A single case of MRI-guided focused ultrasound ventro-oral thalamotomy for musician's dystonia

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Musician's dystonia (MD) is a type of focal hand dystonia that develops only while playing musical instruments and interferes with skilled and fine movements. Lesioning of the ventro-oral (Vo) nucleus of the thalamus (Vo-thalamotomy) using radiofrequency can cause dramatic improvement in MD symptoms. Focused ultrasound (FUS) can make intracranial focal lesions without an incision. The authors used MRI-guided FUS (MRgFUS) to create a lesion on the Vo nucleus to treat a patient with MD. Tubiana's MD scale (TMDS) was used to evaluate the condition of musical play ranging from 1 to 5 (1: worst, 5: best). The patient was a 35-year-old right-handed man with involuntary flexion of the right second, third, and fourth fingers, which occurred while playing a classical guitar. Immediately after therapeutic sonications of FUS Vo-thalamotomy, there was dramatic improvement in the MD symptoms. The TMDS scores before; at 0 and 1 week after; and at 1, 3, 6, and 12 months after MRgFUS Vo-thalamotomy were 1, 4, 4, 5, 5, 5, and 5, respectively. No complications were observed. Focused ultrasound Vo-thalamotomy can be an effective treatment for MD.

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KEYWORDS focused ultrasound; thalamotomy; ventro-oral nucleus; dystonia; functional neurosurgery

DYSTONIA is a movement disorder characterized by involuntary, sustained, and patterned muscle contractions.⁴ Focal dystonia affects one part of the body and includes blepharospasm, cervical dystonia, focal hand dystonia, and other dystonias. Musician's dystonia (MD) is a type of focal dystonia, mainly involving focal hand dystonia, which develops only while playing musical instruments and interferes with skilled and fine movements.¹⁷ Musician's dystonia is more common among musicians in their 30s after years of repetitive and fine movements associated with playing musical instruments.⁹ Approximately 1% of professional musicians are affected with MD during their career.¹ Various treatments for MD are available, including botulinum toxin injections, oral medications, and physical therapy.^{1,14,18} Despite these conservative treatments, more than 50% of affected musicians ultimately give up their professional musical career.¹⁵ For patients with MD refractory to conservative treatments, lesioning of the ventro-oral (Vo) nucleus of the thalamus (Vo-thalamotomy) can cause dramatic improvement of symptoms.⁷ Recently, MRI-guided focused ultrasound

(MRgFUS) thalamotomy, which allows intracranial focal lesioning without an incision, at the ventral intermediate thalamic nucleus (VIM) has been reported to be a safe, effective, and feasible procedure for essential tremor.³ Previously, we reported the effectiveness of Vo-thalamotomy using radiofrequency and Gamma Knife ablation for MD.^{7,8} Here, we report the effectiveness of MRgFUS Vo-thalamotomy for MD. To our knowledge, this is the first case report of MRgFUS Vo-thalamotomy for MD with a successful outcome at the 1-year follow-up.

Case Report

History and Examination

The patient was a 35-year-old right-handed man with involuntary flexion of the right second, third, and fourth fingers, which occurred while playing a classical guitar. He had started playing guitar in his teens. At the age of 26 years, involuntary flexion of the right third finger developed only while playing a classical guitar. Subsequently, involuntary flexion of the right second and fourth fingers

ABBREVIATIONS FUS = focused ultrasound; GKT = Gamma Knife thalamotomy; MD = musician's dystonia; MRgFUS = MRI-guided FUS; TMDS = Tubiana's MD scale; Vo = ventro-oral; Vo-thalamotomy = lesioning of the Vo nucleus of the thalamus.

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FIG. 1. Pre- and postoperative neurological conditions and MRI studies. Involuntary flexion of the right second, third, and fourth fingers while playing a guitar (A). Fluctuation in symptoms was carefully assessed with an MRI-compatible guitar (B). One week after MRgFUS Vo-thalamotomy, improvement in MD symptoms was seen (C). Coronal T2-weighted MR image at 0 (D), 3 (E), and 12 (F) months after MRgFUS Vo-thalamotomy. The lesion at 12 months is still visible. Figure is available in color online only.

developed at the ages of 28 and 33 years, respectively (Fig. 1A). Since the age of 29 years, the symptoms also manifested while using a keyboard and while writing. The patient underwent tendon elongation of the affected fingers, which showed no effect. He was diagnosed with right focal hand dystonia, and MRgFUS left-sided Vo-thalamotomy for right hand dystonia was planned.

