

## Management of a Hemidystonic Patient with Thalamotomy, Campotomy and Cervical Dorsal Root Entry Zone Operation

Gokmen Kahilogullari<sup>a</sup> Hasan Caglar Ugur<sup>a</sup> Ali Savas<sup>a</sup> Ebru Bilge Dirik<sup>b</sup>  
M. Cenk Akbostanci<sup>b</sup> Bulent Elibol<sup>c</sup> Yucel Kanpolat<sup>a</sup>

Departments of <sup>a</sup>Neurosurgery and <sup>b</sup>Neurology, School of Medicine, Ankara University, and  
<sup>c</sup>Department of Neurology, School of Medicine, Hacettepe University, Ankara, Turkey

### Key Words

Dystonia · Dorsal root entry zone operation ·  
Thalamotomy · Campotomy · Pallidotomy

### Abstract

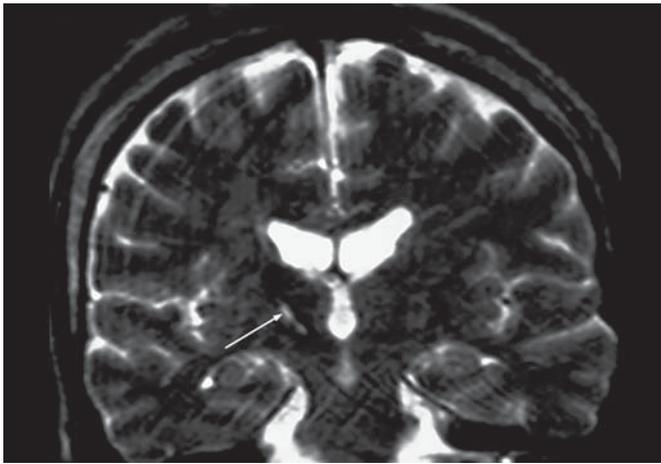
Several medical and surgical procedures have been presented for treatment of dystonia. Thalamotomy, pallidotomy, and campotomy are some of the surgical choices. This study presents a patient with dystonia who underwent a cervical dorsal root entry zone (DREZ) operation after thalamotomy and campotomy. A 23-year-old man who was resistant to medical treatment presented with left hemidystonia. Thalamotomy and campotomy were performed. The patient remarkably benefited from the procedure but dystonic complaints in his left arm continued. A cervical DREZ operation was performed 5 years after the first operation and the dystonic complaints decreased after the surgery. This article presents a new aspect for the treatment of dystonia. Based on the outcomes of the treatment, DREZ operation may be suggested as an alternative surgical treatment for patients with segmental dystonia located in the extremities.

### Introduction

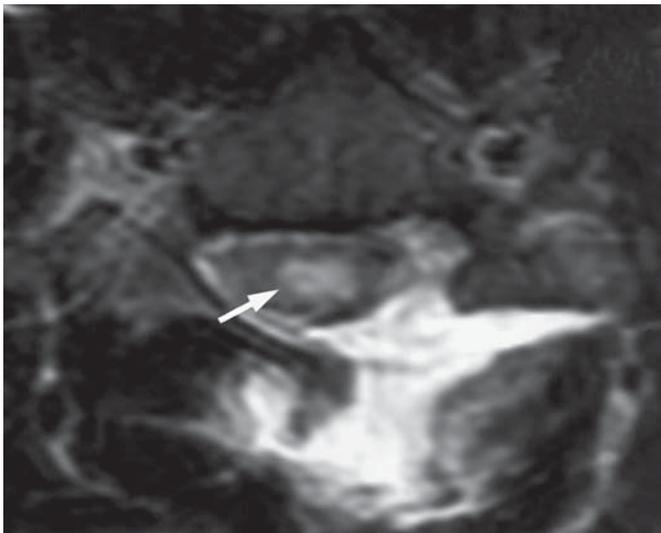
Dystonia may be primary (idiopathic) or secondary. Birth injury, postnatal head injury, encephalitis, meningitis, subarachnoid hemorrhage, stroke, and anoxia are causes of secondary dystonia. Many medical and stereotactic surgical methods have been recommended for the treatment of dystonia. Dopaminergic and antidopaminergic agents, anticholinergics, benzodiazepines, baclofen and local botulinum toxin injections are the medical treatments, while stereotactic thalamotomy, pallidotomy and neurostimulation procedures are the most frequent techniques used for the treatment of dystonia [1, 2].

Treatment of dystonia is still difficult in spite of the medical and surgical choices of treatment. The outcomes of an ipsilateral cervical dorsal root entry zone (DREZ) operation performed on a dystonic patient who was previously treated with contralateral thalamotomy and campotomy are reported.

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**Fig. 1.** Postoperative cranial T<sub>2</sub> coronal MRI showing the lesion after thalamotomy and campotomy.



**Fig. 2.** Postoperative changes on the cervical T<sub>1</sub> axial MRI examination after left C<sub>5</sub>-C<sub>8</sub> DREZ operation.

