

Thesis  
for the degree of Doctor of Medicine  
by  
William Donie; 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's ward; the second and third, when resident physician with Dr Brakenridge; and the last occurred in private practice in Carlisle.

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

I purpose, 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 5<sup>th</sup> 1879.

History: - (reported by his father)

(X) of present illness: - When the patient was a baby, he was fat & 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 & 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; & about the same time his thighs were likewise noted 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.

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

(c) of social surroundings: - He has always been in good circumstances.

(d) of family: - His father & mother are both very healthy. He is the third child. He has one brother & three sisters living, all of whom are quite strong & healthy. A brother died of inflammation of the lungs when fourteen months old.

Present Condition:

Height 3 ft 9½ in. Face plump. Body emaciated. Muscularity poor.

Temperature varies from 98.7° F to 99.8° F.

Nervous and Locomotory Systems: -

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, & the feet extended at the ankle joints in a condition similar to Talipes Equinus. When asked to extend his legs, he works them slowly down the bed by the toes & heels 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 helps himself up with his arms.

The movements at the hip-joints 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 knees 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 *Dalipes Equino-varus*. In both, the *tendo. Achillis* 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; Dorsi flexion not beyond a right angle, Inversion Eversion of the foot normal.

The movements in the upper ex-

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.

This arching of the spine disappears when he is sitting, there is then a slight projection of the spine backward in the lumbosacral 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, he is unable to rise without assistance. In 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 hands & 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, Sternocleidomastoid, Masseter, Erector



Photographs of Walter Taylor

Spines 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, & have an indurated, firm feeling. Muscles of thorax & abdomen are good.

Measurements: -

	- Walter Taylor -	Boy of 10 -
Upper arm (2 inches above olecranon)	5 <sup>5</sup> / <sub>8</sub> in	7 in
Lower arm (2 in. below olecranon)	5 <sup>3</sup> / <sub>4</sub> "	7 <sup>1</sup> / <sub>2</sub> "
Thigh (4 in below a. s. spine)	10 <sup>1</sup> / <sub>4</sub> "	13 <sup>1</sup> / <sub>2</sub> "
Calf. (thickest part)	9 <sup>3</sup> / <sub>4</sub> "	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.

To his courtesy, 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, & 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 Leech, 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 propped up between the back of the chair and the table.

His face became plumper, the temporal and masticatory muscles being hypertrophied.

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

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 3: 1883).

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

The most prominent part of the calves of the legs measured, each,  $9\frac{1}{4}$  inches; the middle of the thighs, each,  $10\frac{1}{4}$  inches; the middle of the upper arms, each,  $5\frac{3}{4}$  inches; and the thickest part of the fore-arms, each,  $5\frac{3}{4}$  in.

The subcutaneous fat was  $\frac{1}{2}$  inch thick over the calves of the legs, and  $\frac{3}{4}$  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 gastrocnemii had more of the pink tinge than the gluteal muscles, and also presented more of the appearance of muscular structure. The erectores spinae muscles 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 gastrocnemii. The scapular muscles were considerably altered, the supra-

spinati 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 muscles.

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 diaphragm muscles, 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 muscles most changed, like the gluteals, scarcely anything but fat cells and bundles of a wavy fibrous tissue could be discovered. In those muscles, less altered like the erectors 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 sarcolemma were much increased in number, but the transverse striation 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. A. H. Young, Pathological Registrar to the Manchester Infirmary, but no evidence of disease could be detected in them.

No changes 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 St Brakenridge's Ward in the Edinburgh Royal Infirmary, August 13<sup>th</sup> 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 & greater. His parents have long noticed, that he fell oftener 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 & 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 mottling of the skin on the leg and buttocks.

Nervous & Locomotory

Systems: - The

patient feels pain after walking in both popliteal spaces & in the hamstrings. 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 gait is from side to side in a sort of rocking & 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 lengthened period than usual. Then, the leg is swung forward from the hip joint, & 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 poor and flabby. The muscles of the fore-arm are poor. The sterno-mastoids 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 inches = 9 inches in full  
(2 in above elbow) from boy of ten.

Fore Arm =  $9\frac{1}{4}$  inches =  $9\frac{1}{4}$  inches  
(2 in below elbow)

Thigh =  $14\frac{1}{4}$  in =  $10\frac{1}{4}$  in  
(4 in below <sup>of ilium</sup> A.S. spine)

Calf =  $11\frac{1}{4}$  in = 9 in.  
(thickest part)

Co-ordination 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 18<sup>th</sup> 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  $5\frac{1}{2}$  years, admitted  
to the Edinburgh Royal Infirmary Feb 25: 1881.

