The Primary Muscular Dystrophies

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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 investigation 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 secondary to some lesion of the Nervous System, central, or peripheral.

These distinctive characters are:

1. Changes in the muscular system, which do not correspond with any known nerve distribution; the muscles or groups of muscles affected undergoing slow but very sure progressive loss of power.
2. Sensation is not impaired.
3. The electrical reaction of the diseased muscles is not changed qualitatively, but only quantitatively. Their electrical irritability is
much reduced to both the galvanic and radian currents, but there is never any reaction of degeneration.

(v) The sphincters are not affected, nor are the laryngeal, pharyngeal and diaphragmatic muscles.

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

Having thus distinguished them as a group from other groups of diseases, considerable more 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, exhibiting the symptoms of various types in differing proportions. In whatever way we may sub-divide 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. Atrophy Muscularis Progressiva. Probably the subdivision of the group suggested by Erb is the best that has as yet been placed before us, from a scientific point of view. It is as follows.

Atrophy Muscularis Progressiva.

I. Cases which occur in childhood.

II. Cases which occur in youth.

I(a) Hypertrophic form. (Ib) Atrophic-hypertrophic.

I(b) Atrophic form. (Iv) Atrophie, 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 Charles Bell, in 1874, Partridge, described cases of what is now recognised as Pseudo-hypertrophic Muscular Paralysis. Menenino, 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 the nervous system.
the cord, or the part of the Nervous System. In 1834, the same observer published the result of another carefully made autopsy, which completely confirmed the views which he had advanced twelve years before, suggested that the degenerative changes in the muscles were possible, due to defective nutrition. In 1831, Duchenne described fully, carefully cases of the Pseudo-Athetotic type, but failed to recognise till 1858 their essentially muscular origin. In 1835, the same observer described a condition of atrophic paralysis, now recognised as the Brain-Scapulo-Humeral type of Landouzy & Dejerine, which he classified 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 1855, Landouzy & Dejerine 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 added, "De l'Atrophie Musculaire," 3rd edit. pp. 695 et., & "De l'Atrophie Musculaire," 3rd edit. pp. 678 et., in Revue de Medicine, Feb.-April 1885. *Meyron, "An Atlas of Diseases," 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.

Leypold, in 1875, wrote a paper in which he proposed to separate the hereditary forms of muscular atrophy from those of the Duchenne type. He pointed out the strong points of resemblance between these hereditary atrophic forms and the pseudo-hypertrophic type. Other important contributions to our knowledge before 1871 were the able monograph by Lowen (1879) on pseudo-hypertrophic muscular atrophy, 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. In 1872, a paper by E. E. published in Paris, in which the first described cases of what is now known as E. E.'s juvenile type of muscular atrophy.

In 1871, E. E. published a treatise (a translation of which has been published by the New Sydenham Society).
Society vol. 148. (1874) in which he established 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 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 12, 10, and 5 years respectively, each of whom exhibits a different stage of the pseudo-hypotrophic type.

Family history after most searching enquiry no trace could be found of any muscular or neuro-muscular disease in any member of the three previous generations. Their father, a previously healthy man, died at the age of 45.
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 and 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 and the right fifth 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 and Auscultation revealed nothing abnormal. Respiration were 24, 22, 12 per minute respectively.
Neither of them had any cough.

Alimentary System: nothing unusual was found except persistent constipation in the elder boy; very bad teeth in the youngest.

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

Urinary System: The urine in all was clear, of normal specific gravity and contained neither albumin nor sugar. A faint trace of sugar was found on applying the yeast test in each case.

