Thomsen's Disease
(Congenital Myotonia)

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by

Arthur Birt M.B. & Ch.M. Edin
of
Berwick, Kings Co., Nova Scotia

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On Thomson's Disease
(Myotonia Congenita)

The advent of a new disease is always of interest, and since 1876, when Dr. J. Thomson of Kappeln, Schleswig, described and established as a separate entity the affection which has since been named deservingly by his name, much attention has from time to time been given to it by different Continental observers. In Britain, however, owing either to the rarity of cases or to failure to recognize them, no instance of this disease was known so far as I can ascertain, until 1887 (Bugara), and with the exception of Hale White's masterly summaries in the Edin. Med. J. Reports, to be referred to later, British contributions to its literature were rather scanty.

I propose to describe a case which I have had very exceptional opportunities of observing, and of which the family history is well known to me, prefacing
The account with a résumé of the literature of the subject up to the present time.

As far back as 1830 Sir Charles Bell hints very strongly at this remarkable affection, and mentions that some people, who can lift heavy weights, etc., have not the proper control of their limbs, so that excitement intensifies the defect. He describes symptoms since recognized as typical of the knee stiffness; the danger of falling, the staggering gait on sudden movement, etc., bearing off on repeated movement; the freedom from pain. But there is no suggestion of rigidity or muscular spasm.

In Benedict's nervous pathology a similar condition is described, but for all practical purposes the history of the disease begins with Thomson's classical account which transmitted "Nervische Krämpfe in willkurlich beweglichen Muskeln in Folge von erhöhter psychischer dispositiones".

Dr. Nössen states members of his family suffered from the complaint, which had existed in the family for five generations.

Nervous System of the Human Body - Char Bell (1st Ed. 1830 p. 164)
J. Nössen - Arch. von Psych und Nervenkrankheiten [Berlin 1876 Bd. V. 5.702]
Benedict - Nerven Pathologie und Elektrotherapie 13th Ed. 1864 2nd Ed. 1874
The characteristic symptoms were:

1. A peculiar tonic cramp-like contraction of the muscles without pain, coming on on attempted voluntary movement after a state of rest, or on changing one type of movement for another.

2. The disappearance of this tonic contraction after repeated similar movements, e.g., in walking.

3. The involvement of voluntary muscles only and those of the limbs more especially the facial, ocular, tongue, and laryngeal muscles being rarely implicated.

4. The presence of a greater or less degree of hypertrophy of the affected muscular groups.

5. The aggravation of the condition by emotion at all.

6. Its association with other neuroses.

7. Its hereditary and practically incurable character.

For reasons which will I think be evident on perusal of my final note to this paper, I need not go into further details of Ramsay's paper, merely drawing attention to the fact that he believed the disease to be a neurosis, and the primary defect to be in the origin of the motor impulses in the cerebrum.
It should be noted that examination of muscle fragments from Homsen's cases gave negative results (possibly due to defective methods of examination).

In 1876, Seeligermüller published a case in which there was hypersensitivity of groups of muscles with skin & papillaries normal, subjective sense of the limbs being asleep, and considerable opposition to passive movement especially in the knee joint. This observer inclined to the view that the defect was in the lateral columns of the cord - a congenital defect in development.

15 years later, Erb in an article entitled "Tonic spasms in muscles capable of voluntary movement," quotes the above mentioned case, and says that this and similar ones depend partly on some congenital anomaly of the nervous system occurring repeatedly in the same family. Further articles by this authority have since appeared adding considerably to our knowledge of the electrical reactions and histology of the affected muscles, to which reference will be made later.


In 1879 Bernhardt recorded a case due to injury, in which the electrical reactions were normal, but that the muscular contractions persisted after the cessation of electrical stimulation. He also noted the wax-like contractions passing from the fingers to the biceps when the muscles were pressed.

Peters. In this same year the phenomena were well described by Peters, and in 1887 Strumpell had a descriptive article entitled "Tonicke Krämpfe in willkürlich bewegten Muskeln" (Pathometria congenita), and in Italy Petrone followed suit.

The next important contribution I can find is that of Westphal and Moelii of being an account of their case shown Jan 9th 1881 before the Society of Psychology, Berlin.

The symptoms were able grouped as follows:

1. Continued immobility of muscles on standing up after sitting. E.g. if the patient have been seated along time, on attempting to rise, he can only with apparent difficulty bend...

