Effects of ventro-oral thalamic deep brain stimulation in a patient with musician’s dystonia: illustrative case

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BACKGROUND Musician’s dystonia is a task-specific focal hand dystonia characterized by involuntary contraction of muscles while playing a musical instrument. Current treatment options are often insufficient.

OBSERVATIONS We present the effects of ventro-oral thalamic deep brain stimulation in a patient with musician’s dystonia. The patient was a 67-year-old pianist with musician’s dystonia who underwent deep brain stimulation with the ventralis oralis anterior and posterior nuclei of the thalamus as targets. The Tubiana and Chamagne rating scale was used to evaluate the effects of stimulation. The outcome was evaluated independently by four clinicians in a blinded manner at 3 months postoperatively. There was a distinct reduction of symptoms during stimulation. At 15 months postoperatively, the beneficial effect remained. No lasting side effects were observed.

LESSONS Further studies are warranted to evaluate the safety and long-term efficacy of this treatment modality.

https://thejns.org/doi/abs/10.3171/CASE22569

KEYWORDS musician’s dystonia; deep brain stimulation; focal hand dystonia; ventralis oralis anterior; ventralis oralis posterior

Dystonia has been defined as a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both.1 Focal dystonia is dystonia limited to a single body region. Examples of focal dystonia are writer’s cramp, blepharospasm, cervical dystonia, cranial dystonia, and limb dystonia.2 Musician’s dystonia (MD) is a form of task-specific focal dystonia that occurs only while playing a musical instrument.2 The prevalence of MD is 1% to 8% among professional musicians.3,4 Since this disorder is often misinterpreted as an orthopedic condition, this prevalence may be an underestimate. The dystonia usually occurs in the dominant hand and in the muscles controlling fine movements of the digits. For pianists in particular, it affects the fourth and fifth digit of the right hand in 90% of cases.5 Mean age at onset is generally the mid-30s.5 The etiology of MD is not completely understood but is probably multifactorial. Overuse and compulsive working behavior in combination with predisposing factors such as genetics and biomechanical characteristics are typically associated with the onset of MD.6 The interaction between these predisposing variants is thought to lead to dysfunctional or maladaptive brain plasticity.6

Current treatment options are often insufficient to save a musician’s professional career. First-line treatment modalities include physical therapy and botulinum toxin injections. A more invasive option is stereotactic ventro-oral thalamotomy, with 12 cases first described for writer’s cramp by Taira et al. in 2003 and 15 cases described for MD by Horisawa et al. in 2013.7,8 The ventral lateral nucleus, including the ventro-oral complex (Voc), of the thalamus is known to be an effective target for the treatment of writer’s cramp.7,9 Herein, we study the effects of ventro-oral deep brain stimulation (DBS) on MD in a professional piano player.
Illustrative Case

History
A 67-year-old right-handed professional pianist experienced involuntary movements of his right hand during piano playing. The symptoms had first developed at the age of 62, and his condition slowly deteriorated. It started out as a slight hesitancy of the fourth digit and progressed to a dystonic posture with extension of the second and fifth digit and flexion of the third and fourth digit present only while playing the piano and not while writing. His brother, a professional violinist, had to quit playing violin at the age of 28 because of MD. Our patient was diagnosed with MD 3 years after the onset of symptoms and was unable to continue public performances. He reported that this seriously affected his quality of life. Electromyography-guided botulinum toxin infiltrations at different specialized centers, trihexyphenidyl (2 mg three times a day), physiotherapy, and acupuncture were unsuccessful in treating the dystonia. Because all previous therapies had failed, we proposed thalamotomy 5 years after symptom onset. Although the dystonia of the right hand caused him to lose his ability to play the piano, it did not affect his ability to play the violin. He was a well-known professional violinist, had to quit playing violin at the age of 28 because of MD, and he could not play the piano because it required voluntary movements of his right hand during piano playing. The patient was extensively informed about DBS and its investigational nature.

