MYOTONIA ATROPHICA (DYSTROPHIA MYOTONICA).*

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PERSONAL CASES.

CASE 1.—J. S., female, age twenty-one, married, born in the U.S., of Russian-Jewish parents, was admitted to the Neurologic Service of the Montefiore Hospital, complaining of stiffness of the joints of both hands.

Her father is fifty-five years old and in good health. Her mother died at forty-eight of dropsy (?); five sisters are living and well; their respective ages are thirty-one, twenty-nine, twenty-seven, twenty-five, and nineteen years; one brother, age seventeen, is an imbecile and is afflicted with the same disease as the patient; she also has another brother who is twenty-two years old and apparently well.

As far as can be ascertained there is no other history of a similar disease or of any other heredo-degenerative disease in the family.

The patient's birth and development were normal. She denies having had the ordinary diseases of childhood, but was subjected to tonsillectomy and adenoidectomy five years ago. Both the operation and convalescence were uneventful. Menstruation began at thirteen; it was regular but always painful, and would last six or seven days. She was married at twenty, and has never been pregnant.

Present Illness.—Her present illness began when she was six years old, with stiffness of the fingers of both hands. At school she had great difficulty in writing, because she could not move her fingers or turn her hands fast enough. She was always late in carrying out commands, especially when ordered to stop writing and lay her pen aside. She could not release these objects from her hands as fast and as readily as the other children did. She also noticed that in shaking

FIG. 1.—Myopathic facies (long, thin and tapering). Note expressionless face, bilateral ptosis, more marked on right than left. Patch of psoriasis on chest and abdomen.

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hands with a friend she had great difficulty in releasing the latter's hand. Upon making a fist she was unable to open her hand immediately, but was compelled to open it in a slow, gradual and deliberate manner, extending finger by finger. As a result of this disability she claims to have been unable to make much progress in school, and therefore did not reach the seventh grade until the age of sixteen, when she left school.

Physical Examination.—The patient presents a child-like appearance, with a typical myopathic facies—long, thin and tapering (*Fig. 1*). Her voice is high-pitched, thin, weak and monotonous. She is unable to speak for any length of time without tiring easily. In walking, she holds her head forward, and her shoulders droop. There is, however, no disturbance in station or gait, and no loss or diminution of the normal associated movements.

In marked contrast to the rest of her body, her skull is unusually large; its circumference is 49·5 cm.; from glabella to inion it measures 31·5 cm., and from mastoid to mastoid over the parietals, 37 cm.; she is 5 ft. high, and her weight is 110 lb.

There are no ocular palsies, but she shows irregular nystagmoid movements on looking to the extreme right or left, but none on looking upwards. The pupils and their reactions, as well as the fundi, are normal. There is a slight facial asymmetry, the right nasolabial fold being somewhat flatter than the left. There is a bilateral ptosis, giving her face a peculiar, somewhat mask-like appearance. Both masseters and buccinators are definitely atrophied. The tongue protrudes in the midline; there are no fibrillations and no atrophy, but on tapping it with the percussion hammer a definite myotonic reaction is elicited. The cranial nerves are otherwise negative. She has difficulty in rotating the head from side to side, and in the recumbent position she is unable to approximate her chin to her chest.

As she changes from the sitting to the lying posture her head seems to fall backwards; on attempting to rise from the reclining posture she cannot raise her head unsupported. Both sternocleidomastoids and platysmas are markedly atrophied. Owing to the atrophic condition of these muscles, the thyroid gland and cricothyroid cartilages, as well as the pulsating vessels of the neck, are readily palpable, but there is apparently no displacement of the trachea and no enlargement of the thyroid.

Her chest is long and narrow, with unusually prominent supraclavicular spaces. The costal angle is very sharp, with a marked widening of the intercostal spaces. The lungs are normal; the heart is normal in size; a blowing systolic murmur is audible over the pulmonic area, which is not transmitted, and is apparently hæmic in nature.

