Thesis
for the degree of Doctor of Medicine
by
William Done, M.B., C.M.
Carlisle

Pseudo-hypertrophic Muscular Paralysis
Pseudo-hypertrophic Muscular Paralysis

In choosing Pseudo-hypertrophic Muscular Paralysis as the subject matter of my thesis for the degree of Doctor of Medicine, I have been influenced by the fact, that I am in possession of notes of four cases of this disease, which have been under my personal observation.

The first case was under my care, when clinical clerk in Professor Sanders' wards; the second and third, when resident physician with Dr. Brakenridge; and the last occurred in private practice in Berwick.

Post-mortem examinations were obtained in the first and fourth case; the former dying at a far advanced stage of the disease, the latter at a comparatively early stage.

I propose, in the first instance to give an account of these cases; and then to consider the disease itself under various headings.
Case I.

Walter Taylor, age 10 years, residing at Burnley, admitted into the Edinburgh Royal Infirmary, under the care of Professor Sanders, Dec. 5th, 1879.

History:—(reported by his father)

(X) Of present illness:—When the patient was a baby, he was fat and plump. He began to walk when sixteen months old, but was thought never to be as strong on his legs as he should have been, although he used to run about with the other children. He had more falls than the other children. He used to waddle in his walk, that is, he threw his weight first on one leg and then on the other. It was always remarked what a good calf he had.

When he was about five years old, his arms were thought not to be so firm as they ought to be; and about the same time his thighs were likewise noticed to be soft. As he grew taller, he seemed
to have more difficulty in rising after a fall. Little was thought of his condition before he was six years old, his parents thinking that as he got older, he would get all right. During the last three years, he has got gradually weaker. During the last year, his condition has got much worse. No cause can be assigned for his condition.

(6) Of previous health: - His general health has always been good. He had inflammation of the lungs when a year old.

(5) Of social surroundings: - He has always been in good circumstances.

(8) Of family: - His father and mother are both very healthy. He is the third child. He has one brother and three sisters living, all of whom are quite strong and healthy. A brother died of inflammation of the lungs when fourteen months old.
Present Condition:
Height 3 ft 9½ in. Face flabby. Body emaciated. Hairuality poor. Temperature varies from 98°F to 99.8°F.

Nervous and Locomotive System:
Motor functions: — When in bed, he usually lies with his legs curled up. He lies on his back, but his legs are turned on their sides, the left being the lower. The thighs are flexed on the abdomen, and the legs on the thighs, or the feet extended at the ankle joints, in a condition familiar to Habits Squirms. When asked to extend his leg, he works them slowly down the bed by the toes and heel alternately. Sometimes he pushes with his hands placed above the knees. When asked to sit up in bed, he cannot do so, unless he grasps the bed clothes, and help himself up with his arms.
The movement at the hip-joint can be performed both passively and actively, but can be very easily resisted. In like manner, he can flex his knee. When he sits over the edge of the bed with his knee flexed, he cannot extend them beyond seven or eight inches; and if the knees be passively extended, he cannot keep them so by muscular effort.

Both feet are in a condition of Valgus Equino-varum. In both, the tendon Achilles is markedly tense, the tendons of the Tibialis, muscles slightly so, and the Peronei also slightly tense from the position of the feet. Extension at the ankle joint is good, dorsiflexion not beyond a right angle; inversion evasion of the foot normal.

The movements in the upper extremity can be performed; but are
easily resisted. The fine movements of the hand are very good; he can hold a pen, and write fairly well.

When asked to walk, he has to support himself against some object before he can attempt it. He can walk without assistance, and walks about a good deal during the day. In walking, he throws his belly forward, and his shoulders back, thus arching the vertebral column in the lumbar region. His arching of the spine disappears when he is sitting, there is then a slight projection of the spine backward in the lumbo-sacral region.