Diagnostic Assessment
Neuropsychological examination showed normal cognitive performances for his age and education level. There were no symptoms of depression, anxiety, or other psychiatric contraindications for DBS. A panel of 68 dystonia and dyskinesia genes (including TOR1A, THAP1, GNAL, ANO3, GCH1, TH, SPR, SGCE, KMT2B, PRKRA, TAF1, ATP1A3 and ADCYS) was negative. Electromyography and brain magnetic resonance imaging (MRI) were normal. The severity of the MD was evaluated using the Tubiana and Chamagne Scale (TCS).10 This rating scale allows for the assessment of musical performance capabilities in patients with MD and consists of six categories varying from unable to play (score 0) to concert performance (score 5; Table 1). The scale is applied by a clinician and is applicable to all musical instruments. In this case, we scored the patient preoperatively as well as at 3 and 6 months postoperatively before and during stimulation (each time after adjustment of the stimulation parameters). A double-blinded evaluation was conducted at 3 months postoperatively. Stimulation was turned on and off, without the patient's knowledge of the condition. The piano playing was recorded and afterward evaluated independently in a blinded manner by four clinicians with piano experience. At 6 months postoperatively, the scores were assigned by the treating clinicians in a nonblinded manner.

Surgical Technique and Preoperative Findings
The brain electrodes were inserted stereotactically in the left ventralis oralis anterior (Voa) and ventralis oralis posterior (Vop) nuclei, under local anesthesia with intermittent sedation. A Cosman-Roberts-Wells stereotactic frame was fixed to the patient's skull, and stereotactic brain MRI (3 tesla, 1-mm thick slices) was performed. Based on the experience-based advice on thalamotomy targeting in patients with focal hand dystonia (FHD) of Taira et al.8 and based on the atlas of Schaltenbrand and Wahren, a schematic plan for electrode positioning was established preoperatively (Fig. 1). Taira usually makes one extended lesion, using three trajectories at a distance of 3 mm each to cover both the Voa and Vop nuclei.11 Because it may be difficult to cover this entire area with only one electrode, we decided to implant two electrodes, one in the Voa nucleus and one in the Vop nucleus, spaced approximately 4 mm apart. Given that the STar Drive Manual System (FHIC Inc.) of the stereotactic frame provides five trajectories for electrode insertion, each located parallel at 2-mm intervals, we planned one central trajectory at the junction between Voa and Vop nuclei. Electrodes were then implanted 2 mm anterior and posterior to this central trajectory.

Stereotactic planning of the target and trajectory were accomplished based on the peroperative MRI using Brainlab surgical navigation software. The trajectory was adjusted to the patient's individual anatomy (especially the location of blood vessels and the caudate nucleus).

Sedation was stopped once the burr hole was made. After incising the dura, three microelectrodes, each 2 mm apart, were placed for registration in the anterior, central, and posterior trajectories. Recordings were made while the patient played a music scale on the keyboard with his right (dystonic) hand versus not playing at all. This was repeated at an interval of 1 mm, starting at −8 mm from target to target along the three trajectories.

Because no clear differences in recording signals were observed at any point, we decided to use only the most anterior trajectory (Voa nucleus) and the most posterior trajectory (Vop nucleus) for further intraoperative stimulation testing. Stimulation with the macroelectrodes (60 μsec, 130 Hz, up to 8mA) was performed. The patient was blinded and did not know whether the stimulation was on or off or which amplitude was used. He was asked to score the dystonia between 1 and 10. Two neurosurgeons with piano experience evaluated the movement of his dystonic hand. Stimulation parameters were set with an external stimulator to a frequency of 130 Hz and pulse width of 60 μsec. Intraoperatively, an immediate positive effect on the dystonia was observed with stimulation of the Voa nucleus as well as the Vop nucleus. However, toward the end of the surgery, this effect could no longer be reliably evaluated because of patient fatigue. At target −8, target −5, target −2 mm and at the target, the therapeutic window was evaluated (Supplemental Table 1). The only adverse effects perceived by the patient at high amplitudes of stimulation were dizziness and lightheadedness. After determining the electrode position with the best clinical effect and the largest therapeutic window, the two final electrodes (Vercise Cartesia directional leads, 8 contacts, Boston Scientific) were implanted at the anterior and posterior trajectory of the STar Drive Manual System, that is, one in the Voa and one in the Vop.

