MYOTONIA ATROPHICA

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MYOTONIA ATROPHICA has been adopted as a convenient designation for a group of cases characterised by myotonia, a symptom which constitutes the essential feature of myotonia congenita or Thomsen's disease, and by muscular atrophy. It is only within recent years that the association of these two symptoms has received recognition, and it is still uncertain whether these cases are to be classified as examples of Thomsen's disease in which the atrophy is symptomatic, or whether they are to be regarded as constituting a clinical entity distinct on the one hand from Thomsen's disease, and on the other from the myopathies which the muscular atrophy closely resembles both in its distribution and characters. It is proposed in the present communication to report four new cases and to describe the leading features of the condition. The bibliography has been very carefully prepared, and, with the exceptions noted, the original articles and reports of cases have been directly consulted.

CASE I.—S. H., male, aged 27, unmarried, was referred to one of us (E. B.) for an opinion by Dr. Ralston Richardson in November 1912. His sister, who came to the hospital with him, informed us that the patient had always been a little peculiar mentally, and that when he left school at 14 he was only in the fourth standard. During the three following years he appears to have wandered about the country, working chiefly as a miner, but never staying for any length of time in one place. When 17 years of age he entered the Naval Reserve. While in the Service he says he was on several occasions treated in hospital for gastric trouble. A year or two after joining he had a fall from the rigging on to his head, and although the immediate consequences do not appear to have been serious, for he was not confined to bed, the weakness of the hands from which he now suffers was noted soon afterwards. After serving for two or three years he was discharged. His sister informed us that the reason for his dismissal, as shown by his discharge sheets, was "mental disease." Since his discharge he has been doing occasional work, though he has never retained a situation for more than two or three days at a time.

According to his sister's statement he is more irritable and obstinate than he used to be, and is at times difficult to manage. She has noticed, too, that he sometimes drops heavy articles from his hands, and that his speech is thicker than it used to be. These symptoms appear to have been present for several years. There is no history of venereal disease. The patient is a total abstainer.

Family History.—The patient is the youngest of a family of 13.
His father died at the age of 65 (of liver disease), while his mother, aged 73, is alive and well. Both appear to have been healthy people, and, so far as can be ascertained, in no way peculiar. Two brothers are affected with similar symptoms. Their cases are recorded in the present paper. The ages of the various members of the family were obtained from the family Bible, and are as follows:

W. H., m., born 1861, alive and well.
L. H., f., born 1863, alive and well.
C. H., f., born 1865, died at the age of 14, cause ?
J. H., m., born 1867, alive and well.
Jo. H., m., born 1869, died at the age of 14, cause ?
D. H., m., born 1871, died from an accident some years ago.
Da. H., m., born 1873, alive and well.
J. H., f., born 1875, alive and well.
H. H., f., born 1877, died at the age of 17, cause ?
Ro. H., m., born 1879, myotonia atrophica.
T. H., m., born 1881, myotonia atrophica.
R. H., m., born 1883, died at the age of 13 weeks (? spina bifida).
S. H., m., born 1885, myotonia atrophica.
R. H. is said to have died from some injury to the back received at birth. The sister from whom these facts were obtained thinks that the spine was open on the surface towards the lower end. So far as she knows, her mother had no miscarriages.

Apart from the three patients whose cases are here described none of the relatives have, so far as can be ascertained, been affected with any condition resembling myotonia atrophica. L. H., the only other member of the family we have had an opportunity of examining, presents no evidence of the disease.

The eldest brother (W. H.) was operated on for cataract a year or two ago. The remaining brothers and sisters are said to have good vision.

There appear to have been no cases of epilepsy, insanity, or nervous disease in the family.

State.—The patient was admitted to hospital on 6th November 1912. His general muscularity is good. His intelligence is of a low type; he is irritable and quarrelsome. His language is said by his fellow-patients to be of the type of indecency generally attributed to the schoolboy. There are no delusions and no loss of orientation, but he is suspicious, and rather cunning. His memory appears to be somewhat defective, as judged by his contradictory statements regarding his previous health.

The face is expressionless. There is no ptosis, the ocular movements are good, and there is no nystagmus. The right pupil is slightly larger than the left, but both are regular in outline, and react promptly to light. Vision is quite acute. All the intrinsic muscles of the face
are somewhat weak. The patient is unable to close the eyes against the slightest resistance, nor can he raise the corners of the mouth. There is distinct weakness of the orbicularis oris, and the buccinators appear to be weaker than normal. There is no difficulty in chewing. The temporal and masseter muscles contract fairly well, but are somewhat poorly developed; the lateral and forward movements of the jaw are well carried out. The jaw-jerk is not obtained. The upper and lower incisors cannot be opposed, although the jaws meet posteriorly. The palatal arch is high and narrow; the soft palate moves well, and the palatal reflex is present. The patient would not submit to a laryngeal examination. The movements of the tongue are somewhat slow. Articulation is slow and somewhat thick and indistinct.

Both sternomastoids are markedly atrophied and weak. The patient is able, however, with an effort to raise the head when lying on his back. The muscles of the shoulder-girdle and upper extremities are well developed, and their actions, with the exception of the hand movements, free and strong. The grasp on both sides is poor. All the intrinsic muscles of the hand show some degree of weakness. The thumb and little finger cannot be opposed. There is, however, no atrophy. When the hands are firmly closed the patient experiences considerable difficulty in opening them. This is due to inability to relax promptly the flexors of the fingers. The same phenomenon is seen in the interossei, for the patient, after grasping firmly, can only with difficulty separate the fingers while opening the hand. The duration of relaxation varies to some extent with the strength of the contraction, occupying, as a rule, several seconds. After repeated movements the duration of the relaxation is found to progressively decrease. Myotonic phenomena during voluntary movement were not observed in any other muscles.

The muscles of the trunk and lower extremities are fairly well developed and powerful, with the exception of the dorsiflexors of the feet. These are found to be distinctly weak in action, as tested by movements against resistance, though they are not definitely atrophied.

Nowhere is there any muscular hypertrophy, nor are fibrillary tremors present.

Beyond some quantitative reduction in the paretic muscles nothing abnormal was noted as regards the electrical responses. The mechanical excitability of the muscles was not examined in this case.

