & his shoulders.

The movements of the two sides are about equal
The other muscles around the Shoulders appear
to be little if at all affected.
The muscles of the arm are soft rather small
especially the biceps which is more affected
on the right than the left side. Flexion of
the elbow can be completely performed but
very slowly especially on the right side.
The muscles of the forearm are soft but of a
good size. Pronation & Supination can be
satisfactorily performed.
The muscles of the hand. are not changed.
The muscles of the back, abdomen & thorax are
small & thin. Those of the back are so weak
that it is extremely difficult for him to assume
the ~~erect~~ sitting attitude when recumbent.
There is no marked Spinal Curvature.
The muscles of the Lower Extremity. The
Glutei are bulky but flabby. The muscles
of the thighs are of fair size except the Vastus In-
ternus which is enlarged in both thighs. The
Quadriceps Extensor is weak. The general
muscular condition as regards development is
about equal in the two thighs & so it is also as
regards power. The Calf muscles are
somewhat enlarged & flabby the left side

being more affected than the right. The distinguishing feature of this case is the very strong contraction of the flexors of the knees & of the Tendones Achillis, resulting in strong permanent ~~contracture~~ flexion of the knees & extension of the ankles. The result of this is complete inability to stand even with support.
The muscles all react to both Galvanism & Faradism, but their irritability is much reduced to both currents.

E. J. aged 13 years, his mother states, began to walk when about two years old. He first showed signs of losing the use of his legs when between five & six years of age & during the last few months both arms have become much affected. In the early stages of the disease he suffered in exactly the same way as we have seen his youngest brother is suffering at the present time. Examination of the Muscular system shows the following condition:-
The muscles of the head & neck seem to be unaffected except the trapezius which is considerably wasted on both sides.
The muscles of the Shoulders except the Infra-spinatus which is considerably enlarged & the

Deltoid which is slightly enlarged are of small size, especially the Latissimus Dorsi & lower two thirds of the Pectoralis Major which are almost completely gone. The absence of these muscles may be demonstrated by instructing the patient to attempt to adduct the arm from the horizontal position against resistance when no muscle will be seen between the humerus & the pelvic brim.

The arm can only be abducted to a very small degree, slightly more on the left than the right side. He cannot touch his right shoulder with his fingers but he can his left.

The muscles of the arm are all small especially the biceps & brachialis anticus. He can flex his forearm completely but extremely slowly.

The muscles of the forearm are all small. Pronation & Supination can be performed fairly well, better on the right than the left side. The muscles of the hand are not affected.

The muscles of the back are small & very weak. There is marked antero-posterior curvature with prominence of the lumbar vertebrae but no lateral curvature.

The Rectus Abdominis is bulky.

The muscles of the Lower Extremity The Glutei are rather large but very flabby & weak. The muscles of the thigh are in no case hypertrophied. The flexors & adductors are atrophied & weaker in the left than the right leg. The Quadriceps Extensor is very weak in both limbs. The muscles of the Calf though still above the normal size are not nearly so large as they used to be, Atrophic processes having set in. They are firmer than they formerly were owing as we shall see later to the absorption of the fat cells & the contraction of the hypertrophied fibrous tissue. There is marked Talipes Equino Varus. The ankle cannot be flexed passively beyond a right angle.

Till eight or nine months ago he was able to walk & for three months longer he could stand with support. Since then, however, he has completely lost all power to do either, owing to his inability, even when supported, to extend either his hip or his knee.

He can only raise himself from the recumbent to the sitting attitude by a great effort with the assistance of something to pull at.

The knee jerks are abolished.
The reaction of the diseased muscles to both Galvanism & Faradism is exceedingly slow, electrical irritability being extremely reduced.
There is no trace of reaction of degeneration.*
These boys remained under my care for some considerable time, but though electricity & massage were freely used & Tabloids of Thymus Gland were administered daily for some time. No improvement was observed in any of them & all that one could hope was that the progress of the disease may have been to some extent retarded. The eldest boy, however, seemed to get slightly weaker in spite of all treatment.

A little later for a very short time I had under my care two brothers aged 10 & 9 years & their sister aged 7 years, all suffering from the same disease. A description of their condition would be to a large extent a repetition of what has gone before without the more advanced symptoms found in the eldest of the three brothers.

* These boys were shown by Dr. Fletcher Little & myself at the meeting of the Clinical Society, London on January 24th. 1896 & a short report is to be found in Volume XXIX of the Society's Transactions.

The chief interest lies in the existence of the Pseudo hypertrophic type of Muscular dystrophy in a girl, which is a comparatively rare occurrence, boys being affected certainly four, perhaps nearly seven times as frequently as girls. They remained for so short a time under my care that nothing can be said as to the progress of the disease.

At the same time as the first three cases were under my care in the same ward was a young man R. B. aged 28 years, a painter by occupation. His family history showed nothing of importance as regards his father, mother, seven Brothers & two sisters, but he had one brother suffering from a much more advanced stage of the same condition. He was a total abstainer from alcoholic drinks. Four years before he had had a severe attack of Influenza & about two years later definite signs of disease began to manifest themselves, while he was resident in Canada. The first thing he noticed was great weakness of the lower extremities, which used frequently to "give way under him".

