The uncommon syndrome of camptocormia is characterized by a forced abnormal posture of the trunk with flexion in the thoracolumbar spine, which increases during walking and abates in the sitting and, mainly, in the recumbent positions. The syndrome was first described in 1818 by the English physiologist Brodie, but it was the French neurologist Souques, in 1915, who coined the term camptocormie. The term camptocormie or camptocormia is composed of 2 Greek words: the verb "καμπτ-ω" (campt-o), meaning to bend, and the noun "κορμ-ος" (corm-os) meaning trunk. Brodie suggested that this abnormal posture could be caused, primarily, by organic affections, such as a destructive process of the vertebrae or myopathy. Others proposed that the flexion of the trunk was a conversion reaction of soldiers who were unable to cope with the stress of combat and military life during World War I and, perhaps, were predisposed to bent posture by prolonged walking in trenches.

During the past 2 decades, many reports have described cases of camptocormia secondary to PD, lesions in the lenticular nucleus, muscular disorders, myasthenia gravis, amyotrophic lateral sclerosis, and other neuromuscular disorders, as well as paraneoplastic and other neurological conditions. In this paper, we describe 2 cases of severe idiopathic camptocormia due to primary dystonia. The condition was resistant to all appropriate drug treatments in both cases, and the patients were eventually treated successfully by DBS. A similar impressive response to GPi DBS was also observed in a case of idiopathic bent neck (camptocephalia). The reversal of the bent-spine state of these patients by DBS of the GPi raises important issues with respect to the role of the GPi in the maintenance of erect posture and the adoption of bipedalism by humans.

Case Reports

Our 2 patients underwent an extensive battery of tests to establish their diagnosis, including: biochemical blood and CSF examination, brain and spinal MR imag-
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Implanted according to our technique. Surgery was quadripolar (M387–28 and M387–40, Medtronic, Inc.) were or paraneoplastic conditions. 

In both cases, bilateral DBS electrodes (Medtronic quadripolar 3387–28 and 3387–40, Medtronic, Inc.) were implanted according to our technique. Surgery was performed with patients in the supine position under local anesthesia. The symptoms of camptocormia abated in the supine position, and the patients tolerated awake surgery without difficulty. Stereotactic coordinates of the target in the posterolateral ventral GPi were 2 mm in front of the midcommissural point, 4 mm below the intercommissural line, and 21 mm lateral to the midline. Introperatively, microelectrode recordings were used for target confirmation, and macrostimulation was used to test for stimulation-induced side effects. The leads were secured in place using Stimloc (IGN, Inc.) and connected to the implantable pulse generator (Kinetta, Medtronic, Inc.) in the subclavicular area. No perioperative or postoperative complication occurred in either patient. Postoperative MR imaging scans confirmed the correct position of the leads. The particular clinical characteristics and the postoperative outcomes of the 2 patients are described below.

Case 1

This 26-year-old woman complained of a tendency to lean forward 3 years prior to being treated by us. Soon after the onset of symptoms, the forward body flexion worsened; this made it impossible for her to maintain the body’s center of gravity within the “walking base” of the legs and forced her to fall repeatedly forward down to the ground. Despite her efforts, she became unable to stand up or walk unaided even after adopting a flexed body posture. Six months after the onset of symptoms, the patient was able to stand up and walk only with the help of another person or by using her hands to support her trunk against a pole or the wall (Fig. 2A, Video 1). Thereafter, she was practically confined to bed or wheelchair.

**Video 1.** Case 1. Preoperative and postoperative video clips. Preoperatively, the patient was unable to sustain upright posture without support. Ten days postoperatively, the ability to sustain upright posture had been fully restored and the patient was able to walk unaided. Click here to view with Windows Media Player.

Deep brain electrodes were implanted bilaterally in April 2006. Stimulation was initiated on the 3rd postoperative day and was applied in continuous mode and unipolar configuration; the initial parameters were: amplitude 0.5 V, pulse width 90 µsec, and frequency 130 Hz. Two days later (on the 5th postoperative day), the amplitude was increased to 2.2 V. The patient’s trunk began gradually to rise and, within 36–48 hours, her body adopted the normal position (Fig. 2B, Video 1). She kept complaining, however, of her body feeling “heavy.” Two months later, the amplitude was increased to 3.0 V while the other parameters remained unchanged; approximately 6 weeks after that, the patient said the feeling of “body heaviness” had stopped. At most recent evaluation, 44 months after the DBS operation, she remained very well; her body posture remained normal and she was enjoying an excellent clinical and functional outcome. She had a new job and had uneventfully undergone an operation for a pilonidal cyst.