He tends to walk on his toes, and lifts his feet high off the ground. He throws his whole body weight on one foot, and then moves the other leg forward by a movement of circumduction, keeping his toes turned in and the foot extended.
Having thus brought this foot in front of and almost crossing the other, he throws the weight of the body on that leg. When walking, he sometimes falls. She is unable to rise without assistance. He rising off the ground he requires the aid of a chair or such like. He first places his hand on the chair, then gradually works himself up on his knees. He next manages to put one foot flat on the ground, but can do no more without a chair, he cannot move off his hand knees.

Patellar Tendon Reflex is absent.

The sternal portion of the pectoral muscle, the latissimus dorsi are small in bulk. The deltoid is of fair volume. The scapular muscles are good. The muscles of the upper and forearm are small in bulk, particularly the biceps. The intrinsic muscles of the hand are good. Trapezius, sternum must and, trapezius, extensor
Sphincter are all good.
In the thigh, the muscles are good on the posterior surface, poor on the anterior. In the calf, the muscles are apparently increased in size. They have an indurated, firm feeling. Muscles of thorax & abdomen are good.

Measurements: -

- Walter Taylor - Boy of 10 -

Upper arm (above elbow) 5 7/8 in 7 in
Lower arm (2 cm below) 5 3/4 " 7 1/2 "
Thigh (2 cm. above knee) 10 1/4 " 13 1/2 "
Calf (thickest part) 9 3/4 " 9 "

All the muscles responded to the Faradic current.

There were no other points of interest in the case. His sensibilities were normal. The other systems of the body were normal.
Further History:—

The boy left Edinburgh in May or June 1880 and came under the care of Dr John Brown of Burnley.

In his country, I owe a reference to this case in the British Medical Journal (Feb 3: 1883).

In the autumn of 1880, he was quite unable to stand without support, or on an attempt being made to place him in the erect posture, his legs doubled up under him in a perfectly helpless manner.

The boy was subsequently admitted to Manchester Infirmary, under the care of Dr Bech, but no new symptom appeared during his residence there.

On his return home, he gradually became more and more helpless; but was able, almost to the last, to sit on a chair, the body being supported between the back of the chair and the table.
His face became plumper, the temporal and masticatory muscles being hypertrophied.

On April 6th, 1882, he was seized with a severe attack of vomiting and diarrhoea, with great prostration of strength. He died on April 9th.

Post-mortem Examination:—
This was conducted by Dr. Brown, and Dr. James Ross of Manchester, thirty-two hours after death. I quote Dr. Ross's description from the British Medical Journal (Feb 13: 1883).

Post-mortem rigidity was well marked, even the wasted muscle of the upper extremities presented some degree of rigidity.

The most prominent part of the calves of the legs measured, each, 9 1/4 inches; the middle of the thighs, each, 10 1/4 inches; the middle of the upper arms, each, 5 3/4 inches; and the thickest part of the fore-arms, each, 5 3/4 inches.
The subcutaneous fat was ½ inch thick over the calves of the legs, and ¾ inch over the gluteal region, while there was very little subcutaneous fat in the lumbar and dorsal regions, but it was more abundant in the back of the neck. The gluteal muscles were of a pale yellow hue, with a slight pink tinge. The conversion of these muscles into fat was so complete, that almost every appearance of muscular structure was lost. The gastrocnemius had more of the pink tinge than the gluteal muscles, and also presented more of the appearance of muscular structure. The erectors spinae muscle were much wasted, and of a pale colour, but they were by no means so much changed in appearance from healthy muscles as either the gluteal muscles, or the gastrocnemius. The scapular muscles were considerably altered, the supra-
spinalis being almost as much changed as the gluteal muscles. The muscles of the back of the neck were wasted, but presented almost the normal colour and texture of muscles. The latissimi dorsi were thin and pale, and more like fibrous membranes than true muscle.

The brain and spinal cord did not present any abnormal appearances to the naked eye.

The lungs were healthy. The heart was soft and flabby, and its cavities somewhat dilated. The walls of the left ventricle were of a pale yellow colour, and friable.

Portions of the diand muscle, of the sciatic nerve, and the first cord of the brachial, along with the spinal cord, were preserved for microscopical examination.