On admission his temperature was subnormal, his pulse 64 & respirations 20. His

urine was Acid in reaction, S. G. 1010 & contained no deposits, sugar or albumin.
The area of cardiac dulness was increased somewhat. The heart's action was weak but regular. There were no cardiac murmurs.
He had a markedly oscillating gait & found some difficulty in turning round. He could not stand with his legs together, nor with his eyes shut without swaying. The knee jerks were almost lost on both sides. The muscles of the Calf, the Vastus Internus, the Glutei, the Deltoid, the Infraspinatus, & the Triceps were all more or less enlarged & weak in the upper extremity, the Biceps was rather atrophied & weak. The electrical irritability of these muscles was much reduced, their reaction both to galvanism & faradism being very slow. There was, however, no trace of the reaction of degeneration in any of them.
He complained of wandering pains especially in the lumbar region & the lower extremities.
He gradually became worse in spite of the administration of various drugs, including Iodide of Potassium, Arsenite of Sodium, Nux Vomica, Thymus Gland, etc. at various times & the frequent application of electricity & massage & was lost sight of owing to

insubordination, suddenly, so that no record was taken of his state on leaving the hospital.
This seems to me, in spite of some unusual symptoms, to be a case of the Adult form of Pseudo-hypertrophic Muscular Paralysis. The enlarged, weak muscles, with the slow electrical reaction but without any reaction of degeneration, & the occurrence of two cases in the family all seem to point to its inclusion in the group of the Muscular Dystrophies.

From these cases it is possible to compile a fairly complete description of the Pseudo-hypertrophic type of Muscular Dystrophy which may be summarised as follows:-
The disease most commonly comes on about the ages of 5 to 7 years & attacks a far larger proportion of males than females. It tends to attack several members of the same family.
The earliest part noticed in very many cases is that the baby is very late in beginning to walk, but this does not as a rule arouse any suspicion. Before any change can be detected in the size & contour of any of the muscles, one observes a distinct but slowly advancing loss of power. This is chiefly

manifested by the readiness with which the child falls down & the difficulty he experiences in getting up again without help, by the increasing awkwardness shown in going up stairs, & by the curious oscillating, waddling gait which gradually develops owing to the increasing difficulty experienced in raising the feet from the ground. These early phenomena are ~~occasionally~~ mainly due to weakening of the extensors of the hip & the knee.

After a varying length of time curious changes are noticed in the outlines of certain muscles. These grow larger in bulk & at the same time often become firmer & harder, when compared with the other muscles which are either unchanged or atrophied. The muscles which are almost invariably affected in this way are the Gastrocnemius & the Soleus in the Calf & the Infraspinatus & Deltoid in the region of the Shoulder. The following muscles are also often hypertrophied, though not nearly so constantly, the Glutei, the Sartorius, the Extensors of the knee, (especially the Vastus Internus & the Rectus Femoris) the muscles in front of the leg, the Lumbar muscles, the Supraspinatus, the Serratus Magnus & the Triceps & very rarely the Biceps.

On the other hand some muscles are almost always distinctly wasted, especially the Latissimus Dorsi & the lower third or even two thirds of the Pectoralis Major which may even be entirely absent. The Biceps which we have already noted as being very rarely hypertrophied is most commonly atrophied considerably & frequently the flexors of the knee & the flexors & adductors of the hip are similarly changed. The muscles of the hand are never, or hardly ever, affected & the same may be said of those of the face & neck, except perhaps the Trapezius which is sometimes atrophied.

The change in muscular bulk is no true index to the power of the muscle, an apparently very large muscle often having scarcely any power at all, e.g. the weakness of the enlarged calves is proved by the absolute inability of the patient to stand on tip-toes after a comparatively short time. From this we see that the large size of the muscles is not due to any real hypertrophy of the muscular fibres themselves, if it were there could be no difficulty in performing such a simple muscular exercise.

Conspicuous amongst the symptoms of this disease are various distortions. These

arise from one of two causes, either as the result of the loss of the balance normally existing between opposing muscles or groups of muscles owing to the progressive weakening of some of them or as the result of contracture in the muscles themselves of the hypertrophied fibrous tissue, which we shall see presently is characteristic of this disease. To the latter class of distortion belongs the extension of the ankle so specially marked in my second case which arises from the strong contraction of the fibrous tissue in the muscles of the Calf. To the former class of distortion belongs the strong flexion of the knees in the same case, the contraction of the flexors of the knee not meeting with its usual opposition owing to the weakness of the Quadriceps Extensor muscle.

Probably the most common distortion, & the one which occurs earliest in the course of the disease is Talipes Equinus, frequently passing on to Talipes Equino-Varus later, resulting from the contraction of the fibrous tissue in the hypertrophied calf muscles. In these cases the heel cannot be got to the ground, nor can the foot be flexed beyond a right angle passively. As the muscles get weaker & walking becomes

more & more difficult, this contraction increases, owing to the loss of the flexion of the ankle involved in walking which tends to some extent to counteract the tendency to contract.
Another very common distortion is Spinal Curvature. Two forms of this are to be seen. An antero-posterior curvature with the concavity in the lumbar region, a Lordosis, is often seen when the patient is in the erect attitude This condition existed in my two elder boys as long as they were able to stand but cannot be observed at all now. It seems to be due to weakness of the extensors of the hip which allows of the slipping forward of the pelvis which carries with it the lower lumbar vertebrae. In order to keep the Centre of Gravity over the feet this has to be compensated by holding the upper part of the trunk far back. This curvature usually quite disappears when the patient is sitting owing to the pelvis being supported on the Ischial Tuberosities & therefore no longer tending to slip forward & is frequently replaced by an exactly opposite one, Kyphosis, in which the lumbar vertebrae are prominent. This is seen as a permanent condition

in the eldest brother.