**Case 2**

This 21-year-old man developed the first symptoms 6 months prior to his admission to our unit in the form of a sensation of his head being heavy and difficulty in maintaining it in the normal upright position. Within a week of the onset of these symptoms, the head fell forward markedly, to the extent that the chin was in contact with the chest. During the second week, his trunk became severely bent, and this posture resulted in his arms adopting a parallel position to the legs, almost approaching the floor, where they could potentially support his body and
prevent his fall down to the ground (Fig. 3A and B, Video 2).

**VIDEO 2.** Case 2. Preoperative and postoperative video clips. Preoperatively, a severe fixed bent spine prevented the patient from keeping the erect posture without support, and he was confined in a wheelchair. Fifteen days after GPi DBS, the camptocormia had fully subsided and the patient’s stance and walking pattern appeared completely normal. Click here to view with Windows Media Player.

Deep brain electrodes were implanted bilaterally in June 2006. Stimulation was initiated on the 4th postoperative day and was applied in continuous mode and unipolar configuration; the initial parameters were amplitude 0.5 V, pulse width 90 µsec, and frequency 130 Hz. Even at these settings, the patient reported that his body felt “lighter,” perhaps because of the micropallidotomy effect. Four days later, on the 8th postoperative day, the amplitude was increased to 2.5 V. At these settings, the patient’s trunk came to the upright position, but the neck forward flexion and the head drop improved only slightly. Two days later, on the 10th postoperative day, the amplitude was increased to 3.0 V. The other parameters remained unaltered—that is, pulse width remained at 90 µsec, and the frequency at 130 Hz. The bent neck was corrected and the patient’s head returned to the upright position within the next 12 hours (Fig. 2C, Video 2). As of this writing, the duration of follow-up has exceeded 42 months. The patient has returned to a normal life with a full range of daily activities, including his studies as a university law student and sports activities such as swimming and jogging.

**Discussion**

As an increasing number of dystonia sufferers are being treated by means of DBS, new forms of the disease that are likely to respond favorably to electrical stimulation are being identified. In the peer-reviewed literature, we found only 11 cases of camptocormia in which the condition was treated successfully with DBS; all of them were secondary to PD or tardive dyskinesia. In all these cases, the time for recovery, following DBS, ranged from 3 to 6 months. The 2 cases of idiopathic camptocormia reported in the present article are of particular interest because both patients experienced a complete restoration of erect posture within 10 days following DBS of the GPi. The known causes of secondary dystonia and the possibility of either somatization disorder, according to DSM-IV-TR, or psychogenic dystonia were ruled out in both patients. To the best of our knowledge, idiopathic camptocormia has not, to date, been an indication for DBS treatment. In the face of the striking response of idiopathic camptocormia to DBS that we observed, it is interesting to review potential mechanisms underlying the effects of DBS in primary compared with secondary camptocormia.

**Idiopathic Versus Secondary Camptocormia: Pathophysiological Basis of the Response to DBS.** Over the past decade, 2 lines of thought have prevailed in the interpretation of mechanisms underlying camptocormia secondary to PD. The first theory associates camptocormia with peripheral mechanisms—mainly, myopathic changes in the antigravity muscles associated with trunk extension. The second theory considers camptocormia as a focal action dystonia of the spine—that is, rigidity of the axial flexion muscles with a weakness of the erector spinal muscles, integrated into the broader PD phenomenology. In particular, the dystonic posture overlaps with and is superimposed upon the postural abnormalities typically present in PD. The underlying disrupted mechanisms may involve the striatum, its projections to the reticulospinal tract, or the thalamus and other interconnected brainstem areas involved in the control of posture, such as the pedunculopontine nucleus. The above-mentioned theo-
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Fig. 4. Coronal and axial MR images showing the DBS electrode tips lying at the posteroventral GPi, bilaterally.

ries are not contradictory and might be linked, as focal myopathy of the paraspinal muscles might be secondary to a primary segmental dystonia of the spine.5,20