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(3) "Reihe allen, Gebel, 9. 1881.
(5) "Arch. de Neurol.," Sept. 1892.
2. Persistent contraction after more violent muscular efforts or after a single powerful effort e.g. sudden flexion of forearm.
3. Inconvenient contractions under certain complicated movements e.g. Children will be seized in the midst of playing & suddenly brought to a standstill.
4. The tonic cramp will come on so suddenly that the somatic & physical phenomena can scarcely be separated. For instance one patient, if she ran quickly & caught his foot against a stone would fall flat on face with arms extended & rigidity of whole muscular system.
5. That, as noted by Thomee, the sudden height of the affection or irriationation will greatly exaggerate the symptoms, as in some cases will cold & exposure (all depressants of the nervous system). On the other hand continuous passive movement favours the restoration to normal for the moment.
6. It affects the muscles of the trunk and extremities, the sphincters being free.
The muscles supplied by the cerebral nerves are however subject to the same affection though in a less degree. E.g. here may be stiffness of the tongue in talking, of the masseters in eating, and of the eyelids in movement.

6. The Muscles supplied by the spinal nerves have shown an increase in bulk, and may be inordinately strong, though the increase of power is not usually proportionate to their size.

7. The mechanical and electrical excitability of the muscles is augmented.

8. In 2 cases the excised muscular fragments have appeared fairly normal.

(Nota. This latter statement must be changed in the light of our present knowledge.)

1882 In this year the leading article was that of Najoli describing two cases of the affection. In one of which there was pain over the lower spinal vertebrae, radiating to the iliac bones + worse after walking. Not increased however by pressure or passive movements.

1883. saw several interesting communications on this subject, notably that of Béclat & Marie. They gave a full report of a case in an exhaustive article entitled "Graisse musculaire au début des mouvements volontaires" (Étude du trouble fonctionnel jusqu'à ce jour non décrit en France). Amongst other points they called attention to the prolongation of the muscular contractions. Schöningh in Germany & Engel in America also described cases arising from fright or nervous shocks (a point which would militate strongly against the muscular theory of the disease referred to later). Westphal also showed 2 cases at the sitting of the Berlin medical society in this year.

In 1884, the only reported case appears to be that of Vigorous, entitled "Maladies de Romsen et Paralysie pseudo-hypertrophique".

In this year Erb contributed another article in which he called attention to the reactions of pathological states up to that time, and described the electrical reactions of the muscles. He described also the histological appearance of excised

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(1) J. Béclat et P. Marie - "Arch. de Neurologie" Jan. 1883.
(2) Schöningh - "Berl. klin. Wochenschrift" July 1883.
(4) Vigorous - "Arch. de Neurologie" Vol. 8, No. 24, Nov. 1884.
MUSCULAR FRACTIONS. The points he observed were: 1) The fibres were much wider than normal; 2) they were rounded, not polygonal on cross-section; 3) the interstitial tissue fibres were normal, but there was thickening of the sarcolemma. He also noted and described in the Vasti & other muscles the wave-like contractions (which are characteristic) passing from the Kettuce to anode when the galvanic current is allowed to flow for some time, or the shocks are repeated at rhythmic intervals. So the prolonged tonic muscular contraction with lengthened relaxation, readily obtained on cathodal current, & of the associated neuro-muscular phenomena he gave the name Syntonic Reaction (MyR).

Eilenberg & Melcheriz now attracted attention by publishing the account of another family group of 14 cases. The nervous encumbrance was marked, two of the children all had cramps & convulsions during tetanic & hemichromatic vomiting during their schooldays. The daughter had migraine & another, angina attacks with vomiting. The electrical reaction differed a little quantitatively.

Illustrating the muscular condition in Thomson's Disease

(1) Forearm showing the marked muscular development on slender bones.
(2) Showing prolonged relaxation of Vastus Ext. some seconds after removal of rhesophore (My. C.)
(The relaxing muscle is upper outer part of thigh)

(3) Lateral view of thigh relaxed, muscles fully inconspicuous.
(4) Front view of thigh with knee extended, showing iron had tonically contracted elsewhere.
Photographs
Illustrating the Muscular Constitution
Other observers who published cases in 1885 were Peter F. Dalrymple, Delapre, and St Jean. The latter mainly on the pathological aspects of the disease.

1886 Hamilton of New York had an important article in this year on the Homans' symptom complex. He noted the distribution of the recorded cases, and summarized the symptoms. He tried apparently to throw doubt on the entity of T.D., but this is now put beyond controversy.

