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DYSBASIA LORDOTICA PROGRESSIVA, DYSTONIA MUSCULORUM DEFORMANS—TORTIPELVIS¹

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On the basis of four cases and because the literature is silent on the subject Oppenheim² attempts to separate from the illy defined and extensive genus of "tics," "myospasms," "choreas," "athetoses," "spasmodic hysterias," a nosologic variety under the name: "Dysbasia Lordotica Progressiva—Dystonia Musculorum Deformans."

He characterizes this condition as follows: It occurs in children and young adults of the Jewish race, and consists essentially of (*a*) a deformity around the pelvis, and (*b*) of tonic and clonic myospasms of the musculature around the pelvic girdle associated or not with similar twitchings of other muscles.

The difficulty begins frequently with slight symptoms in the upper extremities; but wherever it begins the lower extremities are permanently and severely affected—the musculature of the thigh, pelvis and lumbar part of the vertebral column.

In reclining position most of the symptoms disappear, except a variable degree of deformity around the pelvis. Attempts to stand—particularly to walk—activate the condition.

¹ Read before the New York Neurological Society, December 5, 1911.

² "Über eine eigenartige Krampfkrankheit des kindlichen und jugendlichen Alters (Dysbasia Lordotica Progressiva, Dystonia Musculorum Deformans)," *Neurologisches Centralblatt*, October 1, 1911.

A marked lordosis of the lower dorsal and upper lumbar parts of the vertebral column, with inclination of the pelvis and particu-

FIG. 1.

lar prominence of the buttocks, are, as it were, the stigma of the condition (Fig. 1).

The most characteristic disturbance is noticed in the gait of

these patients. Oppenheim calls it the most curious thing he ever encountered, surpassing the deformities, distortions and compensatory motor efforts seen in cases of poliomyelitis and dystrophy. The gait is very bizarre, resembling the locomotion of a quadruped; best described as "monkey gait," "dromedary gait."

At first sight the suggestion of a hysterical nature of the condition is very strong, and indeed this diagnosis is most frequently made.

During locomotion the attitude changes continuously, getting clownish in character; the patient soon shows evidences of strain, the face flushes, the pulse rate is increased, and profuse perspiration sets in.

In the recumbent position most of the deformities disappear, particularly the lordosis, but the legs even in the supine position do not get entirely relaxed. In all Oppenheim's cases the upper extremities were involved also, but to a far less extent. Signs of organic damage to the nervous system are absent. Variations in the tendon reflexes are noted; but these are transient and not uniformly present.

Oppenheim is not willing to look upon this condition as a neurosis, but is convinced that it has a definite anatomic basis, the identification of which he confidently expects. He believes, as the name implies, that we are dealing with a disturbance of the coördination of muscle tone, with a state of essential hypotonia.

According to Oppenheim the only mention that the literature contains on the subject is found in a communication by Ziehen,³ who describes an analogous state and calls it "tonic torsion neurosis."

The etiology of the disease is unknown; it runs a chronic, progressive course, and treatment has no influence. Various methods show transient benefit. In one of Oppenheim's cases the symptoms disappeared after metallotherapy for a time. In one of my cases reëducation helped for a time, and so did intraspinal injections of magnesium sulphate in two for a shorter time.

During the last few years four similar cases came under my observation which I believe it worth while to place on record.

CASE I. The first case was presented before this Society at the meeting of October 6, 1903, by Dr. Leszynsky, to whom I am indebted for the following notes.

³"Allgemeine Zeitschrift f. Psychiatrie," LXVIII, *Neurol. Centralblatt*, 1911, S. 109.

