

DYSTONIA MUSCULORUM DEFORMANS

WITH ESPECIAL REFERENCE TO A MYOSTATIC FORM AND THE
OCCURRENCE OF DECEREBRATE RIGIDITY PHENOMENA.

A STUDY OF SIX CASES *

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Dystonia musculorum deformans is not an uncommon disorder. It is well known for its kinetic phenomena, and most, if not all, of the case reports consist of analysis of the peculiar movements which characterize the clinical syndrome. Thus far spontaneous abnormal involuntary movements, torsion spasms, disturbances in tonus and possibly a racial peculiarity have come to be looked on as the criteria for the diagnosis of the bizarre entity. This clinical conception of the disorder, originally formulated by Ziehen and Oppenheim and so closely followed by subsequent observers, stresses only the hyperkinetic phenomena. But we have found cases in which these phenomena are present only to a slight extent or almost entirely absent, and yet we believe that they form part of the disease.

It is the object in this paper, besides recording six hitherto unreported cases of dystonia musculorum deformans with atypical and unusual features, to call attention to the existence of a myostatic variant of the disorder. But far from attempting to create a new clinical entity, we wish to correlate it with the well-known syndrome. This myostatic or postural variant is indeed a part of the disorder and always coexists with the kinetic disturbance of which it is only a complement. It is, in other words, a fleeting postural phase, observed between the waves of movement, become permanent. The occurrence of large and small fragments of decerebrate rigidity in typical cases of dystonia, to which we wish to call especial attention, further emphasizes the postural or myostatic disturbance in the condition. But, whereas in most cases the kinetic disturbance obscures or overshadows the postural element, there are some in which the latter is plainly evident and a few others in which it is dominant to such an extent that

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one begins to doubt whether the pictures are part of the same syndrome. It is our conviction that they are. The following case reports and their discussion will illustrate these points.

REPORT OF CASES

CASE 1.—A case of dystonia musculorum deformans of the kinetic type beginning in the right hand and, up to the present, involving the musculature of both upper extremities, neck and head, with a fragment of decerebrate rigidity phenomena.

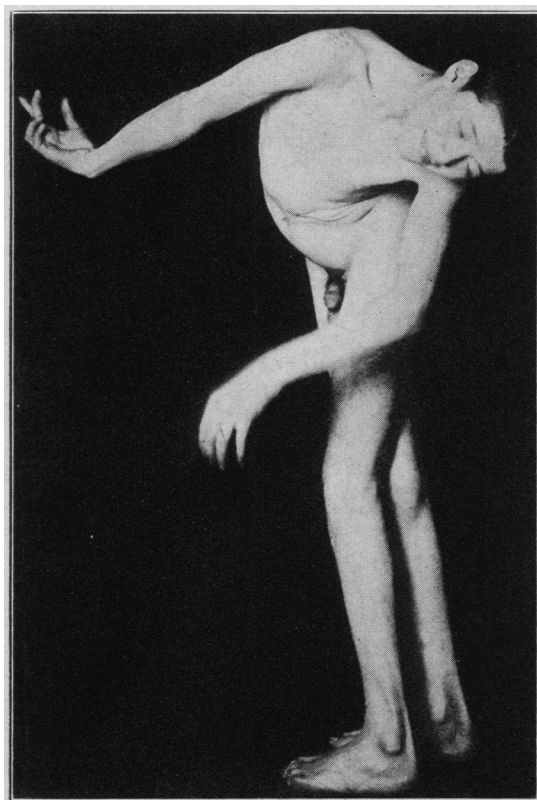


Fig. 1.—A case of dystonia illustrating segmental involvement and a fragment of decerebrate rigidity limited to the right upper extremity.

History.—B. M., a boy, aged 15 years, born in the United States, a Jew of Russian parentage, about three and one-half years ago (1918) noticed difficulty in using his right hand, especially in writing. Soon after it was observed that his left upper extremity was held in a peculiar position, namely, flexed at the shoulder and elbow and adducted. About eighteen months after the onset, the patient's head and body began to bend over to the left side. During the course of his illness there gradually developed peculiar defensive movements of his left upper extremity, uncontrollable movements of the head and turning movements of his body.

Physical Examination.—The gait was normal. There was a marked involuntary torsion spasm involving the musculature of the head, neck, chest and left upper extremity. The head was rotated and markedly tilted to the left, the chin almost resting on the left shoulder. On extending the right upper extremity, the hand was turned with the palm outward, that is, overpronated, and the index finger was hyperextended. This constitutes a fragment of decerebrate rigidity, the so-called pronator sign of Wilson (Fig. 1). There was a coarse rhythmic tremor of the extended hand. While these movements appeared to come on in rhythmic waves, they were practically continuous in one part or another.

