A CASE OF DYSTONIA MUSCULORUM DEFORMANS

By C. C. BELING, M.D.

P. F. Female. Born December 10, 1897, in New York City, of Hebrew parents, both natives of the town of Vilna, Russia.

Family History.—The family history is good. The parents are both living and well. Patient has two brothers and four sisters who are in normal health. Two children, twins, died soon after birth. At ten years of age a maternal cousin developed hemiplegia, following typhoid fever.

Personal History.—Patient was born at full term, developed normally and was considered the brightest and healthiest in the family. It was noticed by the parents that at about 7 years of age she was constantly placing the right foot upon the left, and that soon after the right foot began to turn inwards.

In December, 1904, a week before this peculiar condition was first observed she is said to have been confined to her bed and the house with a fever and sore throat for a couple of days, after which she resumed attendance at school. At this time she was seen by a physician, who said that the ankle was out of place and the foot should be put up in plaster. Four weeks later after the plaster cast was removed, the right foot and lower extremity showed a greater degree of torsion. She placed the outer side of the foot on the ground in walking and standing. The gait was also peculiar. The entire right lower extremity gradually twisted inwards and was carried backwards. Two years later the left lower extremity was gradually affected, the thigh was strongly flexed upon the abdomen and the leg upon the thigh; the foot was extended and turned inwards.

About six or seven years ago she spent varying periods in different hospitals in New York, but her condition did not improve at all.

Four years ago she was put up in plaster in an Orthopedic Hospital in Newark. The plaster cast was applied to the trunk and lower extremities and kept on for a period of three months. On its removal she had abrasions and ulcerations on her back and around the knee joints. No improvement resulted. Soon after she was discharged from this hospital deformity of the spine was noticed. The body began to turn and twist. Twitching jerking movements were marked in the lower extremities. These movements extended to the body and the right arm. Finally the left arm and the neck were also affected. The face was never involved.

In 1908 the parents took her to Sharon Springs for a course

of sulphur baths. During this period she was able to sit up and take her food, using her hands well.

In June, 1909, she was treated at the Newark City Hospital for a period of three weeks. A diagnosis of bilateral athetosis was made at this time. The electrical reactions showed no degenerative characteristics. The patient was examined and X-Ray pictures taken under anesthesia. I regret that the notes then

FIG. 1. Roentgenograms (taken by C. F. Baker, M.D.). Showing slight tilting of pelvis with deformity of neck and upper end of shaft of left femur, with inward rotation.

made at the hospital are unavailable. The radiograms show very well the extent and character of the deformities: the skoliosis, the bony displacement and the torsion of the femur.

PRESENT CONDITION

Attitude. Sitting.—Patient sits in a peculiar bunched-up contorted position, the neck is thrown back, the left elbow joint is strongly flexed and used as a point d'appui, the left wrist is hyperextended, the forearm is in semi-pronation, the hand is used as a support for the head, the fingers are strongly flexed upon the thumb.

The right upper extremity; the forearm is in semipronation

but not to such a degree as the left, the pronator teres and flexors being mostly contracted. She grasps any convenient object with the right hand to further steady herself.

The right lower extremity is flexed at the knee and slightly rotated outwards at the hip; the leg is carried backward until the foot touches the left side of the pelvis.

The left thigh is extended; the knee slightly flexed, the foot in hyperextension and the toes markedly flexed. The right gluteal region is very prominent. Lordosis is hardly present; skoliosis is very marked; the curve is directed toward the left in the dorsal region, the body is strongly flexed upon the lower extremities.

By supporting her left elbow upon the sofa and placing the left hand upon the chin she is able to obtain some rest for a few minutes.

In sitting the lordosis disappears, but the kypho-skoliosis persists presenting a deformity which suggests a camel's hump.

Movements.—In the sitting posture, when patient is at complete rest the movements are very slight at times, while at other times they are more marked. These movements are of a tonic and clonic character. They affect all the extremities, the neck and the body, but the face is free. During the last six months the movements have improved a great deal. They are generally less all over the body, especially in the legs. The right upper extremity has improved to such an extent that she is now able to draw somewhat.

The characteristic feature of the case is the extreme tonic spasm, which persists almost continuously, but varies greatly in intensity. As a result of this spasm very severe distortion of the extremities, neck and trunk have taken place. With this tonic spasm, there are to be noted clonic movements and movements of a jerky character, at times resembling those of chorea and at other times like those of tic. Added to all these voluntary movements complicate the picture.

