THE SURGICAL TREATMENT OF DYSTONIA MUSCULORUM DEFORMANS*

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THE severe disability of patients afflicted with dystonia musculorum deformans arouses the urge to give them relief even at the cost of other neurologic sacrifices. Unfortunately, the etiology of the disorder is too poorly understood at present to permit a specific attack upon brain areas responsible for the abnormal motion.

The bizarre, yet stereotyped movements do not characterize a disease, but constitute a syndrome for which no specific pathologic lesion is known. The syndrome is considered to operate primarily via the extrapyramidal system. The abnormal involuntary motor activity is regarded as a release phenomenon, the result of loss of the inhibitory action of higher controlling centers. The motions persist too long to be reasonably attributed to an irritative lesion.

Dystonia consists of slow, long-sustained turning movements of the head and trunk, with rotation of especially the proximal parts of the extremities. In addition to these torsion movements, there are slow, sustained tensions in the platysma, shoulder muscles, pectorals and leg and foot muscles. Particularly long-sustained, simultaneous tensions of opposing muscle groups characterize dystonic movements. Physiologically, they resemble those of athetosis, but are longer sustained.

This report concerns the results obtained by surgical interruptions of pathways at several levels in the central nervous system for relief of severe dystonia in 3 children.

CASE REPORTS

Case 1. D.Y. was first seen at the age of 7 years. For a year, she had suffered from spasticity of both arms and from cramping spasms of the feet causing pain that made her cry at night. Within 6 months dystonic movements became pronounced.

Over the next 5 years the dystonia progressed. The head retracted, and the extremities were maintained in semiflexion and were relatively fixed. The facial muscles were not involved, but those of the trunk and extremities forced the body into grotesque postures. These movements were extremely painful and the child was reduced to an emaciated state. Extensive drug therapy was ineffective and she was admitted to the Children’s Hospital at the age of 12 years to permit attempts at surgical relief, especially of her pain.

Operations. Encouraged by the results obtained in cases of unilateral Parkinsonism, on July 11, 1946 we first partly severed the anterior limb of the right internal capsule by the method of Meyers and Browder. This did not alter the patient’s condition.

On July 22, 1946, the right area 6 was removed, sparing area 4. This abolished the dystonia and permitted weak voluntary motion of the hand. The right-sided dystonia increased, especially in the foot.

Fearing the effect of a bilateral cortical resection, since the child was intelligent, we sought control of the right side in an attack at the cord level. On July 30, 1946 the right anterior cord fasciculus was cauterized (Putnam’s method) at C1–2 level. This left the child comfortable. The arms and legs were maintained flexed. Right grip was possible and the arm could be extended without eliciting dystonia. Involuntary movement was present in the right leg. Passive extension of the left arm resulted in a strong, coarse tremor and in flexion of the extremities. The neck still retracted to the left moderately under tension or emotion. A more complete denervation of the left side seemed desirable.

On Aug. 28, 1946 the right area 4 was excised above the level of lip response to stimulation. This resulted in a left hemiplegia, without tremor or reflex spasm.

In the next 9 months all gains were preserved; the child gained 20 pounds, was pain-free, friendly and comfortable in a wheelchair existence. Involuntary movement followed use of the right leg, which was flexed at knee and thigh. In an attempt to relieve this, the right pyramidal tract was severed at the 3rd dorsal level on May 12, 1947.

Course. Two years after the last operation (May, 1949) the 15-year-old girl was happy and pain-free. She was 50 pounds heavier than at the start of her surgical “career.” Her I.Q. still measured 110 and she was a freshman in the Crippled Children’s School. She subsequently has been graduated. Her head was tilted slightly to the right. The spine was in marked scoliosis. She could stand in braces, but could not walk. No dystonia nor resting tremor was present. Right grip was satisfactory and she could write legibly but slowly.

Within the last 3 years, coarse tremor and some dystonia have appeared in the right arm and have progressed until the extremity is almost useless. A left pedunculotomy is being considered to improve this.

Comment. This child has been observed over a period of 10 years. Primarily to relieve pain, successive surgical sacrifices were made to include finally: The right cortical areas 4 and 6, the right extrapyramidal tracts (anterior cord fasciculus) at C1–2 and the right pyramidal tract at D3 level. These resulted in abolition of the dystonia at the cost of a left hemiplegia and a right hemiparesis. The end result was happily accepted by the patient and her relatives.

Case 2. J.N. was a 9.5-year-old boy, admitted to the Children’s Hospital on April 25, 1949. The patient was one of twins, the sister being normal. He weighed 3 pounds (1.4 kg.) at birth and lived in an incubator for 3 months. During infancy and after, he had difficulty in eating, eye movements were uncoordinated and his extremities displayed involuntary movements. Despite 4 years of intensive cerebral palsy management his condition deteriorated. For 8 months he had suffered frequent painful attacks of opisthotonos and flexion of the knees and hips.