The spontaneous movements of the left upper extremity consisted of protraction of the arm together with flexion of the forearm; this movement was very similar to that assumed in defending one's face. In the upper extremities the dystonic spasm was revealed in an alternating hypertonia and hypotonia. It is curious to note that walking on all fours was accomplished much better than the motor disturbances of the upper extremities would seem to permit. Further neurologic examination was negative.

Discussion.—The disease began in one upper extremity, which is rather unusual. Other writers, especially Hunt,¹ have called attention to the frequency with which the disease begins in the lower extremity. This patient also shows an exquisite fragment of decerebrate rigidity in the right upper extremity, although the picture is mainly of the myokinetic variety. Finally, the movements are thus far limited to the head and neck, upper extremities and upper part of the trunk. It is quite possible, in view of the comparatively short duration of the illness, that it will extend downward and involve the rest of the trunk and lower extremities; but for the present the disease has assumed a segmental character. Attention is called to this distribution because a segmental delimitation has previously been pointed out in striatal diseases, especially in paralysis agitans.

CASE 2.—*A case of dystonia musculorum deformans of the kinetic type with the occurrence of both fragmentary and almost complete decerebrate rigidity phenomena.*

History.—J. L., a boy, aged 11 years, born in the United States, a Jew of Russian parentage, whose parents were first cousins, and who had three other children living and well, had had the disease for three years. It was noted that his left foot gradually began to "turn in" involuntarily when he walked; this became progressively more marked, and one year later his right foot also began to "turn in and to drop." In July, 1921, in the course of two days, there developed frequent spasmodic contractions in the left lower extremity, which made sitting difficult. The spasms steadily increased in severity and frequency. In September, 1921, the patient developed severe spasms in his hands. In August, 1921, walking became impossible on account of the muscular spasms and fatigability. In December, 1921, sitting became impossible, and the patient became bedridden. For the last two months the patient has been lying on his left side with the head turned to the left. Frequent spasms in the left upper extremity and the muscles of the back have also developed. For the last two months the right lower extremity has been held almost constantly in the extended position.

1. Hunt, J. Ramsay: The Progressive Torsion Spasm of Childhood, *J. A. M. A.* **67**:1430 (Nov. 11) 1916.

Physical Examination.—The patient was unable to walk or stand. The position of election was the reclining one, with the body inclined to the left and the head bent forward and to the left. The left knee joint was in flexion, the right in extension. Both feet were in hyperplantar flexion, the right more

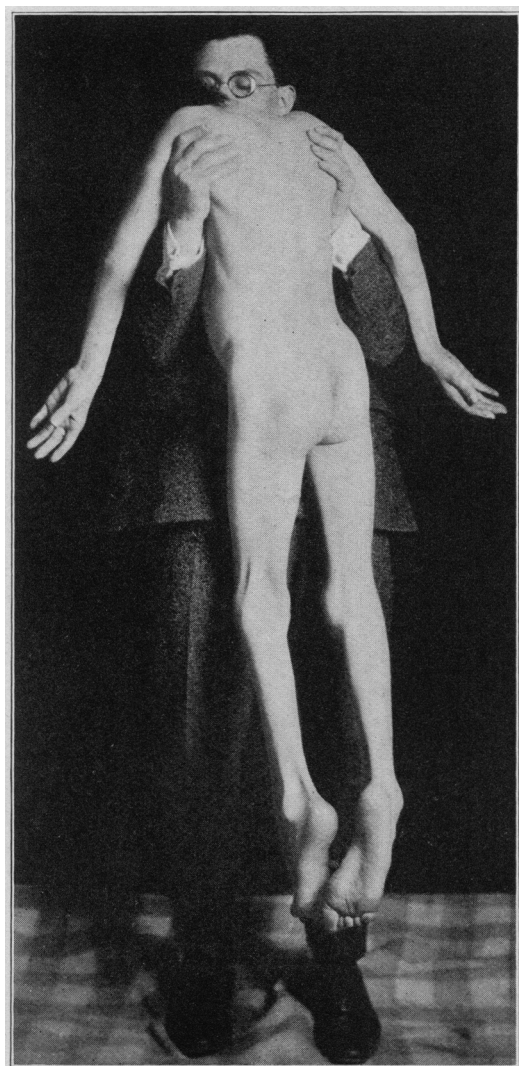


Fig. 2.—Almost total decerebrate posture in a case of dystonia musculorum.

so than the left. Occasionally the left upper extremity was extended with the palm turned outward. The movements came on in wavelike fashion and seemed to be induced by slight stimulation, such as stroking or pinching the skin. He frequently assumed an attitude of opisthotonos with the right lower

extremity markedly extended and the back in extreme lordosis. The abductors of the thigh, flexors of the leg and to a slight extent the adductors of the right thigh, were in almost constant tonic contraction.

