Camptocormia treated with bilateral pallidal stimulation

Case report

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The authors report the neurological, neurophysiological, and neuropsychological effects of using chronic bilateral pallidal high-frequency deep brain stimulation (DBS) in a case of disabling camptocormia.

Deep brain stimulation electrodes were implanted stereotactically to target the globus pallidus internus (GPi) bilaterally. Local field potentials (FPs) were recorded using the DBS electrodes and concurrent abdominal flexor electromyography (EMG) potentials during camptocormic episodes. Videotaped assessments of the movement disorder and neuropsychological evaluation before implantation and at 6 months after initiation of pallidal stimulation were recorded.

There was significant functional improvement following chronic pallidal stimulation, and some improvement was noted in neuropsychological scores. The GPi FPs showed temporal correlation with EMG-recorded rectus abdominis potentials. There were no treatment-related adverse effects.

The authors have found that chronic pallidal stimulation was safe and offered functional benefit in this severely disabling condition. The physiological studies may help further the understanding of the pathophysiology of this rare entity.

**KEY WORDS • camptocormia • pallidal stimulation • field potential •
deep brain stimulation**

Camptocormia is a rare disorder involving involuntary truncal flexion induced by sitting or standing.²⁸ It was reported as a psychogenic illness occurring among soldiers in World Wars I and II,^{9,29} and was subsequently regarded as a psychiatric illness.^{2,20} Recently it has been reported to occur in association with various other neurological conditions,^{15,23,35,37} including idiopathic Parkinson disease.^{5,6} In its severest manifestations patients are literally “doubled over” which renders most normal activities impossible or difficult. In some reported cases the disorder has been responsive to electrotherapy or to corticosteroid medications, whereas in others the disorder is refractory to all the various attempted treatment strategies.¹⁰

Chronic high-frequency DBS has been shown to improve the functional status in a number of movement disorders of varied causes.^{3,13,14,24} These include disorders in which the clinical manifestations are similar to those seen in camptocormia, especially conditions such as focal dystonia.^{3,11,14,27,33} With the development of relatively safe, image-guided stereotactic, nondestructive chronic im-

planted brain stimulation procedures in recent years,⁷ it may be scientifically and clinically valid to attempt to apply the principles and techniques of stereotactic functional neurosurgery to such a severely disabling and intractable entity as camptocormia. We have studied in detail a case of camptocormia neurologically, neurophysiologically, and neuropsychologically and present the beneficial effects of bilateral DBS of the medial GPi.

CASE REPORT

History. This 39-year-old man was referred to our clinic, which specializes in the management of movement disorders, for consideration of neurosurgical intervention to treat disabling episodes of truncal flexion. In June 1995, 4 years prior to referral, he had developed involuntary movements of the trunk, face, and limbs. Neurological examination demonstrated no underlying disease, and MR imaging of the brain revealed normal findings. His psychiatric history (ICD 10) was of generalized anxiety disorder (F41.1), emotionally unstable personality disorder (F60.3), and recurrent depressive disorder (F33). An initial diagnosis of tardive dyskinesia was made in view of prior treatment with neuroleptic medications during ad-

Abbreviations used in this paper: DBS = deep brain stimulation; EMG = electromyography; FP = field potential; GPi = globus pallidus internus; MR = magnetic resonance.

missions for deliberate self-harming behavior. These included haloperidol, droperidol, chlorpromazine, and lithium. Further neuroleptic therapy was avoided, and by 1997 there had been gradual improvement in the facial and limb movements, but he continued to experience disabling involuntary flexure of the trunk. Treatment with propranolol, sodium valproate, clonazepam, tetrabenazine, and clozapine had no effect.

Examination. In 1999 he underwent evaluation in our clinic. No neurological deficits were demonstrated while he lay down; however, sitting or standing provoked involuntary flexure of the trunk beyond the horizontal plane. When he attempted to correct this, he provoked a bobbing movement and severe lumbar pain (Videotape segment 1). This combination of posture-related truncal flexure and pain is often described as camptocormia (in Latin, “bent spine”) in the few cases reported in literature.^{5,10,21} A diagnosis of camptocormia was made accordingly.

