Campotomy in Myoclonia*

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The involuntary, brief, rapid, rather irregular jerks called myoclonic are found in a great variety of conditions; as far back as 1903 several hundred cases had been summarized. However, little has been described as to treatment for this condition. Hassler, et al., abolished the jerky movements by coagulation of the thalamic endings of the brachium conjunctivum. We reported the beneficial effect of lesions of Forel's field H (campotomy), and Laitinen carried out subventrolateral thalamotomy (at 15.5 mm posterior to the anterior commissure and 11.5 mm lateral to the midline) in patients with severe progressive myoclonus epilepsy. Favorable effects on contralateral ataxia, intention tremor, and gait were observed, and for several months the frequency of the grand mal seizures was also reduced while the effect on the myoclonia was less marked.

Because all these communications covered only short-term observation periods, it may be appropriate to report here the long-range effects of campotomy. Since our previous publication we have had the opportunity to observe that case for 6 years after unilateral and 4 years after bilateral operation. We have also treated two additional cases surgically; one of these has been observed for 4 years and the other for half a year after a unilateral lesion of Forel's field.

None of the cases disclosed a familial background or history of encephalitis; however, the first case had meningitis at the age of 6 years. All three patients were males, the ages being 19, 25, and 30 years respectively. The abnormal jerky movements involved primarily the neck, shoulder, and upper limb musculature. Involuntary grunting or "woofing" noises were heard in the first two cases and usually occurred synchronously with myoclonic movements of the neck, upper limbs, and diaphragm. Palatal myoclonus was not a feature of the disease in any of the cases. All these involuntary movements increased with excitement and decreased with sleep. The EEG did not reveal dysrhythmia in any of the tracings. Electromyograms indicated a marked postoperative reduction in the myoclonic jerks. No neurological complication was noted after surgery, and all these patients are presently gainfully employed.

The technique of campotomy and the coordinates have been described previously. In brief, after visualization of the 3rd ventricle with Pantopaque, the sheath of the electrode is introduced perpendicular to the intercommissural line, 11 mm anterior to the posterior commissure for a length of the intercommissural line of 25–27 mm. For a shorter intercommissural distance (23–24 mm) the lesion is placed 10 mm anterior, and for a longer intercommissural distance (28–29 mm) 12 mm in front of the posterior commissure. The puncture canal reaches the level of the intercommissural line 6–7 mm lateral to the median plane. It should also be mentioned that oculomotor disturbances have no longer been observed since the puncture has been performed more anteriorly, namely, at a point corresponding to the midpoint of the intercommissural line. It has proved important to repeat roentgenograms after the electrodes have been inserted into the brain in order to check their position in relation to the intercommissural line so that corrections in their depth can be made, if necessary. Care must be taken to avoid the corpus subthalamicum in view of the danger of producing hemiballistic movements.

Case Reports

Case 1. A 19-year-old white man was admitted to Temple University Hospital on
April 23, 1962, because of grunting and jerking of the neck muscles of 8 years' progression. When 6 years old he had recovered from an attack of "spinal meningitis."

Examination. The involuntary grunting noises were associated with myoclonic movements affecting primarily the neck, left shoulder and arm, and diaphragm. Myoclonic jerks of the abdominal muscles on the left side appeared synchronously with the abnormal vocalization. The movements of the larynx were recorded at 54–56 per minute, while movements of the diaphragm ranged from 50–52 per minute. The movements ceased during sleep and were less pronounced when he was relaxed. When the movements were severe, the head would be jerked backward by the posterior cervical muscles. The objective neurological findings otherwise were normal.

Operation. On September 27, 1962, a right campotomy was performed under local anesthesia.

Postoperative course. Following the operation, there was a marked reduction in the abnormal movements described above. The head was held normally, without jerking, and there was a decrease in the intensity and frequency of the myoclonic movements of the left shoulder and arm (Fig. 1). The disturbing loud grunting noises subsided almost completely. He returned to his former occupation as a leather worker and was gainfully employed until January, 1964, when jerking movements appeared on the right side involving particularly the neck and shoulder muscles. The left side was still tranquil. The EMG's (Fig. 2) likewise showed irregular rhythmic contractions of the neck and shoulder musculature on the right side. The left side was normal.

The onset of right-sided movements started 4 months prior to readmission to the hospital and began gradually with a twitching of the muscles of the right shoulder girdle and the muscles of the right side of the neck, particularly the platysma. The grunting noises returned; at first they were faint, but they became progressively louder so that they annoyed others near him. Nevertheless, he continued to work. He sought admission to have the campotomy performed on the opposite side.