There is no tenderness or rigidity of the abdominal wall and there are no visible or palpable abnormal masses. The skin is dry and smooth, and shows a slight loss of its normal elasticity. There is a normal distribution of the hair, which is, however, quite sparse in quantity; she shows a psoriatic eruption over the anterior and posterior chest walls, over the extensor surfaces of the large joints, and over the sacrum.

The muscles of the upper limbs are poorly developed; muscle power in

the lower extremities is better than in the upper. When she is ordered to open either hand after making a fist, there is noted great difficulty in initiating the movements necessary to perform this act; at first there is a gradual extension of the little finger, followed by a slow extension of the index finger, then of the middle finger, then of the ring finger, and finally the thumb is slowly abducted, until the entire hand is opened. When the biceps humeri, the deltoids, the tongue, the muscles of the thenar and hypothenar eminences, and the calf muscles are struck, a 'worm-like' or 'wave-like' contraction of considerable duration is noted over the area tapped. The musculature of the thighs, legs and pelvis is apparently normal, but perhaps slightly underdeveloped. There are no deformities noted.

The uterus is subnormal in size, the cervix is situated in the axis of the vagina, and the adnexa are not palpable.

The tendon reflexes of the upper extremity are elicited with the utmost difficulty, while those of the lower are equal and lively. There is no evidence of pyramidal tract or cerebellar involvement, and no sensory or sphincteric disturbances. Her blood pressure is 90 systolic, 45 diastolic, and equal on both sides.

The electrical reactions are normal, except in the sternocleidomastoids and thenar and hypothenar eminences, where the muscular relaxation is prolonged and continued for a number of seconds after galvanic stimulation is stopped. Stimulation of the hamstrings gives rise to a contraction wave passing from cathode to anode. All muscles react normally to faradism.

An x-ray photograph of the skull shows the bones of the vault to be very much thickened; the sella turcica is normal in size, but in the anterior portion of the fossa there is a shadow of a dense calcareous mass. There are no evidences of increased intracranial pressure. The bones of the extremities and spinal column are normal.

Psychically the patient is dull, apathetic and somewhat inattentive; there is some evidence of mental deterioration; judgment is impaired. She is apparently a high-grade moron; her moral sense, however, is unimpaired. It would seem that her mental state is one of deterioration rather than deficiency. She claims that her sexual libido is markedly diminished.

Laboratory Findings.—Urine: sp. gr., 1020; alkaline in reaction, yellow, turbid, no albumin, and no sugar; triple phosphate crystals and squamous epithelial cells are present, but no casts and no erythrocytes. Blood examination: 4,200,000 erythrocytes; hæmoglobin, 70 per cent. (Sahli); small lymphocytes, 20 per cent.; large lymphocytes, 10 per cent.; eosinophiles, 0. Blood: Wassermann, negative. Spinal puncture yielded a clear fluid, under normal pressure; globulin, negative; Fehling's reduction, negative; cells, 0. Wassermann reaction and colloidal gold curve, negative. Bárány tests, negative.

Blood Chemistry.

Urea nitrogen	.	.	.	.	.	11.9 mgm. per 100 c.c.
Uric acid	.	.	.	.	.	1.7 " "
Sugar	.	.	.	.	.	96.0 " "

Sugar Tolerance Test.

Before breakfast	83 mgm. sugar per 100 c.c. blood.
One hour after breakfast . . .	177 " " "
Two hours after breakfast . . .	145 " " "
Three hours after breakfast . . .	127 " " "

Metabolic Rate.

Respiratory quotient.	1st hour, 0.805	..	2nd hour, 0.76
Basal metabolic rate.	1st ,, 0 per cent.	..	,, 2 per cent.

CASE 2.—C. G., age seventeen, is a brother of the patient described under Case 1. He appears undernourished, underdeveloped, and presents a typical myopathic facies (*Fig. 2*); his speech is slow, lacking in intonation, with a definite nasal quality to it. In walking he holds his head forward and droops his shoulders; there is no disturbance in any of the associated normal movements, and except for the shuffling of his feet along the floor, his gait is normal.