Every muscle of the body, even those
which had appeared almost normal to the naked eye, had undergone extensive changes. In the muscle most changed, like the pleurae, scarcely anything but fat cells and bundles of a wavy fibrous tissue could be discovered. In these muscles, less altered like the Erectores Spinae, the fat cells were much less abundant, but the muscular fibres were separated from one another by an interstitial connective tissue, consisting of parallel fibres, in the midst of which numerous elongated nuclei and cells were imbedded. The muscular fibres themselves were greatly altered; they were, as a rule, atrophied, some of them being greatly reduced in size. The nuclei of the sarcolamina were much increased in number, but the transverse striae remained well marked, even in fibres otherwise much altered. The fibres
themselves did not appear to have undergone fatty degeneration. A large number of the fibres of the cardiac muscles were atrophied, and were in many places widely separated by interstitial connective tissue.

Numerous sections of the spinal cord were made by W. R. H. Young, Pathological Registrar to the Manchester Infirmary, but no evidence of disease could be detected in them. No change could be detected in either the sciatic nerve or the first cord of the brachial plexus.
Case II

John Reid, age 10, a schoolboy, residing at Whitburn, admitted into Dr. Braemridge's Ward in the Edinburgh Royal Infirmary, August 13th, 1881.

Complains of having had a difficulty in walking for some time previous to his admission.

History. (a) of present attack: -
The patient commenced to walk at ten months, but has been always rather weak on his legs. This weakness gradually grew greater of greater. His parents have long noticed, that he fell often more than other children, and that he could not rise easily from the floor. They can trace the complaint to no reasonable cause.

(b) of previous health: - This has been good. He has had measles and scarlet fever.
(c) of family: — Father or mother are both healthy. The patient is the eldest of five children, the remainder of whom are healthy. No history of nervous disease in the family.

(d) of social circumstances: — He has a comfortable home.

Present Condition. The patient is a sturdy looking boy of the usual height and weight. There is no emaciation.

There is some blotching of the skin on the leg and buttocks.

Nervous & Locomotory System: — The patient feels pain after walking in both popliteal space, & in the shanks. Sensory phenomena are normal. Skin and organic reflexes are normal. Knee tendon reflex is absent in both limbs.
The patient lies in bed with his limbs extended. He can easily draw them up, and to resist this action, a certain degree of force is required; less force in resisting the pushing down of the legs. The left leg is not so strong as the right. His arms are fairly strong. He has perfect command over them. When asked to sit up in bed, he does so without much difficulty. When sitting, with his legs hanging, he can extend them in a straight line with his thighs; and if put in that position, he retains it without much apparent effort. Extension at the ankle joint is also easy. If, when standing, the patient be asked to sit down, he slowly goes on one knee, rests a hand on it, and lets himself down. If now he be asked to get up, he turns himself round, rests one knee
and one hand on the floor, and with
the other hand, he raises himself
by some piece of furniture.
To pick up an object from the
floor, he bends his body forwards,
puts one hand on the corresponding
knee, and with the other hand
slowly takes the object, and then by
an effort of the first hand he
pushes himself up.

In walking, the shoulders are thrown
back, the abdomen forwards, thus
increasing the hollow of the back.
The foot is from side to side in a
sort of rocking or shuffling manner.
The legs are separated, the foot is
lifted rather high from the floor, and
the heel is raised first from the ground,
the toes being in contact with the
ground for a more long treading period
than usual. Then, the leg is moving
forward from the hip joint, so the
foot comes nearly flat on to the ground.
When he is sitting, there is no hollow in the back.

The pectoral, latissimi dorsi, and deltoid muscles are well developed.

The muscles of the upper arm, particularly the biceps, are weak and flabby. The muscles of the forearm are poor. The sternomastoids are thin. The masseters are rather strong.

The erector spinae seems good.

The muscles of the thigh are bulky. The buttock is large. The calves are abnormally large.