J. G. (Fig. 2), born in the U. S., 12 years of age, Christian, is the sixth and youngest child in the family and was born at full term by forceps delivery, without suspended animation. Mother died seven years ago, cause unknown. Father is alcoholic. Other children in good health. Patient has never had convulsions. He was always of average intelligence and learned to talk and walk early. Nothing abnormal was ever noticed, and he attended public

FIG. II. Case I.

school. He always showed great muscular development and was unusually strong for his age. He seemed perfectly well until fourteen months ago, when he began to walk in a peculiar manner with his body bent forward. Soon after he told his family that a boy had kicked him in the back. He also stated at the hospital that he had seen an old man in the neighborhood who walked in

a similar way, that he had imitated him, and then began to walk likewise. But his relatives when questioned had never heard of this before. This statement was not made by the patient spontaneously, but was evidently elicited as an affirmative reply to a leading and suggestive interrogation, and is therefore of questionable value.

He was admitted to the Lebanon Hospital March 9, 1903, and found to be in good general health. In recumbency, or while lying on his abdomen, the trunk and extremities remained in a horizontal and relaxed position. When he attempted to assume an erect posture or to walk, the trunk became flexed on the thighs almost at a right angle, and the head semiflexed on the thorax. In this attitude he walked about groping with his hands, but he managed to get around with considerable ability without injury.

Upon examination both lower extremities became rigid when placed either in extension or flexion, and the knee jerks were unobtainable by any form of reinforcement, as there seemed to be an excess of cerebral inhibition. His muscular power and development were remarkably good, and a thorough investigation of the nervous system and of the vertebral column, etc., proved absolutely negative as to evidence of organic disease. Several days later, the knee jerks were successfully demonstrated. After remaining in the hospital 16 days he cried and begged to go home, attempting to run away, and was discharged completely recovered, being able to walk in a normal manner. During his sojourn in the ward it was often noted that he could walk erect when he thought he was unobserved.

He remained well for several months, running about the streets like other boys. He was readmitted to the hospital July 1 in the same condition as at first described, but of a more exaggerated degree. His grotesque attitude and the result of examination were practically the same as at the first admission, but more force was required to overcome the flexion and hold him in an erect position. Pulse has ranged from 84 to 100. T. and R. normal. Urine of late has shown on excess of indican. Blood: Hmg. 90 per cent.; no leucocytosis.

Recent Examination.—Lower extremities: Muscular power and resistance in both good. Motility in all joints normal; occasional slight transient rigidity of extensors during manipulation; both knee jerks +, left ++ with ankle clonus fairly well marked but of a spurious type. (This is not always demonstrable.) No clonus on right; both Achilles reflexes present, left +; both plantar reflexes absent; abdominal and cremasteric reflexes normal; no objective sensory disturbance; visual fields not tested; no complaint of pain or sensitiveness over vertebrae; during sleep nothing abnormal has been noticed. He can suspend himself by the hands and by the weight of the body maintains an erect attitude for

several minutes. There is no evidence of spasmodic flexion while in the dorsal or prone position.

It will be observed that the sudden flexion of the pelvis on the thighs only takes place in attempts at standing, the condition corresponding somewhat with that described by Charcot many years ago as a flexion type of *astasia abasia*. For want of a better name we must call it *hysteria*. (In hospital about six months—no improvement.)

The same patient was shown a second time before this Society on May 5, 1908, by Dr. J. R. Hunt, who said:

The patient, 15 years old, commenced to show clonic muscular contractions of the trunk muscles in January, 1903. At that time he showed a tendency to stoop and bend forwards; this gradually became more marked, and finally his body became fixed a large part of the time in this bent position while sitting, standing or walking. Various methods of treatment were tried without benefit, and the patient was finally subjected to a long-continued course of psychophysical therapeutics by Dr. R. B. Kruna, with decided improvement, as he was now able to get about in a fairly comfortable manner. It was because of this very considerable improvement and the method of treatment employed that Dr. Hunt brought this case before the Society. He regarded it as an unusual type of *myoclonia*, a form of *tic convulsif* involving the muscles of the trunk. There were no evidences of any organic diseases of the nervous system, and at no time were there any stigmata or crises suggesting an hysterical origin. Mentally the boy was bright and intelligent and there were no impulsions, obsessions or phobias, such as not infrequently accompany the generalized forms of *tic convulsif* in early life.

