DYSTONIA MUSCULORUM DEFORMANS ALLEVIATED BY CHEMOPALLIDOThALAMECTOMY AND SUBSTANTIA NIGRALYSIS

CASE REPORT

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Dystonia musculorum deformans, as the name implies, is generally a crippling and progressive neuromuscular disorder. It results in grotesque postures and finally in permanent skeletal changes, such as rotokypnoscoliosis. It is primarily a disorder of childhood and adolescence and therefore the more devastating. With the use of chemopallidectomy and chemothalamectomy introduced by Cooper,¹ the majority of such patients can be greatly improved if not "cured" of their dystonic syndrome.

In our 6 cases, 4 patients have been completely relieved and 1 considerably improved, and 1 has received no benefit from surgery. One patient required bilateral thalamic lesions which incidentally were done simultaneously; in a second patient, bilateral pallidal lesions were made at 4 separate operations; in a third, bilateral pallidal and unilateral thalamic lesions were produced in 3 operations; a fourth has had unilateral thalamic lesions on 2 occasions with 75 per cent improvement and soon will have a pallidal lesion made. The fifth patient has been the most difficult to treat, requiring bilateral pallidal and unilateral thalamic lesions done in 6 operations. And, in addition, destruction of the left substantia nigra by a new surgical technique—substantia nigralysis.²

This particular case is worthy of detailed review.

CASE REPORT

Dystonia musculorum deformans alleviated after 7 operations over a 21-month period by left chemopallidectomy and substantia nigralysis, and right chemopallidothalamectomy.

A 38-year-old Jewess of Russian descent, had gradual onset of involuntary and uncontrol-

able turning of the head to the left in September, 1956. Within a month she noticed a "pull-

ing sensation" in the right arm which soon developed into dystonic posturing and spasms causing inability to hold on to small objects. By December the axial musculature and right lower extremity had become similarly involved with dystonia. It became necessary for her to wear a cervical brace constantly and use a cane to get about.

Examination. The patient was an oriented, intelligent individual with a passive aggressive disorder of character. The cranial nerves functioned normally. In spite of the cervical brace which she wore constantly the head tilted and twisted to the left. It was impossible to rotate it beyond the midline because of increasing spasms of muscles. The right arm was held in a slightly flexed hyperpronated posture with marked flexion of the fingers. Passive supination caused aggravation of the dystonic spasms and pain in the muscles. Moderate scoliosis of the thoracic spine was accompanied by severe lumbar lordosis, most prominent when the patient was in an erect position. The right hip and knee remained slightly flexed with the foot strongly everted. This posture was exaggerated when the patient walked. Although the muscle tone in the right side was increased, no abnormal reflex changes were elicited on either side. Dystonia was not observed in the left extremities. Except for some mild hypalgesia on the right side sensory findings were normal.
From psychiatric examinations and Amytal interviews over a week’s period it was concluded that the patient suffered from dystonia musculorum deformans rather than any primary psychogenic disorder, thus substantiating the opinion of those neurologists who had examined her.

Laboratory studies including complete blood count, urinalysis, serum electrolytes and cerebrospinal fluid disclosed no abnormalities. An electroencephalogram was normal. The electromyogram (using disc electrodes) demonstrated fairly constant, extremely high potential activity on the right side.

Operation 1. On Oct. 31, 1957, using transtemporal approach under general anesthesia, two plain Cooper cannulae were placed in the left globus pallidus. The first was in a coronal plane at the posterior wall of the foramen of Monro; the second 5 mm. behind the first. Following injection of 0.3 cc. of 1 per cent procaine into the second cannula the dystonic symptoms abated completely. Therefore 0.8 cc. Ethocel* was injected fractionally over an hour. Subsequently there developed a mild right facial weakness and loss of memory lasting about 5 days. The torticollis and spasms in the right hand and foot returned on the 8th postoperative day. An additional 0.2 cc. of Ethocel in the posterior cannula relieved the dystonia except in the right foot. This procedure was again accompanied by mild facial paresis and loss of memory for 3 days. On the 15th postoperative day 0.3 cc. of 1 per cent procaine injected into the anterior cannula abolished her residual dystonic symptoms; 0.5 cc. of Ethocel was then injected over a 50-minute period.

On discharge from the hospital on Nov. 16, 1957 neurological findings were within normal limits except for slight right facial weakness.

Course. After leaving the hospital she did well, even returning to her former work as a secretary for a few days. Then, during the 2nd week in December, 1957, dystonic symptoms developed on the left side, with torticollis to the right. Except for the fact that the left foot had become strongly inverted rather than everted, as had been the case on the right side, the neurologic findings were as described during her first hospitalization. An electroencephalogram on Dec. 27, 1957 was normal, of the low fast-voltage type.

