

Original Articles.

ON MYOTONIA.¹

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The disease, first known through the description of the Silesian physician, Dr. Thomsen, and since then called after him, is an affection scientifically so remarkable and interesting that I may be pardoned for again bringing this subject before you. Nor would I do so now, were it simply for the purpose of recording another case of Thomsen's disease (myotonia congenita); for, while the affection must be one of infrequent occurrence, scarcely more than 50 cases having been published, yet all of these cases resemble each other to such an extent, that one typical case may fittingly be looked upon as a paradigm of the rest.

The subject must, however, to-day be looked upon from a broader point of view than that of mere casuistics.

The history and literature of Thomsen's disease up to 1886 will be found in Erb's monograph, that from 1886 to 1889 in his article in the *Deutsches Archiv*, and that from 1889 to 1894 in the article by Suesskand, published in the *Zeitschrift für klinische Medizin*, vol. xxv. The only case of unusual interest published since then is the one with autopsy by Dejerine and Sottas, and to this I shall have occasion to refer again.

Our knowledge of this affection is now so well defined that the term Thomsen's disease, or myotonia congenita, embraces a clearly circumscribed clinical entity,

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and constitutes a picture to which nothing can possibly be added; nevertheless, it seems to me that the time has come when we must limit this name to the actually congenital cases, and not expand it to include the acquired ones, or, possibly, even such as have the myotonic disorder and myotonic reaction, but in other respects differ to a greater or less extent from cases of Thomsen's disease. In view of the cases which I herewith publish, I think it proper to assign all such cases to the one clinical category of myotonia, and to then subdivide this class into various groups.

If we analyze the well-known picture of the disease, we find it made up of the following components:

1. *The Etiology.* Here the most important factor is, beyond a doubt, heredity, either as a direct transfer from the ascendant, or only indirectly by inherited disposition; atavistically, as in Weichmann's case, or where collateral branches are affected, as in the cases of Knud Pontopidan and Bernhardt; or the disease may occur as a family type, without direct heredity.

2. *The myotonic disorder of movement;* i. e., the occurrence of tension, stiffness and tonic spasm in the voluntary muscles at the beginning of intended movements.

3. *The myotonic reaction,* which is made up of normal mechanical, faradic and galvanic excitability of the motor nerves, and an increased mechanical, faradic and galvanic excitability of the muscles. Here with the galvanic current only closure contractions are obtainable, and these are as strong with the anode as with the kathode; the contractions are always slow, tonic and prolonged.

In many muscles strong faradic currents produce irregular undulating contractions and stabile galvanic currents, rhythmical contraction waves which follow one upon the other.

3. *Hypertrophy of the muscles.*

4. *Absence of all symptoms pointing to gross involvement of the nervous system.*

Of the published cases of myotonia congenita, the large majority correspond absolutely to these requirements. Cases which show symptoms of organic disease of the central nervous system, with myotonic disorder, but without myotonic reaction, such as Dana's, can at once be excluded from the group of myotonias. A large minority, however, show certain smaller deviations from the typical picture. Aside from variations in intensity and extensity of the disease, all or only some muscles being severely or slightly affected, the most frequent deviation consists in the affection being neither hereditary nor congenital. Such cases have been described by Seligmüller, Peters, Weichmann, Rieder, Vigoroux, G. Fischer, Erb and Suesskand.

The following history is that of a typical acquired case:

Case I.—W. J. G., of Wheeling, W. Va., was referred to me by Dr. J. Schwinn of that city, with the diagnosis Thomsen's disease, and was presented at the New York Neurological Society on Nov. 2d, 1897, as a typical case of this disease.

Patient is 28 years of age, was born in Ireland, and came to the United States in 1884. He is a railroad brakeman by occupation.

Family History.—His grandparents lived to an old age. His father died at the age of 70, cause unknown; his mother is living and well at the age of 69. She had nine children, of which two are dead. The others, five sisters and one brother, are healthy.

The patient himself was rather delicate up to his 14th year; otherwise, with the exception of an attack of measles, and one of whooping cough, he was perfectly well. He never noticed any difference between himself and his schoolmates in regard to physical strength, and was able to participate in all out-of-door sports and games. Psychically, he says he was rather timid, and perhaps slightly backward. At the age of 18 he had an attack of typhoid fever without complications, which kept him in the house for five weeks. When he went out for the first time, he experienced a severe painless cramp in the calves of both legs, more marked on the right side. This cramp lasted about two minutes. During the next two weeks it recurred several times each day, and his legs felt somewhat weak. He then complained of feeble sexual powers with frequent nocturnal emissions, but was well and gained rapidly in weight

and strength. During this time his muscles increased in size, so that they seemed to him unusually large, and now that he is questioned, he thinks he was somewhat stiff during all this time.

