Deep brain stimulation for treatment of hemichorea-hemiballism after craniopharyngioma resection: long-term follow-up

Case report

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Hemichorea-hemiballism is a rare movement disorder that has various causes. In treatment-resistant cases, both thalamic and pallidal functional procedures have been shown to yield beneficial results. Until now it has not been clarified whether the thalamus or the pallidum would yield a superior outcome.

After resection of a craniopharyngioma in this patient at the age of 49 years, hemichorea-hemiballism developed, with a latency of several weeks. Because the patient was greatly impaired by the movement disorder, she underwent implantation of deep brain stimulation (DBS) electrodes in the thalamic ventralis intermedius nucleus and the posteroverentral lateral globus pallidus internus. Although both pallidal and thalamic stimulation could suppress the movement disorder, the voltage needed was clearly less with thalamic than with pallidal stimulation. At the last available follow-up 25 months postoperatively, complete subsidence of hemichorea-hemiballism was achieved with long-term thalamic stimulation.

Long-term DBS therapy is an efficient treatment modality for refractory hemichorea-hemiballism in the long run (> 2 years). A bifocal (thalamic and pallidal) target paradigm allowed selection of the optimal stimulation site. Thalamic DBS was more favorable with regard to energy consumption. (DOI: 10.3171/2011.6.JNS101388)

KEY WORDS • craniopharyngioma • deep brain stimulation • hemichorea-hemiballism • pallidum • thalamus

We used a bifocal (thalamic and pallidal) target paradigm to select the optimal stimulation site in a patient with hemichorea-hemiballism caused by resection of a craniopharyngioma. Furthermore, long-term follow-up for more than 2 years is provided.

Case Report

History and Examination. This 52-year-old woman with a 3-year history of right-sided hemichorea-hemiballism was referred to our department for functional neurosurgery. At the time of admission, she had involuntary, predominantly choreic movements of her right leg consisting of variable semipatterned flexion-extension movements of her foot and toes, and also of her right arm, affecting mainly the hand. There was no facial involvement. She could not suppress the involuntary movements, which were absent during sleep. Results of the remainder of the neurological examination were normal; there was no spasticity and no weakness on her right side. In par-
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ticular, the choreic movements of her leg interfered with walking.

At the age of 49 years, she had received a diagnosis of craniopharyngioma, which became manifest with temporal hemianopia. Initial MR imaging studies showed a tumor in the suprasellar region that was more pronounced on the left side (Fig. 1). The tumor was removed via a frontotemporal craniotomy at another hospital. Postoperatively, the patient noted a sensation of heaviness of her right limbs, whereas the hemianopia improved markedly. Approximately 3–4 weeks later, she first noted involuntary movements of her right extremities, which increased over the following days. The MR imaging studies showed a small enhancing lesion located in the left GPe (Fig. 2). The movements consisted of large rotatory excursions affecting the leg more violently than the arm. The movement disorder was classified as hemichorea-hemiballism, and treatment with pimozide (7 mg per day) was initiated. In addition to pimozide, tetrabenazine was prescribed. During the next few months, there was continuous improvement of the movement disorder. The patient, however, did not tolerate medication because of progressive loss of initiative and increased fatigue.

After medication had been tapered off, there was a recurrent aggravation of the movement disorder, which then, however, had more choreic than ballistic elements. Furthermore, there was slight intermittent dystonic dorsal flexion of the right big toe. The patient also experienced mild dysesthesias of her right foot.

Operation. At the age of 53 years, the patient was admitted for DBS to our department. She was scheduled for bifocal thalamic and pallidal implantation of DBS electrodes to define the optimal target for stimulation, according to guidelines published elsewhere. She underwent implantation of quadripolar DBS electrodes (3387; Medtronic, Inc.) in the left posteroventral lateral GPi (target coordinates: \( x = 20 \) mm lateral to, \( y = 3 \) mm anterior to, and \( z = 4 \) mm below the AC-PC midpoint) and in the left thalamic Vim (target coordinates: \( x = 13 \) mm lateral to, \( y = 4 \) mm posterior to, and \( z = \) at the level of the AC-PC midpoint) with CT-guided stereotactic surgery in the awake state.