Locomotor 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 she has not been able to walk as far as she used to do without fatigue, that the drag of her feet is 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 Caly, which are enlarged. And even in a lesser degree the Infraspinatus, Teres major, Pectoralis major, which are similarly affected. There is a marked tendency to palpable equines. When asked to rise from the floor he goes through a series of manual movements which is very characteristic of this disease. He first turns over on to his face, flexes his knees, works to rise on to his hand and knees, then extends his knees, works to rise on to his hand and 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 to extend his hip.
Whenever he stands with his legs widely separated in order to provide himself with as broad a base as possible it is observed that he has a well marked lateral curvature of the spine. He also experiences great difficulty in going up a stairs overcome it some what as follows. Supporting himself on the balusters raising his body with the arm thus occupied, he swings one leg laterally, onto the chair 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 successive step. The difficulty experienced in rising from the floor in going up stairs is mainly due to weakness of the extensors of the hip and knee which results in difficulties in raising the foot from the ground to clear even small obstacles. Hence even in its earliest stages this disease is characterised by a sinister 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
more of the right calf are less responsive than
more 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 de-
scribed as existing in his younger brother.
About eighteen months ago he 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 change.
The Infraspinatus & Deltoide are large & soft.
The Subscipium Dorsi & Pectoralis major espe-
cially in its lower part are much atrophied.
He can raise his arm to a right angle & can
reach 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 arms are soft and 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 but thin. Those of the back are so weak that it is extremely difficult for him to assume the correct sitting attitude when recumbent.
There is no marked Spinal Curvature.
The muscles of the lower extremity, the Glutei are bulgy but flabby. The muscles of the thighs are of fair size except the Vastus Internus which is enlarged in both thighs. The Biceps 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 of flabby.
being more affected than the right. The distinguishing feature of this case is the very strong contraction of the muscles of the knee and of the tendons Achilles, resulting in strong permanent contraction of the knee and extension of the ankles. The result of this is complete inability to stand even with support.

The muscles all react to both Salvarsan and Paradime, but their irritability is much reduced by both currents.

E. J. aged 13 years, his mother states, began to walk when about two years old. The first showed signs of losing the use of his legs when between five and 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 conditions:

The muscles of the head and neck seem to be unaffected except the temporalis which is considerably wasted on both sides. The muscles of the shoulder, except the infraspinatus which is considerably enlarged,
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 and 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 brachii and anterior. He can flex his forearm completely but extremely slowly.

The muscles of the forearm are all small. Pronation and 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 but 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 Shin
lei are rather large but very flabby & weak.
The muscles of the thigh are in no case hypertro-
phied. The flexor adductors are atrophied
weak in the left than the right leg. The
extension is very weak in both
limbs. The muscles of the calf though
still above the normal size are not nearly
as large as they used to be, Atrophic process
having set in. They are firmer than they for
merly were bowing 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 but 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 knees.

He can only raise himself from the recum-
ent 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 and tracodium is exceedingly slow; electrical irritability being extremely reduced. There is no trace of reaction or degeneration.

These boys remained under my care for some considerable time, but though electricity and massage were frequently employed, no improvement was observed in any of them till that one could hope was that the progress of the disease may have been in 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 and 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 Mr. Pechen Little myself at the meeting of the Clinical Society, London on January 27th, 1876. A short report is to be found in Volume XXIX of the Society's Transactions.
The chief interest lies in the existence of the peculiar hypertrophic type of muscular dystrophy in a girl, which is a comparatively rare occurrence, boys being affected certainly more, 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 P. B., aged 28 years, a fruitery by occupation. His family history showed nothing of importance as regards his father, three

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

effects 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. This
The urine was acid in reaction, S. G. 1010 contained no deposit, 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 marked, recumbent gait and found some difficulty in turning round. He could not stand with his legs together, nor with his eyes shut without staggering. The knee-jerks were almost lost on both sides. The muscles of the Calf, the Vastus Intermedius, the Glutei, the Biceps, the Infraspinatus, the Triceps were all more or less enlarged. There was a lack of extension of the Biceps in the upper extremities. The Biceps was rather atrophied. The electrical irritability of these muscles was much reduced, their reaction both to galvanism and to the current being very slow. There was, however, some of the reaction of degeneration in all of them.

He complained of wandering pains especially in the lumbar region and the lower extremities. He gradually became worse in spite of the administration of various drugs, including Iodide of Potassium, Arsenite of Sodium, Nux vomica, Ipecacuanha, and other remedies. At various times the frequent application of electric current was lost sight of owing to
insubordination, suddenly, as that no record was
taken of his state on leaving the hospital.
This seems to me, in spite of some unusual symp-
toms, to be a case of the Adult Form of Pseudo-
paralytic muscular Paralysis. The en-
larged, weak muscles, with the slow electrical
reaction but without any reaction of degenera-
tion, the recurrence 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-
paralytic type of muscular dystrophy which
may be summarised as follows:—
The disease most commonly comes on about the
age of 5 to 7 years attacks a far larger propor-
tion of males than females. It tends to attack
several members of the same family.
The earliest fact 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 or contour of any of the mus-
cles, 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 unsteadiness 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 mainly due to weakening of the extension of the hip and the knee.