Dr. Richard B. Kruna, in demonstrating his method of treatment in the case shown by Dr. Hunt, said that when the boy first came under his observation the body was bent forward to an angle of about 80 degrees to the legs; at the same time the spine showed lateral flexion and rotation, as well as the neck. The *tic spasms* from which he suffered had developed during six months, and had gradually increased in force. The boy had no pain, but in walking the spasms pulled his trunk and head downwards until he was practically doubled up, with his head near his knees. In this position he would brace himself, with one hand upon his knee and the other upon some object near him, such as a table, chair, etc., or upon the ground. With these disadvantages, his gait was of course irregular and jerky, yet surprisingly fast ("monkey-gait"). A careful examination of the patient showed nothing abnormal organically, excepting an increase of indican in the urine and ankle-clonus of the left foot.

To find relief from this distressing condition, which had forced him out of school and rendered him an object of ridicule and even

fright, he spent over four years at various orthopedic and general hospitals. Every resource of orthopedic surgery was tried, including a variety of plaster-of-Paris jackets, steel braces, extension under anesthesia, and finally an operation, consisting in division of the rectus and ileo-psoas muscles. All these were without effect, as was also an injection of magnesium sulphate solution into the spinal canal. The latter caused temporary paralysis, but did not cure him. (Fig. 3, Case I.)

The patient was referred to Dr. Krupa at Dr. J. Fraenkel's suggestion over two years ago for psychophysical treatment: this has been faithfully carried out, and the patient was now able to stand and walk erect, and in the course of another year, the speaker said, his condition would probably be entirely normal. At present, he was able, by voluntary muscle action, to antagonize every tic contraction, but this balancing activity had not yet been developed to the point of being automatic, as it normally should

FIG. III. Showing Case I after Magnesium Sulphate Injection.

be. The speaker then explained briefly the psychical régime which was part of the psychophysical therapeutics, which included the development of the patient's intellectual and moral strength, the training of his powers of initiative and inhibition, regularity and system in habits, etc.

The physical training consisted in medical gymnastics, starting from an analysis of the tic attitude into its components, the elementary muscle-spasms. Each of these tic elements had to receive a remedying antagonistic muscle-exercise. At the onset, the patient was not able to make the antagonist of a tic-muscle contract by itself. This required a series of assisted exercises. In these the patient was helped by an assisting force, outside of his own muscular effort, to overcome the tic-spasm. The percentage of efficiency of the patient's own efforts would show increase according to their repetition, and the assisting force was correspondingly diminished. Finally, the assisted exercises became unnecessary and were replaced by unassisted ones. The

first landmark of progress was the ability to overcome every tic element by its antagonistic action, and every tic attitude by its counter attitude; the second, identical with reestablishment of the normal condition would be reached when these voluntary antagonistic actions, by untiring exercise, had become automatic.

Dr. William M. Leszynsky, who had presented the same patient at a meeting of this Society several years ago, said that Dr. Kruna was certainly to be congratulated upon the brilliant result he had obtained from his psychophysical method of treatment. When Dr. Leszynsky had him under his care there was some improvement after a similar method of treatment in the hospital, but within five months he relapsed into his original condition. He was then sent to the Hospital for Ruptured and Crippled, where he was fitted with various kinds of apparatus, and several muscles tenotomized without relief. He was finally transferred to the Montefiore Home. The diagnosis of the case at that time was hysteria, and his physical condition was associated with a peculiar mental attitude which seemed to point to some form of degeneracy.

This patient came under my observation simultaneously with the two subsequent ones in 1905. I can add little to the description so far reported. Both cases (cases I and II) we treated by intraspinal injection of magnesium sulphate. The effect upon the deformity and spasms was certainly striking, although temporary and very transient. I refrained from repeating this procedure because the reactions were, at first, alarming: fever, embarrassment of respiration, and slight coma.

In discussing these cases at the time with Dr. Dana we were impressed then by their individuality, and I thought they might be designated as cases of "tortipelvis."

Very lately when I saw J. G. again his condition was practically the same. While in my office he showed the most bizarre contortions, particularly on the left side of the body. He walked out in a "quadruped" fashion, alighted from his bicycle with dexterity and alacrity, and evidently found his way through the crowded city traffic.