Microelectrode recording was used to refine the target in the GPi. We measured and recorded the firing rates of 16 neurons along the trajectory in the GPe and GPi. The GPe neurons showed a mean firing rate of 41 Hz. The mean firing rate of 12 GPi neurons was 53 Hz (range 11–81 Hz). A typical GPi neuron is shown in Fig. 2. We did not record any response to passive movements of the contralateral extremities. There was no microlesional effect after insertion of the DBS electrodes—either in the pallidal or in the thalamic target. Prolonged intraoperative testing of the different electrode contacts showed that thalamic stimulation completely suppressed the contralateral movement disorder at less than 2 V with bipolar stimulation (130 Hz; pulse width 210 μsec), whereas complete suppression could only be achieved at a threshold of 5 V with pallidal stimulation. It was therefore decided to use the thalamic DBS electrode for long-term stimulation; this was connected to a Soletra pacemaker (Medtronic, Inc.), which was implanted subcutaneously in the left subclavicular region. The GPi electrode was left in place, and its distal end was isolated in a subcutaneous pouch in the retroauricular region. There were no intraoperative or postoperative complications. Postoperative CT scans showed accurate placement of both electrodes in the GPi and the Vim (Fig. 3). Contacts 1 and 2 were placed within the sensorimotor part of the GPi. Contact 0 of the thalamic electrode was placed at the AC-PC level.

The initial settings for thalamic stimulation were as follows: 0.3 V, 130 Hz, 210-μsec pulse width, and bipolar stimulation, with contact 0 negative and contact 3 positive. At this amplitude, choreic movements were completely suppressed.

Postoperative Course. On follow-up evaluation 9 months postoperatively, the movement disorder was still completely suppressed. At 19 months postoperatively, there was a sudden reoccurrence of the choreic movements after the patient changed an electric bulb. Interrogation of the pacemaker revealed that it was switched off.
After resetting the stimulation, the choreic movements disappeared within minutes, at an amplitude of 0.8 V.

At the last available follow-up at 25 months, with long-term thalamic stimulation the right-sided choreic movements were no longer present. To assess the choreic movements we used the Abnormal Involuntary Movement Scale. The preoperative score was 15, which decreased to 1 (only patient’s awareness, no distress) at the 9-month follow-up, and was found to be the same at the 25-month follow-up visit.

**Discussion**

Although it is well known that hemichorea-hemiballism may be improved by both thalamic and pallidal procedures, our report is the first to demonstrate that thresholds for effective stimulation might vary in an individual patient. Although both pallidal and thalamic stimulation could suppress the movement disorder in our patient, the voltage needed was clearly less with thalamic than with pallidal stimulation. The efficacy of low-voltage thalamic stimulation was also confirmed on long-term follow-up, and there was only a mild increase, up to 0.8 V, with continuous, long-term stimulation. That this was not a placebo effect was confirmed after accidentally switching off the stimulation, which resulted in recurrence of hemichorea-hemiballism. Given consumption of battery energy, which increases by the square of the voltage, thalamic DBS appears to have a more favorable overall profile than pallidal DBS in hemichorea-hemiballism.

In the lesioning era of functional stereotactic neurosurgery, experience was obtained with pallidal and with thalamic radiofrequency lesioning, and both were shown to be effective. The largest series on thalamotomy was published by Krauss and Mundinger. A mean follow-up duration of 11 years was available in 13 patients. Twelve patients showed substantial improvement from thalamotomy with only a few side effects, including transient hemiparesis and mild persistent dystonia in 3 patients. Seven patients were free of hyperkinetic movements, and the other 5 had minor residual and predominantly hemichoreic hyperkinesias. The lesion in the ventrolateral thalamus was combined with a lesion in the zona incerta in 13 patients. A beneficial effect of thalamotomy has also been reported in other case series and case reports providing follow-up from 9.5 to 48 months. Comparable results were reported after pallidotomy, with almost all patients showing improvement at follow-up evaluations ranging between 9 months and 27 years. Furthermore, it appears that the risk profile was similar.