Lateral Curvatures are not uncommon and arise from weakness of the Spinal muscles which is greater on one side than on the other.

Electrical Reaction. The diseased muscles behave in every case towards electricity in the manner stated at the beginning of this paper to be characteristic of the whole group. They react to both the Galvanic & the Faradic currents but the electrical irritability is reduced in proportion to the extent of the changes in the muscles. This of course arises from the fact that only those muscular fibres can contract which still retain some of their muscular qualities.

The Knee-jerk in early cases is little, if at all reduced but as the Extensors of the Knee become more & more wasted it becomes progressively weaker & in advanced cases may entirely disappear.

Sensation is always unimpaired & the Sphincters, laryngeal, pharyngeal & diaphragmatic muscles are practically always unaffected.

Mental Development is as a rule satisfactory.

The Prognosis in the Pseudo-hypertrophic type is invariably unsatisfactory. Every year a progressive disability is to be expected exceedingly few reach adult life. Very few patients survive the loss of the power of standing for more than six or seven years. The prospect is distinctly more favorable in those cases which do not develope until adult life. The disease may never reach such an advanced stage as it does in the younger patients. Gowers* does mention one case typical of this variety, who is over forty years of age, has never been able to run & has remained ~~stationary~~ stationary for at least the last eight years probably longer.

Having described thus the Hypertrophic variety of Progressive Muscular Dystrophy, I propose now more briefly to sketch the other varieties which by many writers are classed together for descriptive purposes under the designation Idiopathic or Primary Muscular Atrophy.

* Dis of the Nervous System 1899 vol. I p. 582.

Atrophic Varieties of Primary Muscular Dystrophy.

These types are of much rarer occurrence than the Hypertrophic variety. They are characterised by muscular wasting from the outset with very rarely any enlargement of muscles. Weakness & wasting generally manifest themselves at the same time in the diseased muscle. They attack the two sexes in about equal numbers. (a) Those cases in which the facial muscles are primarily involved (the Facio-scapulo-humeral type of Landouzy & Déjérine & the Atrophie Musculaire Progressive de l'enfance of Duchenne) This ~~cases~~ type in the great majority of cases comes on ~~at~~ a very early age, but a considerable number of cases have been described which have not developed till adult life & even as late as middle life. The outstanding characteristics of this type are the peculiar changes which take place in the contour & expression of the face, as the result of weakness & wasting of certain of the facial muscles, which has earned for it the term "the Myopathic face."

Very commonly the first-muscles to fail are the Zygomatici & the Orbicularis Oris. As a result of this we find that the naso-

labial furrow is lost & a characteristic change takes place in the smile. Usually, in smiling, the angles of the mouth are drawn upwards & outwards but in this condition there is often nothing more than a faint upward movement owing to the action of the levatores Anguli oris, & labii superioris, or a slight stretching by the action of the buccinators. The affection of the Orbicularis Oris results in the lips being habitually separated to some degree & the lower lip is more or less prominent. The patient can neither pout, nor puree his lips as in the act of whistling. He has no power of blowing out his ~~lips~~ cheeks, as in the process of blowing a trumpet or other wind instrument; owing to the affection of the lips, the pronunciation of the labials is often very imperfect. In some cases the first thing that is noticed is that the child sleeps with his eyes open, owing to the weakness of the Orbicularis Palpebrarum & even if he can close them, the very slightest opposing pressure will make it impossible. Occasionally the weakness & wasting of these muscles is so extreme as to produce a condition of Exophthalmos in some exposures to

to the wind produces a large overflow of tears. Frequently the frontales are diseased & the forehead is then absolutely smooth & cannot be gathered into wrinkles, or into a frown. The muscles of the tongue larynx & pharynx are never involved, the buccinators very rarely & the Ocular muscles never. Gowers,* however, describes a case in which an affection of the facial muscles, corresponding to that described above, was accompanied by paralysis of the ocular muscles. The patient was a female aged 27 years. The ocular paralysis began gradually when she was 24 years old & increased until all power was lost of moving either eye upwards, the left eye inwards & the right eye outwards, & all the other movements were weakened. The internal ocular muscles were normal. The eyelids drooped slightly. The facial changes followed these ocular changes & in this case consisted chiefly in weakness & wasting of the Zygomatici & Orbicularis Oris. Later still the arms became feeble & the flexors of the hip almost powerless. There was no visible change in the nutrition or electrical irritability of the muscles. & the knee jerk was normal.

* Dis. of the Nervous System 1899 Vol I. p. 587

The question is whether this was primarily a case of central or muscular disease. If, as he thinks, it was central, then these peculiar facial changes cannot be looked upon as confined to Primary Muscular Atrophy. If it was muscular, then the ocular muscles are sometimes affected.

At a later period the disease spreads to the region of the shoulder & upper arm & very striking Scapulo-humeral changes are produced When these are fairly advanced the Shoulders become sloping, the neck broad & the upper angle of the Scapula may often be seen projecting between the tip of the shoulder & the line of the neck. When the arms are extended in front, we get the marked characteristic "winging of the Scapula", when, as is very commonly the case, the Serratus Magnus is diseased. The muscles which are most often attacked first are the Biceps, Triceps, Brachialis Anticus & Supinator Longus. The Latissimus Dorsi & lower one or two thirds of the Pectoralis Major are nearly always much wasted & are often completely wanting. Sometimes the whole of the Pectoralis Major & even the Pectoralis

Minor are attacked. Owing to the weakness & wasting of these muscles, as well as the Trapezius & Rhomboids, which are affected in most cases, the characteristic looseness of the shoulders arises. This results in the child seeming as if he were going to slip through your hands when you lift him up with your hands in his axillae. The shoulders are raised to the level of the ears, owing to the Scapulae not being bound down as they usually are by these muscles, which have become weak & wasted. This looseness is seen in all cases of Primary Muscular Dystrophy to some extent.