About two years after this attack of typhoid, he began to complain of stiffness of his legs, especially noticeable when climbing a ladder, or stepping into a car. He also noticed that after a prolonged rest the stiffness in his muscles was always decidedly increased, and that after the first few moments the stiffness gradually disappeared and he again had full use of his limbs. At first only the muscles of his legs were affected, and these slightly, then gradually nearly all of the voluntary muscles became involved.

During the time of this progression, and before his arms were seriously affected, his legs became so bad that in 1893, when he jumped from a car, the muscles would cramp up so that he would fall; then he could get up only by using his hands as a support. If in walking or running he stubbed his toe, his leg would stiffen so that he could not lift his foot from the ground. It would, as he expresses it, "stick to the earth," and then he would fall. Since a year he has complained of a similar condition of stiffness in his hands, so that when turning the wheel of the car brake, it would require a long time before he could loosen his grip and straighten his fingers. This was accompanied by a feeling of tension and stiffness in the arm; also when his arm was forcibly extended he could not bend it again until the spasm ceased.

Recently the neck muscles, the face muscles and tongue have become affected, though not to so marked a degree. He also feels a "stiffness" when he closes his eyes, and at times he feels his right eye "catch" when he turns his eye outward. He has no pains and feels perfectly well. The trouble is worse at certain times than at others; some days he seems all right, but he does not think that the weather has any influence upon his condition. He has often wondered at the fact that though his muscles seemed to increase a great deal in size, his strength was not correspondingly great, in fact, he thinks he has become weaker.

Status.—Medium height. Panniculus adiposus poorly developed, in contrast to the muscular development which is extreme, giving the man the appearance of an athlete. Though all the muscles show an exceedingly strong development, the muscles of the legs, forearms and arms are especially large. The calves, when contracted, measure 42 cm. in circumference at their largest part.

The accompanying photograph gives a good illustration of their appearance. The gross power of the muscles is feeble in comparison to their size. Aside from the muscular disorder,

no symptom of disease of any organ can be discovered. Sensory disturbances are absent; the superficial reflexes are normal. Triceps and Achilles tendon reflexes are not obtainable. The knee-jerk is very well marked, in the beginning even exaggerated, but becomes exhausted after repeated blows, so that at the end of a prolonged examination it is no more

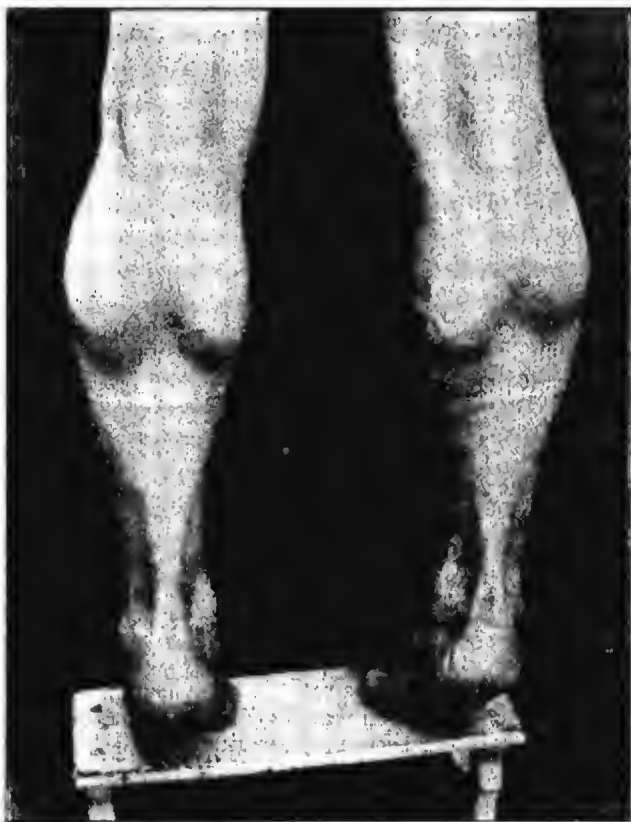


FIG. I.

obtainable, reappearing, however, in its increased state after a rest of half an hour or more.

Very pronounced is the myotonic disorder of all the muscles. When the lids are firmly pressed together, they cannot be opened at once, although he makes great effort to do so. Only after a time is he able to open his eyes. The spasm in

these, as well as in all other muscles, lasts from 15 to 25 seconds.

The internus of the right eye seems somewhat affected, inasmuch as the eyeball cannot at times be brought to the inner canthus or at times having been brought to that position, cannot promptly be returned to its normal position of rest.

The pupils react promptly to light and accommodation, the visual field is not restricted and the fundus is normal.

This condition of spasm is present in the tongue and in the muscles of mastication.

In the tongue, however, we find that the spasm occurs only when the intrinsic muscles are exerted, but that the genio-glossus is free, inasmuch as no spasm occurs in protrusion of the tongue. So also the disorder of the masticatory muscles is limited to the masseters, while the pterygoids (lateral movements) are free. The masseters stand out hard and rigid like bars of iron, when the teeth are forcibly closed. The spasm, contraction and after-duration, is also present in the muscles of the face, but it is not so marked as elsewhere.