The knee-jerks and ankle-jerks are present, though feeble. The arm-jerks cannot be elicited. The epigastric and abdominal reflexes are very active. The plantar reflexes are present, and of the flexor type. Sensibility is unimpaired. The sphincters are unaffected. There is nothing to note regarding the other systems. The testes were not examined.
Case II.—Ro. H., male, aged 33, unmarried. The patient states that as a young boy he was as strong and as well developed as other boys, and that when he left school at the age of 14 he was in the fifth standard. When 16 years of age he had an attack of pneumonia, and was subsequently more or less confined to bed for five months. On recovering from this illness, regarding which we have been unable to obtain any further information, he says he was greatly reduced in weight. For a year afterwards he was very weak, and he has never altogether recovered his strength since. He was laid up again with pneumonia eleven and four years ago. It is now thirteen years since he did any regular work. Prior to this he followed a number of occupations, being employed as a light porter, a shop assistant, and a cleaner on the railway at different times, but never worked for any length of time in one situation. His inability to obtain regular employment appears to have been due to his mental temperament rather than to physical disability. Ro. H. appears to be morbidly conscious of his dignity, and, as his younger brother (T. H.) expresses it, "he would never stand being put upon."

Four years ago the patient says he was bitten on the arm by a horse, and it was after this that he first noticed weakness in this region. Subsequently weakness in the right arm and hand developed. Unlike his brothers, Ro. H. has never noticed any stiffness in the hands. The voice is, he says, thicker than it was. He has no difficulty in opening the eyes in the morning. The legs, he says, are as strong as ever they were, and he can walk long distances without fatigue. His brother T. informs us that he sometimes boxes with him, and that, although easily knocked out, Ro. H. is very quick on his feet. Within the past year the patient has noticed a marked diminution in sexual power.

State.—The patient's mental condition is best described by the expression "a little weak." His general muscular development is very poor. The physiognomy is characteristic, the facies presenting a very similar appearance to that met with in the Landouzy-Déjerine type of myopathy. The hair, which is fine and straight, appears to grow abnormally far off the forehead. The growth of hair on the face is poor. The face, which is expressionless and immobile, appears to be unduly elongated, the temporal and masseter regions being somewhat hollowed, while the mouth is kept slightly open. The upper eyelids cover the cornea to the edge of the pupil, while the lower lids hang somewhat, the sclerotic being visible below the corneal margin. The ocular sphincters are very weak, for even when the patient attempts to firmly close the eyes the lids are not completely approximated. After keeping the eyes closed for half a minute they are promptly opened on command. The patient cannot wrinkle the forehead, and when asked to raise his upper lip in order to expose the teeth the
Three brothers suffering from myotonia atrophica described in the text. From left to right—Ro. H. (Case II.), T. H. (Case III.), and S. H. (Case I.). Note the appearance of the face and atrophy of the sternomastoids.
Case II. (Ro. H.).—Showing the myopathic facies and absence of the sternomastoids.

Fig. 1.

Case II. (Ro. H.).—Showing the peculiar poise of the head on the trunk.

Fig. 2.

Case II. (Ro. H.).—Showing an attempt to forcibly close the eyes and raise the upper lip.

Fig. 3.

Case II. (Ro. H.).—Showing an attempt to flex the head against resistance.

Fig. 4.
movement is a very feeble one. There is, too, some weakness of
the buccal sphincter.

The pupils are of medium size, the right being a shade smaller than
the left. They are regular in outline, and react promptly to light. There
is no evidence of cataract. The right upper lid droops slightly. Although
there is no noticeable strabismus when the eyes are in the
position of rest, when the patient looks to the right the right eye
does not move outwards to the full extent, while on looking upwards
it tends to deviate slightly to the right. This defective movement of
the eyes is said to have been present for many years. There is no
diplopia and no nystagmus. The masseter and temporal muscles,
though somewhat small in bulk, contract fairly well. When the patient
is asked to clench the jaws and then open the mouth he does so
promptly. Articulation is distinct, but the voice is low-pitched, and
there is a suggestion of nasal intonation.

Dr. J. S. Fraser, who kindly examined the throat, reports: "There
is paresis of the soft palate and a persistent infantile epiglottis." As
regards the larynx, he says: "On phonation the cords do not come
quite together, and, on asking the patient to take a deep breath, they
do not separate widely; in other words, while they do move inwards
to some extent on phonation, and slightly outwards on inspiration, the
movements are decidedly less than normal." The palatal arch is un-
usually high and narrow. Swallowing is unimpaired, and the tongue
movements are quite satisfactory.

The neck presents a very striking appearance. The thyroid and
cricoid cartilages, the thyroid gland, and the carotid arteries are
quite superficial. The omohyoid muscles stand out prominently, while
there is a deep hollow above the inner end of either clavicle. These
appearances are due to the almost complete absence of the sterno-
mastoid muscles. When the patient lies on his back and attempts to
raise his head he is quite unable to do so.

The shoulders are somewhat sloping, the right a little more so
than the left. The scapulae are distinctly winged, their posterior
borders lying practically parallel with the spine. This deformity is
due to absence, or extreme atrophy, of the lower and middle thirds
of the trapezius muscle. The upper part of the trapezius acts well, and
is not atrophic. The rhomboids, too, show no wasting. The serratus
magnus, pectoralis major, supra- and infra- spainus, teres major, and
latissimus dorsi muscles are unaffected. The erector spinae is some-
what poorly developed. Abduction and adduction at the shoulder
and flexion and extension at the elbow are well carried out. The
forearms are markedly wasted, both anteriorly and posteriorly, and
there is marked weakness of the extensors and flexors of the wrists and
fingers, and of the intrinsic muscles of the hand. The flexors of the
fingers show the same difficulty in relaxation observed in the case of
S. H., though not to the same degree. In no other muscles of the body
is there evidence of myotonia on voluntary movement.

The muscles of the trunk and lower limbs are poorly developed,
but all the movements are satisfactorily performed against resistance,
with the exception of dorsiflexion of the feet. This movement can
be overcome without much difficulty. There is slight relative wasting
of the anterior tibial muscles.

The gait presents no abnormality. Nowhere is there any muscular
hypertrophy, and there are no fibrillar tremors. Practically all the
muscles of the body, with the exception of the face and sternomastoids,
show a decidedly increased excitability to direct mechanical stimulation.
In the thumb muscles, flexors and extensors of the forearms,
masseters, tongue, supra- and infra-spinati, the anterior tibial and calf
muscles, the period of relaxation is markedly prolonged, the time
varying from 3 to 10 seconds. This is particularly noticeable in the
tongue, calves, and anterior tibial muscles.

The knee-jerks are present and rather diminished. The ankle-
jerks are elicited with difficulty. The plantar reflexes are of the
flexor type. For several years the patient has suffered from cold
hands and feet. Sensation is unimpaired, as are the sphincters. The
abdominal and thoracic viscera present no evidence of disease. The
testes are distinctly atrophied, while the pubic hair is remarkable for
its absence of curl.