The Deltoids are usually normal. Occasionally they are supposed to be rather enlarged, (this, however, is probably only by comparison with the greatly atrophied muscles around them in most cases) very rarely they are somewhat atrophied. The same remarks hold good as regards the Infraspinatis, Supraspinati, & Subscapulares & Sterno-Mastoids.

These muscular changes naturally result in the defective performance of some of the movements of the Shoulder-joint, according to the muscles most attacked. In many cases for instance the hand can only be raised to the

top of the head by swinging it to the back of the neck then climbing up the back of the head.

In the forearm, with the exception of the Supinator Longus which, we have already seen, is generally attacked early, the muscles generally escape. Rarely the long extensors & flexors show some weakness with, or without, slight wasting. Very rarely the muscles of the forearm have been very extensively diseased. As a rule the muscles of the hand escape, but occasionally the ~~hand~~ interossei become affected, as in a case reported by Erb.*

The muscles of the back, abdomen & thorax, often escape altogether & are hardly ever attacked till very late in the course of the disease. The muscles of the neck also suffer late.

In the lower Extremity the Glutei, the extensors of the knee, the flexors of the Hip & the adductors of the thigh are most often atrophied but the muscles of the leg are rarely attacked. When they are, however, the atrophy is usually pretty general.

Electrically the muscles show the qualities already stated as being common to all types of Primary Muscular Dystrophy. Thesis no 2 Translation p. 239.

*Prog. Musk. Dystrophy (Neo Sydenham Soc.

reaction of degeneration.
Sensation is not affected.
Distortions occur, as in the Hypertrophic variety, but are rarely of any considerable degree, & are mostly due to contraction of the muscles less affected by the disease. Lordosis, in the erect attitude, often replaced by Kyphosis in the sitting position, is produced probably by the same mechanism as has been previously described. As the Atrophy increases the outlines of the parts affected becomes much altered.

The above, without the description of the facial changes, is a complete picture of the second subdivision of the Atrophic form of Primary Muscular Dystrophy, commonly called the Scapulo Humeral variety.

The course of this variety is very variable. In some the progress is very rapid it only takes ten or twelve years, or even less, to attain its extreme development. On the other hand patients have been known to live to over seventy years of age Sometimes the atrophy is confined to the part in which it begins eg the face. In others it does not pass to the limbs

till some years after the facial attack, or vice versa, & even then the patient may be able to continue his work for several more years. In some cases the disease seems to come to a complete standstill when the period of development is over, though up to that time it may have been making steady progress. The disease we see then may last for any length of time from ten, or less, to fifty, or more, years. It is never directly fatal. Death results from intercurrent complications, most commonly of a pulmonary character, unconnected with the disease itself. In the severe rapidly spreading cases, death most often results from Phthisis.

Finally we have, what is generally known as Erb's Juvenile Type, a form of Muscular Dystrophy which usually comes on during the second decade of life, generally about puberty. It is a familial disease which attacks both sexes alike. The condition of the muscular system corresponds so closely with that described as existing in the Scapulo-humeral Type that many writers describe the two

together. Besides the usual time of its onset, the only point that need be noted is that in this type the deltoids, Infraspinati, Supraspinati occasionally the muscles show a greater tendency to hypertrophy than they do in the Scapulo humeral type. The outstanding feature of the typical Clinical picture of this variety is the extreme smallness of the arm & thigh when compared with the well developed fore arm & leg. The disease progresses very slowly.

Such are the four most clearly distinguishable types of Progressive Muscular Dystrophy. That many cases of an intermediate character exist has been already stated. Thus a case is sometimes seen in which one half of the body exhibits the hypertrophic variety & the other half the Atrophic, or Juvenile, variety. All sorts of combinations of the various types have been seen. It is not uncommon to find different members of the same family suffering from different varieties of Muscular Dystrophy & this is a very strong argument in favor of their essential unity from a Clinical point of view.

It is now necessary to turn our attention to the Aetiology, Pathological Anatomy, Pathology, Diagnosis & Treatment of the Disease.

Aetiology As to the causation of the myopathies very little of a definite character can be said. It is a congenital condition affecting, in the case of the Hypertrophic Variety males chiefly, in the proportion of at least four, possibly as much as seven, to one; in the case of the Atrophic varieties both sexes about equally. The majority of cases come on in infancy, or early childhood, though the disease is merely potential at the time of birth. The juvenile form does not come on till the second decade & the disease may not manifest itself until adult life. That the cause of the muscular changes probably lies in some congenital defect in the muscles themselves not in any gross anatomical lesion of the Cord will appear evident when we have considered the Pathology of the disease. It is not so easy to say, however, that the changes are not secondary to some trophic changes in the Cord.
Parental consanguinity has been supposed to be a factor in the causation but it is very

doubtful whether it has any influence except when it has become very powerful, by repeated intermarriages for one or two generations. A determining cause of onset is often some acute disease, muscular weakness first manifesting itself during Convalescence.