In the movements of the arms, hands, and legs, this myotonic disorder is most apparent. This is particularly noticeable in the hands when any object is tightly grasped, and in the legs in going up stairs or in jumping from a low stool.

The thorax muscles are implicated with excessive movements; thus when he expands his chest to its maximum capacity, he is not at once able to empty it, but must allow several seconds to elapse. The muscles when uncontracted convey no different sensation to the examiner's hand than does a normal muscle.

Mechanical Excitability.—The mechanical excitability of the trunks of the motor nerves (n. facialis, plexus brachialis, n. ulnaris, n. peroneus) is not increased.

Mechanical Excitation of the Muscles.—All the muscles showed the formation of the marked lasting furrows, upon excitation with the percussion hammer. When stronger blows were employed, an idiomuscular mound was formed in certain muscles (biceps, pectoralis) which remained so long and was of such consistency, that it could be distinctly palpated and manipulated. The total contractions of the muscles showed no after-duration, were very marked upon the first blow, then with each succeeding blow grew less and less, until finally they could no longer be obtained. Even when they were thus lost, the local and fibrillar contractions were obtainable to their maximum extent.

The electrical examination showed normal faradic and galvanic excitability of the motor nerves. The muscles with medium faradic currents showed slow, lazy, lasting contractions (20 to 25 seconds). Single opening shocks, no matter how strong, produced only short quick contractions.

The muscles showed certain quantitative and qualitative changes to the galvanic current. In nearly all of the muscles a contraction could be obtained with less current than is normally the case; for instance, in the triceps a Ka. C. C. is produced with $\frac{1}{2}$ M. A. of current. An C. C. is obtained with about the same amount of current as Ka. C. C., or in some muscles with a very little more. Even with minimum currents the An. C. C. shows a certain amount of slowness; as soon as the current is increased, both contractions, Ka. C. C. and An. C. C. show marked slowness, tonicity and after-duration.

In this patient I was also, after various attempts, able, during the passage of a strong steady galvanic current, to obtain the peculiar rhythmical contraction waves as described by Erb; a large flat electrode being fastened to the back of the shoulder, the other electrode placed in the palm of the hand or upon the flexors of the forearm, and the current of at least 20 M. A. closed. The primary result is a lasting tonic contraction of all the muscles of the arm. Then after about half a minute, slow wavy contractions set up and pass from the Ka. to the An, one wave following another at intervals of 1 to 2 seconds. In the lower extremities I was also able to obtain these waves, but could not satisfy myself of their direction from the Ka. to the An. These waves were not always demonstrable. Their production always required strong currents and a great deal of manipulation of current and electrodes.

Examination of the blood showed nothing abnormal. Hæmoglobin, 80 per cent.; red blood corpuscles, 4,360,000; white blood corpuscles, 7,450.

For purposes of microscopical examination two pieces of muscle were excised from the left quadriceps, and one piece from the left biceps. These pieces were kindly prepared for me by Dr. F. Schwyzer.

Immediately after excision each piece of muscle, in order so far as possible to prevent permanent shortening, was fully extended and thus fixed by means of a skewer, both ends of which were pointed. Each end of the muscle piece was slipped over the corresponding end of the skewer, and thus extended, the pieces were placed for hardening in a mixture consisting of formalin 1 part to 4 parts of physiological salt solution; they were then transferred to alcohol, finally embedded in celloidin, cut and stained. When the pieces were transferred from the formalin to the alcohol, it was noted that one of them had torn away from its fastenings and was very much contracted, while the others remained fastened and extended. This accident, as will be seen later, proved to be rather fortunate. The specimens were stained with iron hæmatoxylin (nuclear stains), and after preparation were compared with specimens taken from normal muscle and with others from myotonia congenita.

A careful examination of all the specimens showed that we had two distinct pictures before us, both of the transverse as well as of the longitudinal sections.

In the one transverse picture (Fig. II.) we found muscle fibres more or less polygonal in shape, the nuclei in a few of the fibres somewhat increased in number, and the interstitial tissue also somewhat augmented. The diameters of the fibres in these sections were as follows: In 100 fibres, the smallest fibre meas-

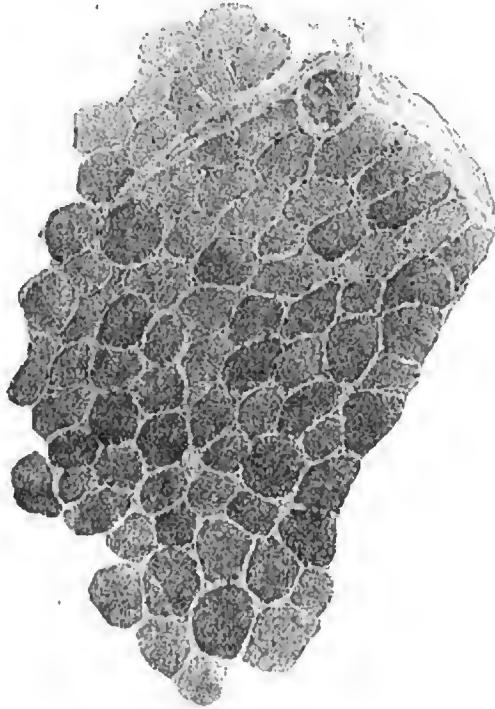


FIG. II.