If the boy was suspended by his upper extremities, he assumed a position typical of decerebrate rigidity: the forearms were extended on the arms, the fingers somewhat flexed at the proximal metacarpophalangeal joints, the forearm rotated outwardly (pronator sign), the lower extremities hyperextended and the back lordosed, forming an opisthotonic arc. However, the head was not retracted but hung limply forward with the chin pointing to the left (Fig. 2). There were no movements in the face or head musculature; the sternomastoids occasionally manifested mild tonic contraction. While there was no true pelvic distortion, there was a tendency to torsion toward the left. The erector spinae groups were in a state of constant contraction and occasionally the muscles of the back of the neck participated in the spasm. The muscles were in hypertonic contraction, but this could be overcome by passive movement, even to the point of hypotonia. Both the fingers and the hands

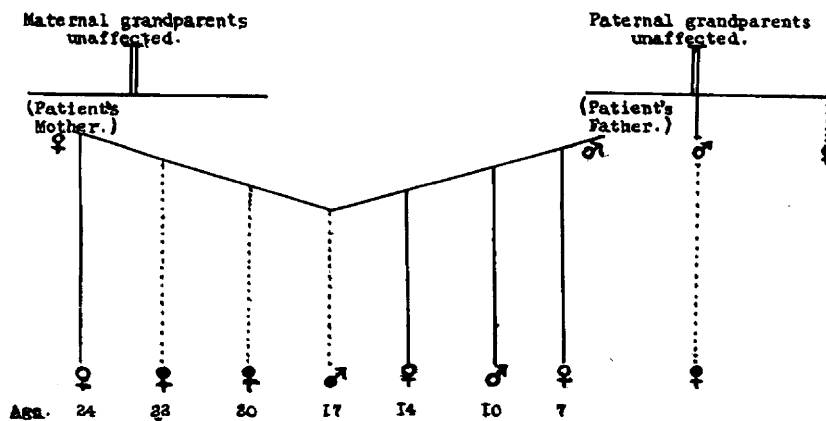


Fig. 3.—Familial incidence of dystonia musculorum deformans in Case 3. The black symbols indicate those affected with the disease. * indicates the patient.

were definitely hypotonic. There was no adiadosokinesis, no ataxia or dysmetria.

The deep reflexes were difficult to elicit. The abdominal and cremasteric reflexes were present. There were no sensory changes. The cranial nerves were normal, except for the existence of a true nystagmus on extreme gaze to the left.

Discussion.—The history of the development, the age of onset, the race of the patient and the abnormal involuntary movements stamp the case as a typical dystonia musculorum deformans, in spite of the fact that there is as yet no torsion of the pelvis. The unusual features are the postural decerebrate rigidity phases. Even as he lies on his back there are rhythmic waves of movement. (We have observed this rhythm in some of the other cases, and it is described in most of those reported in the literature.) The spasms throw the right lower extremity in extension and both uppers in extension, pronation and outward rotation. These movements represent momentary

phases of decerebrate rigidity phenomena. When the boy is suspended, all four extremities and the body assume the typical attitude which occurs on experimental section of the midbrain, with the exception of the position of the head. The occurrence of this phenomenon points to the anatomic delimitation of the lesion in dystonia, even if it does not strictly localize it.

CASE 3.—An unusual case of dystonia musculorum deformans with (1) a remarkable familial incidence, (2) speech disturbance, (3) a remission of the hyperkinetic phenomena with a resultant (4) myostatic residuum, (5) a hemidystonic distribution, and (6) a fragment of decerebrate rigidity.

History.—L. P., a woman, 21 years of age, born in the United States, was a Jewess of Russian parentage. A remarkable familial tendency to the disease was noted (Fig. 3). The illness began at the age of 12. Her gait became "dragging" and difficult. Later her upper extremities became involved. There was no history of precedent acute infection. The physical examination recorded by Dr. I. Abrahamson on April 5, 1920, revealed a bedridden patient with a typical dystonia with severe involvement of the body, head and neck and all extremities. The speech was dysarthric, bulbar in nature. In January, 1921, improvement was noted. The patient was quieter and able to take a few steps without support. In August, 1921, it was noted that the left hand still showed the rhythmic clonic movements, but there was a tendency to extension and pronation (fragment of decerebrate rigidity). The patient was able to walk fairly well. General dystonic movements were still in evidence. In February, 1922, there was marked remission of all movements with considerable general improvement (Fig. 4). She showed some dystonic movements in the left upper extremity with the rhythmic flexor-extensor movement of the hand at the wrist. The left foot was in equinovarus contracture and also exhibited movements, resulting in a left hemidystonia. The trunk and right side disclosed no movements but assumed the postural condition momentarily observed in the dystonias during the brief interval between the waves of movement; that is to say, that which is the fleeting static element in the kinetic disturbance has become a permanent postural attitude in this patient's remission.

