Stereotactic Thalamotomy for Task-Specific Dystonia

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Abstract: Background: Task-specific dystonia (TSD) is a focal dystonia producing abnormal posture of the fingers or the hand, occurring during selective motor activities that involve repeated use. Conventional medical management and botulinum toxin fail to provide relief in all patients. Thalamotomy or DBS have been used as surgical treatment in patients not responding to medical treatment.

Methods: Five patients (all male; age, 18–47 years) with refractory focal hand dystonia underwent ventrooralis (Vo) thalamotomy using stereotactic techniques. Preoperative video recordings, Writer’s Cramp Rating Scale (WCRS), and Symptom Severity Scores (SSS) were evaluated at the baseline and at a follow-up date, which ranged between 8 and 44 months (average, 26 months).

Results: All the patients had difficulty in performing their most common tasks. Duration of symptoms ranged from 6 months to 12 years. All patients obtained immediate postoperative relief from dystonic symptoms, and the effect was sustained during the follow up period. The WCRS improved from a mean of 20.4 before surgery to a mean of 2.4 at the last follow-up, whereas the SSS improved from a mean of 29.8 before surgery to a mean of 12 at the last follow-up. All patients were able to carry out their specific task with little or no difficulty. There were no surgical complications, morbidity, or mortality.

Conclusions: Vo thalamotomy is a safe and effective procedure providing successful symptom relief in patients of TSD.

Task-specific dystonia (TSD) is a movement disorder that is characterized by the contraction of antagonist muscles while performing a specific task. The most common forms of this are the writer’s and musician’s cramp. It is a focal dystonia that often involves highly skilled, repetitive movements. It is classified under focal and action-specific dystonia. It occurs because of a dysfunction of the pallido-thalamo-cortical pathway. Symptoms of dystonia can be improved by interrupting this loop at a certain anatomical point. Although clinical manifestations in focal hand dystonia are typically unilateral in the beginning, the disorder is considered as a bilateral dysfunction of the basal ganglia. This is demonstrated by the fact that up to 25% of patients develop bilateral focal hand dystonia if they switch to the previously unaffected hand. Prevalence of writer’s cramp has been reported as being between 16.3 and 68.9 per million. Nonsurgical treatments such as botulinum toxin have been tried, but the therapeutic effects are not so remarkable.

Stereotactic surgery has been used to treat dystonia. Thalamotomy, pallidotomy, and DBS all have been applied to treat various forms of dystonia, including TSD. The first stereotactic surgery for writer’s cramp was reported in 1969. The ventral oral nucleus (Vo) and/or ventral intermediate nucleus (Vim) of the thalamus are the common surgical targets. All reported series are from Japan, this being the first series of 5 patients being treated by thalamotomy from outside Japan.

Patients and Methods

Between August 2012 and June 2015, 5 patients (all male; age, 18–47 years) with medically refractory TSD underwent Vo nucleus thalamotomy at our institute (Table 1). All patients were carefully diagnosed as having focal hand dystonia by history and clinical examination. Three patients had dystonia while...
writing and 2 had dystonia while percussing “tabla” (Indian drum). Patient 1 had the longest duration of writer’s dystonia (see Video 1). Since 3 years before the surgery, he had started using his left hand; however, when the left hand also started showing signs of dystonia, he sought surgical treatment. Patient 5 had musician’s dystonia, associated with playing tabla. However, this was also observed during typing on the keyboard of a computer (action similar to that involved in playing drums). Preoperative video recordings, Writer’s Cramp Rating Scale (WCRS) for dystonic posture and latency of dystonia, and Symptom Severity Scores (SSS) were evaluated. The procedure was explained to patients in detail. Informed consent for the surgery was obtained from the patient and relatives.

All surgeries were performed under local anesthesia using a CRW stereotactic system (Integra LifeSciences, Plainsboro, NJ). Vo nucleus of thalamus was targeted on CT scan based on the coordinates published by Taira and Hori.3 This was 13.5 mm lateral, 2 mm posterior to the mid anterior commissure/posterior commissure (AC-PC) point, and at the level of AC-PC plane. Intraoperative stimulation was performed using a 1-mm diameter and 2-mm exposed tip electrode. Stimulation of the target would elicit improvement in dystonia and facilitate the action like writing or playing drums. The acceptable limit for eliciting any capsular response was 2 V at 5 Hz and 100 μsec. The improvement in the symptoms was recorded using stimulation at 100 Hz and 100 μsec. Patients were asked to perform the same task that produced their dystonia (i.e., either to write or play the drums). This was followed by a test lesion at 45°C for 60 seconds. If there were no side effects, we would proceed with the lesioning at 70°C for 60 seconds. The electrode was withdrawn and another lesion was made using a similar protocol. A total of two or three lesions were performed based on the electrical stimulation response and clinical improvement. No lesion extended below the AC-PC plane (Fig. 1). A check CT scan was performed to evaluate the lesion and look for any hemorrhage.