Operation 2. On Jan. 2, 1958, a Cooper cannula was placed by the temporal approach into the right globus pallidus 1 cm. behind the foramen of Monro. Injection of 0.6 cc. of 1 per cent procaine failed to relieve the contralateral dystonic symptoms. The cannula was removed.

Operation 3. This was performed on Jan. 7, 1958. Two cannulae were placed in the globus pallidus in a position similar to those for the left hemipallidectomy, namely in the coronal plane of the foramen of Monro and 5 mm. posterior to it. Immediately following the injection of 0.3 cc. of 1 per cent procaine through the anterior cannula the dystonic postures and spasms ceased. Accordingly, 0.5 cc. of Ethocel was injected over a 25-minute period. The patient had some difficulty in recognizing her physicians and friends and in addition showed lack of affection toward her sons. Dystonic movements began to recur in the left hand. These were abolished by an additional 0.5 cc. of Ethocel. However, the patient experienced severe loss of memory for recent events, which cleared in 72 hours. Eight days after the last injection dystonia had recurred to a moderate degree. Instillation of 0.5 cc. of 1 per cent procaine through the posterior cannula abolished these recurrent symptoms. Therefore, 1.0 cc. of Ethocel was injected over a 60-minute period. She became confused and disoriented. This mental clouding cleared over a 7-day period. However, mild dystonia then returned to the left side.

The patient was discharged from the hospital on Feb. 2, 1958, improved, but with definite relics of the dystonia and some blunting of affect. Voluntary muscular strength and sensation were normal. The deep tendon reflexes on the left were somewhat increased over the right side and a positive Babinski’s response was present on the left.

Course. The patient was re-admitted to the hospital for the 3rd time on March 23, 1958, because of increasing symptoms of dystonia in the left extremities and torticollis to the right. The symptoms were particularly prominent in the left leg and foot and lumbar spine. The

* Supplied by Ciba Pharmaceutical Company; a solution of 8 per cent celloidin and 95 per cent ethanol.
right side remained free of recurrent symptoms. In addition she noted some continued difficulty in concentration and memory of recent events. However, she was quite well oriented, alert, cooperative, but depressed over her physical disability. The verbal performance IQ test was 117 as compared to previous 112. Her performance IQ was 82, which represented a decrease since the previous psychological testing done in January. The serial 100-7 test was performed slowly but correctly and the Babcock test sentence could be repeated without error. She seemed to have difficulty in those tasks requiring prolonged attention and concentration.

The dystonia was of moderate degree on the left side, causing hyperpronation of the forearm, flexion of elbow and fingers, exaggerated lumbar lordosis, inversion and plantar flexion of the left foot and mild torticollis to the right side. A mild degree of hypesthesia and hypalgesia with hyperactive reflexes and a mildly positive Babinski’s response were present on the left side.

Operation 4. A transfrontal right chemopallidectomy was performed on March 26, 1958, employing the balloon-type of Cooper cannula. The cannula appeared to be accurately placed. There was immediate abatement of dystonia with distention of the balloon during surgery. Symptoms recurred when the balloon was deflated but again disappeared on re-inflation of the balloon; 0.5 cc. of alcohol-Pantopaque mixture (4:1) was injected 24 hours later. The patient experienced a recurrent gross deficit of memory, particularly for recent events. This cleared in 3 days. Because of return of dystonia in the back and left foot, an additional 0.25 cc. of alcohol-Pantopaque was injected on April 3 and April 9, 1958. Each time the dystonia disappeared but was accompanied by severe loss of memory for recent events and inability to recognize those who were caring for her.

A final injection of 0.6 cc. of Ethocel was given on April 14, 1958. The effect was only transient. At the time of discharge from the hospital on April 28, 1958, mild dystonia had recurred causing semilunar foot and exaggerated lumbar lordosis. Although the patient was depressed she was quite oriented and able to recall recent events and recognize her family, nurses and physicians.

Course. The 4th admission was on June 11, 1958, because of return of severe dystonic spasms and postures on the left side similar in every way to those during her 2nd hospitalization in December, 1957. The patient was oriented and quite able to carry on a normal conversation. She had some difficulty with serial 100-7 test; and recall of recent events and her recognition of persons and places were definitely impaired. The reflexes had returned to normal but the left-sided hypesthesia and hypalgesia had recurred. A mild left facial weakness persisted. A preoperative electroencephalogram was normal except for some rare scattered 5 to 6 c./sec. waves anteriorly.

Operation 5. On June 13, 1958, a right chemothalamectomy was performed. The cannula seemed to be placed accurately in the nucleus ventralis lateralis of the thalamus. Following inflation of the balloon the dystonia disappeared except for some inversion of the left foot. On the 1st and 7th postoperative days, 0.5 cc. of alcohol-Pantopaque mixture (4:1) was injected. The patient did not become disoriented or confused following the injections as she had done in the past after chemopallidectomies. However, mild transient left pyramidal signs developed, including left facial weakness, increased deep tendon reflexes and a positive left Babinski’s sign.