CASE II. For the abstract of the history of this patient I am indebted to Dr. J. Wachsmann of the Montefiore Home.

J. S. entered the Montefiore Home November 3, 1905; discharged March 12, 1907; 24 years old on admission. Family history negative. Previous history: born in Washington, D. C. Jewish. Measles at 3. Malaria contracted in Spanish-American war. Suffered for one year from it. Habits, good. Occupation, electrician. No venereal infection. (Figs. 1, 4, 5.)

Present illness began two months after onset of malaria (5 years ago), with tremor of left toes, exaggerated by emotion. Within one year muscles of left lower extremity became rigid. Two years after onset ankle became inverted. Deformity could

be overcome actively in sitting or recumbent posture, but not in standing. About same time body began to arch backwards. Lost 30 lbs. in weight. Anorexia, constipation. No pain. Eyesight failing for the last five months. Complains of cramps in muscles of lower extremities.

FIG. IV.

January 29, 1906. Father was once paralyzed in all four extremities, but made perfect recovery. Uncle on father's side was affected with tic of left eyelid.

Physical examination November 3, 1905. Gait: as if patient

FIG. V. Showing Case II after Magnesium Sulphate Injection.

were losing his equilibrium. Long steps. Walks on outer edge of left foot. Deformity reducible—no contracture. Reflexes normal. No shortening. About $\frac{1}{2}$ inch atrophy in left thigh. Joints free. Electrical reaction negative. Eyes: pupils dilated $R > L$ —react. Throat: pharyngeal reflex present. Sensibility: (1) Hypalgesia, distribution of both nervi cutanii femoris; (2) hypalgesia of left side of face.

February 8, 1906, 6 c.c. of mg. sul. 25 per cent. solution injected into spinal canal with result as follows (Fig. 4): (1) Stands perfectly straight; (2) gait more steady; (3) pseudo-contracture much diminished; (4) cannot pass urine voluntarily. Some anesthesia of tip of penis. Examination of urine, blood, gastric contents and feces negative.

February 2, 1906. Examination of excised piece of muscle shows no atrophic changes, but possibly some parenchymatous degeneration.

The patient was discharged in better physical condition, but the disease unchanged.

CASE III. School boy, eleven years of age, born in the U. S., Christian. One year ago last January was perfectly well. Began to walk lame in left leg two weeks after a fall down stairs leading from one story to another. He was not hurt at the time, and walked as well as ever immediately after the accident. He complained of getting tired, but never of pain. Lameness grew progressively worse for about 4 to 6 weeks, then stationary. Walked as though loose from the hip-joint, and throwing leg out to the side, but not on the side of his foot; has been able to walk and run during his illness.

Had a hip and leg spica plaster cast put on in February (20) by Dr. Ashley of New York. He walked better with the cast, no longer threw his leg out.

In March, 1910, left arm began to show symptoms—could not button clothes as well, and was awkward in movements with left hand, but can play baseball. Two weeks ago tremor in arm was first noticed, although patient had complained of jerky, nervous feelings in both leg and arm for some time before. During period when he wore cast he began to walk on outer side of foot. No atrophy noticed at any time. No pain at any time. Very seldom had headache, and these very mild and "probably from stomach." No vomiting or dizziness. Eyesight good. Good appetite. No cough. No chills or fever. Was in best of health all his life. No paresthesias. Nocturnal enuresis for 3 years at intervals. No incontinence of stool.

Father and mother alive and well. Sister one, well and strong. No hereditary history, all disease denied, 9 month's infant, normal birth. Fourteen months walked. Talked right time. Teeth at right time. No convulsions. Control of bladder before two years old. Measles 4 years old. Three or four years ago had

FIG. VI. Case III.

chills and fever, one part of day quiet with malaise, other part felt bright. Doctor called it "intestinal fever." Was constipated. Was in bed a week. Perfect recovery. Education, good scholar.