Discussion.—In this patient, the onset of whose dystonia dates back to childhood, the condition began in the lower extremities and became progressively worse, until she exhibited a violent form of the typical kinetic disturbance. Added to that she showed a rare involvement of speech (bulbar dysarthria). So far as we are aware, only two other cases referred to in the literature reveal this disturbance (those of St. Bernstein² and Wimmer³). After two years, during which she became bedridden, a gradual remission set in. It may be of interest to mention that in August, 1920, she had a streptococcus sore throat, and in April, 1921, an acute follicular tonsillitis. One may speculate on the possible bearing of these infections on the remission. The quite remarkable familial history is also worthy of emphasis, in view of the familial tendency of some of the other striatal syndromes, especially since recent criticism tends toward the grouping of numerous clinical entities of a supposedly common basal ganglion origin. Further, although only few dystonic elements are left, they are practically limited to one side—hemidystonia. But

2. St. Bernstein: Ein Fall von Torsions Krampf, Wien. klin. Wchnschr. **25**: 1567, 1912.

3. Wimmer, A.: Etudes sur le syndrome extra-pyramidaux. Spasme de torsion progressif infantile (syndrome de corps strie), Rev. neurol. **28**:952 (Sept.-Oct.) 1921.

the striking feature of this residual state is the static or postural attitude, an attitude to which we also wish to direct attention in some of the succeeding cases and which we designate the myostatic variant, or counterpart, of the myokinetic syndrome.

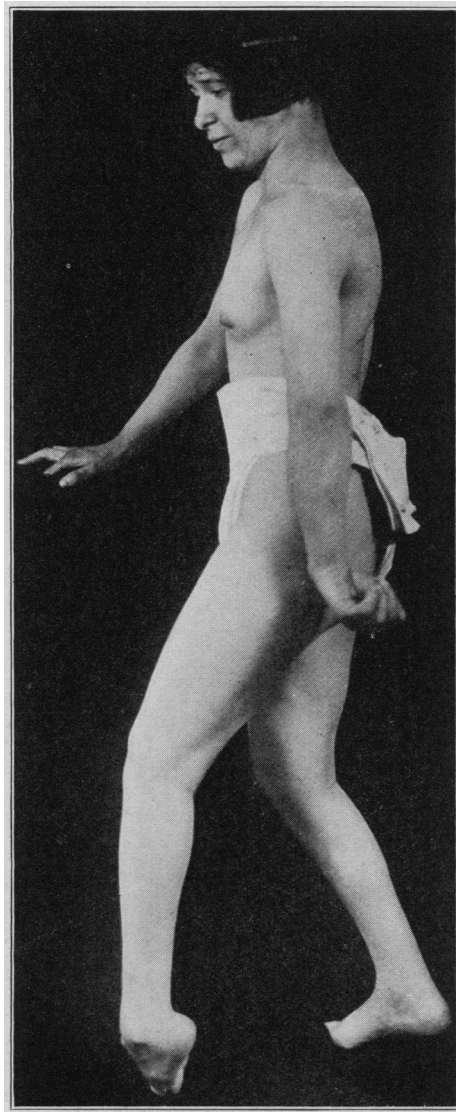


Fig. 4.—Formerly a kinetic type of dystonia, now a myostatic variant exhibiting a hemidecerebrate posture.

CASE 4.—*A case of dystonia musculorum deformans, illustrating the myostatic or postural form, with few hyperkinetic phenomena.*

History.—P. T., a man, aged 20 years, born in Russia, a Jew of Russian parentage, whose illness began in 1913, at the age of 11, one year prior to

the onset fell from a height of about 12 feet, alighted on his feet but sustained no injury. One year later there developed a "drop foot" on the right side, associated with a turning outward of the lower extremity on walking. Three years later his hand began to turn outward, and he found difficulty in using it. About three years thereafter the left lower extremity became affected. The condition has remained stationary to the present day.

Physical Examination.—The patient walked with a peculiar gait, in which there was considerable twisting of the pelvis; the vertebral column was thrown forward, producing a marked lordosis; the lower extremities were rotated outward (everted) and in progression the trunk was bent forward and the lower extremities were flung about awkwardly in movements of hyperextension and hyperflexion (Fig. 5). A tendency to hyperadduction produced a scissor-gait effect. The abnormal movements and position of the spine and pelvis largely disappeared when the patient crept "on all fours." Despite the dystonic gait and the tortipelvis, the patient was remarkably free from tonic spasms when at rest. Except for an occasional tremor of the right upper extremity and clonic plantar flexion movements of both feet, there was a paucity of abnormal involuntary movements. There was no play of hypertonia and hypotonia in the affected muscles. A slight degree of hypotonia was present at the wrists.