Case III.—T. H., male, aged 31, a lithographer by occupation and
unmarried, is a brother of the two patients whose cases have just been
described. At the age of 17 he had pleurisy, for which he was tapped.
So far as he remembers he has had no other illness. He denies syphilis.
After a seven years' apprenticeship he commenced, eight years ago,
regular work as a lithographer. About this time (eight years ago)
he noticed for the first time that when he lifted a heavy weight or
when he turned the key which fixes the lithographer's stone in posi-
tion his fingers felt stiff, and he had difficulty in opening the hands.
This stiff feeling in the fingers, which he says is more pronounced
in cold weather, has persisted up to the present time, and is now,
he thinks, rather more evident than it used to be, though it has
never impaired his working capacity. Since it is only present in
the morning when he begins to work, disappearing completely in the
later part of the day, he is in the habit of fixing his stone in the
evening, in order that it may be ready the following morning.

Two years ago he noticed that his speech was rather thick. His
foreman remarked on it, and advised him to consult a throat specialist,
by whom he was told that he had a narrow, contracted throat. His
speech is, he says, no worse than it was at that time. Apart from the
stiffness in the hands and the thickness in speech he has noticed
nothing amiss. His arms, he says, feel quite strong, as do his legs.
He is fond of boxing, and says he is as nimble on his feet and as quick with his hands as ever he was.

State.—T. H. is a fairly muscular man. There is a lack of facial expression, which is very evident when he speaks. The patient is practically bald on the top of the head, in front of the line of the ears, and there is considerable thinning behind and at the sides. The hair is fine and straight. On the face it is almost confined to the upper lip and to a limited area at and below the point of the chin. The pupils are equal and regular; they react a little sluggishly to light; there is no ptosis, and the ocular movements are satisfactory. A few nystagmoid jerkings are observed on lateral deviation of the eyes. Vision is acute. The patient can elevate his eyebrows slightly, but cannot wrinkle the forehead. The ocular sphincters are very feeble. When he is asked to keep his eyes firmly closed for half a minute and then open them the upper lids are promptly raised. The masseter and temporal muscles are not wasted, and contract well. The incisors cannot be opposed. Articulation is thick and somewhat indistinct, as if the patient was speaking with something in his mouth. There is no definite nasal intonation. Dr. J. S. Fraser reports:—“There is paresis of the soft palate. The epiglottis is infantile, and tilted back over the larynx. Otherwise the larynx is normal.” The tongue movements are satisfactory. Deglutition is unaffected. The sternomastoids are much atrophied. All the other muscles of the shoulder girdle and upper arm are well developed. The forearms are relatively less muscular, though not notably wasted. The movements of extension and flexion at the wrist are distinctly impaired. The grasp is somewhat feeble, as is the movement of extension of the fingers. The small muscles of the hand are certainly weaker than they should be; there is, however, no distinct wasting. The patient cannot approximate the thumb and little finger. When he closes the fist firmly and is then asked to open the hand there is some difficulty in relaxing the grasp, which passes off if the movement is persisted in.

The thoracic and abdominal muscles act well.

When the patient lies on his back and attempts with his arms folded across the chest to sit up he is unable to do so, for the head falls back on account of the weakness of the sternomastoids; if, however, the head is supported, he can raise himself to the sitting posture without difficulty. The dorsal muscles are well developed. There is no winging of the scapulae and no atrophy of the trapezius.

The development of the lower limbs is not so good as that of the upper extremities; it cannot be said, however, that the muscles are distinctly atrophied. The quadriceps on either side, indeed, appears to be rather well developed, and the vastus internus especially is unusually prominent. All the movements of the lower extremities
are well carried out, with the exception of dorsiflexion at the ankle. This latter movement can be overcome without much difficulty. There is marked increase in the excitability of the muscles to direct mechanical stimulation. Prolongation of the period of relaxation was found in the muscles comprising the thenar and hypothenar eminences, the flexors and extensors of the forearms, masseters, tongue, sternomastoids, trapezius, quadriceps, anterior tibial and calf muscles. This was most pronounced in the tongue and anterior tibial muscles. No fibrillary tremors were observed. The knee-jerks are present, though less active than normal. The right ankle-jerk is difficult to elicit, while the left cannot be obtained. The plantar reflexes are of the normal type. The testes appear to be normal. There is no loss in sexual power.

Summary.—Three brothers, aged 33, 31, and 27, are the youngest living members of a family of 13. None of the other members have been, so far as has been ascertained, affected with similar symptoms, nor is there a history of any condition resembling myotonia atrophica, of nervous disease, or of cataract among the relatives. A brother born between the two youngest patients, who died aged 3 months, was very probably the subject of spina bifida. All the patients, but notably the eldest and youngest, are of a degenerate type. The symptoms of the disease first attracted attention in each instance during the third decade, and have progressed very slowly, if at all, since they were first noticed. The clinical features are almost identical, differing only in degree. Thus in each instance there is a myopathic facies, pronounced atrophy of the sternomastoids, more or less weakness of the muscles of the forearms and hands, and slight loss of power in the dorsiflexors of the feet, accompanied by slight wasting of the corresponding muscles and diminution of the tendon jerks. In all of the cases there is some difficulty in relaxing the grasp, but in no muscles, with the exception of the fingers, is there definite evidence of myotonia on voluntary movement. There are no fibrillary tremors and, with the exception of the thigh muscles in the case of T. H., no suggestion of muscular hypertrophy, nor is there any sign of cataract.

Case IV.—L. L., female, aged 38, single, a cook, was first seen in April 1912. The patient states that she was a seven-months child, and that, although she has never been very robust, she has not suffered from any serious illnesses. Since girlhood she has been troubled with megrim, but of recent years the headaches have become much less severe. While she was a schoolgirl her mother used to remark that she ran
Fig. 1.
Case IV. (L. L.)—Showing the facial appearance.

Fig. 2.
Case IV. (L. L.)—Showing the drop foot. The atrophy of the vasti interni and anterior tibial muscles is not well shown.

Fig. 3.
Case IV. (L. L.)—An instantaneous photograph taken during the effort of opening the hand after forcible closure of the fist.

Fig. 4.
Case IV. (L. L.)—Showing an attempt to flex the head on the trunk. Note the wasting of the sternomastoids.
differently from other children, although she could run as fast as girls of her own age. Her mother tells her that she used to throw her legs out in a peculiar way, and she remembers that she was often chaffed about the way she ran. Her hands, she thinks, have never been very strong, for she remembers as a young girl that her mother used to scold her when she was shaking carpets, because she would often let go the carpet she was holding.