Duration of illness is  $3\frac{1}{2}$  years.

He is the eldest of three children. There is  
phthisis in both of his parents' families.

He has been brought up in good circumstances  
or has had no previous illnesses.

The patient commenced to walk at four-  
teen 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 suppurated  
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. Talipes varus of the right foot has come on gradually, being first noticed nearly three years ago.

State on admission into Edinburgh Infirmary: -  
He is a bright, intelligent boy, fair complexion. Muscularity is poorly developed. On the forehead, to the left of middle line, is a scar, produced by a blow, when he was eighteen months old.

Locomotor 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 gastrocnemii 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  $\begin{cases} R & \sim 5 \text{ inches} \\ L & \sim 5 \text{ inches} \end{cases}$

Healthy Child.  $\begin{cases} R & \sim 6 \frac{3}{4} \text{ inches} \\ L & \sim 6 \frac{3}{4} \text{ inches} \end{cases}$

11. Circumference of middle of fore-arm: -

J. J.  $\begin{cases} R & \sim 5 \frac{1}{2} \text{ inches} \\ L & \sim 5 \frac{1}{2} \text{ inches} \end{cases}$

H. C.  $\begin{cases} R & \sim 6 \frac{1}{2} \text{ inches} \\ L & \sim 6 \frac{1}{2} \text{ inches} \end{cases}$

III. Circumference of thigh 6 in below A.S. space

J. J.  $\begin{cases} R & 9\frac{1}{2} \text{ inches} \\ L & 9 \text{ inches} \end{cases}$

H. C.  $\begin{cases} R & > 11\frac{3}{4} \text{ inches} \\ L & > \end{cases}$

IV. Circumference of leg  $2\frac{1}{2}$  in below head of fibula

J. J.  $\begin{cases} R & > 7 \text{ inches} \\ L & > \end{cases}$

H. C.  $\begin{cases} R & > 12 \text{ inches.} \\ L & > \end{cases}$

There is well marked Talipes Equinus 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, & getting on his hands & 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 noop 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 further history of this case is unknown.

#### Case IV.

John Hodgson, age 13 years, schoolboy,  
born in Carlisle, residing at Bellevue,  
near Carlisle, first seen October 28, 1884

He complains of pain in the calves of his  
legs, of weariness in walking, &  
he sometimes falls when walking.

Duration of symptoms is 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 great tendency to obesity. His  
maternal grandmother died insane.

(note. Since the patient's death, two  
of the 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  $3\frac{1}{2}$  years,  
had on two occasions, curious

phenomena, which disappeared on the passage of a round worm. On the first occasion, these resembled in a great degree Tubercular meningitis. On the second occasion, the muscles of the left calf & left thigh were in a condition of spastic contraction. She had a waddling gait & rose off the ground in a very similiar manner to a case of Duchenne's paralysis).

(b). of previous health etc; -  
no previous illnesses. Good surroundings.

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 forwards. Plumb line from neck falls behind the buttocks.

In rising from the ground, he pushes himself up by placing his hands on his knees; but can, if asked to

rise without doing so, rise by a great effort.

The thigh measured  $13\frac{1}{2}$  inches in circumference 4 inches below the great trochanter.

The right calf measured  $11\frac{5}{8}$  inches, the left  $11\frac{1}{2}$  inches at their greatest circumference.

The calf muscles are larger in proportion to the other muscles in the body, & have a hard fibrous feel even when relaxed.

The patellar tendon reflex is slight.

All the muscles react to the 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. Cod Liver Oil.

Towards the end of December 1884,  
he took Enteric Fever, from which  
he died January 8<sup>th</sup> 1885.

Post-mortem Examination: -

The intestine showed usual lesions  
found in Enteric Fever. There was  
considerable engorgement of the lumen  
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 Gowen 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 Progressive muscular Atrophy.

Gowers 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  
cerebrale". Subsequently, he gave it  
a double name, "Paralyse pseudo-  
hypertrophique ou myo-sclérotique".  
Jaccoud styled it, "Sclérose musculaire  
progressive"; Foster, "Paralysis with  
apparent muscular hypertrophy";  
and Seidel, "Atrophia musculorum  
lipomatosa. 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. Gowers in 1886, gives the  
proportion as 7 males to 1 female

in all the published cases; and in his own cases 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 always

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 Gowen's monograph; three brothers and uncle; two brothers, sister, and two cousins (brothers); four brothers, uncle, and aunt.