Pathological Anatomy. It is extremely rarely that one finds post mortem any of the muscles actually larger than normal. Occasionally, however, such has been the case even the muscular fibres themselves have, under the microscope, proved to be hypertrophied. More commonly, however, the muscles are either much atrophied, or, if they have been previously hypertrophied, they have returned to their normal size.

A portion of the enlarged Calf muscle was removed from the leg of my second boy, at the time that a tenotomy was performed in the hope of restoring some amount of walking power to him. Under the microscope this showed: -

(1) Muscular fibres fewer in number than normal & many of them smaller in size. Many were not of uniform size throughout their length being much narrowed in places.

Some were but little altered, none were seen which were distinctly enlarged.
The transverse Striation of the fibres was preserved throughout, but had become very faint in many places, especially where the muscle fibres had become particularly narrowed. This may be due in some cases to a process of granular degeneration, but probably in the majority it is simply a case of fading. The nuclei were increased in number very.

(11) Between the muscular fibres were broad bands of multinucleated fibrous tissue in which considerable numbers of fat cells had been deposited. Around more fibres which had been little, if at all, changed the fat cells were much greater in number, the fibrous tissue less in amount, than was the case around more fibres which were greatly disorganized.
To the naked eye the muscle looked greyish in color & of a fatty nature.

The above is a fairly typical picture of a section of a Pseudo-hypertrophied muscle, as seen under the microscope. Such a section may show fibres in all stages of Atrophy, or Hypertrophy, or in a normal condition, with the Transverse Striation normal or in various

stages of fading, according to the state of the fibre. In some cases the fibres themselves may show fatty, or waxy, degeneration, or vacuolation, but these changes are comparatively rare. This description of the muscle fibres holds good in all the types of Progressive Muscular Dystrophy. In the same muscle all stages of degenerative change may be found, & different muscles in the same patient often exhibit very different appearances.

In muscles which have become Pseudo-hypertrophic, between these fibres are found strands of hypertrophied fibrous tissue, multinucleated & interspersed with a larger or smaller number of fat cells. As the disease advances & the Pseudohypertrophy tends to pass into atrophy, the fat cells gradually diminish in number & may ultimately completely disappear. In these cases the Interstitial Tissue is practically entirely fibrous & the muscle fibres suffer very much more injury as the process advances. This is the condition of the majority of those muscles which undergo primary atrophy. Whether a muscle undergoes Hypertrophy or Atrophy

seems in short to depend as a rule on the presence of excess of fatty tissue, or fibrous tissue respectively. In some muscles which undergo Atrophy, however, the principal change to be observed is often simply a narrowing of the muscular fibres, with gradual fading & ultimate disappearance of the transverse striation, without any formation of fat, or fibrous tissue. Such is the case with the Latissimus Dorsi & Pectoralis Major. This would lead us to the view that it is a primary affection of the muscular tissue due to loss of vital energy, owing to interference with the trophic nerve supply to the muscle.

One important feature in the microscopic appearance of a diseased muscle, which has been specially noted in a fair number of cases, is the large number of muscle-spindles which have been seen in the section. In normal muscle they are not very conspicuous, but, owing to the Atrophy of the muscle fibres in this disease, the relation between the two has become altered & the Spindles stand out prominently. The significance of this fact has not been properly determined as yet. On this subject Dr. L. E. Batten* says:-

* Encyclopaedia Medica Vol 8. p. 196.

"It is known that these structures are connected with nerve fibres of a sensory nature, which pass up in the posterior roots, & it is also known that in such conditions as Infantile Paralysis, Progressive Muscular Atrophy, these muscle spindles do not atrophy. Now if it could be shown conclusively that these muscle spindles exist unaltered in muscle which otherwise had undergone complete atrophy then it would seem probable that the disease must depend on some lesion situated at a point where the course of the motor & sensory fibres lay apart, i.e. somewhere above the Posterior Root Ganglion. The question is not, however, easily answered, for it has been shown by Sherrington* that the muscle spindle only very slowly undergo atrophy after its nerve supply is cut off, & again the number of cases of myopathy in which special attention has been directed to this point is very limited."

The motor nerves have never been found changed microscopically. One observer has ~~observed~~ stated that he has found slight changes in the end muscle plates of the nerves, but, as far as I am aware, this has not been confirmed by any one else. If such

*Journal of Physiology Vol XVII p. 237

be proved to be the case, the disease may be found to be of the nature of a tropho-neurosis due to failure of nutrition in these terminal structures. In a great majority of cases the Spinal Cord has been found practically normal, but occasionally slight degenerative changes of an irregular character have been found, as in the case described by Gowers & Lockhart Clarke* in which the Cervical & Dorsal regions were normal with the exception of here & there slight accumulations of the products of degeneration at the bottom of the fissures which are probably due to perivascular erosions common in all ages. At the last dorsal segment, however, there was an area of granular disintegration in the intermediate grey substance on each side in front of the Posterior Vesicular Tract. This part was unusually translucent for half a centimetre in vertical extent & in the centre of this area the disintegration had produced an actual cavity across which the fibres of the Cerebellar Tract ran unchanged. The anterior grey matter was unchanged, however, & Gowers thinks the changes were merely associated with, not the cause of, the symptoms (Dis. of Nerv. Syo. 1899 Vol. I, p. 379)

* Med. Chir. Transactions Vol LVII p. 247

On this Subject Poore says:* -

"On one important point, however, there is unanimity, viz. that the motor cells in the front horn are not affected in Pseudo-Hypertrophic paralysis. Lesions, degenerative & inflammatory, have been found in the cord, but they have not been definite, or the same in different cases & in view of the fact that the records of patients who have died of this disease are generally the cords of helpless & deformed invalids with distorted Spinal Columns, observers hesitate to link together the spinal & muscular lesions, as cause & effect."