The stiffness of the hands she first noticed after an attack of influenza some seven or eight years ago. The first occasion on which she observed this symptom was after she had been given gas for the extraction of some teeth. After recovering from the anaesthetic she found that she had difficulty in relaxing her grasp of the dentist's chair. The patient cannot say whether the weakness of the hands which now exists was present prior to the stiffness, or whether it was of later development. About three and a half years ago she began to find that she was easily tired on walking. The weakness of the legs has progressed until at the present time she cannot walk more than half a mile, and only then if she takes frequent rests. For several winters she has suffered from cold feet and hands, the fingers at times becoming white and dead.

Family History.—So far as the patient is aware, none of her relatives have suffered from difficulty in walking or weakness of the hands. Her father died of brain fever, following typhoid, at the age of 33, some three months before her birth. Her mother, who is 71 years of age, is in good health. She has a brother, aged 42, and a sister, aged 40, both of whom are well; a half-brother and half-sister, aged 34 and 30 respectively, both enjoy good health. No brothers or sisters have died. None of her relatives have to her knowledge suffered from any form of nervous disease.

State.—The patient was admitted to hospital on 20th April 1912. She is a well-nourished woman, and her general muscularity is fairly good. There are no obvious stigmata of degeneracy. Intelligence is good, and her memory is up to the average. Articulation is a little thick, and at times somewhat nasal. The pupils are equal in size, and their reaction is prompt. The movements of the eyes in all directions are perfect, and there is neither ptosis nor nystagmus. The face is expressionless, and all the facial movements are poor. The forehead can be wrinkled feebly. When the patient closes the eyes as firmly as she is able they can be opened with the greatest ease. On waking in the morning it is interesting to note that she often experiences difficulty in opening them. On closing the eyes as forcibly as possible and keeping them closed for some seconds an appreciable interval elapses before she can raise the upper lids. The masseters and temporals, though somewhat poorly developed, contract fairly well. On asking the patient to close her jaws and keep them clenched firmly for half
a minute she is afterwards able to open the mouth promptly when asked to do so. The jaw-jerk cannot be elicited. The palate moves somewhat feebly. On several occasions some few weeks ago fluids regurgitated through the nose while she was drinking. The palatal reflex is present. Dr. Logan Turner, who kindly examined the larynx, was of opinion that there was slight weakness in the abductor movements of both vocal cords. There is no difficulty in swallowing. The movements of the tongue are distinctly slow. The sternomastoids are very markedly atrophied, and their movements correspondingly feeble. The muscles of the shoulder-girdle are well developed; the prominence of the infra-spinati is indeed somewhat striking. The upper part of the trapezius, the deltoid, teres major, latissimus dorsi, and biceps are very well developed, and the movements which they take part in are effectively carried out. The musculature of the forearms is not so good as that of the upper arms, but there is no pronounced atrophy either here or in the hands. Extension and flexion at the wrist can be overcome without much difficulty, while there is even more pronounced weakness of the extensors and flexors of the fingers. There is, too, distinct weakness of the small muscles of the hands. After clenching the fist firmly the difficulty experienced in opening the hand is very striking, from 15 to 20 seconds elapsing before the fingers can be fully extended. When the grasping movement has been several times repeated the ease with which relaxation occurs rapidly increases, until before long the movement of extension is performed quite freely. That the disability to extend the fingers freely is due to defective relaxation of the flexors is obvious, for these muscles are felt to be in a state of contraction. The same phenomenon is also present in the extensors of the fingers, for when the fingers are extended against resistance and the patient then attempts to close her hand she has considerable difficulty in doing so. The muscles of the back and abdomen are quite strong. All the movements of the lower limbs are powerful, with the exception of dorsiflexion of the feet and extension at the knee and hip. The first-named movement is very feeble, while the movements at the knee and hip are only slightly impaired. There is distinct wasting of the vastus internus on either side, and the anterior tibial group of muscles shows some atrophy, though not to the degree one would expect from the weakness of the corresponding muscles. The patient walks with a steppage gait, raising especially the knee on the right side. The knee-jerks are feeble, and only elicited on reinforcement. The ankle-jerks cannot be obtained. The plantar reflexes are flexion. Sensation is unimpaired. The thoracic and abdominal viscera are healthy. Menstruation is regular.

Historical Résumé, with Special Reference to Incidence.—The occasional co-existence of muscular atrophy and myotonia, the
characteristic symptom of Thomsen’s disease, was first emphasised by Hoffmann, who in 1900 collected 12 instances from the literature, to which he added two personal observations. He classed these cases in two groups. In the first he placed 5 cases (Dana; Déléage (2); Bettmann; Hoffmann) in which the association might be regarded as coincidental, a view adopted by subsequent writers; in the second he included the remaining cases (Jolly; Pelizaeus (3); Kornhold; Schoenborn; Noguès and Sirol; Hoffmann (2)), which were all characterised by a more or less bilateral, symmetrical, progressive atrophy unaccompanied by fibrillary tremors. Although the distribution of the atrophy showed considerable variation, Hoffmann laid stress upon the great similarity between the 2 cases he had personally observed. Both of these patients exhibited (a) a myopathic facies; (b) pronounced atrophy of the forearms, particularly of the flexor muscles; and (c) of the sternomastoids. He further indicated that the wasting resembled more closely the myopathic than the myelopathic atrophies. The atrophy and myotonia must, he argued, be related, for the association was too frequent to be accounted for by coincidence. In his opinion the myotonia was probably the primary disease, the atrophy being symptomatic.

Isolated cases reported by Longard (1898) and Bernhardt (1899) as examples of Thomsen’s disease with muscular atrophy, in both of which there was slight wasting of the forearms, seem to have escaped Hoffmann’s notice. Gaupp in 1900 described a case in which, in addition to some atrophy of the upper limbs (notably the forearms and neck), the patient was the subject of infantilism. Frohmann the same year presented a patient in whom the trapezii and interossei were atrophied, and who at the same time exhibited nystagmus and Rombergism; while Schott in 1901 reported a case in which there was some flattening of the thenar eminences and interosseal spaces, speech being somewhat indistinct and the facial movements weak.

Rossolimo’s contribution (1902) is of special interest, since the Russian observer was the first to employ the term myotonia atrophica. The case which he reported closely resembled the two described by Hoffmann, the atrophy involving the face, masseters, forearms, extensors and adductors of the thighs and muscles below the knees. Although Schoenborn and Frohmann had previously noted the microscopical appearances presented by excised portions of muscle, the histological changes met with in the present case are more fully described and well depicted. According to Rossolimo,
the atrophy in the muscle fibres is to be regarded as a stage in the hypertrophic process, and the muscular wasting a symptom or variety of the myotonia.