He presents a bilateral ptosis; the scleræ are blue. The pupils are equal and regular, and react normally to light and accommodation; fundi and ocular movements are normal.

There is definite wasting of both masseters, temporals and buccinators, giving rise to the characteristic tapering facies (*Fig. 2*). The left nasolabial fold appears somewhat flatter than the right; the lower lip droops, so that there is a constant dribbling of saliva from the mouth. The tongue protrudes in the midline; no atrophy, tremor, or

FIG. 2.—Myopathic facies. Note similarity in length of face and chin to that of sister in *Fig. 1*.

fibrillation is present, but on tapping it with the percussion hammer a very marked myotonic reaction is obtained, the lateral margins of the tongue presenting wave-like indentations from one-eighth to three-eighths of an inch in length. The cranial nerves are otherwise negative. There is marked atrophy of the sternocleidomastoids and platysmas.

The muscles of the upper extremities are poorly developed, especially those of the forearms. The biceps, deltoids, and small muscles of the thenar and hypothenar eminences present the typical myotonic reaction when tapped with the percussion hammer.

The patient has no difficulty in making a fist, but when he attempts to open it he does so by extending slowly, first the index finger, then the middle

finger, then the ring and little fingers, and finally the thumb is extended and abducted. This phenomenon (myotonus) disappears only after prolonged and repeated attempts, after which he can readily and 'smoothly' make a fist and open it.

The muscles of the lower extremities are fairly well developed, but there is some weakness of the flexors of both feet. The myotonic reaction can also be elicited in the gastrocnemii. The tendon reflexes of the upper extremities cannot be elicited, while those of the lower extremities are present and equal on both sides. The external genitalia are poorly developed. There are no disturbances of sensibility, no evidence of cerebellar involvement, and no sphincteric disorder.

The examination of the heart, lungs and abdomen is negative. The skin

FIG. 3.—Lateral view, showing hatchet-like facies of case 1.

FIG. 4.—Lateral view, showing hatchet-like facies, case 2.

is dry and smooth, with some loss of its normal elasticity. The distribution of the hair is normal, but it is scanty in amount. Psychically the boy is a high-grade imbecile; emotionally he is non-affective, extremely unreliable, and unable to adjust himself to circumstances. His attention cannot be maintained for any considerable length of time. (Unfortunately, we could not persuade this patient to be admitted to the hospital for laboratory and *x-ray* studies.)

SUMMARY OF PERSONAL CASES.

Both patients then present: A tapering facies with a hatchet-like appearance on profile (*Figs. 3 and 4*), a bilateral ptosis, amyotrophy of the masseters, buccinators, sternocleidomastoids and platysmas; increased muscle irritability of the biceps, deltoids, thenar and hypothenar muscles, and the gastrocnemii, associated with myotonus; genital hypoplasia; slight mental defect, viz., mental deterioration in

the sister, and feeble-mindedness in the brother. We therefore regard both of them as cases of myotonia atrophica or dystrophia myotonica.

DISCUSSION.

Ever since Rossolimo¹³ described myotonia atrophica discussion has been going on as to whether there is such a clinical entity, or whether

FIG. 5.—Posterior view, showing atrophy of the trapezii, left more marked than right: patch of psoriasis on back.

the condition is merely a symptom-complex. Numerous syndromes resembling myotonia, in Rossolimo's sense, have been described in the literature. Most of these cannot be properly classified. There are reported numerous cases of tetany, epilepsy, tabes, amyotrophy, syringomyelia, multiple neuritis, etc., in which the myotonic reaction could be elicited in some of the muscles. This has always led to a discussion whether, in these instances, the myotonia is a symptom or a complication of the principal disease, or whether it is a different but distinct clinical entity associated with the disease in question. Some writers regard myotonia atrophica as a clinical type of myotonia congenita or Thomsen's disease. It presents, however, some clinical features which distinguish it from genuine Thomsen's disease, and this is probably the reason that Rossolimo and others have insisted that the condition is a separate affection. Lewandowsky and others regard myotonia atrophica as merely an atypical form of myotonia congenita, and, according to Pelz, 12 per cent. of all cases of myotonia congenita show muscular atrophy.