Pathology At one time, not many years ago, all the types of this disease were supposed to have a spinal origin arising from changes in the ~~motor~~ cells in the Anterior Grey matter. Consideration of the results of microscopic examination given above would certainly lead one to suppose that any changes which may have been found in the cord are so inconstant & so slight that they can have no causal relation to the disease, but are probably only secondary & indirect results of the disease.

*Trans. of Duchenne's Works (New Sydenham Soc.) p. 191 Editorial Note

Duchenne* who first carefully described the facio scapulo himeral type sought to prove that it was of spinal origin due to atrophy of the Anterior horn cells of the Spinal Cord. He considered that their trophic functions only were affected not their motor functions. This belief prevailed until Landouzy & Déjérine+ in 1886 demonstrated that it did not depend on, nor was it associated with, any demonstrable lesion of the cord. These facts have already been mentioned in the historical sketch at the beginning of this thesis.

The facts given in describing the Pathological Anatomy necessarily lead one to the conclusion that this is a disease primarily of the muscles themselves, as far as one can judge from the material which is as yet at our disposal.

As the result of careful observations on the development of the muscular system of the foetus[a], it has been demonstrated that the muscles which are as a rule most affected are those which developed earliest in foetal life. This apparently due to some defect

*New Sydenham Society, Translation p. 6 et seq.

+Revue de Médicine 1885 pp. 81 & 257.

[a] Rabinski & Onanoff. Soc. de Biologie Feb 11th 1888

in the developmental processes which take place in the germinal tissues which goes to form muscles. Accepting this theory, it is not surprising to find that in some cases this disease is associated with similar developmental defects in the Nervous system, such as epilepsy & intellectual weakness.

No proof exists of this disease being acquired in a healthy normal organism.

The defect would appear to lie in the maternal & not the paternal part of the embryonal tissues from the fact that the disease has been known to appear in several children by the same mother but by different fathers e.g. the case reported by Duchenne* in his work on localised electrisation.

To decide how the changes in the muscle fibres take place is a matter of great difficulty & has never yet received a final solution. The predominant fact under the microscope is, as we have seen, the overgrowth of fibrous tissue with or without the deposition of fat cells. Whether, however, these changes primarily arise from this overgrowth, or, as seems to me more probable, this overgrowth is secondary to morbid processes of a congenital nature taking place in the muscular fibres is a question upon which

*New Sydenham Soc^{y} translation p. 184.

much difference of opinion has existed & still exists.

Duchenne*, in his excellent paper on Pseudo-hypertrophic Muscular Paralysis, summarises the results of his investigation thus†:-

"(i) Hyperplasia of the Interstitial Connective "Tissue with production of more or less fibrous "tissue is the fundamental anatomical "lesion of the muscles in Pseudohypertrophic "Muscular Paralysis."

"(ii) This Hyperplasia is present in all the "~~affected~~ paralysed muscles when they begin to increase "in size." (Hence he named the disease Myo-sclerotic Paralysis.)

"(iii) The increase in size of the muscles is in "direct proportion to the hyperplasia of the "interstitial fibroid & connective tissue".

"(iv) The increase of fibrous & connective tissue "is usually associated with a small number "of fat vesicles, or, as has been observed in "Germany it may be replaced by a large num-"ber of fat vesicles. This latter condition ap-"peared to him to constitute the most advanced "stage in the change of the Interstitial Connec-"tive Tissue".

"(v) The transverse striation of the muscular"

* New Sydenham Soc. translation pp. 176. et seq.

"fibres is preserved, in most of them, in all their
"length but it becomes very faint & scarcely
"apparent. At the points where the transverse
"striation has disappeared are seen longitu-
"dinal markings & sometimes when these be-
"come effaced the sheaths of the sarcolemma
"appear to contain fat cells, which however
"are in reality derived from the surrounding
"connective tissue & which otherwise differ
"from the fatty granules which are charac-
"teristic of fatty degeneration of the muscles."
"(VI) The hyperplasia of the interstitial connec-
"tive tissue appears generally only in the second
"stage of the disease & appeared to him to be pre-
"ceded by a congested condition of the muscles
"which occasions also a slight increase in
"their volume. At this period (first stage of
"the disease) the transverse striation is often
"exceedingly faint."

While Duchenne considers now that the funda-
mental anatomical lesion is the Hyperplasia
of the Interstitial Connective Tissue, Erb
on the other hands looks upon the primary
essential changes as taking place in the
muscle fibres themselves & not in the interstitial
Connective tissue. He believes that the

alteration in the Connective Tissue may at the utmost begin at the same time as, but more probably follows after, the changes in the muscular fibres & that last of all the lipomatous tissue appears in the overgrown connective tissue. He concludes thus*:-

"According to our present knowledge the "cause of the morbid changes in the muscles "is somewhat as follows: First, there are "alterations in the muscular substance itself. "These hypertrophy & their nuclei proliferate "they swell up, taking more of a rounded form, "& fatty subdivision goes on; peri fascic "there is slight increase & proliferation of "nuclei in the connective tissue. At an early "period, however, muscles here & there atrophy, "& this process quickly extends more or less, "gradually gaining the upper hand, & finally "leading to complete disappearance of the mus "cle substance. Along with it goes on a very "great increase in the amount of connective tis "sue, with proliferation of the nuclei, thicken "ing of the vessels & so forth. In this tissue, "sooner or later, fat cells make their appearance, "leading, it may be, to the most extreme forms "of fatty degeneration. The muscular fibre

*New Sydenham Society translations vol 148. p 259

"has now wholly, or about wholly, disappeared
"The final result of the whole process is one of two
"forms. It may be an atrophic sclerosing lipo-
"matosis, in which there is a good deal of pure
"connective tissue without much fat, the
"original volume is much reduced, or it may
"be an hypertrophic lipomatosis in which
"there is little else than fat, while the original
"volume is either maintained or exceeded".