More important still was an able summary by Hale white of our knowledge up to that date on some recent advances in our knowledge of Homans' disease. This observer often summarizing the literature, recorded cases and symptoms, gave a clear and able account of the electrical reactions of the muscles, the microscopic appearances of the fibres.

That part of his summary relating to the muscles may be here given:

1) Muscles are increased in bulk.
2) No corresponding increase in strength.
3) On attempted voluntary movement, the

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(1) Arch de Neurologie No 29, 1885
(2) Union Med. Paris 1885 XXXV, 50-52
(3) Centralbl f Neuroh. Leipzig 1885 VIII, 122-126
(4) Medical Record, New York, Jan 23, 1886
(5) Trans. Am. Phys. 7, 1886
Later period usually prolongs (this is modified later): then comes a second contraction lasting 5–30 seconds, then sometimes a rapid relaxation. This peculiarity constitutes a myotonic contraction.

4. The above is more marked the longer the interval of rest, or after repeated contractions the difficulty gradually wears off.

5. Mechanical stimulation of nerves:
   - Diminished excitability (of my case AB)
   - Faradic or galvanic stimulation of nerve = slight myotonic contractions or being rapidly repeated.

6. Mechanical stimulation of muscle:
   - Increased excitability with long myotonic contraction. Galvanic stimulation usually gives closing contractions, KSZ is generally the best giving a good myotonic reaction, but the exact order is variable.
   - Strong faradic current, on closing, gives a good MyR. On opening gives a very slight contraction.

7. Stimulations rhythmically succeeding each other we get the wave-like contractions from cathode to anode.
Other cases by Hammond of New York and 
Fisher also appeared this year, and an 
article by Delmas. "Maladie de Thomson," which 
is of interest. He also issued another monograph 
1887. Saw the first 2 cases exhibited in England 
Buggard showing two brothers - typical cases 
along with Dr. Geo. Henschell he has the 
distinction of first demonstrating the 
disease in Britain (The diagnosis of my 
own case was made by myself and confirmed 
by medical friends before these cases were 
shown). They were described in The Lancet. 
In Germany an article appeared by Michelisen 
"Zur Kasuistik der myotonie congenita oder 
Thomson'sche Krankheit."

1888. Jacoby of N.Y. in this year wrote an 
able article on the disease. He inclined 
to the "Muscular Rhythm" as regards its pathology 
a point which will be discussed later. 
Michelisen showed one case before the 
Psychical Society of St. Petersburg, in which 
the disability was greatly diminished under 
Low pitch & alcohol. He found lengthening of latent 
period (3 in his traceage)

1888. Bernhardt, one of the original workers at T.D. published a paper on its pathology in this year.

1889–90. The leading papers of this year were those of Hale White in the Guy's Hospital Reports. A very complete description of a case, with myographic tracings, electrical reactions, and a general discussion of all the points of interest — in fact a most able and careful study of the disease. As I have already referred at some length to White's previous article on this subject, I need hardly go into details. His account of the electrical condition agrees very closely with that of my case and represents probably the most typical state of affairs. The rarity of the disease is shown by his statement that only Buggard's cases had been known at meetings in England up to Jan 37–90, and that in an appendix to his monograph stated that 46 cases only had been recorded up to that date. Another article that attracted attention was that of Wising, who described a Swedish case.

1890. In this year Cook Riversen described
a case in England, and Dreschfeld another
which was shown at Manchester to the medical
Society. The cases appear to have been pretty
typical, and Hughes rothers published
American cases some typical some not.
Rinner called attention to two cases in which
the eye muscles were affected (this is well
known in my own case also).

Although other papers might be mentioned
they do not chronicle any advance in
our knowledge of the affection, indeed
to Hansen himself, leprous, Marie
especially. So we are indebted for
the gift of all we know about this
remarkable disorder, which their researches
have established as a separate entity
in the list of diseases.

I shall only refer to one other article
viz that of Dejérine (4) Pottas describing
an Autopsy (so far as I am aware the
only autopsy) in a case of T.D.
It was a man of 32. who died of
Acute Nephritis. There was no abnormality

(2) Dreschfeld. ibid. 1890. I. 429.
of the Nervous System detected. They gave a careful account of the histological appearance of the muscles and described the vacuolation and interstitial growth of connective tissue as being a later degenerative stage of the affection. They found that the hypertrophy commences its most marked in the muscles that are most exercised and come to the conclusion that it was a primary myopathy of functional origin.