Transverse section of extended muscle. (Oc. 4., Obj. A. Zeiss.)

ured 60 microns; the largest, 100 microns. Of these 100 fibres, 90 per cent. measured less than 90 microns, and only 10 per cent. were found to measure between 90 and 100.

In the second series (Fig. III.) of transverse pictures we find the single fibres mostly with rounded edges, the nuclei and interstitial tissue decidedly increased, and almost all of the fibres very much larger than in the other specimens. Meas-

urements here, again of 100 fibres, showed the smallest to measure 75 microns; the largest, 195 microns in diameter. 33 per cent. measured between 75 and 100; 60 per cent. between 100 and 150 microns, and 71 per cent. between 150 and 195 microns. The longitudinal sections again revealed the same variations in different sections, some showing the same characteristics as those of the second series of transverse sections, others not.

In addition it was seen that in the one set all the muscle

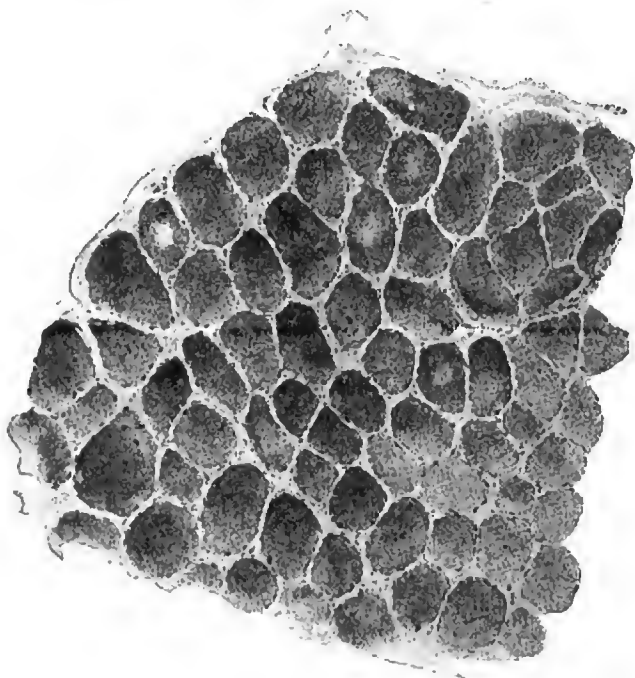


FIG. III.

Transverse section of non-extended muscle. (Oc. 4., Obj. A. Zeiss).

fibres were parallel, straight, with linear contours, while in the other there was no such regularity; the fibres not being straight and parallel, but convoluted. Furthermore, the transverse striation was here indistinct and delicate, the entire fibre being more homogeneous, and in some of the fibres only a longitudinal striation being discernable.

In short, our two sets of specimens show sections which in the one set closely approximate those from normal muscle,

while in the second they correspond fully to what has heretofore been looked upon as characteristic of Thomsen's disease.

Inasmuch as all the specimens were taken from the same individual, at the same time, and treated in precisely the same manner, this difference must be due to the accidental breaking away from its fastenings of the one piece of muscle; in other words, that piece of muscle which was allowed to contract to its limit showed the characteristic changes of Thomsen's disease, while the other not entirely contracted pieces gave sections which appeared almost like normal muscle. To this we shall refer again.

This case is as typical a case of Thomsen's disease as can well be found. The myotonic disorder, the myotonic reaction, the hypertrophy of the muscles, with their disproportionate weakness, all make the diagnosis unmistakable. Yet the absence of heredity, and especially the occurrence of the affection after an attack of typhoid fever at the age of 18, render it imperative to make use of some other designation than that of myotonia congenita.

The hypertrophy of the muscles coming on, as it did, at the same time as the myotonic disorder, and not having preceded it, shows the entire symptom complex to have been an acquired one. There is not the slightest ground for the assumption that any symptom of the disease existed prior to the attack of typhoid.

The result of the microscopical examination of the muscles is also of great interest, and it must cast a doubt upon the changes thus far described by myself and others as being pathognomonic of Thomsen's disease.

Another acquired case, but one which deviates very much from the congenital type, is the following.

I will curtail the history as much as possible.