Erb, however, while thus describing the history of a diseased muscle, is not altogether disposed to believe that the disease is primarily an affection of the muscles. He very strongly inclines to the opinion that it is, on the other hand, the outcome of functional disturbances in the cord, consequently of the nature of a trophoneurosis, in contradistinction to the gross anatomical lesion of the cord which produces Progressive Spinal Muscular Atrophy. He says*:-

"The considerations which weigh with me are
"various. The muscles depend for their nutri-
"tion to a very large extent on trophic nerve cen-
"tres; the localisation of this atrophy frequently
"follows in a noticeable way the exact course
"of the nerves in a plexus, or the disposition of the

*New Sydenham Soc. trans. vol CXLVIII p. 262 etc.

"centres in the central organ, & occasionally we
"find a case of spinal amyotrophy presenting an
"almost exactly similar arrangement. Here-
"ditary influence is of great importance; men-
"tal aberrations are common among the patients
"& other neuroses often occur in their families.
"Further, even in the undoubtedly spinal cases,
"such as Acute Anterior Poliomyelitis, similar
"morbid changes (hypertrophy, proliferation
"of nuclei, division of fibres) both in the mus-
"cles & the connective tissue have been pointed
"out by W. Muller[a], Déjérine-Huet[b], Joffroy-
"Achard[c] & Hitzig[d]. In Pseudo-hypertrophic cases
"malformations & changes of a minor kind in the
"Spinal Cord have been met with. When I con-
"sider these facts, & bear in mind further
"the results of Heubner's[e]" (an undoubted case of
Dystrophy with extensive atrophy of the large
cells of the anterior horns of grey matter) "&
"Fohmaier's[f] observations" (a similar case but
with less marked changes) "I cannot avoid the"

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a Beitr. zur pathol. Anatomie u. Physiol des menschl. 1871. Rückenmarks

b Archives de Physiologie 1888 p. 386.

c Archives de Médicine expériment. I 1882 p. 64

d Ueber spinale Dystrophien. Berl. Klin. Woch. 1889 Nr 28

e Ein paradoxer Fall von infantiler progress. Muskelatrophie Wagner Festschrift Leipzig 1887

f Ueber progr. Muskelatrophie. Deutsch. Med. Woch. 1886 Nr 23 & 24

"The suspicion that after all the affection may be de-
"pendent on the nervous system. It is tempting to sup-
"pose, as I formerly expressed it, that we have to do with
"a kind of tropho-neurosis, having its origin in the tropic
"centres of the cord - a disturbance of the function
"of these centres which finds its expression in the
"very complicated muscle changes of the di-
"sease. While on this supposition there are, as a
"rule, no coarse nerve changes, now & then, after
"the affection has lasted a long time or been very
"intense, such a change does become visible.
"The idea is inevitable that if something like
"this is the case the relations between dystrophy
"& spinal amyotrophy will turn out again to be
"close & intimate. The latter would represent
"an affection of the tropic centres that from the
"very first is a distinct coarse anatomical
"lesion taking effect in a degenerative atrophy
"of the muscles with fibrillar twitchings, reac-
"tion of degeneration, etc. The former would at
"the outset be merely a functional disturbance
"of these centres, conditioned probably by dif-
"ferent causes, & expressing itself as muscular
"dystrophy with all its characteristic symp-
"toms. At the same time there would remain
"the possibility that even this merely functional

"disturbance might in the long run become asso-
"ciated with a coarse lesion of the centres. Many
"things about these affections would agree very
"well with such a supposition, among them the
"occasional occurrence of reaction of degeneration
"in dystrophic cases the occasional initial
"localisation of spinal amyotrophy in the
"shoulder + trunk. But ..... the whole ques-
"tion is by no means yet ready for decision + the
"future alone can lift the veil reveal the finer
"processes that as yet lie hid from us."

This striking suggestion of Erb supported as it is by as many ~~strong~~ arguments is of undoubted importance + may ultimately turn out to be the true explanation of this disease.

The principal objections which present themselves to my mind are:-

(1) The correspondence in the localisation of the lesions with those found in definitely Spinal cases is, as Erb confesses, only very occasionally seen, being absent in the great majority of cases.

(11) In many cases where the functions of several muscles are influenced by a particular segment of the Cord very variable conditions are found some being changed in one way, others in another, + others again not at all. Thus the fourth

cervical segment of the cord is stated to influence the functions of the Deltoid, Infra-Spinatus & Supraspinatus which we have seen are usually either hypertrophied or unchanged; of the Biceps, Supinator Longus Rhomboids which are most commonly atrophied; & of the diaphragm which is practically never attacked. The trifacial nerve through its ophthalmic division supplies the orbicularis palpebrarum which is very commonly attacked in the facial form & the ciliary muscle of the eyeball which is never affected so we might adduce many other examples.

(iii) As has been previously said the changes found in the cord are so irregular as to be negligible from the point of view of causation & occur, as a rule, in advanced & distorted cases. These according to E & B would probably follow after a functional disturbance of the same region.