Since this date I have been unable to find anything of importance, except that one of the Paris theses for 1897 was on this subject. I regret that I was unable to obtain the article for reference of that even the name of the author has escaped me. Some interesting references points about its relationship to other nervous diseases were brought forward.

[Note] Paris, University of, Theses, 1897
My own case, a man of 34 - 5 ft 8 in. in height and 150 lbs weight has the following history. The muscular stiffness was first noticed at the age of 7, when it was supposed to be due to rheumatism and precautions in the shape of heavy underwear &. accordingly taken. Even as a young schoolboy he was noted amongst his fellows for physical strength associated with a degree of clumsiness of movement on initiating any voluntary action after a state of rest. He found himself handicapped at cricket or football by the muscular spasm, often suffering heavy falls & slight accidents as a result of it, e.g. when starting in a race he could take the first step or two, would then be pulled up by the strong spasm in the leg muscles, after a little the stiffness would wear off on continuing to run, and when fairly "limbered up" he was no more inferior to his schoolfellows in speed or endurance. So whilst running he
happened to strike his foot against an
obstacle, he was very oft to fall headlong
without being able to save himself and
with all his muscles stiff and rigid.
The arms were equally affected and
were early noticed to be more muscular
than those of other boys of the same age.
On turning the head suddenly, as in looking
over the shoulder there would be marked
stiffness of the neck muscles, and he
has always worn a larger collar than
most people. The jaw muscles frequently
showed the same tendency e.g. after taking
one bite, he would sometimes be unable for
a few moments to take some another until
the spasm in the muscles went off.
The tongue muscles & those of the eyeball
at times showed the characteristic
prolonged contraction in marked fashion.
Giving rise to thickening & difficulty in
articulation, and to straining of the eyes
in kind of glare. These symptoms passing
off rapidly. The latter one after stroking
the eyelids a little. There appears also
to have always been a similar affection
In the voluntary muscles concerned in the act of mastication. (This is interesting in view of the statement, frequently made, that these are never affected).

The other facial muscles would at times show traces of the stiffness, but this was only noticeable in cold weather.

My patient naturally found these diminutive vagaries very annoying, and there can be no doubt, as Powers remarks, that the complaint casts a gloom over the life of the sufferer.

The symptoms were noticeably improved by warmth and moderate exercise; but were greatly aggravated by any mental excitement, fatigue, or by prolonged cold.

He describes the spasm as having been always preceded by a dreadful sense of numb powerlessness, and has also a vivid recollection of many absurd and ridiculous situations in which his then unrecognized complaint landed him in past years.

The patient has always been very "nervous." He is highly strung, easily exhausted, and never

* A subjective feeling of the limbs being "asleep."

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About what has been on the whole remarkably free from serious illness. I should note that excessive sweating seems to have always been present on anything like serious exerting a symptom I do not recollect seeing mentioned in other cases. It was most marked in head sweat.

Inquiry into the family history shows that the mother, and the only married maternal aunt, transmitted hereditary tendencies to their children. In the one instance strongly, in the other only feebly.

In the case of the strong inheritance, the son (my case) and one daughter were well marked instances of Moro's disease. The one highly nervous, the other partially imbricale (death at 18 with obscure cerebral symptoms—no autopsy) of the other two daughters, one suffered from repeated attacks of Hysteria and one from an afflatus of some Hysteria for some time.

Amongst the children of the maternal aunt, (four sons & a daughter), two sons were affected with a slight form of V. O. showing itself mainly in the lower limbs.
and accompanied by a certain degree of the
characteristic muscular hypertrophy.
No cases were traced on the paternal side
of the family, but my patient's father was of
neurotic type, whilst there was no such
tendency in the Aunt's husband. This fact
perhaps accounting for the more severe
incidence of nervous symptoms in the
children of the former.

Family tree of 3 generations showing incidence of
T.D. other neuroses in patient's family.

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Grandparents

(healthy)

Husband
(healthy)

Daughter Son Daughter Son Daughter
No neurotic Neurotic (?) Neurotic traces of T.D.