The following measurements were taken:

Upper arm: \(7 \text{ inches} \) = \(9 \text{ inches in full} \) (2 in above elbow)

Fore arm: \(9 \frac{3}{4} \text{ inches} \) = \(9 \frac{3}{4} \text{ inches} \) (2 in below elbow)

Thigh: \(14 \frac{3}{4} \text{ in} = 10 \frac{1}{4} \text{ in} \) (4 in below A.S. spine)

Calf: \(11 \frac{1}{4} \text{ in} = 9 \text{ in} \) (thickest part)
Coordination is perfect.
The muscles all responded to the faradic current.
The cerebral & mental functions seem normal.
The other systems are normal.
The patient left the hospital on Nov 16th in the above condition.

I saw the patient two years after the above report was written. The disease was then in a much more advanced state. He was almost confined to bed. He was unable to walk without support.
The further history of this case is unknown.
Case III.

John Jackson, age 5½ years, admitted to the Edinburgh Royal Infirmary Feb 25: 1831. Duration of illness is 3½ years.

He is the eldest of three children. There is no patent in both of his parents' families. He has been brought up in good circumstances, and has had no previous illnesses.

The patient commenced to walk at fourteen months, and learned as rapidly as other children, and at this time was very stout and strong for his age.

At eighteen months, he had a fall, striking himself on his forehead and causing an ecchymosis, which suppurred and had to be opened.

After this fall, he became weaker, and his mother specially noticed that when he fell, he did not attempt to save himself with his arms as other children do. She thinks, that at this time his arms were weaker.
than his legs. Juxta, varis of the right foot has come on gradually, being first noticed nearly three years ago.

Sixth on admission into Edinburgh Infirmary:—

His is a bright, intelligent boy, fair complexion. Muscularity is poorly developed. On the forehead, to the left of midline, is a scar, produced by a blow, when he was eighteen months old.

Locomotory System:—

In standing, the patient's back has all the curvatures increased, the lumbar curve being particularly exaggerated. A perpendicular line, dropped from the shoulders, falls behind the gluteal region. The knees are closely approximated, and the feet wide apart. Without his boots, he has difficulty in maintaining the erect posture.

On sitting up, the lumbar curve
entirely disappears, and the back becomes straight.

The muscular development is poor throughout the body generally. In the arms and thighs, the muscles are small, soft, and flabby. In the legs, the gastrocnemius do not feel abnormally firm, in fact are softer than in a healthy child of the same age.

The quadriceps, extensor femoris of each leg seem somewhat enlarged. The spinal muscles in the lumbar region are large firm in proportion to the other muscles of the body.

Measurements:

1. Circumference of middle of upper arm:
   - John Jackson: \[ R = 5 \text{ inches} \]
   - Healthy Child: \[ R = 6 \frac{3}{4} \text{ inches} \]

11. Circumference of middle of forearm:
   - J. J.: \[ R = 5\frac{1}{2} \text{ inches} \]
   - H. C.: \[ R = 6\frac{1}{2} \text{ inches} \]
III. Circumference of thigh 6 in below A.S. spine
   J. J. \{ R \} 9 1/2 inches.
   H. C. \{ L \} 11 3/4 inches.

IV. Circumference of leg 2 1/2 in below head of tibia
   J. J. \{ R \} 7 inches.
   H. C. \{ L \} 12 inches.

There is well marked Talipes Equinum in both feet, with Talipes Varus in the right foot.

In walking, the legs are somewhat straddled, and at each step, the shoulders are moving to the side of the leg upon which he is supporting himself.

When laid on his back, and asked to sit up, he is unable to do so without rolling round, setting on his hand knees, with his hands still on the floor, he draws up first one leg, & then the other, until he is supported on his hands and feet.
the feet being wide apart. One knee is now bent, and the hand of the same side grasps the thigh just above the knee; and by a lean to the other side, the body is raised, and the opposite thigh is grasped by the corresponding hand. He now straightens himself by "climbing up his thighs," and finally by a great effort, he assumes the erect posture.

When asked to pick an object from the floor, he walks up very close to it, and then, bending his knees considerably, he makes a swoop to one side, picks up the object, and catching the thighs, he straightens himself as above described.