Click here to view videotape segment 1: Preoperative assessment performed in October 1999. The patient has a normal posture supine but is unable to maintain a sitting position without using his arms. When standing or walking, he develops severe truncal flexure that cannot be overcome. (75 MB)

In view of the severely disabling nature of his disorder and its resistance to all medical therapy, a decision was made to implant bilateral GPi stimulators. We based this decision on reports of different kinds of focal dystonias, complex dyskinesias (both parkinsonian and nonparkinsonian), tardive dystonia, and some cases of generalized dystonia treated successfully by pallidal intervention.^{3,7,14,27,30,31,33,34} Informed consent was obtained.

Operation. In October 2000 he underwent computerized tomography/MR imaging-guided stereotactic implantation of bilateral DBS electrodes targeted to the anteroposterior segment of GPi, using the Radionics Image Fusion and Stereoplan.²⁵ The electrodes were initially externalized to test the effects of stimulation. Although no benefit was seen initially, 4 days after stimulation was initiated he reported a reduction in the effort required to maintain a sitting position. A dual-channel implantable pulse generator was subsequently connected and situated subcutaneously for chronic stimulation. We obtained MR images postoperatively to confirm the site of placement of the DBS electrodes (Fig. 1).

Fig. 1. Postoperative axial (*left*) and coronal (*right*) T₁-weighted MR images. The electrodes are identified by areas of hypointensity surrounded by a hyperintense “flare.” Both electrodes are straddling the GPi, one on each side.

SOURCES OF SUPPLIES AND EQUIPMENT

We obtained the pulse generator (Kinetra) from Medtronic (Minneapolis, MN). The Fusion and Stereoplan system was acquired from Radionics (Burlington, MA). Conmed (Rome, NY) manufactured the Neotrode Ag-AgCl electrodes. The amplifier (1902) was purchased from CED (Cambridge, UK). Off-line analysis was performed using Matlab software produced by Mathworks, Inc. (Natick, MA). Neil Hoggarth (Laboratory of Physiology, Oxford, UK) developed the in-house software.

RESULTS

Clinical Assessment

After 1 month of chronic pallidal stimulation, the patient no longer suffered from sudden “spasms” of truncal flexure, allowing him to eat in a normal position. At the latest follow-up examination, 6 months postoperatively, he was able to stand normally and walk in an upright position for approximately 50 ft. He was far more independent and was able to go to places such as the local pub and shops with minimal use of a cane. In addition, he can sit upright without undue effort to maintain his position (Videotape segments 2 and 3). He felt some heaviness around the neck initially in the first few weeks after initiation of stimulation; this has since subsided. He experienced no other adverse neurological effects following the pallidal stimulation. Switching off the pulse generator results in reappearance of his truncal flexion jerks within a variable period ranging from 2 to 12 hours.

Click here to view videotape segment 2: Assessment performed 3 months postoperatively (January 2001). After implantation of bilateral GPi stimulators, the patient’s symptoms gradually improved. He can sit upright without much effort and can stand upright for short periods of time. When walking a short distance, his posture is more normal, with intermittent truncal flexure. Assessment performed 7 months postoperatively (May 2001). Additional improvement is apparent—he requires no effort to sit upright, stands upright for longer periods, and walks with less difficulty while experiencing fewer episodes of truncal flexure. (81 MB)

Neurophysiological Assessment

Local FPs were recorded using the externalized DBS leads placed in the GPi 2 days postoperatively. Simultaneous EMG recording of the contra- and ipsilateral rectus abdominis was performed using neonatal surface disc electrodes. Signals were sampled at 250 Hz, amplified by 1000, filtered below 100 Hz, digitized, and continuously displayed on-line with an adjustable window. We used in-house developed software. Records were obtained at rest and after inducing the myoclonic-like abdominal contractions by asking the patient to stretch his arms forward or to try to stand. Off-line analysis of data was performed. During the analysis, particular attention was paid to potentials at frequencies below 20 Hz.

Analysis of the FP and the corresponding EMG data revealed that there was a clear temporal correlation between the basal ganglia potentials recorded from the medial GPi and the muscular electrical bursts from the rectus abdominis corresponding to the myoclonic-like jerks (Fig.

Fig. 2. Raw traces showing the FPs recorded in the right GPI and the concurrent trace from the left rectus abdominis EMG. The patient had myoclonic-like abdominal flexion at 25 and 55 seconds.