CLINICAL FEATURES.

Myotonia atrophica is characterized by the presence of increased tonus in some muscles, with a flaccid paresis and atrophy in others, in a more or less symmetrical and constant distribution. In contrast to the congenital form, it does

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FIG. 6.—Typical myotonic reaction as shown in both hands. Patient attempting initial movement in opening closed fist.

not appear until the second or third decade of life. Males are more often affected than females. The myotonia generally precedes the atrophy and, as the disease advances, marked atrophies develop in some muscles, associated with myotonia (either in these or other muscles).

The muscles most commonly atrophied are: the orbicularis palpebrarum, the orbicularis oris, the temporal muscles, the sternocleidomastoids, the pectorals, the vasti and the anterior tibial extensors. In some cases the atrophy involves the same muscles as in the typical Erb's type of dystrophy, or in the Aran-Duchenne type of progressive muscular atrophy.

Kennedy⁸ has pointed out a peculiar feature in this form of myotonia, viz., that there is an extraordinary similarity in the appearance of the patients; they all seem to look as if they belonged to the same family—like brothers and sisters.

In typical cases the usual myotonic electrical reaction is modified in that the atrophied muscles show the usual diminished excitability to faradism, and to a lesser extent to galvanism. Occasionally the presence of a myasthenic reaction may be demonstrated. Some cases show in localized muscle groups a modified myotonic response—a slow contraction and slow relaxation to both faradic and galvanic stimulation. A wave of contraction may at times be elicited, with

FIG. 8.—Same as Fig. 6.

a very strong current, passing from cathode to anode. There is no reaction of degeneration present, except in the muscles that have completely atrophied.

FIG. 7.—Palmar aspect of hand, showing typical myotonic reaction (fist almost completely opened).

The condition of the reflexes depends upon the degree of wasting in the atrophied muscles. In pure cases of the disease there are no fibrillary twitchings, no sensory changes and no symptoms referable to the cranial nerves or to the sphincters. Hirschfield recently reported a case in which, in addition to the symptoms of myotonia atrophica, the patient had atrophy of the shoulder girdle and periods of bradycardia. After spinal puncture the pulse dropped to 40 per minute and continued so for a week. Under atropine it rose to 64. The cardiogram showed a slowing of the entire cycle, particularly the ventricular systole. The patient gave a history of previous similar attacks without cause.

Another characteristic feature of the disease is its association with congenital cataract. This association is too frequent to be a mere coincidence. It is rather suggestive that both myotonia and cataract in the same individual are evidences of an abiotrophy. Greenfield noticed there were some families in which some members showed cataract without myotonia, others myotonia without cataract, and still others, myotonia and cataract. The disease has also been found to be associated with other evidences of abiotrophy, such as the loss of hair, genital hypoplasia and absent reflexes with tabetiform degeneration of the cord. In the male, the most frequent of these extramuscular symptoms is baldness, with atrophy of the testicles next in frequency. There is no doubt, however, that the sexual functions are affected sooner or later in a very large number of cases in both sexes, and it is therefore reasonable to believe that the almost constant loss of libido and of sexual power is due to changes occurring in these organs. In some patients the external genitals are infantile. Celibacy and childless marriages are common.

A constant and characteristic feature, to which the disease owes its name, is the myotonia. This may remain more or less generalized, but it may also be limited to only a few muscles. It is characterized by a slow relaxation of the muscle at the end of contraction. The phenomenon is best and most frequently shown in relaxing the hand-grasp. By persistent percussion of the involved muscles a mechanical myotonia is produced. This is most commonly elicited in the tongue and in the muscles of the thenar and hypothenar eminences, although it may be present in many other muscles.