The Patellar tendon reflex is absent. There is no other point of interest in the case.

The patient's history of this case is unknown.
Case IV.

John Hodgson, age 13 years, schoolboy, born in Carlisle, residing at Bellevue, near Carlisle, first seen Oct. 28, 1854.

He complains of pain in the calves of his legs, of weariness in walking, of taking a fall when walking. Duration of symptoms in three years.

History: — (X) of family: — His father is a strong healthy man. Two paternal aunts have died of phthisis. His mother & her female relations have a first tendency to obesity. His maternal grandmother died insane.

(Note. Since the patient's death, two of his younger members of the family have come under my observation. A boy, age 7 years, suffered from pneumonia with a high temperature, became deeply comatose, remained so for about thirty hours, but ultimately recovered. A girl, age 3½ years, had on two occasions, curious
phenomenon, which disappeared on the passage of a round worm. On the first occasion, they resembled in a great degree tubercular headache. On the second occasion, the muscles of the left calf and left thigh were in a condition of spastic contraction. She had a waddling gait and rose off the ground in a very similar manner to a case of Duchenne's paralysis.

(6). Of previous health or: - no previous illnesses. Good nursing.

Present Condition:

He is a well nourished boy.

When he stands, the shoulders are thrown back, there is a hollow in the lumbar region, the belly is thrown forward. ThUMB LINE FROM HECK FALLS BEHIND THE BUTTOCKS. In rising from the ground, he begins himself up by placing his hand on his knees; but can, if asked to
rise without doing so, rise by a great effort.
The thigh measured 13 1/2 inches in circumference 4 inches below the great trochanter.
The right calf measured 11 1/2 inches, the left 11 1/2 inches at their greatest circumference.
The calf muscles are larger in proportion to the other muscles in the body, I have a hard fibrous feel even when relaxed.
The patellar tendon reflex is slight.
All the muscles react to true Faradic current.
The walk is waddling. He inclines to walk on his toes. Both the tendo Achilles are tense. The feet are lifted high off the ground.
The other systems seemed normal.

Treatment: — Faradic current to the muscles three times a week. Glistening oil.
Toward the end of December 1884, he took Enteric Fever, from which he died January 8th, 1885.

Post-mortem Examination:—

The intestine showed unusual lesions found in Enteric Fever. There was considerable engorgement of the lung; with blood. The spinal cord, portions of the calf muscles were removed. The muscles showed microscopically a great increase of connective tissue between the bundles of muscular fibres. The spinal cord was sent to Dr. Brew of London, who found no abnormal condition in it.
Pseudo-hypertrophic Muscular Paralysis

Historical. This disease was first clearly described by Duchenne of Boulogne, in 1861. Previous to this, cases, now recognised as examples of the disease, had been described.

Sir Charles Bell in 1830, Partridge in 1847, Meryon in 1852, and Oppenheim in 1855, all described cases; the last two observers being of opinion that they were identical with Profenius muscular atrophy. Powers wrote an able monograph on the subject in 1879.

At the present time, the disease is well recognised, considerably over two hundred cases have been described, and an account of it is now given in every text-book of medicine.

Nomenclature. Duchenne, at first, named the disease, "Paraplégie hyper-
trophique de l'enfance de cause cérébrale." Subsequently, he gave it a double name, "Paralytic pseudo-
hypertrrophic or myo-sclerotic."
Jaccoud styled it, "Sclérose musculaire propulsive"; Foster, "Paralytic with apparent muscular hypertrophy";
and Seidel, "Atrophia muscularum lipomatosae.
A convenient, although in some ways objectionable term is "Duchenne's Paralysis."

Definition: It is a disease of early life; characterized by loss of power in certain muscles, all of which undergo atrophy, some an apparent hypertrophy; and tending to a fatal termination.