The patient complained a good deal of various symptoms, was introspective, egocentric and was likely to use high sounding phrases. His attitude at times strongly suggested the functional.

Discussion.—The lack of progression, the atypical bizarre gait and attitude, the want of dystonic movements at rest, together with the patient's psychic make-up, speak in favor of a hysterical condition. But further study of the case convinces one of its organic nature and its relationship to dystonia musculorum deformans. For this patient shows the very postural or myostatic phase which may be momentarily observed in all the typical forms of the kinetic syndrome. But, whereas the postural static component is masked by the hyperkinetic phenomena in the general run of cases, in the present instance it has become the dominant feature. Corroborative evidence of the relationship of this syndrome to dystonia musculorum deformans may be found in the history of its development, the age and race of the patient, the torsion-posture of the pelvis, the lordosis, the clonic movements in the feet and the general attitude. The fact that the static phase is partially present in an undoubted case of dystonia musculorum deformans gives weight to the view that where the entire clinical syndrome is dominated by the postural component we are merely confronted with the complementary side of the same picture.

CASE 5.—A postural or static instance of dystonia musculorum deformans (dysbasia lordotica progressiva) with slight kinetic involvement.

History.—H. R., a man, aged 32, a driver, born in Russia, a Jew of Russian parentage, four months after a fall began to feel "stiffness and pulling sensation" in the hamstring muscles of the left thigh, followed by a "drawing sensation" in the left arm and forearm, with a tendency to flexion at the elbow and fingers. At the same time the left side of his face became involved in a similar muscular spasm. In 1912, a laminectomy (seventh cervical nerve to fifth dorsal vertebra) was performed, apparently in the belief that some cord involvement was the cause of his motor disturbance. Prior to the laminectomy alcohol was injected into the left elbow region and left popliteal space, appar-

ently with the unfulfilled hope of controlling the abnormal involuntary movements. Both of these operative attempts have left behind physical signs which are independent of the actual condition, yet serve to mask it.

As this case presents almost in its entirety the myostatic variant of *dysbasia lordotica progressiva* and may arouse doubt as to its proper classification in the group of dystonias, it may be mentioned that Drs. I. Abrahamson and J. Ramsay Hunt independently diagnosed the case (1919) as one of dystonia.

Physical Examination.—The patient was examined in February, 1922. There was frequent play of the muscles of the face, particularly of the upper lip, resembling a slow grimace. There was a slight tilt of the head and overaction of the *platysma myoides*. The right *sternocleidomastoid* was somewhat more

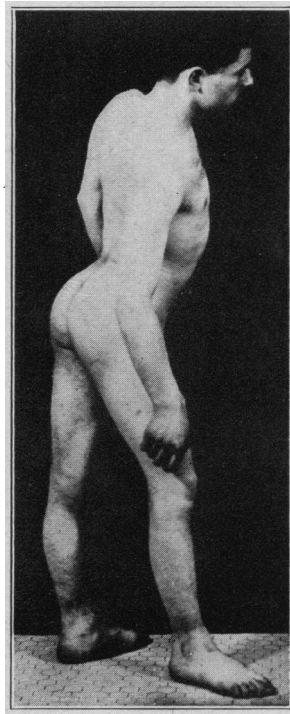


Fig. 5.—The lordotic attitude in a myostatic form of dystonia.

prominent than the left. There were few or no bodily movements with the exception of some in the toes, which appeared to be a cross between a dystonia and athetosis. On attempting to walk the movements of the toes gained in amplitude. On the right there was dorsal extension of the big toe and fanning of the others when the patient lifted his foot from the floor; this movement was of a dystonic type. There was a slow turning of both feet, especially of the right, into an equinovarus position. The lower extremities were held mainly in an extended position; the feet especially in plantar flexion. The right big toe was frequently held in spontaneous dorsal hyperextension. On suspending the patient by the arms both lower extremities were hyperextended at all joints.

The gait simulated a spastic walk with bilateral dropped foot. It was slow, wide, swinging and shuffling with a broad base, and appeared somewhat inco-

ordinate (Fig. 6). Yet there were no equilibratory and no deep sensory disturbances. The left abdominal reflexes were diminished, the lower deep reflexes increased. There was no true Babinski sign, but there were a left Mendel-Bechterew and a Rossolimo sign. There was neither hypertonia nor hypotonia in the right upper and lower extremities. A slight hypotonia was