Absence of demonstrable weakness in the orbicularis oculi seems to be the rarest negative finding, while absence of weakness in the sternocleidomastoids is next in order of rarity. The extensors of the neck are weak, and therefore the head inclines forward, so that the eyes are directed towards the ground. This condition, associated with a drop foot and steppage gait, due to weakness of the muscles of the leg, presents a picture resembling the attitude and gait in tabes.

Speech is nearly always affected. It is low and monotonous, and has a more or less nasal quality. This may be due to either muscular atrophy, or myotonia, or both, for the lips, tongue, soft palate and vocal cords are often weak, and the tongue, and other muscles as well, are frequently myotonic.

PATHOLOGY.

Macroscopically, the muscles may appear paler and clearer than normally, but they are not fatty. Microscopically, in pieces examined during life, there is a very definite enlargement of the muscle fibres. The latter are rounder than normal, the transverse striations are poorly marked and the nuclei of the sarcolemma increased in number. Occasionally atrophic fibres are found scattered between the hypertrophied fibres. There is also an increase of the interstitial connective tissue. Schiefferdecker believes that the increase in the sarcolemmar nuclei is only a relative one. In the sarcoplasm he found granules which are not seen in normal muscle, and which he considers specific for Thomsen's disease. No changes in the central or peripheral nervous system have been found in the uncomplicated cases. In a case reported by Adie and Greenfield¹ there was an increase in the colloid in the pars intermedia of the hypophysis; the suprarenals showed a patchy distribution of the cortical lipoids.

PATHOGENESIS.

Various theories have been enunciated to explain the peculiar condition of the muscle which gives rise to myotonus. Most authors agree with Erb that the cause must be sought within the muscle itself. In favour of this theory are the histological abnormalities found within the muscles themselves. Electrical examination also shows that the abnormal reactions become more marked the more directly the current stimulates the muscle. Whether the abnormal condition of the muscle depends on the primary hereditary malformation, or secondarily on nervous changes, or metabolic disturbances, is not as yet definitely known. Jenssen believes that it is due to a retardation of assimilation processes and a defective excretion by dissimilation processes. During contraction of muscle there occur processes of dissimilation, whose products are continuously elaborated during muscular relaxation, with a simultaneous formation of assimilation processes. Retardation of the latter gives rise to a prolongation of the contraction curve with a subsequent lengthening of the relaxation curve. Levi and Passler assume that the increased irritability of the sarcoplasm is at the basis of the reaction, and that the myotonus is a typical sarcoplasm reaction.

Kramer and Selling¹⁰ agree with Reiss that the myotonic reaction is best explained on the same basis as the reaction of degeneration. The

practical identity of the two phenomena is also admitted by Erb. Taking into consideration the two phenomena as they occur during direct galvanic stimulation of the muscle, it is found that the difference between the reaction of degeneration and the myotonic reaction is merely a quantitative one. The frequent though not constant reversal of the contraction formula as noted in the reaction of degeneration has often been observed in myotonia. The principal difference between the myotonic reaction and the reaction of degeneration is merely that in the former the irritability of the nerve is retained, so that faradic irritability particularly is conserved. The similarity of both reactions becomes in some respects more evident when we take into consideration the partial reaction of degeneration, in which nerve irritability is naturally retained. The relations of both of these forms of reaction to each other allow the assumption that there must exist in both cases a similar pathological change in the muscle, which in the reaction of degeneration is due to a diminution of nerve influences, whereas in the myotonic reaction, in the absence of any demonstrable nerve damage, it must be due to some other cause which has as yet not been determined.