Son
(healthy)

Son Son Son Daughter
T.D. Slight T.D. Slight Neurotic

Son
(healthy)

Daughter T.D.
(Neurotic)

Daughter T.D.
(Slight)

T.D.
(Died without symptoms)

Present Case.
On stripping the patient, it is at once apparent that he is very muscular. This is the more noticeable since the bones are rather slender and he carries not an ounce of superficial fat. The muscular hypertrophy is rather irregular in its distribution, being most marked in the muscles of the neck, back, and arms. In standing there is marked lordosis, and a slight degree of flexion of the elbow, though due no doubt to the greater enlargement of the flexor and hamstring groups being more marked than in the case of his opponents. In the upper limb the deltoid, biceps, triceps are very well developed, while the lower limb the calf muscles although standing out sharply when contracted are not nearly as much hypertrophied as the vasti, hamstrings, and flexors. The intrinsic muscles of the thumb and little finger are unusually well developed, and at the wrist the muscular fibres run far down on the thumb.

Measurements:
- Neck = 14½ ins. 17 in. with muscles contracted
- Wrist = 6½ in.
- Forearm = 11½ ins. relaxed 12½ ins. contracted
- Biceps = 11½ ins.
- Calf = 13 ins.
- Neck = 19½ ins.
- Chest = 33½ ins.
Examining the trunk muscles: the pectorals, when the arms are firmly crossed, stand out in bold relief, and on making the patient lunge forward with extended arm a beautiful demonstration of the serratus anterior is obtained. The lumbo-sacral muscles are very massive, and the intermuscular spinal groove exceptionally deep. Glutei very powerful. The neck is thick, powerful with a remarkable development of the sternomastoids, especially on the right side.

The recti abdominis can be made to show the recti segments very sharply by causing the patient to contract them.

The muscles at rest feel rather flabby, then otherwise, i.e., on sudden voluntary contraction, e.g., flexion of forearms, these concerned in the movement are felt for a few seconds to be of almost iron hardness which rapidly dies down leaving the muscles soft and rather fatigued. Tested by passive resistance, the dynamometer and various other strength trials.

\[ \text{Strength of the muscles} \]

**N.B.** It appears that the patient has never gone in for athletics to any extent, and that the muscles "keep about the same" whether used much or little. It is however the ones most used that show the hypertrophy.
Here is found a capability for short efforts of strength, but fatigue in the muscles exercised rapidly supervenes. The strength is usually proportionate to the bulk of the muscles, but it is more quickly exhausted than natural (i.e., it only lasts as long as the first few powerful tonic contractions).

**Measurement of Neck = 16\%\% in. of Forearm flexed = 13\%\% inch. Of upper thigh = 19\%\% inch.**

The peculiarity of movement is very noticeable. If the patient be made to sit for a few minutes and then told to get up and walk across the room, he rises stiffly and clumsily and takes a step forward, but on attempting to follow it or to be hampered by the prolonged contraction of the calf muscles, and it is only after walking a few yards that the spasm lessens off. In the same way, he can sharply flex the forearm at the word of command, but cannot for a few moments extend it fully again. Even attempt however is done more easily or at least a normal range.
and rapidity of movement is attained.
It is to be noted that the whole of the
muscular system shows a degree of
rigidity on getting up after a state of rest.
It is not only initiating a movement
after rest that induces the prolonged
muscular contractions; change from one
movement to another thus introducing
a new set of muscles. Thus, the patient
walks perfectly in the ordinary way;
but, when directed to "reverse", although
understanding perfectly what is required
he is unable to do so at first, being
brought up short by the muscular
spasm which at once supervenes.
Again, when directed to grasp the hand
of the examiner strongly and then unclasp,
he cannot for a few seconds relax the
grip—a phenomenon which has naturally
proved very embarrassing both to himself
and others on more than one occasion.
The setting of the eyeballs on suddenly
turning them to sideways, and the resulting
stare only passing off on striking
the eyeballs to relax. The symptom can be
Early demonstrations as can also the mischief
of speech or sudden attempting phonation
after along silence. The intercessors appear
to be healthy, writing not being interfered
with.

The accompanying myographic tracings
show well the characteristics of the contraction
in Thomsen's Disease. They represent the typical
stages of the fingers. Contractions repeated
at the word of command as soon as
the preceding relaxation was complete
(as indicated on the tambour).

It will be seen that the contraction is
slow, powerful, and much prolonged (lasting
from 10 to 30 seces or even longer) so that
it is regular in character, while the
period of relaxation is much prolonged
and the relaxation irregular in character.