When discharged from the hospital on June 21, 1958 she walked with slight inversion of the left foot and complained of stiffness of the lumbar spine and left upper extremity.

Course. The 5th admittance to the hospital occurred on Oct. 22, 1958. During the summer months the dystonia had gradually returned to the left upper extremity and was more severe in the leg and foot. In addition, the patient’s head was again twisting toward the right side. She was depressed to the point of being suicidal. Here memory was normal for recent and remote events; she could recall 7 numbers forward and 5 backward and did the serial 100-7 test without error. Her general fund of knowledge and ability to concentrate on a subject seemed normal. The cranial nerves functioned normally except a left hemihypesthesia was found which included her face. The left arm sought two positions, being either extended and hyperpronated with fingers extended, or tightly flexed across her chest with fist clenched. The left
leg was held in extension; the foot inverted with involuntary movements if the position was altered. The back was in opisthotonus and the head was turned strongly toward the right. No dystonic spasms or postures were seen on the right side. The left patellar and Achilles reflexes were somewhat hyperactive and a questionable Babinski's response was present on the left. An electroencephalogram was normal of low-voltage fast type.

Operation 6. On Oct. 31, 1958, using a transfrontal approach, balloon-type Cooper cannulae were placed in the right globus pallidus and right nucleus ventralis lateralis of the thalamus. Inflation of the balloon of the thalamic cannula was followed by immediate cessation of the dystonic movements. Twenty-four hours later 0.5 cc. of alcohol-Pantopaque mixture was introduced and another 0.5 cc. was injected on the 4th postoperative day. Mild transient left facial weakness occurred. The sensory deficit had abated after the first injection. The dystonic movements remained abolished. On the 11th postoperative day the balloon of the right pallidal cannula was distended. The following day 0.5 cc. of alcohol-Pantopaque was injected without incidence.

The patient was discharged on Nov. 13, 1958 completely free of dystonia and without apparent neurologic deficit except for a mild left facial weakness.

Course. The patient remained quite well for 12 weeks. In February, 1959, she had onset of involuntary intermittent spasms of the posterior cervical musculature causing torticollis to the left. Concomitantly, she noticed that the right hand began to shake. Her symptoms progressed rapidly and within a week her head was forced constantly and painfully to the left. Severe involuntary flexion of the right elbow and right hand occurred. In order to overcome and prevent the spasm the patient would lie with her right arm under her body. Soon the dystonia spread to involve her right foot, which became everted. Thus, her clinical picture was quite similar to her original symptoms prior to her 1st chemopallidectomy in November, 1957.

The patient was quite oriented and cooperative. Her memory for recent and remote events seemed normal. She could do the 100-7 test rapidly and accurately; also recall 8 numbers forward and 5 numbers backward; she recalled words at least 10 minutes later. Her stream of conversation was normal and her general fund of knowledge was adequate. However, her personality had changed over the many months of her illness. She was less inhibited and was inclined to relate obscene jokes to the staff. Her friends had observed that she was more careless about her personal habits. At the same time, however, she had become deeply depressed about her recurrent symptoms.

Function of the cranial nerves was normal except for a 30-per cent bilateral loss of 30 decibel by air and bone conduction. The visual fields were normal. Hyposthesia and hypalgiesia were found on the right side. The patient lay with her right hand beneath her right hip to prevent severe involuntary flexor spasms of the right elbow and hand. The flexor spasms were associated with hyperpronation of the forearm. The examiner had great difficulty in overcoming these powerful flexor spasms. The right lower extremity was held in a slightly flexed position at the hip and knee with the foot strongly everted. The muscle tone on the right was increased. When any voluntary movement of the right extremities was attempted, an irregular 3 c./sec. gross tremor of that member would develop. The reflexes on the right side were normal but brisk on the left. Some examiners felt the Babinski's response was positive on the left; all agreed it was negative on the right.

A preoperative electroencephalogram was mildly abnormal with fairly rhythmical basic frequency of 9 to 10 c./sec. appearing primarily over the posterior regions of the scalp but with some scattered 5 to 7 c./sec. activity from the anterior leads in a nonlateralized, nonfocal fashion.

The examiner was hesitant to perform a left chemothalamectomy for fear that the resulting bilateral thalamic and pallidal lesions would exceed the limit of physiological safety of connections between various cortical areas, thalamus, basal ganglia and mesencephalon and cause severe intellectual dysfunction.