### Case Report

A 23-year-old male presented with hemidystonia on his left side. The patient complained of tremors and contractions in his left arm and leg. He had a history of encephalitis in the first month after birth. His complaints had started with tremors when he was 3 years old, and increased in time with the addition of contractions at the extremities. Despite many medical treatments such as dopaminergic and antidopaminergic agents, anticholinergics, baclofen and botulinum toxin injections, his complaints had not improved. Neurological examination revealed severe dystonic contractions and postures on his left extremities and trunk. The dystonic contrac-

**Table 1.** The scores in preoperative and postoperative period based on the Fahn-Marsden Dystonia Scale

	Dystonia general	Dystonia left arm	Disability
Preoperative	48	16	11
After thalamotomy and campotomy	25	16	11
After DREZ operation	22	12	8

tions were frequent and had high amplitudes. Based on the Fahn-Marsden Dystonia Scale, motor general subscale was 48 and disability subscale was 11. Six years ago, a right CT-guided stereotactic thalamotomy and campotomy had been performed (fig. 1) after which the dystonic complaints of the patient decreased on his left side, but the dystonic flexion posture on his left arm continued. The motor score for the left arm was 16 and disability subscale score was 11 (Fahn-Marsden Dystonia Scale). Five years later, through left C<sub>4</sub>-C<sub>7</sub>-T<sub>1</sub> hemilaminectomy, 46 radiofrequency lesions (Type ENA El-Naggar-Nashold Angled Nucleus Caudalis DREZ Electrode, Model RFG-3CF; Radionics, Burlington, Mass., USA) were performed on the left C<sub>5</sub>-C<sub>8</sub> DREZs during 15 s at 75°C. The patient's postoperative course was uneventful. The findings of neurological examination on the 9th postoperative day were compared with the findings of the preoperative period. On postoperative Fahn-Marsden Dystonia Scale, general motor subscale was 22; left arm motor subscale was 12, and disability subscale was 8. The scores have been presented in table 1. Cervical magnetic resonance imaging (MRI) performed after surgery (fig. 2) revealed that the lesions related to surgery were at the appropriate sites of the spinal cord. The patient was discharged on the 10th postoperative day. Active physiotherapy and botulinum toxin injections were performed for the remaining dystonia in the left arm after discharging.

### Discussion

Dystonia may be primary (idiopathic) or secondary. Primary dystonia or dystonia musculorum deformans (DMD) run in families. DMD is a disease of young people, with most cases commencing in childhood, though some do not appear till the teen years; adult onset is rare. The second large group of dystonia patients consists of secondary cases that resemble DMD. These are usually segmental, hemidystonic, or generalized in pattern but differ from primary dystonia in being caused by recognizable insults to the central nervous system. The etiology of our patient was encephalitis like 4 of 30 secondary dystonic patients in Tasker's series. The other causes of secondary dystonias are birth injury, postnatal head injury, meningitis, subarachnoid hemorrhage, anoxia and un-

known causes. Treatment of dystonias is still difficult despite advances in medical treatment. It is also difficult to define surgical indications for dystonia; the clinical picture varies enormously, and there are many different causes. Surgery does not alter the progress of dystonia or the underlying neurological deficits. Thus, no matter how successful the surgery is, the eventual outcome is dominated by the course of the subsequent disease and the underlying neurological deficits. Published series are, however, few and small, and any conclusion must be regarded as tentative. Thalamotomy was made in 54 patients and the outcomes were described [1, 2].

In medical treatment, the first choice is dopaminergic agents in primary dystonia. Paradoxically, the antidopaminergic treatments are effective in primary dystonia. Other medical treatments are anticholinergics, benzodiazepines, baclofen and local botulinum toxin injections [3, 4].

Thalamotomy, pallidotomy and pallidal stimulation are the most frequently used techniques for dystonic patients. Studies reported that half of the patients with primary dystonia improved after pallidotomy; however, thalamotomy and campotomy produced relatively better results [1, 3, 5–7].

Based on the anatomical fact that the outflow from the pallidum to the thalamus runs through the fasciculus lenticularis and the ansa lenticularis and that these two bundles together with ascending cerebellorubrothalamic pathways converge in the field H of Forel, Spiegel and Wycis advocated this area as a stereotactic target. Their target lay 6–7 mm lateral from the midline of the third ventricle, 11 mm anterior to the posterior commissure, and 2 mm below the level of the intercommissural line. The advantage would be that at this level the maximum number of fibers can be interrupted by the smallest lesion. Spiegel and Wycis called the procedure campotomy (interruption of campus Foreli and zona incerta) and reported a more consistent improvement of tremor than rigidity, while bradykinesia had not improved [6, 7].

In 1972 Sindou [8] performed the first DREZ operation on a patient who had painful Pancoast-Tobias syndrome. There are many indications for cervical DREZ operation: spasticity, cancer pain, pain after brachial plexus avulsion or from spinal cord, pain due to peripheral nerve lesions, cervical root avulsion, post herpetic pain and hyperspastic states with pain [9, 10]. Mertens et al. [11] described the importance of radiological anatomy of the spinal cord at the cervical level.

DREZ operation has also been recommended for facial pain [12], conus medullaris root avulsions [13], and

the treatment of pain with spinal cysts in paraplegia [14]. Nashold [15] has described the neurosurgical technique of the DREZ operation and a design of radiofrequency lesion electrodes in the caudalis nucleus DREZ operation [16].

Sindou [17] has claimed that if spasticity is found spread to an entire limb, surgery in the dorsal roots or the DREZ can be performed to decrease spinal cord hyperexcitability. Spinal DREZ operation has been preferred because of its strong hypotonic effect. This operation not only destroys the nociceptive fibers (which take part in spasticity genesis) but also encroaches the myotatic fibers that run in an intermediate position to reach the ventral horn. The importance and mechanism of DREZ operation in spasticity have been described in literature [18, 19]. However, there is no information regarding the effect of DREZ operation on dystonia in the reported series. On the other hand, DREZ operation may be one of the important regions considering the relay effects of dystonic movements.

To our knowledge, this is the first case of DREZ procedure after thalamotomy and campotomy for a dystonic patient. DREZ operation may be suggested as an alternative surgical procedure to patients with segmental dystonia located at the extremities, which is resistant to medical and surgical treatments. Our experience leads us to suggest an additional DREZ procedure for segmental dystonia after deep brain surgery.

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