There is a certain conflict between the voluntary movements and the tonic spasms, which may be noted in the bizarre useless motions that are gone through when the patient attempts to execute any ordinary command. Frequently it is only after much effort that she is able to accomplish the desired act.

The tonic spasms are most marked when the patient attempts to stand or walk. During sleep all movements cease.

She is able to walk only one or two steps without any assistance, but with slight assistance, for example, touching the furniture, she is able to go from one room to another. The gait is bizarre and suggests the walk of a dromedary.

The limbs on the left side are most affected. Abduction of the left foot is impossible, but extension and flexion of the toes is preserved.

Except for the characteristic positions of the upper extremities the movements in them practically cease, when the patient is at rest in bed. The tonic spasmodic movements in the lower extremity still persist, and clonic components are most marked in the left leg. There is a hypertonia in the legs, especially in the calf muscles and anterior groups, but the muscles can be distinctly palpated under the skin. The left foot is in the equinus position, the right exhibits a talipes equino-varus.

FIG. 2. Marked scoliosis of dorsal spine—approximation of ribs on right and separation on left.

At rest the inner hamstring muscles of the left thigh are mostly in strong tonic contraction. This is replaced from time to time by a short tonic contraction of the quadriceps group.

In sitting the lordosis disappears, but the kypho-skoliosis persists.

She is able to rise well from the dorsal position. The right upper extremity is at present the least affected. One occasionally sees jerky movements in it. It is quietest in a semipronate position.

In the prone position the lordosis is increased, especially when the muscles of the back are thrown into spasm, but the skoliosis is not so prominent.

Movements are all present in the ankles although much restricted.

Reflexes.—The knee jerks are present on both sides and are obtainable without reinforcement. The plantar reflexes are flexor in type. Achilles reflexes are obtainable at times. The right Achilles tendon reflex is only weakly present, the left is more active. Both are apparently diminished.

MEASUREMENTS

	R.	L.
Upper arm	8½ in.	8½ in.
Forearm	8¼ in.	8 in.
Wrist	6½ in.	6½ in.
Thigh	12½ in.	12¼ in.
Calf	8 in.	8 in.
Ankle	5½ in.	5½ to 6 in.

FIG. 3. Lateral view of ankle, showing displacement of tarsal bones due to talipes equino-varus.

The measurements do not show any marked difference. They are not as accurate as they may be, owing to the difficulty of measuring due to the constant movements of the limbs.

The cranial nerves are normal, no cerebellar disturbances. No sensory disturbances. Speech is normal. The breasts are well developed. No thyroid enlargement. The thoracic and abdominal organs show no gross lesions apart from their displacement due to the excursions of the torso.

Wassermann test is negative. The blood examination shows a secondary anemia of the chlorotic type.

REMARKS

The history of this case shows that an insidious disease appeared in a hitherto normal girl of 7 years of age, of Russian Hebrew parentage, without any hereditary degeneracy.

Its chief feature consisted in the appearance of a progressive generalized muscular spasm of a tractile and torsion-like character.

The disease began in the right foot and exhibited itself as a disturbance of gait in which hypertonia and spasmodic components were evident. These spasms produced peculiar deformities in the trunk and extremities.

The movements were almost always slow, rather stereotyped and not so graceful and rhythmic as those of bilateral athetosis. Occasionally jerky muscular contractions were observed. The muscular spasms were not under the influence of the will.

In the early stages voluntary movements were not seriously disturbed, but at the present they are very strongly interfered with.

Complicated motor acts, such as rising, walking, eating, etc., have been profoundly affected.

There has been no diminution in the power of the muscles, except for those produced by the contractures.

The movements cease during sleep.

The spasmodic of the right upper extremity has shown a slight degree of regression.

The intellect has been entirely unaffected. The emotional state has been normal. Neither suggestion nor hypnosis has had any influence. There have been no hysterical stigmata.

The disease shows a marked similarity to bilateral athetosis, chronic chorea and degenerative tic.

The characteristic participation of the facial muscles, the peculiar grimacing, noted by Lewandowsky, the combination with it of signs of spastic diplegia, which is a frequently associated phenomenon, the enormous influence of emotional processes, seen in bilateral athetosis, are lacking in our patient.