Since the advent of DBS, this treatment modality has also replaced radiofrequency lesioning for treatment of choreic movement disorders. Whereas pallidal DBS has been used in patients with Huntington disease and cases of neuroacanthocytosis, either thalamic or pallidal DBS was applied in single instances of hemichorea-
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hemiballism.12,22 Nakano et al.21 described improvement of hemichorea-hemiballism with DBS of the ventrolateral thalamus. The movement disorder in their patient developed in the course of severe diabetes, and a lesion in the striatum was found on imaging studies. After a follow-up period of 9 months, sustained improvement of the movement disorder was reported. Hasegawa et al.13 described a case of persistent vascular-related hemidystonia-hemiballism treated with DBS of the GPi. After 15 months of continuous stimulation, hemiballism was not present, but the dystonic posturing persisted.

It has long been thought that hemichorea-hemiballism is almost always related to a lesion in the STN. More recent studies, however, have revealed that lesions outside the STN might trigger the development of this movement disorder as well.7,10,23,28 Exceptionally, intracerebral lesions after neurosurgical interventions for removal of tumors may cause hemichorea-hemiballism.2,18 In our patient, craniopharyngioma resection most likely resulted in occlusion of a small perforating vessel, with a subsequent infarction in the GPe. It is important to note that in monkey models hemichorea-hemiballism could be caused by injection of bicuculline into the GPe and the putamen, although its severity was less than after injections into the STN.6,20

Microelectrode recordings have mostly demonstrated underactivity in the pallidum in patients with hemichorea-hemiballism. The Atlanta group demonstrated decreased firing rates (between 30 and 40 Hz) in the GPe and the GPi, which were significantly lower when compared with those found in patients with Parkinson disease.26,29 Our microelectrode recording findings are in line with these observations. Whether reduced reactivity to passive manipulation of the contralateral extremities is specific or not remains open to further investigation. It was postulated that the decreased neuronal activity in the pallidum might result in decreased inhibition of the thalamus and thereby induce the hyperkinetic movements.23,26 In contrast, the patient reported by Hashimoto30 showed almost normal firing rates, and Tang et al.27 reported similar firing rates in patients with Huntington and Parkinson disease.

Multifocal targeting and stimulation of basal ganglia nuclei is a technology that only recently has received more attention and more widespread use.3,4,5,10,15,25,30,31 Regarding the reversibility of the DBS technique, it offers the possibility to evaluate the efficacy of stimulation in different targets in the same patient. Also, it may be used to combine the effect of stimulation within the same target9,16 or in different targets.23 In our patient, thalamic stimulation appeared to be more favorable than pallidial stimulation. It should be noted, however, that the difference in voltage requirements does not necessarily denote superiority of one target over another, because many factors can influence the voltage required for therapeutic effect.

Conclusions

Thalamic DBS is an efficient treatment modality for refractory hemichorea-hemiballism in the long term. Although pallidal DBS appears to yield the same efficacy at the same risk profile, intrindividual comparison of both targets shows that thalamic DBS is more favorable with regard to energy consumption.

Disclosure

Joachim K. Krauss is a consultant to Medtronic and received honoraria for speaking. Hans-Holger Capelle received speaking fees from Medtronic. Thomas Kinfe was supported by Medtronic for participation in training courses.

Author contributions to the study and manuscript preparation include the following. Conception and design: Krauss, Capelle. Acquisition of data: all authors. Analysis and interpretation of data: all authors. Drafting the article: all authors. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Krauss. Administrative/technical/material support: Krauss. Study supervision: Krauss.

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