2). Fast Fourier transforms were derived to display the power spectra of the FP and EMG data (Figs. 3 and 4). The relationship between the two signals showed significant strength of correlation, or coherence—that is, the mathematical measure of correlation that we used—in the 0.5 to 3.5-Hz range (Fig. 5). The phase relationship between the brain and muscle signals in this frequency range suggests that the GPI signals affect the muscle (Fig. 6).

Neuropsychological and Functional Outcome

We used routine preoperative Oxford Movement Disorder Group neuropsychological evaluation protocol (described in detail elsewhere³⁰), which is composed of three components: 1) a clinical interview screening for any disturbances in mood, appetite, sleep, sensory-perceptual function, intellect, and thought, as well as questioning expectations of surgery, psychosocial circumstances, and any psychiatric history; 2) questionnaire measures of generic quality of life and psychological symptoms; and 3) a comprehensive psychometric assay of cognitive functions (that is, intelligence quotient, speech-language, spatial, memory, attention-executive, and speed of processing functions) (Table 1).^{1,17,26,30,32,36}

Preoperative assessment revealed that the patient's neu-

Fig. 3. Power spectrum in the frequency domain of the FPs recorded in the right GPI. The data have been filtered at below 20 Hz.

Fig. 4. Power spectrum of the left rectus abdominis EMG potentials. Only frequencies below 20 Hz are represented.

ropsychological profile was reasonably well preserved, although his performance on a number of tests (RSPM, phonemic fluency, story and figure recall, and recognition memory) was one standard deviation below his predicted premorbid level of ability. There was more marked and selective impairment demonstrated on tests of attention-executive function. His Total Percentage Disability rating on the Functional Limitations Profile (the United Kingdom version of the Sickness Impact Profile—a generic measure of quality of life⁴ was high (23%), and his score (13) was at a level of “caseness” (> 8–10; that is, where further clinical investigation might be indicated) on the anxiety subscale of the Hospital Anxiety and Depression Scale (Table 1). Postoperative assessment at 6 months revealed clear improvements in the patient's scores in almost all cognitive domains, particularly in tests sensitive to memory/new learning ability, speed of information processing, and attention (Table 1).

DISCUSSION

Clinical Outcome

In this case the neurological problem was complex with essentially two factors contributing to the patient's clinical presentation. The background of serious psychiatric disorder

Fig. 5. Graph showing the coherence between the right GPI FPs and the left rectus abdominis EMG potentials. It shows significant coherence between the brain and muscle activity in the 0.5 to 3.5-Hz range.

Fig. 6. Graph plotting the phase between the GPI FPs and the corresponding rectus abdominis EMG potentials in the coherent frequency range. The curve suggests that the brain signals determine the muscle activity.

der, with subsequent neuroleptic-induced tardive dyskinesia, was one factor that led to our patient's disability. These symptoms, however, had been in abeyance for the

last few years, as seen in his preoperative neuropsychological profile (Table 1). The predominant feature of his present motor disability, which had such a profound debilitating effect affecting his daily life, was the camptocormic restriction on his posture and gait. In recent times camptocormia has been described as a form of segmental dystonia²⁸ and has also been proposed as an extreme manifestation of parkinsonian rigidity or dystonia.⁵ It has been shown that pallidal stimulation improves a wide variety of dystonic conditions,^{27,33,34} as well as parkinsonian rigidity and dyskinesias.^{7,31} Because our patient's symptoms were refractory to medical therapy and were severely disabling, it seemed scientifically reasonable to offer him pallidal stimulation.

In this case the treatment-related benefit was not immediate, occurring gradually over the next 6 months. This has been reported in other cases of dystonia.^{12,27,33} As such, it argues against a major placebo contribution to the response. Perhaps this supports other reports in which the authors propose that camptocormia is a form of segmental dystonia.²⁸ Apart from the motor improvement in posture