After a varying length of time, curiously enough, are noticed in the outlines of certain muscles. These grow larger in bulk and the same time often become firmer and 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, and deltoïd in the region of the shoulder. The following muscles are also often hypertrophied, though not nearly so constant: the biceps, the biceps, the extensor of the knee (especially the vastus internus and the rectus femoris). The muscles in front of the leg, the biceps, the infraspinatus, the serratus magnus, the triceps very rarely the biceps.
On the other hand some muscles are almost always distinctly wasted, especially the Latissimus dorso in the lower third or even two thirds of the Pectoralis major which may even be entirely absent. The Pectoral muscles which we have already noted as being very rarely hypertrophied are commonly atrophied considerably. Frequently the flexors of the knee, the flexors and 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 muscle except perhaps the Trapezius which is sometimes atrophied.

The change in muscular bulk is a true index to the power of the muscle, an apparent increase in 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 tiptoes after a comparatively short time. From this we see that the large size of the muscle 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 stigmata.
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 one of them, or as the result of contracture in the muscles themselves. The hypertrophied fibrous tissues 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 and 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 clefting 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 four back. This curve usually quite disappears when the patient is sitting owing to the pelvis being supported on the Ischiatic Tuberosities therefore no longer tending to cleft forward its frequency 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 electro-stop and the Faradic current, 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 extension of the knee becomes more and more involved it becomes progressively weaker. In advanced cases it may entirely disappear.
Sensation is always unimpaired. The sphincters, laryngeal, pharyngeal, and 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 toes of the lower limbs for more than six or seven years. The prospect is distinctly more favorable in those cases which do not develop until adult life. The disease may never reach such an advanced stage as it does in the younger patients.

*Sorner—does—mention—one—case—typical—of—this—variety,—who—is—over—forty—years—of—age,—has—never—been—able—to—run.—This—remained—stationary—for—at—least—ten—years.—Probably—longer.

Having described thus the Hypertrophic variety of Progressive Muscular Atrophy, 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.

*Six of the Nervous System 1899 Vol. I p. 582.
Atonic Varieties of Primary Muscular Dystrophy.

These types are of much rarer occurrence than the Hypertrophic variety. They are characterized by muscular wasting from the outset with very rarely any enlargement of muscle. Weakness + wasting generally manifest themselves at the same time in the diseased muscle. They attack both sexes in about equal numbers.

(a) Those cases in which the facial muscles are primarily involved (the Facio-Scapulo-Humeral type of Landouzy + Féjérine + the Atonic Musculaire Progressive de l'enfance de Duchenne). This rarest 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 contours + 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 Zygomaticus + the Orbicularis Ovis. As a result of this we find that the naso-
labial furrows is that no characteristic change takes place in the smile. Usually in smiling, the angles of the mouth are drawn upwards and outwards but in this condition there is often nothing more than a faint upward movement owing to the action of the levators anguli oris, labii superioris, or a slight stretching by the action of the buccinator. The action 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 purse 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 action 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 resulting from these muscles is so extreme as to produce a condition of Erythrophalmo ia in some cases.
to the wind produces a large overflow. If seen
frequently the frontale are discussed. The
forehead is then absolutely smooth and cannot
be gathered into wrinkles, or into a frown. The
muscles of the tongue, larger of pharynx are
nevernoised, the buccinator very rarely.
The Ocular muscles never.

* Evers, however, describes a case in which an affection
of the facial muscles, corresponding to that de-
scribed 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 outwards the
right eye outwards, fall the other move-
ments were weakened. The internal
ocular muscles were normal. The eyelids
dropped slightly. The facial changes
followed these ocular changes. In this case
consisted chiefly in weakness consisting of
the zygomatico-orbicularis oris. Later
still the arms became feeble. The flexors of
the hip almost powerless. There was no visi-
ble change in the irritation or electrical irrita-
bility of the muscles. The knee jerk was normal.

The question is whether this was primarily a case of central or muscular disease. If, as he thinks, it was central, then those 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 shoulders, upper arm, and every 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 and the line of the neck. When the arms are extended in front, we get the marked characteristic winging of the scapula, which is very commonly the case, the serratus major is involved.