### TABLE 1. The TCS: a subjective clinician-based rating scale of musical capabilities

<table>
<thead>
<tr>
<th>Score</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Unable to play</td>
</tr>
<tr>
<td>1</td>
<td>Plays several notes but stops because of blockage or lack of facility</td>
</tr>
<tr>
<td>2</td>
<td>Plays short sequences w/o rapidity &amp; w/ unsteady fingering</td>
</tr>
<tr>
<td>3</td>
<td>Plays easy pieces w/ restrictions; rapid sequences stir up problems</td>
</tr>
<tr>
<td>4</td>
<td>Nearly normal playing but avoids technically difficult passages for fear of motor problems</td>
</tr>
<tr>
<td>5</td>
<td>Normal playing; return to concert performance</td>
</tr>
</tbody>
</table>

Based on Taira and Hori, 2003.8
Intraoperative lateral fluoroscopy was used to verify the lead path and location. Thereafter, the neurostimulator (Vercise Genus Rechargeable, Boston Scientific) was implanted prefascially in the left abdomen with the patient under general anesthesia. The postoperative computed tomography scan was fused with the stereotactic MRI plan to verify the correct position of the electrodes. Both electrodes proved to be positioned in the planned targets. Final tip position of the most ventral contact for the Voa was 12.2 mm lateral to the anterior commissure-posterior commissure (AC-PC) line, 2.1 mm superior to the -line, and 0.5 mm anterior to the midcommissural point (MCP). Final tip position of the most ventral contact for the Vop was 12.9 mm lateral to the AC-PC line, 1.7 mm superior to the AC-PC line, and 3.1 mm posterior to the MCP (Fig. 2).

**Follow-Up and Outcome**

Follow-up evaluation was performed at different time intervals (Fig. 3). Both electrodes were on at each interval. The dystonia was scored using the TCS preoperatively and at 6 months postoperatively.
by the treating clinicians. At 3 months postoperatively a double-blinded evaluation was done by a panel of four clinicians with knowledge of playing piano, who scored multiple recordings with DBS on and off. The preoperative TCS score was rated as 1 by the treating clinicians.

On the first day after the surgery, the Voa electrode was tested and activated. Given the tiredness of the patient, we did not test the Vop electrode. After a monopolar review of both electrodes at day 11, the piano playing improved but still lacked speed. In the first 6 weeks after surgery, the patient’s mood deteriorated, according to the patient because there was only a little improvement in the dystonia at that time. He consulted a psychiatrist once, and his mood improved spontaneously after a few weeks.

At 3 months after surgery, a monopolar review of both electrodes was repeated while the patient played the piano (Video 1, Supplemental Table 2). Median TCS scores from the double-blinded evaluation were 2 when stimulation was off and 4 when stimulation was on, while using the most successful stimulation parameters after the monopolar review (Table 2). When determining the therapeutic window, the patient experienced dizziness and lightheadedness at 8 mA of stimulation. When stimulating at 5 mA, there was sufficient beneficial clinical effect without side effects. In the following weeks, the patient consulted three times for further minor adjustments. Final stimulation settings used contact 6 of the Voa and contacts 7 (40%) and 8 (60%) of the Vop. Parameters consisted of a frequency of 255 Hz, a pulse width of 60 μsec, and an adjustable amplitude between 0 and 10 mA.

At 6 months postoperatively, TCS scores were 4, as judged independently by the treating clinicians. The patient was very satisfied, and no additional adjustments to the stimulation settings were required. DBS was turned on continuously, and the patient experienced no negative side effects. He usually reduced the stimulation to 5 mA at night and increased it back to 10 mA in the morning to save battery, but he found that leaving the stimulation on 10 mA at night had no adverse impact on his sleep. When defining the therapeutic window, dizziness and intermittent contractions of the tongue were perceived by the patient at an amplitude >10 mA at the Vop electrode. These symptoms occurred only transiently and disappeared within a few seconds. At the level of the Voa electrode, no side effects were noted when increasing the stimulation to maximal amplitudes. Stimulation of the Vop electrode had the greatest beneficial effect on the dystonia, in contrast to a minor effect when using only the Voa electrode.

At 11 months postoperatively, an augmentation of the amplitude to 17 mA was necessary to maintain the same effect, but with a reduction of the pulse width to 30 μs (only Vop is on, 7-50%, 8-50% c+, 30 μsec, 225 Hz, 17 mA [0-17 mA]).

TABLE 2. TCS scoring results of four independent clinicians with piano experience

<table>
<thead>
<tr>
<th>Fragment</th>
<th>Clinician 1</th>
<th>Clinician 2</th>
<th>Clinician 3</th>
<th>Clinician 4</th>
</tr>
</thead>
<tbody>
<tr>
<td>1, on</td>
<td>4</td>
<td>3</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>2, off</td>
<td>3</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>3, off</td>
<td>2</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>4, off</td>
<td>2</td>
<td>3</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>5, on</td>
<td>4</td>
<td>4</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>6, on</td>
<td>4</td>
<td>3</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>7, off</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>8, on</td>
<td>3</td>
<td>3</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>9, off</td>
<td>2</td>
<td>2</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>

off = stimulation turned off; on = stimulation turned on.