On the other hand points which may be in favour of a trophoneurosis are: -

The apparently unconfirmed observation previously recorded of changes in the muscle end-plates of nerves.

The numerical prominence of spindle cells &c.

served in several cases of diseased muscles in section which corresponds with the appearances which exist in Infantile Atrophy & the spinal cases.

The weight of evidence at present at our disposal, however, must lead us to the conclusion that the disease originates in the muscles themselves, owing to congenital defect in their tissues. This view receives strong support from the coexistence in many cases of similar developmental defect in the Nervous System.

That the changes originate in the muscle fibres themselves rather than in the Interstitial Connective Tissue seems to me to be the more probable theory. None of the Histological observations which have been announced seem to be opposed to the theory & a strong point in its favor seems to me to be the condition already stated to be found in some of the diseased muscles e.g. the Latissimus & Pectorals, in which the fibres undergo simple atrophy without any interstitial change at all.

Diagnosis With all the foregoing facts before us, diagnosis, as a rule, should present very little difficulty.

The enlarged Infraspinatus in combination with the very defective Latissimus Dorsi & Pectoralis Major especially when associated with enlarged weak Calf muscles is of the greatest diagnostic importance in the Pseudo-hypertrophic Variety.

In the Facio-Scapulo-humeral type the peculiar affection of the muscles of the face, especially of the Zygomatici, is characteristic, though perhaps not actually diagnostic.

In the Scapulo-humeral & Erb's Juvenile Types the marked wasting of the Shoulder, upper arm & Thigh in comparison with the well developed fore arm & Leg is suggestive.

The time of onset, together with a greater tendency to hypertrophy of certain muscles, will serve to distinguish the Juvenile from the Scapulo humeral variety.

The oscillating gait - characteristic method of rising from the floor are also of importance in separating them from Spinal cases.

From any Spinal injuries with which the Muscular Dystrophies might be confused,

They may be distinguished by the electrical conditions of the muscles especially the absence of the reaction of degeneration, the absence of fibrillary twitchings of the muscles, the gradual loss of knee-jerk, the irregular distribution of the muscle changes. The slow gradual onset of the disease & its special progress to attack children & to appear in more than one member of a family are also points of considerable importance. Progressive Spinal Muscular Atrophy, for instance, rarely manifests itself before adult life. Another very important fact is that the Muscular Dystrophies commonly begin in the proximal end of a limb & spread downwards, very rarely affecting the muscles of the hand at all. Spinal cases on the other hand usually begin at the distal end in the muscles of the hand & spread upwards to the trunk. In Spinal cases, moreover, the Atrophic Changes are more universal in their incidence.

The nervous affections with which the Muscular Dystrophies might be most easily confused, with their distinguishing features are:—

1) Progressive Spinal Muscular Atrophy

occurs generally in adults. Usually attacks the muscles of the hand first. Muscles show fibrillary twitchings + reaction of degeneration. The muscle reflexes are long retained.
(II) Syringo-Myelia attacks distal parts of limbs first. Shows no hypertrophy. + Reaction of Degeneration is present + Sensory disturbances are prominent.
(III) Chronic Multiple Neuritis in which there are pain + other sensory disturbances, reaction of degeneration + characteristic localisation of the paralysis.
(IV) Progressive Neurotic Atrophy or the so called "Peroneal" type, which though congenital, + a disease of childhood shows atrophy beginning at the distal ends of the limbs with reaction of degeneration, fibrillary twitchings + sensory disturbances.

Treatment Nothing of a satisfactory nature can be said on this subject.
Duchenne* claims to have cured the disease in its earliest stages by faradization combined with Hydropathy + Shampooing.
In most cases little or nothing can be done

* New Sydenham Society translation p. 183.

to check the progress of the disease.

The general health of a member of a myopathic family, who has not yet been attacked by the disease, should be very carefully attended to, & an attempt should be made to protect him from all that tends to depress vitality & so predisposes to the disease. Regular exercise, short of decided fatigue, should be insisted on in such a case.

When the disease has actually manifested itself it would appear that drugs are of no use as curative agents. Many have been tried such as Phosphorus, Arsenic, Iodide of Potassium, Nux Vomica, & Thymus Gland but all with equal non-success. Electric currents of both kinds have been tried with about as much good result. Gibb* thinks that electricity & massage may do good, even produce arrest of the Atrophic forms at any rate, but their course is so irregular that it is almost impossible to say that they have been arrested by any of the means adopted.

The only thing that seems to be of real value is regular muscular exercise. It is of the utmost importance that the patient should

* New Edenburgh Soc. Trans. 1894 p. 265

keep about on his feet as long as he has the least power should be drilled in a regular method of muscular exercise, which should not be too violent, should be regulated in accordance with the strength & condition of nutrition existing in the particular muscle being exercised. In this way more muscle fibres which as yet show no tendency to degeneration may be stimulated to further growth & may attain greater power. It at the same time counteracts the tendency to contraction which exists in the affected muscles. Where strong contraction exists, especially in the Calf muscles, tenotomy should be tried in most cases in the hope of restoring some of the lost power of walking. It had not the smallest effect, however, when tried in the record of my cases. Massage by stimulating the circulation & consequently improving the nutrition of the muscles is probably of some value.

In pronounced cases great care must be taken to protect the patient from catarrh or other intercurrent affections, as they, especially if of a Pulmonary character, usually hasten the fatal termination of the disease