The second contraction shows the same points
but in a less marked degree and
the approach to the normal muscular
contraction becomes closer at
each attempt until the Myotonic (Ref)
character is completely lost. It should
be noted that if complete relaxation be
Not wait for the patient being told to flex & extend the fingers repeatedly as quickly as possible that the second relax contraction begins before the first has completely relaxed. At this, to a less extent, holds good with the second or third. The latent period did not seem prolonged.

**Effect of Mechanical Stimulation of Motor Nerve**

E.g. rolling silmarik between finger & thumb.

Contraction = more ready and relaxation slower than normal = somewhat increased irritability.

**Effect of mechanical stimulation of muscle**

If the Tactus extends be struck several sharp blows in succession with a percussor hammer, clumpy contraction is produced which persists sometimes 25-35 sec before slowly dying away = increased irritability (Here compare photo indicating this slow relaxation in the Vastus ext.)

**Effect of Faradic stimulation of motor nerves.**

(a) Quantitatively = Normal
(b) Qualitatively = Contractions persist a long time, and relax very slowly, and the stronger the current the better this is marked. No opposing contractions. (MyC of Erb)

N.B. MyR = Myotonic Reaction of Erb.

MyC = Myotonic Contraction as shown in Myograph.
Effect of Paralytic Stimulation of Muscles.
Quantitatively normal, unless the current localization is strong enough to produce any contractions when they persist and relax slowly as before. (MyC)

The stronger the current the easier these reactions are obtained. There are only short, lightning-like contractions on opening the current.

Effect of Galvanic Stimulation of Nerves.

In a momentary application of the current nothing abnormal. But if the electrodes are left in contact for a short time, the resulting contraction persists some seconds and relaxes with abnormal sluggishness (MyC) as before... and as before, the stronger the current the more marked this phenomenon. On opening contractions are obtained.

Effect of Galvanic Stimulation of Muscles.
Quantitatively increased. ACC is quite as easily obtained as KCC in thumb muscles, fingers, etc. With weak currents contraction and relaxation is almost momentary, but with stronger ones the contraction is prolonged and relaxation also (MyC) after lasting many seconds. This is more
Marked if the current be allowed to run a short time. When the shocks were given at regular intervals wave-like flickerings were at times noticed passing from the cathode to the anode, as has been described by Erb & other observers.

On two occasions a second very marked contraction wave was noticed after the electrode was removed.

The main point elicited by this examination is, that to both a sudden single stimulation & alternating current, the period of contraction & relaxation of the muscles (especially the last) is much prolonged in Meissonier's disease. Whether the stimulus is voluntary or electrical does not matter.

The contraction is regular, the relaxation irregular.

Dr. Swynge of Bristol kindly excised a fragment from the Vastus externus; and an examination of this gave the following results:

The muscle fibres were considerably larger than normal, appearing as if two or more had been fused together. They measured from 1/50 to 1/400 inch, in D'escott (No. 1/50 - 1/400 Leuci's Anst.) They were rounded t. n. t.

* When the current (galvanic) was allowed to run for some time the same phenomenon was as tonic noted.
The nuclei were more numerous than in normal muscle, but not out of proportion to the size of the fibres. The striation was not quite distinct, but no excess of interstitial tissue was noted (as observed by Erb in one case, in probably a later degenerative stage). The "Achilles' pouches" were more distinct than usual, due to increase in the interfibrillar "protoplasm.

These characteristics of the muscles, electrical and histological, agree closely with the descriptions of Erb, Dale White, Hamilton, Adie, and Cottas and other observers, so that the case is quite a typical one in these respects.

The muscles are free from pain either on handling or during the tonic apparatus. But a complete feeling is uneven and during isometrics, there is no hyperaesthesia or dysaesthesia.

So delayed sensation. Muscular sense appears normal. The knee jerks are equal & ready. Superficial reflexes natural. Special senses show nothing abnormal, except that, along with a moderate degree of myopic astigmatisim, there is the “facing” of the eyeballs.

* M.E. The eyeballs are very prominent, rises the "facing" will
of the dilator muscles of the eye previously referred to. Colour sense & vision fields are normal. The profuse sweating on slight exertion or under emotion has been already noted. The viscera seem healthy. Pulse is large & soft. Occasionally palpitation occurs under trifling causes. Patient has suffered for years from a degree of neurasthenia and his hyper-sensitive nervous system reveals itself in many little ways.