Nothing is known as to the direct cause of the disease. Syphilis, Intemperance, Consanguinity, and nervous 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 legs & falls oftener than other children. At the same time, it is often remarked what a well developed calf the child has. The child, by & 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 props, 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 unaffected. The impairment of power is irrespective, to some extent, of the change in the size of the muscle.

The enlargement of muscles is commonly confined to a few, although in some cases, a very large number of the muscles have been affected. Duchenne describes one case, in which every muscle of the body

was enlarged, & compares the boy to the Farnese 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 muscles were much diminished in size & quite soft. In the other three cases, they were enlarged, & had a firm, fibrous feeling.

The *infra-spinatus* is the muscle, next most commonly affected.

After them, come the Extensors of the Knee, the *Eltai*, the Lumbar muscles, the Deltoid.

The Triceps Biceps are sometimes enlarged, but more frequently diminished in size.

*Latissimi Dorsi* and the sternal portions of the Pectorales are often wasted, the former muscle being sometimes absent or merely represented by a fibrous membrane.

The Trapezius, the muscles of the front of the leg, the Flexors of the knee, the muscles of the fore-arm, the muscles of mastication, and the Tongue are more rarely affected. The heart is occasionally affected.

It is probable that the intrinsic muscles of the hand, and the facial muscles 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 muscles 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 muscles, we have weakness in their action, shown 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 extremities, the depressors of the arm are mostly affected, that is the latissimi dorsi and the sternal pectorales.

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 Gluteus medius, which, in health, supports the body when supported on one leg, & 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

Reason above stated.

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

The various movements have been described 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, & in some cases "climbs up the thighs," at the end of the act, in order to assist the extension of the knee joint, and hip joint respectively.

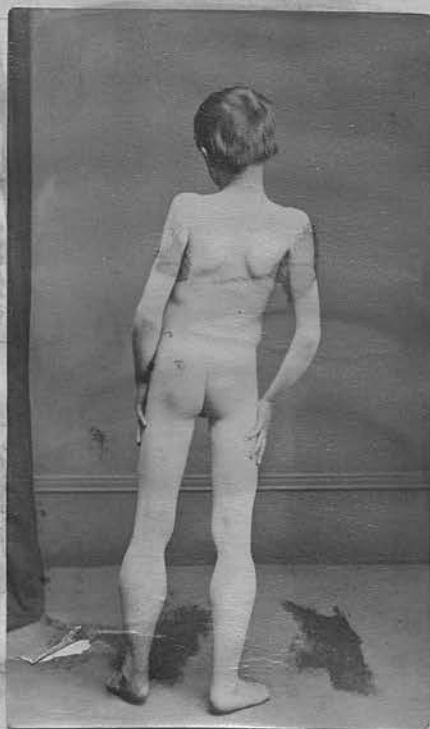
By so doing, the weight of the trunk is transferred from the hips to the knees, and thus a lever of the third order becomes one of the second.

We find various distortions. Thus, Talipes 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. It exists when the patient is in the erect posture,

but disappears when he sits down.  
The shoulders are usually thrown  
far backwards, so that a plumb  
line from them falls well behind  
the buttocks. Duchenne attributed  
this to weakness in the Erector Spinae.  
Gowers is of opinion that it is due  
to the weakness of the hip Extensors



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 rarer symptoms may be mentioned; but are of small importance. Mottling 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 rarer symptoms may be mentioned; but are of small importance. Mottling 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 ~~mean~~ average duration is five to six years. Death is usually from some inter-current 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.

Howes lays much stress on the enlarged infra-spinatus, and the wasted latissimus & sterno-pectoralis.

Prognosis. This is most unfavourable. 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 muscles must be maintained as well as possible by systematic muscular exercises, faradism, kneading, & such like. Iron, Phosphorus, Strychnia, and Cod Liver Oil have been recommended for internal administration.

Pathology. In the muscles, 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 changes have been described.

No changes have been found in the anterior cornua of the spinal cord. A considerable number of cases have been examined in which no changes 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 Gowen calls a "perverted tendency of development inherent in the germinal tissue of the muscular system".

References to works consulted.

Gowers. "Pseudo-hypertrophic muscular Paralysis"  
London 1879

Duchenne "Collected works"

Lydenham Society

Articles in Bramwell's Spinal Cord;  
Gowers Nerve Diseases Vol I; Ross's  
Nerve Diseases Vol II; Hammond's  
Nerve Diseases.