May 3, 1910. Measurements of calves, R. 25.7, L. 26.3. Left calf looks fuller and rounder. No limitation of motion of left leg. Muscular power of left leg about equal to right. Thigh, R. 34.5 cm., L. 31.7 cm., 12 cm. above patella. Muscles of left leg are in almost constant spasm. Least movement causes tonic spasm especially around hip-joint. Muscles feel rigid, hard, especially adductors. Leg cannot, apparently, be flexed on abdomen, due evidently to spasm of extensors. Leg is held somewhat adducted, and rotated inward. Patient walks on outer margin of foot. Left arm is more or less constantly in state of clonic pronator spasm, fingers flex slightly. Can stop by closing hand firmly and making arm rigid. Power excellent R=L.

May 12, 1910. X-ray taken on May 6. Report as follows: There is a slight decalcification of the head and neck of the femur, and periostitis extending from the head to the greater trochanter, along the upper surface of the neck. There is apparently a tilt of the pelvis, and I believe confirmatory plates should be made higher up to determine whether there is any involvement of the spine.

CASE IV. L. R., 12 years old. Jew. Was seen by Dr. Gibney.

Born naturally; when 10 months old fell from a chair and had convulsions until teeth appeared. When three years old was run over right leg. At another time fell from a wagon. Still another time had some bicycle accident. During one year gait became impaired.

Well developed and nourished. Some swelling in region of right groin. Gait and attitude show marked kypo-scoliosis—lumbar. On walking spasmodic adduction—rotation. Is afraid to be alone in the room. Plantar reflexes—Babinsky type.

The histories of these patients have undoubtedly a definite ring of a family likeness. This impression is certainly strengthened by an inspection of the photographs of the cases here reported, and the one taken from Oppenheim's publication. The resemblance becomes striking indeed.

Upon analysis it becomes evident that we are dealing with a condition showing the following characteristics: (1) The symptoms are variable, fugacious and intermittent for a time. (2) Early they show a local predilection. (3) A permanent deformity around the pelvic girdle. (4) Spasmodic phenomena associated with function.

I believe that the analogy with essential torticollis forces itself upon one's attention. Possibly here, as in torticollis, the future

will show that there may be a symptomatic and essential variety. Indeed in my last two cases this thought occurred to me on account of the traumatisms to the hip which preceded the onset of symptoms and the X-ray findings in one case.

Etiologically no definite factors are so far apparent. The disease is not confined to the Jewish race. Malaria was noticed in two cases, and traumatism in two of my cases. In one case an hereditary factor is elicited; hysterical paralysis in the father and tic in the uncle. Apparently the disease develops upon a "tic" "spasmophile" soil, precipitated by various provocative causes.

If the Chwostek phenomenon proves to be as it is claimed, the signature of the spasmophile disposition, the examination for this symptom should be added to the routine examination of all tic cases.

The differences in the severity and localization of the tic symptoms could be explained by (*a*) the difference in the degree of the predisposition, and (*b*) by the degree of the local disposition, in consequence of function or overwork of the given parts, according to the theory of Edinger.

Thus in childhood and youth, when the facial and later the musculature of the pelvis and lower extremities are mostly used, facial and pelvic tics will be more liable to develop; and in later life tics of the shoulder girdle and upper extremities.

Such a widespread constitutional condition it would be difficult to identify with a uniform causative anatomic lesion. The lesion—structural—if found, will have to be interpreted as sequential.

It would seem more reasonable to look for the cause of such disorders in the epithelial bodies and parathyroid glands, in changes of their product and the influence upon the mineral metabolism, particularly the calcium metabolism. The coagulation time of the blood and the study of the calcium metabolism should be made part of the routine examination of the tic cases.

Finally, bearing in mind the fact proven by Jacques Loeb that the orderly, rhythmical function of muscle is dependent upon its being bathed by a serum containing definite proportions of mineral ions, systematic therapeutic efforts along these lines suggest themselves.

Regarding the classification and nomenclature it would seem safest for the present to classify the condition among tics and spasmophilias, and to individualize it by the name "tortipelvis."