Surgery and Postoperative Course

With the patient under local anesthesia, a Cosman-Roberts-Wells stereotactic frame was fixed onto the skull after shaving the entire scalp. The stereotactic coordinates of the Vo nucleus were 1.5 mm posterior, 15 mm lateral, and 1.5 mm superior to the midcommissural point, which corresponded to the center of the coagulated lesion made by our radiofrequency Vo-thalamotomy. The procedure was performed with 3-T MRI (GE) and ExAblate Neuro systems (InSightec). During the treatment, an MRI-compatible guitar was used to assess the patient's dystonic symptoms (Fig. 1B). Repetitive low-power sonications for 10–20 seconds, inducing a temperature of 40°C, were performed to evaluate the side effects. Subsequently, therapeutic sonications for 10–20 seconds, with gradual increases in acoustic power and energy, were applied (13 sonications; maximum temperature 63°C). Immediately after therapeutic sonications, marked improvements were observed in the affected fingers while playing a guitar, without complications (Fig. 1C). Tubiana's MD scale (TMDS) was used to assess the neurological condition before and after MRgFUS Vo-thalamotomy as follows: score 1, subject plays several notes but stops because of blockage or lack of facility; score 2, subject plays short sequences without rapidity and with unsteady fingering; score 3, subject plays easy pieces but cannot perform more technically challenging pieces; score 4, subject plays almost normally but avoids difficult passages because of fear or motor problems; score 5, subject returns to concert performances.¹⁷ Clinical follow-up with TMDS

evaluations was performed at 0 and 1 week and 1, 3, 6, and 12 months after MRgFUS Vo-thalamotomy. Magnetic resonance imaging evaluations were performed immediately (Fig. 1D) and at 1 and 3 months (Fig. 1E) after MRgFUS Vo-thalamotomy. We had attempted to make a lesion with a volume of 300 mm³ on the T2-weighted MRI, and the volume of the created lesion was 348 mm³ (Fig. 1D). The TMDS scores before MRgFUS Vo-thalamotomy and at 0 and 1 week and 1, 3, 6, and 12 months after MRgFUS Vo-thalamotomy were 1 (Video 1), 4, 4, 5, 5, 5, and 5 (Video 2), respectively.

VIDEO 1. Preoperative guitar play. Involuntary flexion of right second, third, and fourth fingers severely interfered in continuous fine movement in guitar play. Copyright Takaomi Taira. Published with permission. [Click here to view.](#)

VIDEO 2. Guitar play at 12 months after MRgFUS Vo-thalamotomy. Marked improvement of dystonic symptoms continued without any deficits. Copyright Takaomi Taira. Published with permission. [Click here to view.](#)

At the 12-month visit, the patient could play a classical guitar without any motor or sensory deficit.

Discussion

The present case showed that MRgFUS Vo-thalamotomy is effective for MD and is a less invasive procedure, which may be preferred not only for aged patients with an atrophic brain who are unsuitable for surgical procedures but also for young patients.

Gamma knife thalamotomy (GKT), which does not require craniotomy, is another less invasive procedure and is useful for treating tremor as well as focal hand dystonia.^{8,13} The long-term effect and safety of GKT have been reported.^{10,12,19} Permanent complications after GKT, including sensory and motor deficits, can occur in approximately 3% of cases and from unexpected lesion expansion.¹⁹ The biggest concern with GKT is the inability to deliver a test

dose to confirm the optimal position for the lesion during the procedure. Recently, a long-term effect of FUS thalamotomy has been reported.² Permanent neurological complications have included sensory (14%) and gait (9%) disturbance.³ However, there are limited reports of cases treated with FUS thalamotomy, and the reported follow-up periods are short compared to those for GKT. The long-term effect and safety of FUS thalamotomy have not been established. Magnetic resonance imaging–guided FUS Vo-thalamotomy produces lesions using thermal ablation and is expected to produce long-term improvement safely, as with radiofrequency thalamotomy.⁷ In the present case, the effects of Vo-thalamotomy persisted for 12 months without any complications. However, symptoms can recur after radiofrequency Vo-thalamotomy because of insufficient or incorrect thermocoagulation that does not cover the entire Vo nucleus of the thalamus. Even with the use of FUS thermocoagulation, symptom recurrence due to insufficient coagulation with a single sonication at peak energy has been reported, with an absence of coagulated lesions on MRI at 3 months.¹¹ To maintain continuous therapeutic effects with permanent lesions, repetitive applications (4–5 times) of therapeutic sonications at the peak energy are required.

Although little attention has been paid to the surgical treatment of MD, several studies have shown the efficacy of Vo-thalamotomy not only for MD but also for other task-specific focal hand dystonias, such as writer's cramp, postapoplectic hemichorea, and paroxysmal kinesigenic dyskinesia.^{5,6,16} Although conservative treatments should be attempted first, in medically intractable cases, Vo-thalamotomy using FUS can be effective.

Additional studies with a larger sample size and long-term follow-up are needed to further elucidate the effectiveness of MRgFUS Vo-thalamotomy for MD.

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Disclosures

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Author Contributions

Conception and design: Taira, Yamaguchi. Acquisition of data: all authors. Analysis and interpretation of data: Taira. Drafting the article: Horisawa. Critically revising the article: Taira, Horisawa. Approved the final version of the manuscript on behalf of all authors: Taira. Study supervision: Taira, Yamaguchi.

Supplemental Information

Videos

Video 1. <https://vimeo.com/277439192>.

Video 2. <https://vimeo.com/277439212>.

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