Unilateral Ventral Intermediate Deep Brain Stimulation Improves Spasmodic Dysphonia
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BACKGROUND
- Essential tremor (ET) is frequently associated with other co-morbid movement disorders such as spasmodic dysphonia (SD), writer’s cramp and other focal dystonias.
- Ventral intermediate (VM) thalamic deep brain stimulation (DBS) is a well established treatment for contralateral limb tremors.
- There is now emerging evidence that voice tremors can also improve with both unilateral and bilateral thalamic stimulation.
- While patients with dystonia clearly benefit from pallidal stimulation, thalamic stimulation may be preferred in cases of dystonic tremor.
- There is limited data regarding the effects of thalamic stimulation on SD.
- We observe have 2 cases of unilateral thalamic stimulation, originally implanted for treatment of tremor, that incidentally improved each patients’ SD.

CASE HISTORIES

Case 1: 59 year old left handed woman with ET and writer’s cramp starting in her 20’s was first evaluated at our center in 1995. Tremors were refractory to multiple therapies associated with persistent disability. After consensus approval, she underwent bilateral VM DBS (3387 leads, Medtronic, MN) placement in January 2011.
- Pre-operative Fahn-Tolosa-Marín (FTM) tremor rating scale (TRS) was 18.5 with normal phonation.
- During follow-up she discontinued right VM stimulation due to stimulation induced side effects.
- In January 2012, left VM IPG depletion revealed recurrent right hand tremors and new voice difficulties consistent with adductor spasmodic dysphonia.
- After IPG exchange her voice symptoms improved with re-initiation of left VM stimulation (Video 1) to original settings (Table 1).
- For further characterization of voice symptoms strobovideolaryngoscopy (SVL) was performed ≥1 hour off stimulation and after 5 minutes on stimulation. (Video 2)(Table 2).
- Appropriate lead location of left VM electrode was confirmed by MRI brain (Figure 1).

Case 2: 68 year old right handed woman with ET for > 20 years, spasmodic dysphonia, restless legs syndrome and migraines was first evaluated at our center in 1999. Her tremors were refractory to multiple therapies associated with persistent disability. After consensus approval, she underwent left VM DBS (3387 leads, Medtronic, MN) placement in January 2011.
- FTM-TRS was 22 with evidence of spasmodic dysphonia pre-operatively.
- With optimization of her stimulation parameters (Table 1) she was noted to have near complete resolution of right hand tremors and marked improvement of SD (Video 2).
- Neuroimaging was not obtained due to sustained benefit from stimulation.
- For further characterization of voice symptoms SVL was performed ≥1 hour off stimulation and after 5 minutes on stimulation. (Video 2)(Table 2).

RESULTS, VIAODS & TABLES

Table 1: Optimized DBS settings

<table>
<thead>
<tr>
<th>Case</th>
<th>Left VM</th>
<th>Left VM</th>
</tr>
</thead>
<tbody>
<tr>
<td>C1</td>
<td>C1+1</td>
<td>C1-2</td>
</tr>
<tr>
<td>C2</td>
<td>C1+2</td>
<td>C1-2</td>
</tr>
</tbody>
</table>

Polarity | Amplitude (V) | Pulse Width (µs) | Frequency (Hz)
--------|--------------|-----------------|----------------
C1+1    | 4.0 V        | 90µs            | 140 Hz         |
C1-2    | 4.0 V        | 90µs            | 180Hz          |

Table 2: Strobovideolaryngoscopy

<table>
<thead>
<tr>
<th>Case</th>
<th>ON DBS</th>
<th>OFF DBS</th>
</tr>
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<tbody>
<tr>
<td>C1</td>
<td>Arytenoid spasm on 2-hour adduction and tension in the false vocal folds</td>
<td>Arytenoid spasm on adduction and tension in the false vocal folds</td>
</tr>
<tr>
<td>C2</td>
<td>Arytenoid spasm on 5-minute vocal spasms and decreased spasm in false vocal folds</td>
<td>Arytenoid spasm and decreased tension in the false vocal folds with marked improvement in sustained phonation</td>
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DISCUSSION
- Our two cases demonstrate improvement of SD with unilateral thalamic stimulation confirmed by SVL.
- There was no evidence of kinetic tremor in either case.
- Kinetic tremor extends through the entire larynx and is not limited to the vocal folds suggestive of essential voice tremor.
- SD is best managed with botulinum toxin injections to the vocal cords. However, this treatment is temporary and can be limited by weakened voice caliber with breathy quality in which cases alternate therapies are needed.
- A single case of spasmodic dysphonia reported to improve with bilateral thalamic stimulation reported. To our knowledge these are the first two cases of incidental SD reported in the literature that demonstrate improvement with unilateral thalamic stimulation.
- The VM nucleus receives somatosensory input from the cerebellum which is implicated in the pathophysiology of ET.
- Functional neuroimaging implicates cerebellar dysfunction in the pathophysiology of SD. Thus it is possible that stimulation of the cerebellar efferent pathways to the thalamus may result in improvement in both spasmodic dysphonia and limb tremor in our patients.
- Bilateral stimulation is traditionally considered necessary to treat midline symptoms, yet in our cases unilateral stimulation yielded substantial improvement in voice in both patients.
- These observations support further investigation of the effects of unilateral thalamic stimulation for the treatment of SD refractory to botulinum toxin therapy.

REFERENCES