A number of case records occur in the literature during the next five years (Jaquet; Schultze; Cassirer; Fuchs (2); Lannois; Berg; Curschmann; Lortat-Jacob et Thaon; Nonne-Sienerling-Hoffmann-Steinert; Batten; Pässler-Steinert (2); Pelz; Fürnrohr (2); Mirallié; Jalaber et Culerre; Kleist; Voss; Ramsay-Hunt (2)). For the most part these reports are contained in the notices of medical societies, and are in consequence brief; the communications of Berg, Pelz, and Fürnrohr are, however, in this respect exceptional. Berg in his dissertation, which the present writers have been unable to obtain, reviews the subject of muscular atrophy in myotonia congenita. Pelz, writing upon atypical forms of Thomsen's disease, gives abstracts of 17 of the cases reported up to that time in which there was associated atrophy. His remarks in this connection are limited to a consideration of the relationship between the myotonia and the atrophy. While admitting that the latter may be symptomatic—a view which, in his opinion, is strongly supported by the circumstance that in the majority of cases the myotonia had long preceded the atrophy, while in no instance reported up to that time had the reverse been noted—he tentatively suggests the possibility that these cases may constitute a type of disease distinct from myotonia congenita on the one hand and from the myopathies on the other. Fürnrohr, in his paper, reviews the literature, and from his study arrives at certain conclusions which may be briefly summarised as follows:—

(1) There is a "myotonia without tone," the myotonia showing itself in the electrical and mechanical myotonic reaction in the muscles, even in the absence of demonstrable myotonic phenomena, during voluntary movements. (2) The myotonic manifestations, as a rule, long precede the atrophy. (3) Single or numerous muscles may be affected by the atrophy, no law for the localisation of the atrophy having as yet been determined. (4) The small muscles of the hands and feet are affected frequently by the atrophy, in the localisation of which the occupation of the patient may under circumstances have a certain influence. (5) Under appropriate treatment there may be a certain amount of improvement.

Two important papers appeared simultaneously in 1909, the one by Batten and Gibb, the other by Steinert. The English observers draw attention to the frequency with which a similar
distribution of the atrophy is met with, these cases constituting a type which they think worthy of recognition. The characteristic features of the condition which they describe are “weakness of the facial muscles (myopathic face), atrophy of the sternomastoids, atrophy of the vasti of the thighs and dorsiflexors of the feet, and a slow relaxation of certain muscles after contraction. The more forcible the contraction, the slower the relaxation.” Among 29 recorded cases which they were able to find, they give abstracts of 15 which more or less presented these features, as did 5 new cases which they here describe. These cases they analyse as regards familial characters, etiology, sex, age of onset, distribution of the atrophy and the myotonia; and, referring to the atrophy, they suggest “the possibility of there existing a form of myopathy differentiated clinically from the classical types as much by the distribution of the wasting as by the presence of the myotonia.” They make no reference to the cases which they exclude as failing to conform with the type which they describe.

Steinert in his exhaustive monograph deals with 26 collected cases and 6 personal observations, 4 of the latter having been already reported by previous observers (Nonne-Sienerling-Hoffmann; Berg; Pässler (2)). The 5 cases referred to by Hoffmann in which atrophy seemed to be coincidental are not included in this list. Pelz’s case, in which there was weakness but no definite atrophy, is also excluded on account of the absence of wasting, although Steinert has no doubt that it belongs to this group. Upon analysing this material he points out that involvement of the forearms, sternomastoids, and face, a distribution which was present in all the patients he had personally observed, is highly characteristic of the muscular atrophy which occurs in association with myotonia. These cases do not, however, in his opinion, constitute a distinct type, but rather a type of muscular atrophy in myotonia congenita. In some of the reported instances the atrophy was widespread, in some it was limited to the upper and lower extremities, while in others the arms were alone affected. The last-named cases, in Steinert’s opinion, probably represent an early stage of the condition. Among other points to which he directs attention are the occasional occurrence of congenital deformities, of atrophy of the testes, of vasomotor phenomena, notably a tendency to cold hands, and of baldness. The second part of Steinert's paper is devoted to an account of the anatomical findings in one of his cases, the only case which has up to the present time been examined post mortem.
Within the past three years a number of cases have been reported (Turney (2); Steinert; Greenfield (3); Hirschfeld; Kennedy and Oberndorf (2); Pemberton (2); Grund (4); Farnsides; Griffith; Tetzner). Greenfield's observations are of special interest, for his patients, and those described by Turney, of which he also gives a detailed account, come of a family several of whom suffered from congenital cataract.

The present writers, after excluding 5 cases in which the atrophy was probably a coincidental occurrence in the course of Thomsen's disease (Hoffmann), have found 61 cases of associated myotonia and atrophy, making 65 in all when those reported in the present communication are included. These cases are in the writer's opinion all to be regarded as examples of the same condition, differing only in the extent and degree of the atrophy. Strong arguments in support of this contention are:—Firstly, that myotonia is a symptom which has only been met with in Thomsen's disease and in a few isolated cases of syringomyelia (Rindfleisch), apart from the two cases under discussion; and secondly, that the atrophy which these patients present is characterised by distinctive features, in that it is more or less symmetrical, very slowly progressive, and affects similar parts.

Batten and Gibb, as we have seen, were the first to demonstrate that in many cases in which myotonia and muscular atrophy are associated the distribution of the wasting is strikingly similar. While admitting the importance of this observation, the writers are at the same time inclined to think that it is, in some respects, undesirable to differentiate as a definite type cases which appear to be identical with those in which the atrophy is similar in its features though less extensive.

Upon analysing the 65 cases above referred to in relation to this point, we find that it is possible to class them in three groups, in accordance with the distribution of the atrophy. We would, however, again repeat that this division is, in our opinion, an arbitrary one, and is merely adopted here in order to demonstrate the relationship between cases in which the wasting is localised and others in which it is widespread.

**Group A.**—Observations by Noguès et Sirol; Hoffmann (2); Rossolimo; Fuchs (obs. I.); Berg-Steinert; Cassirer; Nonne-Siemerling-Hoffmann-Steinert; Lortat-Jacob et Thaon; Pässler-Steinert (2); Mirallié; Jalaber et Culerre; Kleist; Fürnrohr (obs. II.); Ramsay-Hunt (2); Steinert (3); Farquhar Buzzard; Batten and Gibb (4); Turney (2); Pemberton (2); Kennedy and Obern-
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dorf (2); Hirschfeld; Greenfield (3); Grund (4); Griffith; Farnsides; Tetzner; Bramwell and Addis (4).—In this group are included 45 cases which closely resemble each other, in that all exhibit the myopathic facies and wasting of the upper extremities, while in the great majority there was in addition atrophy of the sternomastoids and of the limbs.* It is interesting to note that with one exception—Greenfield's second observation—all the cases reported since 1909 which the writers have been able to find (23 in number) fall into this class.