Many investigators, especially Naegeli, Hauptman and others, have endeavoured to solve this problem by attributing the muscular phenomena of this disease to certain biochemical processes. The change in metabolism and the trophic disturbances due to dysfunction of the glands of internal secretion, so commonly found in this disease, have been utilized by various authors in support of their theories. The evidence thus far adduced in substantiation of these claims is not conclusive. Curschmann,⁴ Steiner and others believe in the old hypothesis, as announced by Erb, that the inner secretory as well as the dystrophic phenomena have a common cause in the central nervous system. Gregor and Schilder have also shown by their 'action-current' phenomena in myotonic muscles that the myotonic contractions are due to central influences. Stocker, from a study of a case of paralysis with myotonus, concludes that myotonia is only a symptom, for the localization of which he implicates the lenticular nucleus. He is supported in his contention by Ake Barkman,² who believes that the cause of myotonia is to be found in the extrapyramidal system. He calls attention to the fact that Söderbergh, Thomalla and others have insisted that the tonic spasms which sometimes precede the active movements in Wilson's disease bear a close resemblance to myotonic crises. The same author believes that as in myxœdema the athyreosis may give rise to disturbances of nerve function (according to Söderbergh, to cerebellar symptoms), so in myotonia the central nervous system may be affected by toxins as a result of disturbances of internal secretion. Unfortunately, thus far, none of these writers nor anyone else has been able to substantiate any of these claims anatomically. Hoffmann believes that

myotonia atrophica is a condition in which myotonia is a primary disease and the muscular atrophy is a secondary process. Jendrassek, on the other hand, sees no necessity in assuming such causal relationship between these two conditions. He believes that they are both forms of degeneration which are frequently associated in the same individual.

Findlay ⁶ believes that the disease is primarily one of the muscles, but the nervous system plays an important rôle in its production, because—

1. The creatin and the creatinin metabolism points to an unusually efficient muscle and not a degenerated one.

2. During voluntary movements myotonic reaction develops promptly, but during involuntary movements no myotonic reaction is observed.

3. The condition gradually disappears on repetition of movements, but not on repetition of a direct electrical stimulus.

4. The mental state of the patient has a decided influence on this condition.

Symptoms resembling myotonia have been produced in animals by veratrin and creatinin poisoning. Some observers have been inclined to attribute the disease to autointoxication. The generally accepted view, however, is that the disease is a form of myopathy, which is to be included among the heredo-degenerative diseases or abiotrophies.

Recently Weiss and Kennedy ¹⁵ have reported a case of myotonia in which, after a period of thyroid medication (6.8 gm. thyroid and 9.6 mgm. thyroxin, in fourteen days), followed by the administration of atropin (0.075 mgm. per kilogram of body weight), they found distinct improvement in the myotonus. There was improvement in the muscular movements, with absence of myotonus and fibrillary twitchings. This effect, however, was only temporary, lasting about a day.

In our Case 1, we administered thyroid extract over a prolonged period, and later thyroid extract combined with benzyl benzoate and atropin, with no results whatever. But after a definite course of physical exercises, inducing prolonged and repeated movements of the hands, forearms, arms, legs, and thighs, we found temporary improvement in the myotonia of the patient. It would not only temporarily disappear after physiotherapy, but even when the patient was properly urged and commanded to carry out a definite movement normally, such as opening the fist, thus sustaining Findlay's view that the mental state has a decided influence upon this condition.

A critical review of all the aforementioned factors makes it obvious that the problem of the pathogenesis of this disease has not been solved. By far the most pronounced feature of the condition is its hereditary character. The symptoms and course would seem to suggest that at the basis of it is some form of degeneration—a form of abiotrophy. We

feel that the pathogenesis of myotonia atrophica will remain an obscure chapter in medicine ; at least, until more reliable data can be furnished us by embryologists, pathologists, and biochemists.

CLINICAL VARIETIES.

(a) *Myotonia acquisita*.—This is a type of myotonia simulating Thomsen's disease, but often associated with traumatism and infections. The type is therefore acquired, not congenital, and is usually transient and curable. Its pathology and pathogenesis are unknown.

(b) *Pseudomyotonia hemiplegica*.—These are conditions of hemiplegia, with additional features such as myotonic reactions, with involvement of the arms and legs, but without atrophies or hypertrophies. This variety is probably due to vascular degenerative lesions involving the midbrain and extrapyramidal systems. These are most likely the cases that Stocker and Barkman and others have in mind when they attempt to attribute myotonia to disease of the lenticular nucleus. We doubt, however, whether these cases can be regarded as cases of myotonia atrophica or dystrophia myotonica in the sense that most clinicians use these designations.