D. L., male, 40 years of age, pianist by occupation, consulted me first about eight years ago. The family history is unimportant. Patient himself was perfectly well until his 26th year, when he met with an accident, slipping and falling. He at that time fell forward and struck flat upon his palms, over-extending both hands. This was followed by considerable

pain and some swelling in both wrist joints, but in about 10 days he had apparently entirely recovered, so that he again began to practice upon the piano, and, as was his custom, played from 6 to 10 hours daily. He then, after several days of such practicing, overstretched his right hand; this was immediately followed by a painless abductor cramp of the hand muscles, so that he could not bring his fingers together until he rubbed and warmed them, when the cramp passed away.

In the course of the following six months such cramps occurred whenever he forcibly stretched his fingers, i. e., abducted them, the left hand becoming similarly affected. He gave up piano playing, but without beneficial result, for the trouble grew constantly worse. Soon the cramps affected other muscular groups, finally involving all the muscles of the upper extremities, and occurring upon any forced movement.

The spasm is especially noticeable when he attempts to grasp any object tightly; then his hand clings to the grasped object, so that he cannot for the time being relinquish it.

He is worse in winter than in summer; cold water increases the liability to spasm, and he, therefore, always washes his hands in warm water. He has never complained of his legs troubling him.

Examination showed patient to be of strong and healthy appearance. With the exception of the muscular ones, no disorders of any kind are discoverable. The muscles of his arms, shoulders and neck are almost athletic in their development; the legs also are unusually large, but do not attract attention in the same measure as do his arms. The patellar tendon reflexes are exaggerated; the mechanical excitability of the muscles of both thighs is remarkably increased, a slight blow producing a quick contraction of the entire irritated muscle; tonicity is not present. Electrical reactions are normal.

The arms, in comparison to their development, are very weak; extension and flexion of the forearm upon the upper arm can very easily be prevented by comparatively slight opposition. A dynamometer, which under pressure by the average man indicates 150 for the left and 200 for the right hand, registers only 60 and 80, respectively, upon maximum pressure by the patient. Myotonic motor disorder is found to exist in all the muscles of the hands, arms and shoulders. In these same muscles, as well as the chest and neck muscles, the myotonic reaction to mechanical and electrical excitation was plainly demonstrable. The nerves (plexus brachialis, n. accessorius and n. ulnaris) show normal reactions to both forms of excitation. Rhythmical contraction waves were never obtainable.

A few months ago I sought an opportunity of again examining this patient; he was then in a condition similar to that just described, but it was evident that the disease had made some progress. His entire upper body, with the exception of the abdominal and faeial muscles, shows the myotonic disorders. So long as patient is careful to use his hands and arms only for ordinary movements, he has no trouble of any kind, but every sudden, violent exertion, or any occupation necessitating a quick maximum contraction of any of the above muscles or museular groups, as well as sudden exposure to cold, brings on a spasm which is myotonic in character.

The myotonic reaction is present at all times.

His legs, with the exception of increased knee-jerks and hyperexcitability of the muscles to mechanical excitation, show no disorder.

Notwithstanding that this case differs materially from the typical picture of Thomsen's disease, in coming on after injury and overstrain in a man of 26, who had previously been in good health, and in affecting only the upper part of the body, and leaving the abdomen and legs entirely free, it must, nevertheless, be classed as a case of myotonia.

Whether or not it may be looked upon as a variety of Thomsen's disease is a question which is of no import, yet it is, aside from its acquisition, not any more atypical than the case of Martius and Hanseemann, which Erb considers as such a variety, notwithstanding that the disorder occurred only temporarily under the influence of cold, was limited to the upper extremities, and showed no myotonic reaction during the free intervals.

It is to this class of cases that the term myotonia acquisita should be allotted, and, in my estimation, this term should be restricted in its use to cover only such cases. The term myotonia now describes a special form of spasm, characterized, as we have seen, by the myotonic motor disorder and the myotonic reaction; for all cases not showing at least these phenomena it would be better to make use of some other designation.

Thus Talma and Fürstner have published cases of

"myotonia acquisita" which certainly have nothing at all in common with the congenital form of the disease, as the name would seem to indicate. Talma's cases represent a series, showing an acquired tendency to spasm, occurring chiefly upon intended movements, the muscles involved, on account of their hyperexcitability, being easily thrown into a state of tonic spasm by various irritants. These spasms, however, increase in intensity the longer the muscles are used, while the reaction, which Talma describes as being similar to the myotonic reaction, shows upon careful interpretation nothing more than an abnormal hyperexcitability to mechanical and electrical irritants. In Fürstner's case, also, the myotonic reaction was not present, mechanical and electrical examination producing effects which were not totally dissimilar to this reaction.

It would, I believe, be better to designate all cases which show simply tonic spasm accompanying or following active movements, when unaccompanied by the other myotonic signs, by the term which Seligmüller has suggested, namely, *intention spasm* (a spasm occurring upon intended movements).