Operation 7. Substantia nigralysis was decided upon. This was performed on July 30, 1959 with the patient in a sitting position. A small left occipital craniotomy was fashioned.
The occipital lobe was then elevated exposing the posterolateral aspect of the brain stem. Using a modified Grass depth electrode holder, a bipolar needle electrode was inserted into substantia nigra to a depth of 5 mm. (Fig. 1). The electrode entered the brain stem at the junction of the superior and inferior colliculus in the lateral mesencephalic sulcus. It was directed tangentially to the peduncle. The patient was allowed to awaken. Her right extremities immediately assumed dystonic postures and spasms. One mA. of direct electrolytic current was passed for 1 min. The polarity was reversed and again 1 mA. of current was given for 1 min. The patient had no change in the dystonia, therefore the electrode was inserted 3 mm. deeper at the same site. Another mA. of current was passed for 1 min. in either direction and the dystonia promptly disappeared. Normal strength was retained.

**Course.** The patient had a remarkably benign course. She was immediately oriented and alert after operation and showed no evidence of receptive or expressive dysphasia. At no time did loss of memory or periods of confusion develop. Function of the cranial nerves remained as before surgery. The right hemihypesthesia disappeared. The visual fields were unchanged. Voluntary movements on the right were free, rapid and strong without abnormal posturing or dystonic spasms. Passive movement of the extremities showed normal tone except for a slight catch resistance when the right forearm was extended. The gait was normal. The deep tendon reflexes were brisk but equal and Babinski’s sign was now positive bilaterally. The plantar responses became flexor on the 7th postoperative day.

The patient was no longer depressed. She seemed to take a genuine interest in her personal
appearance and was not as inclined to tell “off-color” stories. She was anxious to have help that would allow her to return to some type of self-supporting occupation. Neurological find-
ing 8 weeks after surgery were normal. The patient continued to be free of her dystonia musculorum deformans.

DISCUSSION

Substantia nigralysis is designed to destroy the majority of the substantia nigra nucleus throughout the mesencephalon and lower diencephalon. In order to accomplish this it is necessary to make a continuous lesion from the level of the inferior colliculus to the medial geniculate body. The bipolar electrode is inserted tangentially to the cerebral peduncle in the lateral mesencephalic sulcus to a depth of 8 or 9 mm. at the inferior colliculus and 10 or 11 mm. at the superior colliculus. A modified Grass depth electrode holder has been used to control the position and depth of the ele-

trode. The exposure of these brain-stem landmarks is readily accomplished by elevating the occipital lobe and splitting the tentorium. The patient is placed in a sitting position and is awake during the electrolysis so that the extremities can be moved voluntarily upon request. The electrolytic current is passed for short periods of time in order to assess its effect.

At the present time the pathophysiological mechanisms that cause dystonia musculorum deformans are unknown. Stimulation of the pallidum and thalamus at various parameters during the course of the 5th and 6th operations of the present case failed to enhance or decrease the dystonia. Using this surgical technique of sub-

stantia nigralysis, direct stimulation and recording from various depths in the sub-

stantia nigra can be carried out. The results of stimulation of the substantia nigra with the extremities at rest, or during voluntary and passive movements of the ext-

remities may be of help in future cases in determining the particular role these nuclear masses play in the basal-ganglia motor system in various hyperkinetic syn-

dromes.

The results of surgical treatment in a series of cases of dystonia musculorum de-

formans confirm reports by Cooper and others that lesions produced stereotaxically in the globus pallidus and/or the nucleus ventralis lateralis of the dorsal thalamus will generally improve, if not completely relieve, the patient’s symptoms. The pres-

et case illustrates that a lesion produced in the substantia nigra may also relieve a patient suffering from this disorder. From this one experience, however, it is not pos-

sible to say whether or not a lesion must be present simultaneously in either the thalamus or the globus pallidus. Meyers et al. have reported that a lesion in the lat-

eral aspect of the substantia nigra produced by use of focused ultrasonic waves will alleviate rigidity and tremor of parkinsonism and athetosis. The substantia nigra may become a useful target for dystonia, as well as parkinsonism, because disorienta-

tion and states of confusion which may accompany pallidal or thalamic lesions should not occur.

SUMMARY

A case of dystonia musculorum deformans successfully treated over a 21-month period by a combination of right chemopallidothalamectomy and left chemopallidectomy and substantia nigralysis is presented. Three other patients have been com-

pletely relieved and 1 considerably improved by surgery. The substantia nigra is proposed as an additional surgical target for dystonia musculorum deformans.
ADDENDUM

Since this manuscript was submitted for publication, the patient reported (Oct. 20, 1960) that she is feeling well, and has remarried. She walks well, but uses a cane because of occasional dizziness. She has no abnormal movements or postures of her limbs, trunk or neck and is perfectly able to do her entire housework in addition to caring for her disabled husband, who suffers from multiple sclerosis.

REFERENCES