N.B. There is a certain degree of pain between of hands after having now described my case I shall attempt to shortly review the theories of the disease put forward by various observers. It must be confessed at the start that the pathological material is of the scantiest. The only autopsies I know of is that of Dejerine & Brottes already referred to. Besides which in about 20 or 30 cases fragments of muscle have been examined histologically.

The four chief theories are these:

(1) That it is a psychosis, interchangeable with the neuroses & hysteria & epilepsy.

The muscular hypertrophy being secondary.

(Thomsen, Hamilton, Dejerine &)

(Thomsen, Hamilton, Dejerine &)}
This was Hornemann's own view, and appears to be much the same as that of Hamilton of New York who regards it as a hysterical nature dependent partly on insufficient nervous inhibition and partly upon an unstable emotional state interfering with volition. He explains the myotonia as due to a repeated reflex spasm made possible by a diminution of the inhibiting influence of the upper level over the multipolar cells of the cord.

This view undoubtedly derives strong support from the fact that T.D is often (even generally) associated with other neuroses in the same family or in the patient. That emotion and other depressants of the nervous system affect the symptoms so markedly, and thereby lead to the conclusion that this is the most tenable position.

3) That it is a primary myopathy (Lydon). This observer considers it an altered muscular condition of the muscular tissue congenital in origin. The points put forward for this view are (1) the frequent alteration in the electrical excitability (2) Ringer-Laudalbury's experiments

on frogs. He found that certain salts such as sodium phosphate are capable of producing a similar muscular spasm in the frog that the spasm persists both after the division of the nerve or after the intramuscular nerve endings have been paralyzed by curarum. This however does not prove that the muscular derangement is the only factor—that it is not so as is suggested by the facts that it has arisen in adults from Koch & R. Stuyan (Bernhardt & Schmätzlil & Engel).

Other modifications of this muscular theory are those of Jacoby, who assumes that there are too many sarcomatous elements in each fibre of which there is no proof; and of Bernhardt, who thinks it is due to a faulty nutrition of muscular fibres. He quotes Sellmann's observations on new-born animals, in which it was shown that the ascending limb of the myographic curve is prolonged to the descent of the curve very slow; an analogy has also been drawn with the condition in fish and rabbits, in which there are two kinds of muscular fibres—red & white.


differences in their contraction curves. 
Wolff, Baller & Leriche others consider it a congenital error in muscular tonicity, judging from (1) the extension of tic to almost all the voluntary muscles.
(2) Their transitory character. (3) The phenomenon produced by electrical stimulation.
Leriche thinks it more probably due to an inhibition of the sympathetic or vasomotor influence on the vessels of the muscles arresting the removal of the chemical products of muscular action thus interfering with nutrition & muscular movements. That the muscles in fact are in a state approaching rigor mortis where in myosin is developed.
This ingenious view has not of course any proof advanced in support of it, and, along with all the others which attempt to fix the primary fault on the muscles seems to take far too little cognizance of the associated mental condition, the now proven fact that the neuroses can almost invariably be traced in the family. Again in my own & in several other recorded cases it has been expressly noted.
that the muscles show no increased firmness, but are on the contrary rather flabby than normal between centres when at rest. By that it is due to a developmental defect in the pyramidal tracts of the cord (Destombes) or to a local spasticity of the innervation centres in the cord. Other observers, but the experiment and fact of the observed facts of T. D. both show rigidity apart from obvious nerve centre lesions. The point (cf. negative results of examination of nervous system by Bégurier and Bottes) argues strongly against the cord theory. This is also no exaggerated undue jerk or other "spastic" signs.

(C) That it due to an abnormal functional state of the nervous system & muscles - congenital in both. Overaction of the muscles being accompanied by overaction of spinal cells of the cord & perhaps also of the pyramidal cells of the cortex. (Gowers and others) as Cook & Swinburne offer the following explanation - after a period of rest during which the motor cells have acquired their maximum force.

The voluntary impulse causes an excessive discharge of motor energy which, if not stopped by the action of other cells, sets up a second and succeeding discharge in the same cells, thus the muscle is again stimulated to contraction before it has had time to relax from the first contraction, that is, after the cells have been thus relieved from their excess of energy, they become more stable and are then able to work smoothly or without fault. The same condition might be applied to the muscles of the body.

This it has been objected that the rigidity in R.D. is not a tonicus (which results when there is a continuous flow of stimuli to the muscle) for in this case, it is with the cessation of the stimuli that the chief effect comes out.