Etiology. The disease is very much more common in males than in females. Foster in 1886 gives the proportion as 7 males to 1 female
in all the published cases; and in his own case, as 33 to 10.
Further, in females, the symptoms in many cases are slight, and the disease advances slowly.
It is a disease of early life. In one-third of the cases, the first symptoms have appeared when the child is beginning to walk. In more than three-fourths of the cases, symptoms have been noticed before the age of ten.
Poore has described some cases where the symptoms were noticed later, at the ages of 24, 26, 28, 37, and 40 respectively.
It tends to affect several members of the same family. Thus in 220 cases, 102 were isolated, 118 were in 39 families. Thus Gowen describes cases where six were affected in one family, four brothers and a maternal uncle and aunt.
The antecedent cases have been alway
traced in the mother's family.

Sex itself has little influence in the grouping. By way of example, the following groups may be mentioned, being taken from Tower's monograph: three brothers and uncle; two brothers, sister, and two cousins (brother); four brothers, uncle, and aunt.

Nothing is known as to the direct cause of the disease. Syphilis, intemperance, consanguinity, and heredity, diseases have nothing to do with it. It is as common, or perhaps more common, among the better classes as among the poor.

Symptoms. The onset of the disease is very gradual; the child is weak in the leg or falls oftener than other children. At the same time, it is often remarked what a well-developed calf the child has.

The child, by and by, begins to waddle
in its walk, to stand with the legs far apart, and to have difficulty in
rising from the ground.

The symptoms may advance until the
child becomes almost absolutely helpless,
unable even to sit on a chair
without support, as in the first
case cited.

The symptoms are all due to the
change, which takes place in the
muscles, which are sometimes
enlarged, sometimes diminished
in size. Some muscles are un-
affected. The impairment of
power is irrespective, to some extent,
of the change in the size of the
muscle.

The enlargement of muscles is com-
monly confined to a few, although
in some cases, a very large number
of the muscles have been affected
Sacheville describes one case, in
which every muscle of the body
was enlarged, & compare the boy to the "Faunus Hercules."
The muscles of the calf are enlarged in nearly every case; & have also, in common with all the enlarged muscles, a hard, fibrous feeling. In Case III quoted, the calf muscle, were much diminished in size & quite soft. In the other three cases, they were enlarged, had a firm, fibrous feeling. The infraspinatus is the muscle, kept most commonly affected. After them, come the Extensor of the Knee, the Glutei, the Lumbar Muscle, the Deltoid.
The Triceps Biceps are sometimes enlarged, but more frequently diminished in size.
Latissimi Dorsi and the sternal portions of the Pectorals are often wasted, the former muscle being sometimes absent, or merely represented by a fibrous membrane.
In Jaye's junctus, the muscles of the joint of the leg, the flexors of the knee, the muscle of the forearm, the muscles of mastication, and the tongue are not rarely affected. The heart is occasionally affected. It is probable that the intrinsic muscle of the hand, and the facial muscle are never affected. Hammond describes a case in which the left side of the face was hypertrophied but this was probably due to an enlargement of the muscle of mastication of that side. I believe that a case, in which all the facial muscles were reported to be hypertrophied, is described by Dr. G. S. Gerhard in the Philadelphia Medical Times, Oct 16 1875.

As a result of these changes in the muscle, there is weakness in their action. There is in various degrees in the lower extremities, the weakness
is greatest in the flexors of the hip; but as a rule, in advanced cases, all the groups of muscles become affected. In the upper extremity, the depressors of the forearm are mostly affected, that is the latissimi dorsi and the external pectorals.

This muscular weakness is manifested in various ways. Equilibrium is maintained with difficulty; therefore the patient straddles when he stands in order to enlarge the base of support. In walking, the gait is waddling, so as to bring the centre of gravity over the foot which is on the ground. Duchenne has shown that this is due to the weakness of the Glutæus head, which, in health, supports the body when supported on one leg, or rotates the pelvis alternately on each limb in the act of walking. When weak, the body has to be thrown further over to either side, for the
As was above stated,

the manner in which patients go up
stairs, and rise from the floor or
a chair, are very characteristic.