The muscles which are most often attacked first are the Pectoralis Major, Biceps, Brachialis Anterior, Supinator Longus. The Risorius and two or three others of the Pectoralis Major are nearly always much wasted. Rarely, often completely wanting. Sometimes the whole of the Pectoralis Major even the Pectoralis
Minor are atrophied. Owing to the weakness, wasting of these muscles, as well as the Trapezius and 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 axilla. The shoulders are raised to the level of the ears owing to the Scapulae not being forced down as usually by these muscles, which have become weak and wasted.

This looseness is seen in all cases of Primary Muscular Dystrophy to some extent.

The Scapulae are generally 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 Deltoides, Supraspinatus, Infraspinatus, Subscapularis, Teres-Major, and so forth.

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

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

The muscles of the back, abdomen, thighs, often escape altogether and 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 Erectors, the extensor 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. Observations Prof. Musch. Dystrophy (New Sydenham Soc. Trans. P. 239).
reaction of degeneration.
Sensation is not affected.
Distortions occur, as in the hypertrophic variety, but not rarely of any considerable degree, and mostly due to contraction of the muscles less affected by the disease. Contractis, in the erect attitude, often replaced by Clypsis, 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 Atrophy, commonly called the Scapulo-Humeral variety.

The course of this variety is very variable. In some the progress is very rapid, not only taking ten or twelve years, or even less, to attain its extreme development. In the other hand, patients have been known to live 10 or 20 years. Sometimes the atrophy is confined to the part in which it begins, the face. In others it does not pass to the limbs.
All alone 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 and never directly fatal.

Death results from intercurrent complications, most commonly, if 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 the 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 show a greater tendency to hypertrophy than they do in the fibrous-tissue type. The outstanding feature of the typical clinical picture of this variety is the extreme smallness of the arm and thigh when compared with the well-developed fore arm and 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 a 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 Astrology, Pathological Anatomy, Pathology, Diagnosis & Treatment of the Disease.

Astrology

As to the causation of the majority of these 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 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 necessary, however, that the changes are not due to secondary or some hypertrophic 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 rare to find post-mortem any of the muscles actually larger than normal. Occasionally, however, such has been the case when 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, may 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 an amputation 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 and many of them smaller in size.
2. Many were not of uniform size throughout their length, being much narrowed in places.
Some were but little altered, more 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 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 also.

(11) Between the muscular fibres were broad bands of multinucleated fibrous tissue in which considerable numbers of fat cells had been deposited. Around these 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 these fibres which were greatly disorganised. 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 pseudohypertrophied 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, interpenetrated with a larger or smaller number of fat cells. As the disease advances, the pseudo-hypertrophy tends to pass into atrophy, the fat cells gradually diminish in number and 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 these 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 out until ultimate disappearance of the transverse striation, without any formation of fat, or fibrous tissue. Such is the case with the Retinaculum Dors. Delt. or Loculitis muscle.

This would lead us to the view that it is apparently affection of the muscular tissue due to loss of vital energy, owing to interference with the motor 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-splendrels 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 splendrels stand out prominently. The significance of this fact has not been properly determined as yet.

On this subject Dr. E. B. Rattan* urges—

"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 Paralytic Progressive Muscular Atrophy, these muscle spindles do not atrophy. How 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 spindles 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 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 anyone else. I have...
be proved to be the case, the disease may be found to be of the nature of a hyperviscous due to failure of nutrition in those 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 Cowen and Clarke in which the Cervical and 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 erosion 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 fact was unusually translucent for half a centimetre in vertical extent from the centre of this area. The disintegration had produced an actual cavity, which the fibres of the Cerebellar Tract ran unchanged. The anterior grey matter was unchanged, however, Cowen thinks, but the changes were merely associated with, not the cause of the symptoms (Dis. of Nerv. Syst., 1879, Vol. V, p. 173).
On this subject Poore says:—

"One important point, however, is one permanence, viz., that the motor cells in the posterior 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 cords 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 causal results.

Pathology. At one time, not very 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 as incidental as the existence of such that they can have no causal relation to the disease, but are probably only secondary indirect results of the disease.

* From Pachenie Works (New Sydenham Soc.) 12/19.

Editorial Note
Duchenne was first carefully described the Facio-Scapulo-Humeral 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 and not their motor functions. This belief prevailed until Landouzy & Dejerine 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 me to the conclusion that this is a disease primarily of the muscles themselves, as far as one can judge from the material which is at yet at our disposal. As the result of careful observations on the development of the muscular system of the foetus, it has been demonstrated that the muscles which are as a rule most affected are those which developed earliest in foetal life. It apparently due to some defect.