Second examination. With the exception of the grunting noises and the myoclonia involving the platysma muscle on the right side and the muscles of the right arm and shoulder girdle, the neurological examination was essentially negative. The speech did not appear to be altered, but periodically the patient would emit a loud grunting noise or a noise similar to a "bark." These noises would occur at irregular intervals, without warning; the abdominal muscles and the body jerked at the time the noise was emitted. No pathological reflexes were elicited, and the deep tendon reflexes were normal throughout. There was no evidence of any weakness or paralysis on either side. Perception of pain, temperature, and light touch was normal. The coordination tests failed to reveal cerebellar dysfunction or ataxia, and the gait and station were normal.

![Fig. 1. Case 1. EMG's of muscles on the left side of the neck (1–5) and the left shoulder (1–3) before (A) and after (B) right-sided campotomy. Intervals between vertical lines correspond to 1 sec. (From Spiegel, Wycis, et al., J. Neurosurg., 1963, 20:871–881.)](image-url)
Second operation. On March 19, 1964, a left campotomy was performed.

Second postoperative course. Immediately following surgery, the patient was alert and conscious. The grunting noises had ceased, and the myoclonic movements of the right side of the body were practically absent, except in rare instances when an occasional twitch would be seen in small groups of muscles of the neck, perhaps once or twice over a period of 5 minutes. His speech was unaffected. When he was discharged on March 27, 1964, there were practically no involuntary movements on the right side of the body, and the grunting noises had completely stopped. Nor were there any abnormal movements on the left side after the right campotomy. There were no neurological complications immediately after operation or at subsequent observations. He returned to his former occupation.

After a lapse of 4 years we were fortunate in locating the patient for reexamination (6 years after the first operation). He was married and had one child. He was steadily employed as a maintenance manager. He no longer had any involuntary myoclonic jerks except for a few mild contractions of the right platysma muscle, and the grunting noises had not returned. The EMG's (Fig. 3) repeated on February 23, 1968, showed only a few bursts of irregular rhythmic activity from the right platysma muscle. None was recorded from the left side.

Case 2. A 25-year-old man was admitted to Temple University Hospital on January 5, 1964, because of grunting noises and jerking of the head. At the age of 4 years he had developed a torsion spasm of the neck musculature which caused the left ear to be drawn to the left shoulder. At that time he underwent a corrective surgical procedure (the exact nature of which is unknown) which straightened his head but which was followed by lightning-like jerks of the muscles of the neck and both shoulders. The head would be flexed acutely forward and the shoulders drawn upward; the head would then come to rest in a position of a slight tilt to the left. The irregular jerks were accompanied by a grunting, jerky speech which was worse during excitement. There was no history of infection or convulsive seizures, and the family history was irrelevant.

Examination. There were frequent uncon-
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6 YRS. POST-R. CAMPOTOMY (1962)
4 YRS. POST-L. CAMPOTOMY (1964)

RIGHT PLATYSMA

(UNIPOLAR RECORDING)

LEFT PLATYSMA

1-SEC.

Fig. 3. Case 1. EMG's of right and left platysma muscles 6 years following a right-sided campotomy and 4 years after an additional left-sided campotomy.

trollable jerkings; these arrhythmic movements were most pronounced in the head and neck muscles with tilting of the head to the left. The shoulder was elevated more on the left side than on the right. Palatal myoclonic movements were not seen, but the speech was jerky and characterized by frequent emission of grunting noises. The rapid muscular contractions would cause the arms to be extended with the shoulders elevated. Simultaneously the head flexed acutely downward. All of these movements were exaggerated by excitement. He walked with a widened gait. X-rays of the skull disclosed some developmental asymmetry of the occipital condyles.

Operation. On January 9, a right campotomy was carried out under local anesthesia.

Postoperative course. There were no complications except for a very mild left facial relaxation which lasted but 1 week. When the patient was discharged 8 days later, there were no involuntary jerking noted on either side. The left shoulder which had been higher than the right before surgery was now lower. The voice became more distinct and without the grunting character. The EEG tracing was normal before and after surgery. The EMG's before campotomy revealed irregular bursts of electrical activity in the upper limb muscles on both sides, while, following surgery, occasional bursts were noted in the left forearm and right shoulder only. The EMG recorded 1 year later (Fig. 4) and 4 years later revealed similar findings. Four years following surgery he was still symptom-free and without disturbing muscular movements. His speech was normal and he was employed in the merchant marines.

Case 3. A 30-year-old white man developed electric-like irregular jerks of both upper and lower extremities and the neck 4 years prior to admission; the condition had recently become progressively worse. The movements ceased during sleep. There was no history of prior infection of the CNS or serious illness. The family history was negative.