After a careful survey of the question and a close examination of one's own case, I have found in the psychical theory the most tenable one, and regard it as a disturbance of the psychic-motor areas of the brain, legitimation of which voluntary impulses congenital or, often hereditary, closely allied to hysteria and other neuroses. That there is a defect either in the origination or the
Transmission (or in both) of voluntary impulses, and that the muscular hypertrophy is a secondary condition. I think that some such explanation as that of Cotz's theory would explain the muscular condition as well as the psychical state. The pathological material is of very scanty, however, that it is hardly worth while adding to the already numerous explanations of the disease.

Diagnosis. This is to be made from (a) the history (b) the myotonic reactions of the muscle (especially the cutaneous clonus and Dorsalis) which is also congenital characterized by rigidity of lower limbs; but the rigidity does not pass off on movement. (c) this disease shows exaggerated knee juts & the symptoms of spinal epilepsy. The upper limbs, tongue & are unaffected. T.D. may also be mistaken for Pseudo-hypertrophic paralysis by a careless observer which disease shows (1) true paralysis (2) very large, but very weak muscles, side by
Side with other quite atrophies.

From Lateral Sclerosis it is distinguished by the fact that there is no loss of power, that the rigidity wears off with exercise.

Etc., it should be remembered, called attention to the fact that there are cases not typical in which the myotonic reactions are associated with other nervous condition symptoms. These appear to be very rare, however.

It is important for the sufferer and his friend that the condition should be recognized early. It disqualifies in Germany for the Army and Navy. In England the Indian and other public services (in which riding is an essential accomplishment) would also be out of the question. In fact any pursuit in which the free use of the muscular system is an essential is barred to the unfortunate subject of Thomson's disease. It does, indeed, cast a shadow over life in Gowers' words.

The life of open air muscular exercise, recommended by Thomson, is the best plan. Exercise short of fatigue and

* A warm and dry climate is a huge advantage.

These patients suffer much more than ordinary folks from cold and damp. - the spasm being more marked.
Carefully regulates. In both boys and girls (it is very rare in girls) games such as tennis, cricket, rowing, etc. may be freely encouraged. But riding, swimming in the open, skating may subject the patient to the risk of serious injury, from his inability to save himself from the effects of sudden attacks of the spasm or brought on during the exercise. Thus on one occasion my patient had a narrow escape from drowning on plunging into a lake from an anchored boat—the cold water & effect of the dive causing a prolonged rigid contraction of the muscular system, altogether preventing the use of the limbs.

Here in cases of such cases (schoolmastersing) should have the nature of the trouble explained to them, that the effects of over-pressure physical & mental (which are most injurious) may be avoided carefully. The nervous system should be kept in good condition. Occasional courses of hot baths & massage may help the muscles a little at times of extra stiffness. No hopes of cure should be held out, but
The patient may be told that no change for the worse is to be expected after puberty and every effort used to combat the phlegm, lack of confidences which is often experienced by the sufferer. For the excessive sweating a most annoying symptom of the complaint I have found nothing of any avail.

Note by the writer:
The above is an account of my own case, which was diagnosed by myself in 1881 (before any case had been shown in England). The diagnosis has since then been confirmed by F.R. Eaton of Liverpool and others. After considerable hesitation, I now submit it in the shape of a thesis, thinking it might be of some interest from the fact that this (so far as I am aware) the first time, since Thomson's classical paper that a physician has been in the unfortunate position of being able to describe the complaint as occurring in his own person. For obvious reasons I have refrained from recording the case in the Medical Journals.

Arthur B. B.
In the light of this and other typical cases described we may now attempt a comprehensive definition of this affection:

It is now an hereditary (rarely acquired) functional disease of those parts of the neuro-muscular system which are concerned in the origination and translation of voluntary motor impulses, of indefinite duration and somewhat progressive tendency, with its incidence chiefly in family groups and on the male sex. Characterized by (1) A condition of general cerebral irritability, (2) A sudden spasmodic muscular seizure attending the performance of a voluntary act, (3) An apparent increase in subsidence after the repetition of the act, (4) An apparent increase in the bulk of various groups of muscles, which are chiefly or most frequently the subjects of the cramp-like contractions, (5) A marked increase in electric-muscular irritability, (6) A normal or only slightly excited state of tendinous reflex activity (sometimes sub-normal), (6) An occasional involvement of the certain of the cranial nerves concerned in voluntary movement, (7) An absence of any gross change (Reinnervation) in the N.I. but true overdevelopment of the muscular fibres.