Such intention spasms have been described in connection with a variety of disorders, some having yet other symptoms in common with myotonia congenita, others admitting of no comparison at all with this disease. Such descriptions show that intention spasms may occur in disease of any part of the muscular and nervous system, and are especially often found in hysteria, tetany and occupation neuroses.

A case of this nature, but which showed so great similarity with the myotonic disorder and reaction that I am doubtful as to whether it is not, after all, a "myotonic condition," is the following: Perhaps it would be well to speak of such cases under the name of *myotonia transitoria*. This patient, L. G., was presented at the New York Neurological Society in May, 1892:

Cigarmaker; 34 years of age; Russian by birth. The family history, so far as obtainable, is negative, the father dying at 49 of some acute disease; the mother is living and healthy. The five brothers and sisters are healthy. Patient himself was always well, with the exception of a chancre in 1881. No secondary symptoms. He has been married five years; his wife has had three healthy children and no miscarriages.

About the end of February, 1892, spasms set in in the fingers of both hands; especially affected were the index and middle fingers. The spasm was limited to the flexor muscles of the fingers, and occurred only upon active movement, never spontaneously. Thus, so long as he kept his fingers open, there was no trouble, but as soon as he closed his fingers upon any object, a spasm ensued in the flexors, which prevented him from releasing the object grasped. The trouble gradually increased in extent, soon also involving the wrist.

He has never complained of pain. He comes for treatment because the trouble incapacitates him for work. He is able to do all the work which can be done with extended fingers, such as rolling the cigars on a flat surface, but when it is a question of finishing the point by turning with his fingers, he cannot let go of the cigar on account of the flexor spasm.

Examination, May 1st, 1892.—Muscular system not abnormally developed. The muscles of the hands, forearms and shoulders are, however, large and well formed, while the other muscles of the body are more flabby and smaller. The internal organs are normal. No fever. Urine contains neither sugar nor albumin. Pupils equal; react promptly. Ocular movements free; facial muscles unaffected. Tongue shows no deviation. Smell, taste, hearing, normal.

Upon intended movement the muscles of the hand, when these are closed forcibly, enter into a state of tonic spasm, so that the hand cannot be opened for a number of seconds. This intention spasm, in addition to the flexors of the hand and fingers, involves the adductors and abductors of the thumb and fingers, so that when the fingers are forcibly spread or voluntarily pressed together, they remain in either of these positions without being under control of the will.

These flexion spasms are most marked in the thumbs and 4th and 5th fingers, the 2d and 3d not being so much affected. The adductors and abductors of the thumbs and the deltoids show the following disorder:

Mechanical Excitation.—Quick, sharp blows by means of a percussion hammer produce marked contraction of the entire muscle (except in the deltoid, where only fibrillar contraction is produced). This contraction is slow and tonic in its formation and duration, lasting from 15 to 20 seconds. The same lasting contractions may be obtained by pressing or roll-

ing the muscles under any hard object. The mechanical excitation of the nerves is unchanged.

Electrical excitation with the faradic current showed, with strong currents, slowness and tonicity of the contraction, with an after-duration of 15 to 20 seconds.

With the galvanic current there was found in the same muscles an increased excitability, they reacting to very small currents, and tonicity, with after-duration, being produced by stronger currents (6 M. A.). The first tonic contractions were always obtained with the An. C., and it then required 1 to 2 M. A. more current to obtain a tonic Ka. C. C., while the relationship of the normal quick contractions obtained with minimal currents remained unchanged (Ka. C. C. > Aa. C. C.)

At that time no other muscles showed any changes whatsoever, functional, mechanical or electrical.

Three weeks later the biceps of each arm was found to present the same subjective and objective disorder as the other muscles. This condition lasted unchanged until September, 1892, when it gradually improved functionally, the My. R., however, persisting to the same extent.

In December patient claimed that he was perfectly well, and had been working at his trade uninterruptedly since two weeks. Examination failed to show any functional disturbances, and the muscles reacted normally to mechanical and electrical stimuli.

If we thus apply the term myotonia only to such cases as present the myotonic motor disorder and the myotonic reaction, and relegate all other spasms coming on upon voluntary movements to the category of intention spasms, our classification of myotonia congenita, myotonia acquisita and myotonia transitoria will at once make the class spoken of clear. The diagnosis of myotonia can present no difficulty, and its differentiation from tetany, pseudo-hypertrophy, spastic spinal paralysis, and even from Eulenburg's paramyotonia, need hardly be dwelt upon. In the latter affection not only are the specific myotonic reactions absent, but the muscular stiffness occurs only under the influence of cold, and not in consequence of muscular action.

The result of the microscopical examination of the muscle from Case 1 makes a reconsideration of the pathogeny of Thomsen's disease interesting.

Three theories regarding the nature of the disease, the psychopathic, the neuropathic and the myopathic, have each found adherents.