Examination. The neurological examination was essentially negative except for the myoclonia. The activity in the right deltoid muscle was particularly pronounced and increased over that on the left side. The speech was irregular and stuttering, but abnormal noises were not produced. Synchronous with contraction of the neck musculature the thyroid cartilage would move up and down. Palatal myoclonia was not observed. Liver studies including a scan and kidney function tests were normal.

Operation. On September 28, 1967, a left campotomy was performed. Immediately after producing the first lesion of the campotomy (+ 2 above the anterior-posterior commissure plane) a decrease in the spontaneous activity of the right foot and hand was noted; as the lesions were continued from
above downward (from +2 to −4 below the ac-pc plane) the involuntary arm and neck movements totally subsided without any associated paralysis.

Postoperative course. The patient had no complication following the left-sided lesion of Forel’s field and had a marked reduction of all clonic movements on the right side. When he was examined 6 months later not only was his improvement maintained but it was noted then that there was a decrease of involuntary jerks on the left side as well which were now limited to a few jerks of the platysma and the thenar muscles of the left hand. The patient had been placed on Valium (15 mg daily) which may have played some role in this improvement, although prior to operation Valium in the same quantity did not have such an effect. An operation on the other side is still held in reserve pending future developments.

Discussion

In Case 1, the history of “spinal meningitis” at the age of 6 years was suggestive of inflammatory involvement of the spinal cord and brain stem. In the second case the possibility of unproven impairment of the basal ganglia has to be considered in view of the development of a torsion spasm at the age of 4 years. In the third patient it seems justified to classify the myoclonia as “essential,” since other neurological signs, epileptic phenomena, or EEG abnormalities were absent.

Before we attempt to explain the effect of the lesion of Forel’s field H upon the myoclonic jerks, it seems useful to discuss the basic pathophysiology. Unfortunately, our knowledge in this respect is rudimentary. In a large number of cases, particularly in those studied by Guillain, et al., the inferior olive, the contralateral dentate nucleus, the red nucleus, and the central tegmental tract were affected. It has been hypothesized that lesions of the olivocerebellar-dentato-ruber system or of the central tegmental tract may release the activity of cell groups of the reticular formation. The hypothesis that abnormal discharges are conducted by reticulo-spinal fibers to various segments of the spinal cord and by axons of reticular neurons to nuclei of motor cranial nerves may help to explain the synchrony of the jerks in muscle groups innervated from different segments of the neuraxis. The weakness of this hypothesis lies in the experience that lesions of the dentate nucleus do not always induce clonic jerks, either in animals or in humans.

This suggests that other factors in addition to lesions in the triangular area outlined by Guillain, et al., may have to be present for myoclonic jerks to develop. One such
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factor seems to be the latent period necessary between the lesion of the olivo-cerebel- lo-dentate system and the appearance of the myoclonia as pointed out by Faure-Beaulieu and Garcin. A further factor is indicated by Milhorat's experiments in Rhesus monkeys. In these experiments, excision of the mesial thalamus lowered the myoclonic threshold, apparently by releasing facilitatory systems originating in the lateral thalamic nuclei. Lateral thalamectomy eliminating these facilitatory impulses abolished the clonic movements. In this connection experiments by Spuler, et al., in cats are interesting. They found that the tremor elicited by stimulation of the mesencephalic tegmentum may be facilitated or modified by stimulation of either the ipsilateral or contralateral ventrolateral area of the thalamus. This effect was still demonstrable after the fibers of the internal capsule originating in the frontal lobe and motor cortex and the pallidofugal fibers had degenerated. Furthermore, stimulation of Forel's field was also able to increase a tremor induced by stimulation of the mesencephalic tegmentum. From lesions in Forel's field, Johnson and Clemente could trace degenerating fibers into the mesencephalic tegmentum.

These experimental data suggest that facilitatory impulses from the ventrolateral thalamus and impulses passing through Forel's field (whose origin must still be analyzed) may reach the reticular formation and facilitate its activity. This may help explain why lesions of the ventrolateral thalamus (Hassler, et al.) as well as lesions of Forel's field (present observations) may reduce or abolish certain types of myoclonic jerks. We should like to emphasize that it would be premature to generalize from these observations and to expect a beneficial effect of campotomy in all the various types of myoclonia, particularly where the EEG findings point to a cortical involvement. Here further studies are necessary.

Summary

Three cases have been described in which the beneficial effect of lesions of Forel's field upon myoclonic syndromes could be observed for periods of 6 years, 4 years, and ½ year respectively. The possible pathophysiological mechanisms involved have been discussed.

References