The various movements have been de-
scribed in the cases quoted, and
the photographs below show a boy
rising from the floor.
In rising from the floor, the patient places his hands on his knees, or in some cases "climbs up the thigh" at the end of the act, in order to assist the extension of the knee joint, and hip joint respectively. By so doing, the weight of the trunk is transferred from the hips to the knee, and thus a lever of the third order becomes one of the second.

We find various distortions. Thus, Telafer Equinus is exceedingly common, and is due to the primary contraction of the calf muscles; a contraction which occurs both in the length and breadth of the muscles. Fixation of the knee joint and elbow are more rarely met with. Spinal curvature, antero-posterior with the concavity backward, is very commonly met with. If exists, when the patient is in the erect posture,
but disappears when he sits down. The shoulders are usually thrown far backward, so that a plumb line from them falls well behind the buttocks. Duchenne attributed this to weakness in the Erector Spinae. Powers is of opinion that it is due to the weakness of the hip extensors.
so that the pelvis and lower lumbar vertebrae incline forwards; a compensatory backward inclination of the dorsal spine being necessary to preserve the centre of gravity.

The patellar tendon reflex is at first normal, then diminished, and finally disappears, according to the loss of power in the knee extensors.

The electric irritability of the muscles is unaffected at first, and then becomes lowered to both forms of electricity. There is no reaction of degeneration.

There are no other nervous disturbances. Some rare symptoms may be mentioned, but are of small importance.

Blistering of the skin; rise of temperature in the calves; difficulty in passing urine; mental dullness; epileptic fits, are among such.
so that the pelvis and lower lumbar vertebrae incline forward; a compensatory backward inclination of the dorsal spine being necessary to preserve the centre of gravity.

The patellar tendon reflex is at first normal, then diminished, and finally disappears, according to the loss of power in the knee extensors.

The electric irritability of the muscles is unaffected at first, and then becomes lowered to both forms of electricity. There is no reaction of degeneration.

There are no other nervous disturbances.

Some rare symptoms may be mentioned, but are of small importance. blotting of the skin; rise of temperature in the calves; difficulty in passing urine; mental dullness; epileptic fits, are among such.
Progress. According to Hammond, the average duration is from six to six years. Death is usually from some intercurrent affection, most commonly some lung affection.

Diagnosis. This is easy. The paralysis with enlargement of the calf muscles, the peculiar gait, the "hollow back", the mode of rising from the ground, are all very characteristic. Powers lays much stress on the enlarged infraspinatus, and the wasted biceps minor & sternopectoralis.

Prognosis. This is most favourable. Duchenne claimed recovery in two cases, but the subsequent progress was not ascertained. Some improvement has been recorded in early cases from the use of faradism. The earlier the development, the more rapid is the course.
Treatment. - Treatment is of little avail. The general health must be attended to, and cold exposure avoided. The nutrition of the affected muscle must be maintained as well as possible by systematic muscular exercises, paradox, kneading, touch, etc. Iron, Phosphorus, strychnia, and cod liver oil have been recommended for internal administration.

Pathology. In the muscle, there is a growth of connective tissue surrounding the muscular fibres. There are also fat cells between the fibres. The muscular fibres are more or less atrophied. The motor nerves are unaffected. The spinal cord in some cases has been found unaffected; in others slight and irregular change have been described.
No changes have been found in the anterior cornea of the spinal cord. A considerable number of cases have been examined in which no change in the spinal cord could be observed. In the two cases cited in which post-mortem examinations were made by competent observers, no changes could be detected.

As negative results are more important than the discovery of changes in the cord, the production of which may be secondary or even in some of the cases post-mortem, and the significance of which is difficult to explain, it is fair to conclude that the disease is not spinal in its origin.

In all probability, the disease is one primarily of the muscles, what Rivers calls a “perverted tendency of development inherent in the seminal tissue of the muscular system.”
Reference to works consulted.


Duchenne "Collected Works"

Lydecker Society

Articles in Bramwell's Spinal Cord.

Gowers, Newcom Disease Vol I; Ron's

Newcom Disease Vol II; Hammond's

Newcom Diseases.