Group B.—Observations by Kornhold; Schoenborn; Lannois; Fünrohr (obs. I.); Voss.—This group comprises 5 cases, all of which exhibited atrophy in both upper and lower extremities, the features and distribution of the atrophic paresis in these regions resembling that met with in the first group.

Group C.—Observations by Jolly; Pelizaeus (3); Longard; Bernhardt; Frohmann; Gaupp; Schott; Jaquet; Fuchs (obs. II.); Schultze; Curschmann; Pelz; Greenfield (obs. II.).—The cases included in this group are 15 in number, the atrophy in all but one case being localized to the upper extremities. Details regarding the distribution of the atrophy in these cases are as follows:—

Case I.—Jolly.—Atrophy of the right thenar eminences and to a slight degree of right forearm and right hand. Reaction of degeneration in muscles of thumb.

Cases II., III., and IV.—Pelizaeus.—Deltoids and thenar eminences; reaction of degeneration in thumb muscles. Two sisters appear from the brief report to have had myotonia and atrophy, the latter having a similar distribution.

Case V.—Longard.—Very slight atrophy limited to the forearms; some muscles markedly hypertrophied.

Case VI.—Bernhardt.—A slight degree of weakness and atrophy of forearms, hands, and finger muscles.

Case VII.—Frohmann.—Almost complete atrophy of lower two-thirds of both trapezius muscles, also atrophy of interossei. This patient showed nystagmus and Rombergism.

Case VIII.—Gaupp.—Muscles of forearms, small muscles of hands, triceps, and supinator longus; infantilism (?)

Case IX.—Schott.—Interosseal spaces shrunken and thenar

* One of Batten and Gibb's patients presented the myopathic facies and atrophy of the sternomastoids, with no involvement of the limbs. This case has been classed in Group A. A brother of the patient presented the typical features of myotonia atrophica.
eminences flat; face asymmetrical and expressionless, knee-jerks and ankle-jerks increased.

**Case X.**—*Jaquet.*—Atrophy of posterior part of deltoids, and some flattening of both anterior and posterior aspects of forearm.

**Case XI.**—*Fuchs* (obs. II.).—Very slight atrophy limited to the upper extremities; fibrillary tremors.

**Case XII.**—*Schultze.*—Atrophy of forearms.

**Case XIII.**—*Curschmann.*—Atrophy of radial extensors of right hand and extensors and abductors right thumb, also of the abductor pollicis and extensors of left thumb. Knee-jerks absent.

**Case XIV.**—*Pelz.*—Slight weakness of arms, though no definite atrophy.

**Case XV.**—*Greenfield* (obs. II.).—Some weakness of dorsiflexors of left ankle, but very little obvious wasting. Two sisters and a brother have typical myotonia atrophica.

It is possible that in some of the cases included in this last group the association of the atrophic paresis with the myotonia was "accidental." This may perhaps be so in the cases described by Jolly and Pelizaeus (obs. I.), in both of which the reaction of degeneration was present in the muscles of the thumb; in Fuch's second case, in which fibrillary tremors were observed; and possibly in the case reported by Frohmann, in which nystagmus and Rombergism were noted; but the writers are inclined to think that with these possible exceptions the relationship of the atrophy and myotonia is to be regarded as similar to that which exists in the cases classed in the first two groups, and that to attempt to separate them therefrom by any sharp line is undesirable. It is of interest to note, that two sisters and a brother of Greenfield's patient, in whom the atrophy was limited to the dorsiflexors of one foot, presented a typical picture of the disease.

**Features of Myotonia Atrophica.**—Myotonia atrophica is a rare condition. Of the 65 cases found in the literature by writers, 36 have been reported in German, 17 in English, 6 in French, and 6 in American publications.

A variety of names have been employed to denote these cases, viz.:—Acquired myotonia (Jolly); Thomsen's disease with muscular atrophy (Frohmann, Noguès and Sirol); partial myotonia congenita (Gaupp); partial myotonia with muscular atrophy (Schott); atrophic form of Thomsen's disease (Cassirer); myotonia with muscular atrophy (Lannois); muscular atrophy with the electrical reactions of Thomsen's disease (Lortat-Jacob et Thaon); combination of myotonia and muscular dystrophy (Nonne);
atypical form of Thomsen's disease (Pelz); amyotrophic myotonia (Päßler); facio-scapulo-humeral myopathy with myotonic symptoms (Mirallié; Jalaber et Culerre); muscular atrophy with slow relaxation of muscles (Kleist). All recent writers have adopted myotonia atrophica, a term first introduced by Rossolimo, as the most appropriate designation.

**Etiology and Mode of Onset.**—Males are much more often affected than females; thus among 62 cases there were 51 men and 11 women.

The age at which the condition develops is often difficult to ascertain with certainty, since the symptoms are as a rule so insidious in their onset. This is especially true of the myotonia, which in its slighter degrees may exist for many years without causing any appreciable inconvenience to the patient. As Batten and Gibb point out, in the majority the earliest indications are noted during the third decade. This was so in all of our cases. In quite a number of instances, however, notably in several of those in which the atrophy was confined to the upper limbs, the symptoms developed from the 16th to the 20th year, while several patients stated that they had noticed some weakness of the hands since childhood. Three patients first exhibited evidence of the disease after the age of 40.

The myotonia has without doubt preceded the atrophy by a number of years in the great majority of cases, although occasionally the latter has been the first symptom to engage attention (Rossolimo; Hunt; Batten and Gibb; Kornhold; Pemberton). Difficulty in relaxing the grasp is indeed almost always the earliest symptom to attract notice. Stiffness of the tongue was the first symptom in cases observed by Schott and Steinert (obs. I.). The paresis, when it appears, affects the upper extremities, notably the forearms and hands, almost invariably in the first instance, very exceptionally the lower extremities. In one case (Hoffmann) atrophy of the face was the earliest sign.

The tendency for myotonia atrophica to affect several members of a family is the one feature of outstanding etiological significance. Thus three of our patients were brothers; in one family a brother and four sisters (Greenfield-Turney), in another a mother, two sons, and a nephew (Grund), suffered from the disease. Other instances are recorded in which two brothers (Päßler; Hunt) and a brother and a sister (Hoffmann; Batten and Gibb) were affected. The mother, grandmother, and a brother of Pelz's patient are reported to have been troubled with "muscular stiff-
ness," while the mother of Fearnside's patient, dying at the age of 70, and two sisters, are said to have had the same difficulty in walking. Steinert met with a patient whose father and brother were affected with myotonia without atrophy. The sister of a case reported by Turney, and the mother of one of Batten and Gibb's patients, had difficulty in relaxing the grasp, while the father of Nogues and Sirol's patient is stated to have had some difficulty in chewing.