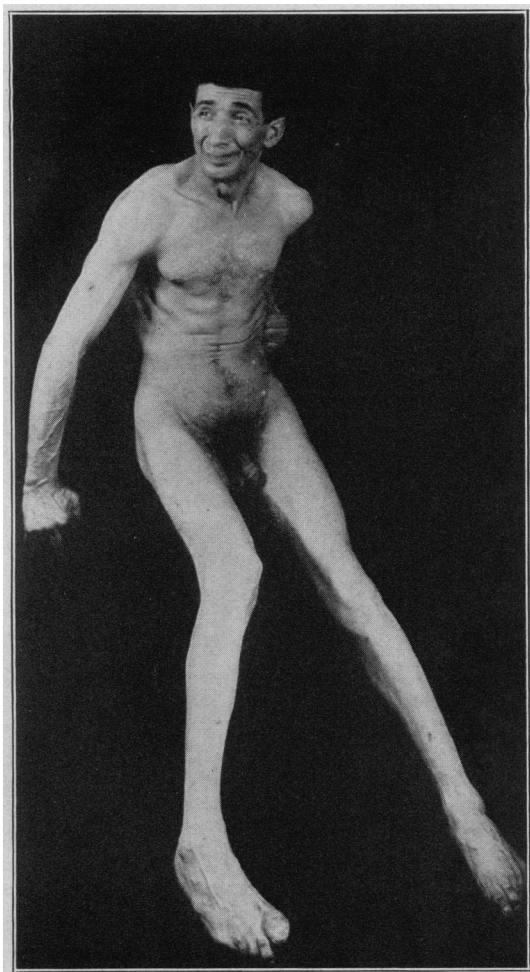


Fig. 6.—A typical instance of the myostatic variant of dystonia musculorum. Note the spontaneous Babinski sign and the grimacing facial expression somewhat like that seen in progressive lenticular degeneration (Wilson's disease).

noted in the left lower extremity. There was flexor contracture of the fingers of the left hand into the palm. The left interossei were atrophied. Marked hypertonia was found in the left upper extremity, which was held flexed at an angle of 90 degrees at the elbow. The speech sounded as though the words were uttered through articulated teeth and partly closed lips. The labials

were especially interfered with because of the overaction of the lower facial muscles and platysma. The face appeared spastic, as if a smile were frozen on it.

Discussion.—There are a few physical findings in this case which complicate the picture, but it appears that they have been brought about by surgical interference and therefore do not form part of the syndrome. The contracture of the left hand, the atrophied interossei and the fixed hypertonic attitude of the left upper extremity are due to the therapeutic attempts which were made to control the abnormal movements. But the general posture and the few abnormal involuntary movements leave no doubt that the patient represents the myostatic variant of the hyperkinetic syndrome. His gait especially resembles to a marked degree that of the patients in Cases 3 and 4, and indeed that of every case of dystonia if one could subtract the abnormal movements. Additional corroborative evidence that the case belongs to the myostatic form may be found in the fact that the lower extremities assumed the hyperextended decerebrate postural attitude when the patient was suspended in the air with support under the axillae.

The speech disturbance was not of a dystonic character, but resembled more that encountered in progressive lenticular degeneration (Wilson's disease). This may be of some significance in linking together the two syndromes. The spontaneous dorsal extension of the big toe, while not a true Babinski sign, is worth remarking, and will be further alluded to in the next case.

CASE 6.—*A case of dystonia musculorum deformans of the kinetic type revealing (1) fragments of decerebrate rigidity, (2) a paralysis agitans-like tremor of right thumb and hand, (3) a dorsal extension (Babinski) of the left big toe.*

History.—E. H., a young woman, aged 19, born in the United States, a Jewess of Russian parentage, had had a tenotomy of the left Achilles tendon in 1918. The illness set in gradually at the age of 8, first in the right foot and then in the right hand and upper extremity; later in the right foot; and finally the left upper extremity became affected.

Physical Examination.—The gait appeared somewhat spastic. She walked on the toes of the right foot and on the outer margin of the left sole. Both feet were inverted, the left more than the right. The right lower extremity tended to cross in front of the left. While sitting there were observed involuntary inversion of the left foot and occasional extension of both legs on the thighs. Now and then a spontaneous dorsal extension of the left big toe occurred. On extending the arms forward and above the head, the right underwent alternating supination and pronation with a tendency to eversion of the palm.

There were involuntary flexion and extension movements at the right elbow. At times there were small oscillations of the right thumb and hand in a manner closely simulating the tremor of paralysis agitans. The movements in the left upper extremity were much less marked. At times there was over-extension of the left wrist and involuntary flexion of the fingers, together with flexion and extension at the left elbow. There were no involuntary movements of the head and neck and no torsion of the back or pelvis.

The reflexes of the upper extremities could not be elicited owing to the presence of the constant involuntary movements. The knee and Achilles' tendon reflexes were present and equally active. The abdominal reflexes were present and equal. On plantar stimulation there was marked dorsal extension of the left big toe and fanning of the others. This seemed to be an actual reflex response,

although a spontaneous dystonic Babinski movement also occurred. The cranial nerves were normal except for a slight bilateral horizontal nystagmus.