(c) *Paramyotonia congenita*.—In 1886 Eulenberg⁵ described a peculiar heredofamilial affection of the muscles, which he termed paramyotonia congenita. This is characterized by a tonic spasm occurring in certain voluntary muscles of the face, orbicularis palpebrarum and orbicularis oris, so that the individual is unable to open his mouth or eyes for a period of fifteen minutes or longer. This spasm usually comes on in cold weather and is not influenced by excessive exertion. Warmth has a tendency to diminish such attacks. The muscles of the neck, of deglutition, as well as those of the limbs, may be involved. The characteristic symptoms of the disease make their appearance at birth and are sometimes found coincidently with Thomsen's disease, in members of the same family. (We have not been able to find in the literature an authenticated record of a case of this disease in America ; we have no personal experience with it, and merely reproduce here an abstract of Eulenberg's original description for the sake of completeness.)

DIAGNOSIS.

The diagnosis of generalized myotonia scarcely presents any difficulties. Some cases of *pseudohypertrophic muscular dystrophy* may bear a superficial resemblance to myotonia. The typical disappearance of the myotonus after repetition of the movements, the presence of the myotonic reaction in the latter, and the peculiar distribution of the muscular hypertrophy in the former, will make the diagnosis clear. The absence of pyramidal tract symptoms and the presence of the

myotonic reaction will serve to differentiate the condition from *spastic spinal paralysis*. Partial myotonia, e.g., of the forearms, may simulate the *chronic tonic form of tetany* and *occupational cramps*. A careful history and the elicitation of the myotonic reaction will be sufficient to establish the correct diagnosis. *Family periodic paralysis* is another condition which may cause some confusion. The paralysis, which may last for hours or days, is irregular in its periods of occurrence, and is followed each time by a complete recovery; this, combined with the absence of the myotonic reaction, will be diagnostic. *Pure myokymia* and *atrophic myokymia* are, as contrasted with pure myotonia, merely syndromes expressing an acute or subacute condition of irritability of the muscles. They are characterized by the presence of muscle waves, flickers and twitchings. They are acquired conditions usually due to exogenous factors and strictly localized in connection with a marked or rudimentary neuritis or myositis. They are, therefore, frequently encountered in cases of overexertion, trauma, infections and compression of the nerve roots. Lately their presence has also been reported in cases of epidemic encephalitis. Simultaneously with the fasciculo-fibrillary muscle wave in myokymia there may arise many phenomena of irritation, the so-called 'myotonoid' after-effects, true muscle hypertrophy, muscle tension, cramps, mechanical and electrical phenomena. All of these, however, bear only a superficial resemblance to myotonia. The *pseudospastic paresis of hysteria* may also very rarely and superficially resemble myotonia. In the atrophic form, however, the differentiation from other forms of wasting sometimes depends entirely on a careful electrical examination, especially of the muscles which are not atrophied.

PROGNOSIS AND COURSE.

The prognosis is favourable as regards life, but unfavourable as regards cure. The disease is a chronic and progressive one. The cases which have been reported as improved or cured were undoubtedly atypical forms following trauma or infection. The not uncommon transition from the ordinary form of myotonia to the atrophic form makes the prognosis as to life less favourable.

TREATMENT.

Much can undoubtedly be done by continued exercise of the will and by regular gymnastic exercises and massage. Cold baths must be avoided, and strenuous gymnastic exercises have a tendency to hasten the transition to the atrophic form. Frink reports good results following the administration of thymus extract in one of his cases. Some authors have advocated the use of strychnine with and without the various glandular extracts. We ourselves have had occasion to treat these cases in the wards of the Montefiore Hospital with all the measures advocated,

but, unfortunately, we could find no method of treatment that has had the slightest favourable effect on the progress of the disease.

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