An acute illness preceding the onset of symptoms had been noted in a few instances, e.g. an acute arthritis of the knee-joint (Rossolimo), an attack of acute rheumatism (Buzzard), an infective fever of undetermined nature (Lorat-Jacob et Thaon), an attack of pneumonia (Fünnrohr; Mirallié; Jalaber et Culérre), ulceration of the cornea (Turney, obs. II.). Exposure to cold was blamed in one case. A trauma was followed by symptoms in two cases; thus Tetzner's patient first observed stiffness in the hands five weeks after a severe accident in which the clavicles and two ribs were broken, while a patient of Kennedy and Oberndorf's, six weeks after a fall on the right knee, found that the corresponding leg was weak. Curschmann and Jolly were inclined to think that occupation had played a part in determining the localisation of the atrophy in the cases which they described.

Symptoms.—The physiognomy presented by patients with myotonia atrophica is well figured by a number of authors (Nogues and Sirol; Rossolimo; Lannois; Batten and Gibb; Pemberton; Greenfield; Griffith; Tetzner).

The Distribution and Character of the Atrophy, as has been mentioned, varies. In the majority the face, limbs, and sternomastoids were involved, in a few the limbs were alone affected, while in some cases the wasting was limited to the arms and hands. Summarising the general distribution in the 65 cases referred to we find that the upper limbs were implicated in all but two (Batten and Gibb; Greenfield), the face in 45, and the lower limbs in 40.

The facial appearance is repeatedly likened to that seen in the Landouzy-Déjérine type of myopathy. In some cases the weakness, which affects especially the ocular and oral sphincters, was so extreme that the eyelids could not be opposed.

The sternomastoids were atrophied in 37 cases, in a number of which, as in our second patient, these muscles are said to have been absent or practically absent. In every instance in which
the sternomastoids were atrophied the face was also affected, while in one patient (Batten and Gibb) these parts were alone involved.

The upper limbs were implicated in all but 2 cases. The muscles of the shoulder-girdle comparatively seldom suffer. Atrophy of the trapezius was, however, noted in 11 cases, in 3 of which the lower two-thirds of the muscle was absent. Winging of the scapula, present in 4 cases, was, in all, as in our case, apparently due to paresis of the trapezius. The supra- and infra-spinati were markedly atrophic in 4 cases, the latissimus dorsi in 2. The upper arm muscles showed wasting in 25 cases, the deltoids being specially mentioned in 10. The atrophy was usually more pronounced in the triceps than in the biceps in those cases in which these muscles were involved. Wasting of the forearm musculature is very constant. These muscles appear, indeed, to have been more or less wasted in all but 11 cases, while in 14 they were the only muscles of the upper extremity to be affected. The atrophy, when pronounced, was, as a rule, more marked along the ulnar aspect of the limb, the flexors being usually weaker than the extensors. The intrinsic muscles of the hands presented some degree of atrophy in 15 cases.

The lower extremities were involved in 40 cases, in all of which, with the exception of Greenfield’s patient, the upper limbs also suffered. When the thighs were involved, as was the case in 16 instances, it was always the anterior muscles, and, as Batten and Gibb pointed out, especially the vasti, the rectus femoris commonly escaping. In two cases respectively the adductors and glutei were notably affected. The muscles below the knee were implicated in 34, while in 14 the atrophy in the lower extremities was limited to this region. In 15 the wasting was confined, or practically confined, to the anterior tibial group, while in 7 the peronei are especially referred to. Ten patients showed a steppage gait. Atrophy of the calves is seldom noted, and when present was always associated with a more pronounced change in the muscles on the front of the leg. In Steinert’s second case the muscles of the back were much wasted.

The masseters and temporalis were the seat of atrophy in 17 cases. Three instances of consequent dislocation of the jaw have been reported (Steinert, obs. I. and II.; Tetzner). Some defect of movement of the vocal cords was noted in cases referred to by Steinert (obs. I.); Batten and Gibb (obs. III.); Griffith; Bramwell and Addis (obs. II. and IV.). Ptosis was present in four instances.
Griffith's patient complained of diplopia at times. The pupils were unequal in 2 of our cases. Abolition of the light reflex has not been observed. Palatal weakness, though seldom specifically mentioned, would appear to be not uncommon, judging from the not infrequent reference to nasal speech. Nasal regurgitation was noted by Tetzner, and was also present in one of our cases. Cassirer's patient had some difficulty in swallowing. In two cases (Steinert, obs. IV.; Lortat-Jacob et Thaon) atrophy of the tongue is described, while in Griffith's patient the tongue felt flabby and could not be voluntarily hardened.

The degree of muscular atrophy varies greatly. In cases reported by Noguès et Sirol, Fürnrohr (obs. II.), Fuchs (obs. I.), and Steinert (obs. I.), the wasting was both extensive and pronounced. Again, in some cases in which the atrophy was quite localised it was also marked, while in others it was comparatively slight. Although sometimes unilateral, and this applies more particularly to cases in which it was limited in extent, the wasting was, in the great majority of cases, symmetrical. Fibrillary tremors are referred to in only 4 cases (Voss; Fuchs (2); Lortat-Jacob et Thaon). The electrical excitability in the wasted muscles presents merely quantitative changes. Two writers, however, refer to the presence of a true reaction of degeneration (Jolly; Pelizaeus), while a partial reaction of degeneration was met with by Schoenborn. A myasthenic reaction has also been observed (Steinert, obs. I.). Muscular hypertrophy, present in six cases, involved in one several muscles (Longard), while in others it occurred in the thenar eminence (Cassirer), the anterior tibial muscles (Batten and Gibb), the calves (Griffith), and the infra-spinati (Greenfield; Bramwell and Addis).

The Myotonia.—An essential symptom of the condition manifests itself in (a) a slow muscular relaxation after contraction, and (b) a slow, tonic, persistent contraction when the muscles are struck or stimulated electrically. Exceptionally the myotonia is widespread and pronounced, as in Griffith's case; far more often it is comparatively slight, and in many cases is only obvious to the patient in some slight difficulty in relaxing the grasp. Even in the absence of evidence of myotonia during movement the symptom cannot be excluded unless the characteristic response to mechanical and electrical stimulation is also absent. Embarrassment during mastication, in moving the tongue, and in opening the eyes in the morning was complained of by several patients in whom no objective muscular difficulty was observed beyond an inability to open
promptly the closed fist. The myotonic response to mechanical stimulation was very beautifully observed in the tongue in one of our patients. In this case, as in several of the reported instances, the myotonia was only noticeable in the early part of the day, while this patient also assured us that the stiffness was more pronounced when the hands were cold.