Discussion.—The following facts are worthy of note: First, the restriction of the kinetic disorder to the extremities, giving an appendicular distribution. This emphasizes once more the possibility of the segmental involvement in the disorder, a point alluded to in connection with Case 1. Second, the presence of fragments of decerebrate rigidity in the right upper extremity and in the feet discloses the static component. Third, the paralysis-agitans-like tremor of the right thumb and hand is of particular significance, as it indicates a probable involvement of the efferent pallidal mechanism of the corpus striatum. The presence of this tremor may furnish another clue to the localization of the lesion. Fourth, attention should be directed to the presence of the Babinski toe phenomenon, which is even more marked in this man than in the preceding case.

SUMMARY AND DISCUSSION

The six cases summarized in this paper show a number of features which hitherto have not been associated with dystonia musculorum deformans. It is quite difficult to explain the nystagmus in three of the cases, unless it is assumed that the ocular movements are part of the dystonia. Although this is somewhat far-fetched, it may be pointed out that irregular movements of the eyeballs have been observed in violent choreas. So, too, it is difficult to interpret the presence of the Babinski sign in Case 6 without other signs of involvement of the pyramidal tract. Further, the fanning of the lesser toes and the dorsal extension of the big toe is occasionally observed in the same patient during the spontaneous movements. Case 5 also exhibits this spontaneous dorsal extension and fanning. One is tempted to speculate on the significance of the Babinski phenomenon and to question whether it is really primarily a pyramidal tract phenomenon or an unexplained striatal release mechanism.

The speech disturbances in two of the cases are also unusual. Two other cases reported in the literature showed similar disturbances. Wimmer's recent report mentions difficulty in speaking, but the statement is made that this was not a dysarthria. The speech was explosive; there was poverty of words and sometimes echolalia. There is no reason why the speech muscles should not be involved in dystonia in view of their undoubted striatal and cerebellar innervation. In this connection one may again instance chorea in which dysarthria is not uncommon. If an attempt is made to correlate dystonia with lenticular degeneration, it is not surprising that there is speech disturbance, although this is of quite a different type in the latter disease. The speech disturbance in Case 5 was somewhat reminiscent of Wilson's syndrome.

In many of the cases which we have described, and in numerous others reported, there is something wavelike or rhythmical to the movements. Rhythm in general is such a universal phenomenon and

in animal movement so primitive that special emphasis should be given to its presence in dystonia. Very likely it represents a reversion to a lower segmental characteristic of movement. In this connection it may be once more pointed out that two of the cases showed an actual tendency to segmental distribution of the affection.

The racial element, which, though very common, is not universal, was found in all of our patients. So, too, there was an apparent yet noteworthy functional tinge to some of the cases. It is well known that many of the patients in the early stages of the disease are considered hysterical. Case 1 bore the burden of that diagnosis for one year and Case 4 is still reminiscent of it, and yet there is no doubt as to the organic nature of the condition. Wilson's illuminating remarks on decerebrate fragments in hysteria may serve as possible explanations in this connection.

We called attention to the occurrence of decerebrate rigidity phenomena in the cases herein recorded. Wilson ⁴ made passing mention of them in dystonia, but he did not follow up his observations. His allusion, however, suggests a static component in the hyperkinetic disorder. The importance of the demonstration of this combination, as well as the dissociation, lies in the fact that it furnishes clinical evidence for the accepted notion of the physiologic unity of posture and movement. Conversely, there is sufficient physiologic evidence to support the view that there does exist a closely interwoven static counterpart of the kinetic form of the disease. The conception of movement as recently elaborated by numerous observers ⁵ permits the inclusion in one entity and proves the basic relationship of two apparently dissimilar clinical pictures, namely, the myostatic and the myokinetic variants of dystonia.

This enlarged conception is not altogether novel, at least from the point of view of correlating several clinical entities which are probably based on one anatomic substratum. Strümpell originally postulated an amyostatic syndrome in which he included Westphal's pseudo-sclerosis, Wilson's lenticular degeneration and paralysis agitans. At the same time that a special attempt was made to define the clinical syndromes of dystonia lenticularis (Thomalla) and double athetosis, attention was called to their common underlying physiologic mechanism and anatomico-pathologic substratum. The conviction has gradually arisen that the numerous clinical entities are not quite so capable of sharp delimitation as was originally thought. Attention was even drawn to the fact that disease of the liver may occur not alone with

4. Wilson, S. A. K.: On Decerebrate Rigidity in Man and the Occurrence of Tonic Fits, *Brain* **43**:220, 1920.

5. Goodhart and Tilney: Brady Kinetic Analysis of Somatic Motor Disturbances, *Neurol Bull.* **3**:295 (Sept.-Oct.) 1921.

lenticular degeneration but with dystonia musculorum deformans as well, thus showing that they are related in another way. Further, it was shown that far from being limited to the corpus striatum, the lesion in dystonia may be very diffuse. A recent review of the clinical syndromes of the corpus striatum by Lhermitte⁶ lent but little clarity to the situation and only served to show that the same kinetic disturbance may be due to different lesions and that the same lesions may give rise to different kinetic disturbances. In other words, in the present state of our knowledge, it is futile to attempt too sharp a definition of the clinical or pathologic syndromes.