The recordings were made 3 months postoperatively, and the clinicians were blinded as to which fragments the DBS was turned on or off. The median score is 2 with the stimulation turned off and 4 with the stimulation turned on. Stimulation parameters as indicated on the overall timeline in Fig. 3.
At 15 months postoperatively, the patient was satisfied with the DBS, as he could play concerts again (TCS score 5). He only switches the stimulator on during piano playing, and with this strategy he has a very good effect at 13 or 13.5 mA. However, when he keeps the stimulator on at night, he needs 17 mA the following day to obtain the same effect.

**Patient Informed Consent**

The necessary patient informed consent was obtained in this study.

**Discussion**

Functional and structural changes in the brain are observed in healthy musicians. However, in patients with MD, this plasticity of the brain when learning new tasks becomes maladaptive. Presumably, this is due to excessive training of repetitive movements in combination with other predisposing factors. These maladaptive neuronal structural and functional changes have been observed in the motor and sensorimotor cortices, as well as in the basal ganglia. For example, Kita et al. found that MD is associated with disruption of basal ganglia resting-state functional connectivity in the putamen. In turn, this influences the function of connected networks, such as the basal ganglia-thalamo-cortical loops. It is believed that interruption of these abnormally functioning loops by stimulation or lesioning of the Voa and Vop nuclei can improve dystonic symptoms. Therefore, the Voc has been identified as a proper target for thalamotomy as well as DBS in the treatment of different types of dystonia.

Lesional surgery of the Voc for patients with medically refractory FHD was introduced by Siegfried et al. in 1969. A recently published retrospective study by Horisawa et al. included 171 patients with task-specific FHD who underwent unilateral ventro-oral thalamotomy and provided class IV evidence concerning the feasibility and effectiveness of ventro-oral thalamotomy. DBS for FHD is less commonly performed, but several groups have obtained results similar to those with thalamotomy. These studies are case reports or case series with a total of 10 patients suffering from writer’s cramp. Fukaya et al. compared thalamic Voc DBS to globus pallidus internus DBS by implanting two electrodes in a patient with writer’s cramp. Thalamic Voc DBS turned out to be superior in this specific case.

To date, there are no comparative studies between DBS and lesional surgery for the treatment of FHD. In lesional surgery, there are no hardware-associated complications. According to a systematic review including 8983 patients, the overall risk of developing a hardware-associated complication after receiving DBS in general is 11.75%. Because the onset of MD is generally at a young age, the risk of hardware-associated complications after receiving DBS in general is 11.75%. The onset of MD is generally at a young age, the risk of hardware-associated complications after receiving DBS in general is 11.75%. Because the onset of MD is generally at a young age, the risk of hardware-associated complications after receiving DBS in general is 11.75%. The onset of MD is generally at a young age, the risk of hardware-associated complications after receiving DBS in general is 11.75%. 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Disclosures
Dr. Nuttin reported serving as the Medtronic Chair for Neuromodulation and Boston Scientific Chair for Neuromodulation for Stroke, outside the submitted work.

Author Contributions
Conception and design: Nuttin, Poncelet, Smeets, Visser-Vandewalle, Vandenberghe. Acquisition of data: Nuttin, Poncelet, Smeets, Vandenberghe. Analysis and interpretation of data: Nuttin, Poncelet, Smeets, Vandenberghe, Peeters, Van Bogaert. Drafting the article: Nuttin, Poncelet, Smeets, Visser-Vandewalle. Critically revising the article: Nuttin, Poncelet, Smeets, Vandenberghe, Peeters, Van Bogaert. Reviewed submitted version of manuscript: Nuttin, Poncelet, Taira, Vandenberghe, Peeters. Approved the final version of the manuscript on behalf of all authors: Nuttin. Administrative/technical/material support: Nuttin, Smeets. Study supervision: Nuttin, Taira.

Supplemental Information
Videos
Video 1. https://vimeo.com/849497246

Online-Only Content
Supplemental material is available with the online version of the article. Supplemental Tables 1 and 2. https://thejns.org/doi/suppl/10.3171/CASE22569.

Previous Presentations
Poster presentation of the abstract at the annual Belgian Society of Neurosurgery meeting, March 26, 2022, Leuven, Belgium.

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