Against the psychopathic theory stands the entire weight of clinical evidence, which need not again be reviewed.

The myopathic theory, on the other hand, has had a great deal in its favor, especially the proofs deduced from anatomical findings. These are:

1. The hypertrophy and rounding off of the muscular fibres.
2. The increase of sarcolemma nuclei.
3. The diffuseness, indistinctness, and even loss of the transverse striation, and,
4. The minute changes which I have described as consisting in a massing together of the sarcous elements, and a correspondingly coarse appearance in some parts of the muscle, while in others these elements are minute, scarcely perceptible and widely separated from each other.

These anatomical findings are unfortunately all based upon the examination of pieces of muscle excised from the living body. The only autopsy recorded is the one described by Dejerine and Sottas,¹ which, while furnishing satisfactory proof to the reporters themselves of the purely myopathic nature of the disease, must impress every one else as being useless for purposes of argument.

Certainly many of the changes described in this report may be due to serous infiltration of the connective tissue and to secondary changes in the muscular parenchyma, thereby nutritionally produced. Furthermore, the cerebrum and cerebellum were not carefully examined, so that, even allowing that the muscular disintegration be not due to serous infiltration, its primary nature still re-

¹ This patient died of an acute nephritis with uræmic symptoms, and, aside from the muscular changes, the autopsy showed a marked serous infiltration of the entire body, hydrothorax, hydropericard and œdema of the lungs.

mains unproven. This autopsy, therefore, being unavailable as proof in support of the myopathic nature of the disease, we are again confined to that proof which may be derived from pieces of muscle.

The value of this proof has been seriously affected by the investigations of Oppenheim and Siemerling concerning the influence of excision upon pieces of muscle taken from the living body. Text books on histology, and different articles upon various topics, give the measurements of muscular fibres taken from the corpse as varying from 20 to 70 microns. The average fibre thus obtained measures 50 microns. Oppenheim and Siemerling found the following measurements in muscles excised during life:

1. Normal case; average, 69 microns; maximum, 106 microns.
2. Normal case; average, 93 microns; maximum, 121 microns.
3. Hysteria; average, 74 microns; maximum, 146 microns.
4. Hysteria; average, 69-93 microns; maximum, 140 microns.
5. Traumatic neurosis; average, 93 microns; maximum, 140 microns.

Inasmuch as the irritation due to excision causes considerable contraction of the excised pieces, as a result of which the primary fibres become shortened and correspondingly broadened, Oppenheim and Siemerling excised pieces of muscle from the adductor femoris of a living rabbit (a) simply; (b) after preliminary fixation upon a staff, so that they could not contract; (c) stretched upon a staff. The fibres of class a were all round, and measured from 46 to 99 microns in diameter; those of class b were all polygonal, and had a diameter of 22 to 66 microns, while those of class c measured from 9 to 33 microns. In addition to this preponderance of size and change in form of the simply excised fibres, they also revealed the presence of a greater number of nuclei.

The description of the extended and non-extended fibres from my case, therefore, conclusively proves that these observations may be directly applied to the human body, simply recognizing that my fibres were taken from a case in which the mechanical contractility of the muscles was very much augmented, and, therefore, the figures obtained must be higher than those obtained through similar excision of normal muscles.

Finally, we must draw the conclusion that the muscular changes hitherto considered pathognomonic of Thomsen's disease are merely the result of mechanical excitation of the muscles, and simply corroborate the clinical fact of their hypercontractility.

Admitting, however, the fact that the muscular changes found appear to be of a secondary origin, the existence of a functional disorder which allows the muscles to overact in consequence of excitation, cannot be denied. Whether this functional disorder lies primarily in the muscular or central nervous system is a question which cannot yet be answered.

Interesting, and, perhaps, casting some light upon the pathogenesis of this peculiar affection, is its occurrence in the case of W. G., after an attack of typhoid fever.

We know that many of the gross disorders of the nervous system are of post-infectious nature, thus it must be to-day acknowledged that multiple sclerosis is in very many instances the result of an antecedent acute infection.

That severe infectious diseases, as typhoid and diphtheria, through the toxins produced by their microbes, deleteriously influence the very impressionable nervous system is undoubted; and it has even been shown by Babès that the microbes themselves may migrate into the spinal cord and into the nerve cells, without causing any local lesion.

After the elimination of such toxins or microbes, the entire organism apparently again returns to its normal state, but who can say whether cells so acted upon are

not functionally altered. Certainly it has long been known that the nerve cells of adults who have passed through many sicknesses are not entirely normal, and the changes found have been fully described by Babès; changes which in healthy small children are never found. On the other hand, the nerve cells of adults often show no such changes. This fact can only be explained by the assumption of an inherent weakness in the nerve cells of certain individuals, while those of others possess more power of resistance.