**Results**

Before the operation, all patients had difficulty in performing their most common tasks (e.g., writing, playing table, etc.) with spasms of the involved fingers and dystonic posturing of the hand. Duration of symptoms ranged from 6 months to 12 years. All patients obtained immediate postoperative relief from dystonic symptoms, and the effect was sustained during the follow-up period. Patient 2 had some difficulty in writing noticed at the last follow-up. Patient 5 had difficulty in percussing with one finger of the three involved. However, his condition was better than the preoperative condition. He had anxiety in playing during a performance, but could easily play the drums when alone. The WCRS improved from a mean of 20.4 (range, 16–26) before the surgery to a mean of 2.6 (range, 0–5) at last follow-up, whereas the SSS improved from a mean of 29.8 (range, 27–34) before the surgery to a mean of 12 (range, 10–15) at the last follow-up. All patients were able to carry out their specific task with little or no difficulty (Table 2). There were no surgical complications, morbidity, or mortality.

**Discussion**

The exact pathophysiology of TSD is not known. However, several studies involving morphometric MRI, diffusion-weighted imaging, and functional MRI have revealed a difference between TSD and non-task-specific dystonia.21 Various reports have reported widespread changes in gray matter volume in the hand region of sensorimotor cortex, putamen, thalamus, and cerebellum.22,23 Functional MRI studies performed on the noninvolved side revealed that there was reduced activity in the contralateral anterior putamen/globus pallidus and the ipsilateral anteriorterior globus pallidus.6 This goes to underline the bilateral involvement in patients with unilateral disease, which is again borne out by progression of the dystonia to the opposite hand over a period of time (as illustrated in our case 1).

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**TABLE 1 Clinical details of patients with TSDs**

<table>
<thead>
<tr>
<th>No.</th>
<th>Age (Years)</th>
<th>Sex</th>
<th>Duration of Symptoms (Years)</th>
<th>Task Affected</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>47</td>
<td>Male</td>
<td>12</td>
<td>Writing</td>
</tr>
<tr>
<td>2</td>
<td>24</td>
<td>Male</td>
<td>2</td>
<td>Writing</td>
</tr>
<tr>
<td>3</td>
<td>45</td>
<td>Male</td>
<td>2</td>
<td>Playing drums</td>
</tr>
<tr>
<td>4</td>
<td>18</td>
<td>Male</td>
<td>2</td>
<td>Writing</td>
</tr>
<tr>
<td>5</td>
<td>27</td>
<td>Male</td>
<td>6 months</td>
<td>Playing drums</td>
</tr>
<tr>
<td>Mean</td>
<td>32</td>
<td>—</td>
<td>3.7</td>
<td>—</td>
</tr>
</tbody>
</table>
TSD is observed in various segments of the society, including surgeons, barbers, musicians, painters, watchmakers, etc. Botulimum toxin is the most common treatment option with modest efficacy, with long-term results showing only 36% patients having sustainable improvement. These patients, in the absence of effective treatment, are at the risk of losing their livelihood. They often start using the opposite hand, but because the disease is bilateral, the other side also starts showing similar disability. Stereotactic surgery with its high success rate should therefore be considered earlier in order to preserve the dignity and profession of the person.

There have been limited publications of stereotactic surgery for TSD, with only two series having an experience of more than 3 cases, both of them being from Japan. Abnormal activity in Vop has been demonstrated in patients with TSD, and the lesioning of Vop in these patients provided relief in their symptoms. Zhuang et al. recorded low-frequency bursts from Vim in patients with dystonic tremors. Relief from TSD after lesioning of Voa/Vop further supports the role of these nuclei in the circuit of pathways for TSD. Ventral oralis complex has been the most widely used target, the other target being Vim. The target selected for this surgery was based on Hassler’s classification of thalamic nuclei and its relationships as demonstrated in the Schaltenbrandt and Warren atlas and the coordinates reported by Taira and Hori. The laterality of the target was altered based on the relationship of the internal capsule, ensuring that it was at least 1 mm medial to the medial border of internal capsule. We started with the coordinate suggested by Taira and Hori and modified the laterality based on the anatomic relationship to the internal capsule and the stimulation-derived response. Our threshold to change the laterality was a capsular response at a frequency of 5 Hz, pulse width of 0.1 μsec, and 2-V voltage.