The reflexes show no constant alteration. Thus in 33 cases in which they are specially referred to, the knee-jerks are described as present or normal in 10, diminished in 9, absent in 8, and increased or brisk in 2, while in 4 cases one knee-jerk was absent. Among 16 cases in which the condition of the ankle-jerks is mentioned, it is noted that in 11 they were absent, in 1 diminished, and in 4 normal or exaggerated. The plantar reflex is always of the normal (flexor) type.

Sensory symptoms have been but seldom observed. Longard's patient complained of severe lightning pains in the legs, while in one of Batten and Gibb's cases (obs. I.) sharp shooting pains in the thighs and small of back were noted. There was extensive anaesthesia in the case described by Lortat-Jacob et Thaon, also in one of Hoffmann's cases, the last-named writer attributing the sensory disturbance in his patient to a complicating hysteria. The slight sensory defect on the outer surface of the thighs noted by Kleist may possibly have been due to meralgia paresthetica, which condition was also noted by Steinert in one case.

The frequency with which cold hands are complained of is somewhat striking (Pässler (2); Rossolimo; Steinert (obs. IV.); Fearnside; Tetzner; Bramwell and Addis (obs. IV.)). Pronounced dermographia was seen by Steinert (obs. III.) in one case. The same author remarks on the frequency with which premature baldness occurs; it was a well-marked feature in four of his cases. Two of our patients presented a remarkably poor development of facial hair. Congenital cataract, as pointed out by Greenfield, may occur in families suffering from myotonia atrophica; this observer met with four instances in the family which he reports. In Fearnside's case cataract was also present, two sisters of this patient suffering from the same condition, while Hirschfeld's patient, a glass-blower by occupation, is said to have suffered from glass-blower's cataract. Congenital deformities are occasionally referred to. Thus Voss has noted a congenital defect of the abdominal muscles; in 2 cases a malformation of the ear is described, while in 2 of our patients the palatal arch was unusually high. Steinert was the first to indicate that atrophy
of the testes is not uncommon. Nine cases have now been reported in which this observation has been made (Pässler (2); Fürnrohr (2); Gaupp; Steinert (2); Pemberton; Bramwell and Addis). Polyuria was present in one of Steinert's cases, while in the case reported by Griffith the pulse-rate was unusually slow.

**Morbid Anatomy.**—Our knowledge of the pathological anatomy of myotonia atrophica is based, firstly, on the examination of excised pieces of muscle in cases reported by Schoenborn, Frohmann, Rossolimo, Lannois, Siemerling, and Griffith; and, secondly, upon a complete post-mortem examination made by Steinert.

Schoenborn found an increase in the size of the muscle fibres in the deltoid exactly similar to that met with in Thomsen's disease, while in the tibialis anticus the fibres were atrophied and there was an increase of the nuclei and connective tissue which, in his opinion, resembled more closely the myopathic than the myelopathic atrophies. Frohmann described both hypertrophied and atrophied fibres with nuclear proliferation. Lannois met with a lesion in the muscles which he says resembled in every detail the classical picture of the myopathies. Siemerling noted marked hypertrophy of muscle fibres with increase of interstitial tissue. Rossolimo was struck with the great variation in the size of the fibres; he, too, remarks on the great nuclear proliferation and increase of interstitial tissue. Griffith observed a considerable increase in size of many of the muscle fibres, with marked increase of the nuclei within the muscle; there was, however, no increase of the interstitial tissue, either fibrous or fatty, in the portions of muscle he examined. Steinert's observation is of great importance, since the case which he records is the solitary instance in which a post mortem has been made. The patient was a man of 44 in whom there was extensive atrophy, most pronounced in the muscles of the thumb, face and jaw, and the sternomastoids. The outstanding abnormal feature was the widespread cirrhosis of the muscles; in addition there was, however, a tabetic-like degeneration of the posterior columns, which the author is inclined to think may not be an accidental complication, but an integral feature of the disease.

**Relation to Thomsen's Disease and Myopathy.**—There is a very close resemblance between myotonia atrophica and Thomsen's disease. The symptom myotonia is almost peculiar to these two conditions; in both there is a tendency for several members of
a family to be affected, while there is reason for believing that myotonia atrophica may, like Thomsen's disease, be transmitted from one generation to another. Further, cases of myotonia atrophica have been reported in which other members of the family have suffered from myotonia without atrophy. Again, the atrophy closely resembles in its features that met with in the muscular dystrophies, although it does not correspond in its distribution to any recognised type of myopathy. The writers are inclined to agree with Steinert, who regards myotonia atrophica as a type of Thomsen's disease.

Prognosis and Treatment.—Time must elapse before any exact expression of opinion is possible as to prognosis, although existing data would seem to show that the condition in itself has little tendency to shorten life. Since atrophy of the testes has been observed in quite a number of cases, testicular extract may possibly be found to be of value.

The writers' acknowledgments and thanks are due to Dr. S. A. K. Wilson, who pointed out to them the nature of the condition in the case of L. L., the first example of the disease which they had personally observed.

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THE SURGEONS AND BARBERS OF EDINBURGH:
THEIR SEPARATION IN 1722.

By CLARENDON HYDE CRESWELL.

With reference to the very interesting "Note on the Incorporation of Surgeons and Barbers," by Mr. R. Scott Moncrieff, which appeared in your issue of December last, may I be permitted to add a few supplementary remarks which may throw further light upon the relations that existed between these anciently-constituted bodies both immediately before and after their separation in 1722.

While a judicious discrimination between loquacity and undue brevity is to be commended, yet it cannot be supposed that the details of this obscure subject are so well known that a knowledge of them, however superficial, can be taken for granted. To be too brief might give rise to misconception, and therefore it is prudent to unravel knots difficult of solution with more care and deliberation than is usually bestowed upon subjects with which the general public are better acquainted.

Mr. Scott Moncrieff in his opening remarks observes, "It must not, however, be concluded from the fact of their official incorporation [the barbers and surgeons] as one body in 1505 that the two crafts came together for the first time in that year, for the deed [the Seal of Cause] shows that for at any rate some time prior to that date they had been closely associated together in the upkeep of ‘an altar situate within your College Kirk of St. Geill in the Honour of God and St. Mungo our Patron.’"