*New Sydenham Society, Tranglation p. 69 et seq.
+Revue de Medicine 1885 p. 81 + 237.
& Rabinski Bonnoff. Soc. de Biologie Feb 11. 1888
in the developmental processes which take place in the germinal tissues which go 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 and 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 electricism.

To decide how the changes in the muscle fibres take place is a matter of great difficulty that never yet received a final solution. The prevalent 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 the overgrowth, or, as seems to me more probable, this overgrowth is secondary to most fibro-proto processes of a congenital nature taking place in the muscular fibres is a question upon which *New York Times* June 8, 1874.
much difference of opinion has existed, and still exists.

Duchenne, in his excellent paper on Pseudo-
hypertrophic Muscular Paralysis, summa-
rizes the results of his investigation thus:

"(1) Hypertrophy of the Interstitial Connective

Tissue with production of more or less fibrous

Tissue as the fundamental anatomical

Stage of the muscles in Pseudo-hypertrophic

Muscular Paralysis."

"(2) This Hypertrophy is present in all the

Paralyzed muscles when they begin to increase

in size. (Hence he named the disease myo-

sclerotic Paralysis).

"(3) The increase in size of the muscles is in

direct proportion to the hypertrophy of the

"interstitial fibrous connective tissue."

"(4) 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 ad
tive

Stage in the Change of the Interstitial Connective

Tissue."

"(5) The Transverse Section of the muscles

* New England Soc. Translation 1876. etc.
"fibre is preserved, in most of them, in all their length but it becomes very faint and barely apparent. At the points where the transverse situation has disappeared are seen longitudinal markings. Sometimes when these become 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 characteristic of fatty degeneration of the muscles."

"(v) The hyperplasia of the interstitial connective tissue appears generally only in the second stage of the disease, it appeared to him to be preceded by a congested condition of the muscles which occasions also a slight increase in their volume. At this period (first stage) the transverse situation is often exceedingly faint."

While Wrsche consider it as the fundamental anatomical lesion is the hyperplasia of the interstitial connective tissue, but on the other hand looks upon the primary essential changes as taking place in the 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 proliferations of connective 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 by proliferation of their nuclei project, they swell up, taking more of a rounded form; splitting and division goes on; just as soon as there is an increase of proliferation of nuclei in the connective tissue. At an early period, however, muscles here the relationship, this process quickly extends more or less gradually gaining the upper hand. Finally, leading to complete disappearance of the muscle substance. Along with it goes on a great increase in the amount of connective tissue, with proliferation of the nuclei, blocking of the vessels and vessels. 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..."
has now wholly, or almost wholly, disappeared.

The final result of the whole process is one of two forms. It may be an atrophic, sclerotic lipomatosis, 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 increased.

To this, however, while thus describing the history of a diseased muscle, it 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 astroplasia, in contradistinction to the gross anatomical lesion of the cord which produces Progressive Spinal muscular Atrophy. Hence:

"The considerations which weigh with me are various. The muscles depend for their nutrition in a very large extent on trophic nerve centres; the localization 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. 1876, p. 262 et seq."
...centres in the central organ, occasionally we find a case of spinal amyotrophy presenting an almost exactly similar arrangement. Here itary influence is of great importance; mental aberrations are common among the patients. The new ones often occur in their families.

Further, even in the undoubted spinal cases, such as Hertz, Anterior Poliomyelitis, similar muscle changes (atrophy, proliferation of nuclei, division of fibres) both in the muscles and the connective tissue have been pointed out by W. Müller, Dégéine, Huet, Joffroy, Reich & Hitzig. In Reuter-hypertrophic cases malformation changes of a minor kind in the Spinal Cord have been met with. When I consider these facts, I begin in mind further the results of Newbner (an undoubted case) Dystrophy with extensive atrophy of the large cells of the anterior horns of the grey matter. "Frohman's observations" (a similar case but with less marked changes) cannot avoid me.

Rückermann


Archives de Physiologie 1888 p. 385.

Archives de Medicine experiment. 1882 p. 64.


Wagner Zeitschrift Ethik 1891.