The main symptom in writer’s dystonia patients of TSD is difficulty in writing, which is slow and changes one’s handwriting to a less legible form. Usually, distal muscles of the dominant hand are the first ones affected. A tight grip of the pen is typical, and hand-wrist flexors are more commonly involved than extensors, even though hyperextension of the distal phalanges or even the fingers has been observed. Three of our patients had dystonia in the right hand beginning within seconds of starting to write and resulted in a tight grip of the pen and gradual abduction of the arm and forearm as they continued writing.

In the patients of musician’s dystonia subset of TSD, the dystonia involves fingers while playing a particular musical instrument. Two of our patients had musician’s dystonia, and this would set in when they would percuss a drum (see Video 2). The percussion action, even if done on a plain surface, would induce dystonia within a few seconds. We noted instantaneous improvement in their symptoms on the operative table after a test and the final lesion. In some patients, we had to perform more than one lesion to achieve complete relief. However, we ensured that we would not cause any pyramidal or speech dysfunction (as evaluated by test lesion).

A review of the reported series has revealed a 16% rate of transient morbidity ranging from hemorrhage to dysarthria. Three of the 25 patients required a repeat surgery followed by relief. In our series, we had no morbidity or mortality. We did not need to repeat the lesion because we tailored the number and the site of lesion for each individual patient.

This case series has limitations in terms of the sample size and in terms of being a retrospective study. The present scores do not reflect quality-of-life impact, and this could be of further help in understanding the broader impact of the treatment.

Conclusions

TSD is a disabling symptom with limited options for medical treatment. The severity of the symptom may force one to change his job/profession. Thalamotomy offers significant relief of symptoms to most of these patients with minimal risks. We would recommend that these patients be offered the option of surgical treatment before they are forced to modify their profession or lifestyle.

Author Roles


P.K.D: 1A, 1B, 1C, 3A
S.S.: 1C, 3A
B.K.: 1B, 1C, 2A, 2B
R.R.: 1C, 2A, 2C

Disclosures

Ethical Compliance Statement: We confirm that we have read the Journal’s position on issues involved in ethical publication and affirm that this work is consistent with those guidelines.

TABLE 2 Results of Vo thalamotomy

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>WCRS</th>
<th>SSS</th>
<th>Follow-up</th>
<th>Results</th>
<th>Recurrence</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>Preoperative</td>
<td>Postoperative</td>
<td>Preoperative</td>
<td>Postoperative</td>
<td></td>
</tr>
<tr>
<td>1</td>
<td>18</td>
<td>3</td>
<td>31</td>
<td>11</td>
<td>Improved</td>
</tr>
<tr>
<td>2</td>
<td>25</td>
<td>5</td>
<td>34</td>
<td>15</td>
<td>Improved</td>
</tr>
<tr>
<td>3</td>
<td>17</td>
<td>0</td>
<td>28</td>
<td>10</td>
<td>Improved</td>
</tr>
<tr>
<td>4</td>
<td>26</td>
<td>0</td>
<td>27</td>
<td>10</td>
<td>Improved</td>
</tr>
<tr>
<td>5</td>
<td>16</td>
<td>5</td>
<td>29</td>
<td>14</td>
<td>Improved</td>
</tr>
<tr>
<td>Mean</td>
<td>20.4</td>
<td>2.6</td>
<td>29.8</td>
<td>12</td>
<td></td>
</tr>
</tbody>
</table>

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Funding Sources and Conflicts of interest: The authors report no sources of funding and no conflicts of interest.

Financial disclosure for past 12 months: P.K.D. has received consultations and grants from Medtronic Inc and Boston Scientific Company.

References


Supporting Information

Videos accompanying this article are available in the supporting information here.

Video 1. This is a preoperative, intraoperative, and immediate postoperative video of case 1 with writer’s dystonia.

Video 2. This is a preoperative and 3-year postoperative video of a patient with musician’s dystonia. Note that the dystonia only affects while playing drums and not while writing.