TABLE 1
*Preoperative and postoperative neuropsychological test scores**

Test	Preop Scores	Postop Scores
overall intellectual ability		
National Adult Reading Test	105	
Standard Progressive Matrices IQ	91	105
speech & language skills		
Phonemic Verbal Fluency	31	37
Categorical Verbal Fluency	17	19
Boston Naming (no. of correct answers)	37	36
speed & capacity of language processing (z score)	0.44	1.56
speech articulation rates	10.8	11
attention-executive skills		
Digit Span (forward/backward)	6/4	8/5
CANTAB		
spatial span (z score)	-0.77	-0.77
RVIP "A" Prime	-2.7	-1.2
SOC (minimum move solutions)	-1.2 (raw score 6/12)	-0.63 (raw score 8/12)
ID/ED (stages reached; ED-shift =8)	-1.8 (raw score 7/9)	-1.8 (raw score 7/9)
spatial skills		
Judgement of Line Orientation	24	28
copy of MCG Complex Figure	31	34
memory/new learning		
Recognition Memory Test		
words	-0.33	0.66
faces	-0.66	-0.33
AMIPB Story Recall		
immediate	-0.89	0.59
delayed	-0.94	0.92
MCG Complex Figure Recall		
immediate	13	20.5
delayed	12.5	17.5
questionnaire measures		
FLP disability (total %)	23	23
HADS		
anxiety	13	14
depression	3	2

* The National Adult Reading Test and the Standard Progressive Matrices IQ have control mean scores of 100 ± 15 (standard deviation); all other values represent raw scores unless otherwise noted. Abbreviations: AMIPB = Adult Memory and Information Processing Battery; CANTAB = Cambridge Neuropsychological Test Automated Battery; ED and ID = Extradimensional/Intradimensional Set Shifting task (a decomposition of the Wisconsin Card Sorting test); FLP = Functional Limitations Profile; HADS = Hospital Anxiety and Depression Scale; MCG = Medical College of Georgia; RVIP = rapid visual information processing (a continuous performance test of sustained vigilance); SOC = the Stockings of Cambridge (an analog of the Tower of London test of planning).

Pallidal stimulation for camptocormia

and gait (Videotape segments 1–3), we also documented improvement in almost all cognitive domains, particularly in tests sensitive to memory/new learning ability, speed of information processing, and attention (Table 1). This finding reinforces the current opinion that camptocormia is not simply a motor disorder. Another important observation that refutes the likelihood of a beneficial response being due to a placebo effect was the variable period of time for the camptocormia-related physical symptoms to recur after stimulation was switched off. After 1 month of DBS, the symptoms recurred within 2 hours of ceasing stimulation. Following 6 months of DBS, it took an overnight period for the dystonic movements to manifest after stimulation was stopped. This variability has been reported in cases in which pallidal stimulation was used to treat other forms of dystonia and has been ascribed to plastic changes or to reorganization of the somatosensory input to the GPi neurons.³³

Electrophysiological Monitoring

Field potential recording allows sampling of abnormal synchronous discharge in a population of neurons as opposed to the single-unit recording more commonly used in functional stereotactic neurosurgery.¹⁸ We believe this permits better understanding of abnormal electrical activity in a functionally cohesive subcortical area in patients with a pathological condition. This is especially true if there is a concurrent recording of abnormal electrical activity from related peripheral tissue—for example, EMB-detected tremor from corresponding muscle groups^{18,22} or cortical electrical activity.¹⁹ We have found that FP recording has improved our ability to target effective sites for chronic DBS.^{18,22} Highlighted by this case is the importance of monitoring of FPs from the deep brain nuclei with simultaneous recording of relevant EMG parameters, to achieve accurate placement of the stimulating electrodes. We found that the brain potentials and the corresponding EMG phenomena displayed a strong temporal correlation, and this was matched by the clinical response observed in the patient. The phase relationship between the basal ganglia and affected muscle electrical activity sheds some light on the possible pathophysiological pathway of this abnormal movement. This along with the results of the coherence study, suggests that abnormal synchronous activity in the medial GPi is closely related to the involuntary truncal flexion and perhaps drives this action. This observation supports the case for an association between GPi stimulation and clinical benefit in this patient. Other authors have also commented on the electrophysiological link with GPi activity in cases of dystonia.¹⁶

CONCLUSIONS

We have successfully treated a complex case of camptocormia by administering pallidal stimulation. We also explored the electrophysiological link between the affected muscles and the GPi in camptocormia. We hope this will add to the existing body of knowledge concerning this condition as well as to the role of pallidal stimulation in dystonic disorders in general.

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