As few cases of dystonia have come to necropsy, Wimmer's fairly typical case may be cited in connection with the rather universal tendency to consider the disease one of basal ganglion, more especially lenticular, origin. On microscopic study Wimmer found cellular degeneration and neuroglial changes in the caudate and lenticular (putamen) nuclei, in the dentate nucleus of the cerebellum, in the thalamus, pons and cerebral cortex. The changes simulated those seen in pseudosclerosis and those found in the striatum were not more marked than elsewhere. Wimmer further quotes Spielmeyer to the effect that one might regard Wilson's lenticular degeneration, pseudosclerosis and torsion spasm as "variations in clinical expression of a pathologic process which is essentially the same."

Hall⁷ has also pointed out that the pathologic process is not limited to the lenticular nucleus, but involves the pons and cerebral cortex as well. He further states that degeneration of the liver may be found alike in progressive lenticular degeneration, pseudosclerosis and dystonia musculorum deformans. Schneider⁸ adds that hepatic cirrhosis may be found in dystonia lenticularis as well as in the syndromes just mentioned. In Wimmer's case, too, the liver was cirrhotic.

It may be of interest to point out that the syndrome under discussion is termed either *dysbasia lordotica progressiva* or *dystonia musculorum deformans*. There is no question that they are one and the same clinical entity. And yet one name stresses the peculiar postural attitude in the gait and the other emphasizes the abnormal movements. The important fact is that these two phases have been noted, although they have not been correlated.

6. Lhermitte, J.: The Anatomical and Clinical Syndromes of the Corpus Striatum, *Neurol. Bull.* **3**:163 (May) 1921. Translated from the *Annales de Medicine* of August, 1920, by Huddleston and Kraus.

7. Hall, H. C. (of Copenhagen): *La Dégénérescence Hepato-lenticulaire*, Paris, 1921.

8. Schneider, Erich: *Torsionspasmus: ein Symptomkomplex der mit Leberzirrhose verbundenen progressiver Lenticulardegeneration*, abstr., *Neurol. Zentralblatt* **39**: (April) 1920.

The occurrence of a parkinsonian tremor in a true case of dystonia (Case 6) may serve as a further link in the chain of striatal syndromes. Paralysis agitans may be used by way of analogy to show that it is quite possible for either a kinetic or static phase to dominate a clinical picture. Just as there are cases of paralysis agitans which are characterized by tremor and others by the loss of associated movements and postural attitudes, so there are cases of dystonia which are made prominent by the abnormal movements and others which are signaled by the postural attitude.

CONCLUSIONS

1. There exists a myostatic variety of dystonia musculorum deformans as contrasted with the usual myokinetic form.
2. Phenomena of decerebrate rigidity may frequently be observed in dystonia musculorum deformans.
3. All cases of dystonia have an underlying postural background, one of the manifestations of which are the phenomena of decerebrate rigidity.
4. The myostatic and myokinetic phases of dystonia, which may be observed in all cases, are capable of dissociation. Either the static or kinetic phase may dominate the clinical picture.
5. The involvement in dystonia may be segmental in character.
6. In dystonia there occur not infrequently signs of other striatal diseases which point to a common anatomic and physiologic relationship.

DISCUSSION

DR. J. RAMSAY HUNT, New York: I should prefer to use the terms "rigid or paralytic type" for the group of cases described by Dr. Wechsler. If I am correct in my conception of the efferent system and its division into a kinetic and a static system, we shall in the future recognize groups of symptoms referable to the static mechanism. The corpus striatum is essentially a kinetic mechanism. When there is a striatal paralysis there follows naturally postural fixations, just as after hemiplegia from lesions of the corticospinal system. Therefore, why should the term "myostatic" be applied to express a paralytic manifestation of a kinetic mechanism?

In other disorders of the corpus striatum, for example paralysis agitans, we recognize a tremor type and a rigid type, and this makes a satisfactory clinical distinction. In my experience with dystonia these rigid or paralytic types represent late stages of the disease not unlike the late paralytic stage of paralysis agitans and athetosis when paralysis and rigidity replace the earlier motor disturbances.

I am, however, in entire sympathy with Dr. Wechsler's division of these two clinical groups, but it seems that it would be better to speak of them as a rigid or paralytic type rather than myostatic.