There can be no objection to the statement that such an inherent weakness may occur hereditarily in many members of a family; just as whole families show an hereditary weakness of brain cells, and become neurasthenics. But this disposition in the majority of cases lies dormant until stirred up by some accidental cause. In the histories of muscular dystrophies the precedence of such an accidental cause in the shape of an attack of measles, scarlet, typhoid is not unusual, and it is not improbable that in the affection under consideration such a productive cause has in many instances also been at work.

It, therefore, seems to me permissible to look upon the disease as due to an embryonal developmental disorder of the nerve cells, consisting in the more or less diminished resistance of these cells to the influence of certain toxic processes, and that these intoxications then are in such predisposed individuals the direct producers of the disease.

Certainly, our knowledge of the pathogeny of this disease must remain obscure until a careful cytological examination of both the brain and spinal cord has been made.

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DISCUSSION.

Dr. Graeme M. Hammond presented three children, two girls and a boy, and all members of the same family, as examples of myotonia congenita. Two other children in the family showed no evidence of this disease, and were apparently normal in all respects. In the three cases shown the disease did not manifest itself until the eighth year of life; previous to that age, the children appeared to be like others, excepting that they were very dull, and made little progress at school; for example, the boy, who is now fourteen years of age, cannot spell words of even four letters. All three children had a peculiar facial expression; they looked depressed, and were subject to violent crying spells at the slightest provocation. One of the girls had had chorea, and subsequently developed the myotonic disorder. The family history was unimportant, and contained no other instances of this disease. All of the children had had chills and fever, and one of the girls still has these attacks occasionally.

The disease in the three cases was entirely confined to the hands and arms. The rigidity of the muscles was great, and the reflexes were sluggish. The electrical reactions were normal, except that tonic contractions were produced, the same as if the muscles had been struck. No muscular hypertrophy was noticed. The palms were well developed, but the strength of the arms and hands was much less than it should have been. The legs were not at all involved, but the knee-jerks in all three of the cases were almost entirely absent. The peculiar facial expression, Dr. Hammond said, was not due to rigidity of the facial muscles. No difficulty was experienced in moving the jaws, and no spasm of the ocular muscles existed. The hand could readily grasp an object, but relaxed with difficulty. The mental deficiency in these cases seemed to indicate some degenerative cerebral change, perhaps in the pyramidal cells of the cortex. Three cases occurring in one family also pointed to some congenital influence.

In conclusion, Dr. Hammond said he did not present these as typical cases of Thomsen's disease, but simply as cases of myotonia congenita.

Dr. Theodore Diller said he had had an opportunity to examine the first patient referred to in Dr. Jacoby's paper, and in that case the symptom complex was even more striking than in the three children shown by Dr. Hammond. In addition to the other symptoms, a distinct spasm of the extrinsic ocular muscles was produced by movement of the orbicularis palpebrarum. The patient could close his eyes tightly, but could open them only slowly, and with a distinct effort. All the reactions which Dr. Jacoby mentioned were

present. The wave of contraction, which various writers have referred to, could not be elicited. Dr. Diller said the case did not impress him as being of psychical origin.

Dr. J. J. Putnam inquired whether there was an abnormal shortening of the muscular belly, as occurs in muscular dystrophy.

Dr. Jacoby replied that such a change in the muscles had not been observed.

Dr. W. G. Spiller said he knew of two or three cases reported in the literature, in which intention spasm was present. Two were cases of syringomyelia, and the third, one of brain tumor. He had been permitted to examine the specimens in the only case of Thomsen's disease in which a necropsy had been obtained—that of Dejerine and Sottas. The lesions in the muscles resembled those described by Dr. Jacoby. The speaker said that from our present knowledge of the histological conditions, myotonia and progressive muscular dystrophy present histologically many features in common.

Erb has ventured the opinion that progressive muscular dystrophy may be due to functional changes in the cord. If this view is correct, the speaker thought it very remarkable that such a striking clinical picture as we see in advanced cases of this disease should result from changes which cannot be detected by the microscope. If Erb's theory regarding the spinal origin of progressive muscular dystrophy is correct, such a theory may be equally true of myotonia.

Dr. Jacoby, in closing, said that under the classification of myotonia only such cases should be included which present certain definite symptoms. In addition to the myotonic disorder and myotonic reaction, there should be an absence of anything pointing to gross disease in the central nervous system. Cases which do not fulfil these requirements should not be called myotonia, but should be classed under their proper designation, or designated as intention spasms. The so-called myotonic changes in the muscles, which have been described by various writers, and regarded as pathognomonic of this disease, have been found, by recent observation, to be not at all pathognomonic; the changes have been found to depend on the extensive shortening of the muscles, due to the method of preparing them for microscopic examination. In the case of Dejerine and Sottas, it was reported that nothing was found in the spinal cord, but the cerebrum and cerebellum were not examined. The changes in the muscular tissues could be accounted for by the fact that these tissues were much infiltrated with serous fluid.