The suspicion that after all the affection may be dependent on the nervous system. It is tempting to suppose, as formerly expressed it, that we have to do with a kind of trophic neuritis, having its origin in the trophic centres of the cord—a disturbance of the function of these centres which finds its expression in the very complicated muscle changes of the disease. While on this supposition these are, as a rule, no nerve 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 to be closer, intimate. The latter would represent an affection of the trophic centres that from the very first is a distinct course anatomical lesion taking effect in a degenerative atrophy of the muscles with fibrillar twitchings, resorption of degeneration, etc. The former would at the outset be merely a functional disturbance of these centres, conditioned probably by different causes, expressing itself as muscular dystrophy with all its characteristic symptoms. At the same time there would remain the possibility that even this merely functional
"disturbance might in the long run become associated with a course lesion of the centres. Many things about these afflictions would agree very well with such a supposition, among them the occasional occurrence of reaction of degeneration in dystrophic cases. The occasional initial localization of spinal amyotrophy in the "shoulder rotator. But, the whole question is by no means yet ready for decision. The future alone can lift the veil and reveal the finer processes that as yet lie hid from us."

This striking suggestion of Dr. Y supported and extended 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 localization of the lesions with those found in definitely spinal cases is, as Dr. Y confesses, only very occasionally seen, being absent in the great majority of cases.

2. 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, not at all. Thus the form
Cervical segment of the cord is stated to influence the functions of the deltoid, infra-spinal, and supra-spinatus, which we have seen are usually either hypertrophied or unchanged. In the first, the tarsal muscle of the thumb is most commonly affected, if the dislocation is practically never attacked. The tarsal muscle, though its arthritic division supplies the obicularis palpebrarum, which is very commonly attacked in the partial form. The ciliary muscle of the eyeball, which is never affected, we might adduce as an example.

So has been previously said, the changes found in the cord are so irregular as to be negligible from the point of view of causation. In advanced and distorted cases, these according to Exx would probably follow after a functional disturbance of the same region.

On the other hand points which may be inferred are:

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

The numerical prominence of spindle-cells...
served in several cases of diseased muscles in
section which corresponds with the appearance
which exists in Infantile Atrophy in the spinal
cases.

The weight of evidence at present is on its
favor, however, must lead us to the conclu-
sion that the disease originates in the mus-
cles themselves, owing to congenital defect in
their tissues. This view receives strong sup-
port from the coexistence in many cases of
similar developmental defects in the Nerv-
ous System.

That the changes originate in the muscles
fibres themselves rather than in the inter-
stitial 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 on any 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 Deltoides, in which the fibres
undergo simple atrophy without any inter-
stitial 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, Parascapulus Major especially, is associated with enlarged weak calf muscles is of greatest diagnostic importance in the pseudo-hypertrophic variety.

In the Locis-Scapulo-Humeral type, the peculiar affection of the muscles of the face, especially the occipitomandibularis in characteristic, though perhaps not actually diagnostic:

In the Scapulo-Humeral type, juvenile type, the marked wasting of the Shoulder, upper arm, Thigh in comparison with the well-developed forearm, still 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 joint characteristic method arising 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 condition 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 and its especial propensity to attack children. It appears 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 and 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 and 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 unretarded.

(i) Syringo-myelia attacks distal parts first. Shows no hypertrophy or Reaction. If degeneration is present sensory disturbances are prominent.

(ii) Chronic multiple Neuropathy in which there are pain to sensory disturbances = reaction of degeneration, characteristic localization of the paralysis.

(iii) Progressive Neuropathy or so-called 'Premonial' type, which though congenital in 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.

Rauchli claims to have cured the disease in its earliest stages by paradigmination combined with Hydrotherapy + Shampooing.

In most cases little or nothing can be done.

*New Edequium Societ. Tr. 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, as attempts should be made to protect him from all that tends to depress vitality. Associations 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 HORSEHORSE, ARENASM, JODIDE OF POTASSIUM, NUX VOMICA, etc. Thymus gland but all with equal success. Electric currents of both kinds have been tried with about as much good result. So it thinks that electricity 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 Sydenham Society 1894*
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 and condition of the particular muscles existing in the particular muscles being exercised. In this way the muscles fibres which as yet show no tendency to degeneration may be stimulated to further growth and thereby attain greater power. It at the same time counteracts the tendency to contraction which exists in the affected muscles. Where strong contraction especially in the calves muscles exist, 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 case of one 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 cataract or other intercurrent affections, as they, especially if of a Pulmonary character, usually hasten the fatal termination of the Disease.