Idiopathic camptocormia can be explained on the basis of models of dystonia originally proposed by Albin et al.,1 DeLong,7 Vitek,29 and Bar-Gad and colleagues,4 and the classical “noisy signal hypothesis” of Marsden and Obeso.17 The currently accepted pathophysiological model suggests that dystonia leads to an aberrant pattern of basal ganglia activity that disrupts local and distant function. Critical to the success of this model remains the assumption that different disease phenotypes are associated with phenotype-specific noisy signals—that is, different patterns of synchronized, disruptive activity.5 Accordingly, functional somatotopy in the GPi is quite possible in that autonomous oscillations of specific relay nuclei clusters regulate specific cervical and spinal movements.3,12,18

In dystonia, the beneficial effects of GPi DBS in altering basal ganglia output and modifying thalamocortical and subcortical activity are usually progressive, with early and late responses. Phasic or mobile dystonic components improve early, while tonic or fixed components respond progressively, over weeks or months, because of brain plasticity; pathological neural reorganization that has developed over the lengthy course of dystonia needs a prolonged period of stimulation to be modulated therapeutically. The above limitations may be important in understanding the latency of response to GPi DBS in the reported cases of secondary camptocormia, with a range from 3 to 6 months.18,19,25 In those patients, bent spine constituted an endophenotypic dystonic abnormality23 (part of a wider abnormal clinical phenotype), in which the basal ganglia dysfunction is part of a process of aberrant synaptic reorganization that has evolved over the course of the primary disease; consequently, a prolonged period of time is needed for the benefit of GPi DBS to become apparent.

The 2 cases presented in this article highlight idiopathic camptocormia as a new emerging indication for DBS of the GPi. In these cases the following 3 features are of particular interest: 1) the purity of clinical phenotype (bent spine with no other dystonic symptoms or underlying pathology); 2) the quick response to DBS; and 3) the low stimulation voltage required for restoration of normal erect posture. On the basis of the prevailing pathophysiological model of dystonia,7 the above characteristics of our cases suggest that idiopathic camptocormia may be caused by an abnormal oscillation (“noisy signal”), highly confined in a GPi cluster that is critical for the maintenance of body extensor activity, upright posture, stance, and walking; this highly localized “local” disorder is not associated with either spatial extension of abnormal synchrony within the basal ganglia or cortical remodeling.

The GPi and the Evolution of Erect Posture. In addition to the 2 cases of idiopathic camptocormia presented in this report, a third patient, a 52-year-old woman, suffering from treatment-refractory idiopathic camptoccephalia (head drop) has been treated in our unit by means of bilateral DBS of the GPi with excellent results.22 The positions of the electrodes were carefully analyzed by digital fusion of the postoperative MR imaging scans with the preoperative stereotactic CT scans in all patients. In all cases, the stimulating pole was located near the posteroventral GPi (Fig. 4). Clearly, brain regions such as the interfastigial cerebellum and the bilateral midbrain tegmentum have an important role in the maintenance of erect posture and bipedalism.13 However, none of these structures has been a target for DBS and, to the best of our knowledge, restoration of erect posture has only been achieved by DBS of the GPi. The positive response to GPi DBS in secondary camptocormia18,19,23,25 idiopathic camptoccephalia,22 and the idiopathic camptocormia presented in this report suggest that the posteroventral GPi is involved in the maintenance and regulation of erect posture.

Many different evolutionary developments have facilitated the adoption of erect posture by humans6 as a response to a changing and challenging environment. As the anatomy of the primitive human body adapted to meet the requirements of erect posture and bipedalism, it is highly probable that the brain “rewired” itself to adapt to the requirements of a changing body and new forms of mobility. In that process, remodeling and generation of particular oscillations in the GPi might have been an essential requirement for the appropriate balancing of extensor and flexor activity to maintain the erect posture. In dystonic patients, dysregulation of the GPi cluster responsible for erect posture could induce an electrophysiological regression to a less evolved GPi function such as that existing before the adoption of bipedalism. The therapeutic benefits of DBS, as presented in this report on dystonic camptocormia and in our previous article on dystonic camptoccephalia,22 may lead to a better understanding of the significance of the electrophysiological activity of the globus pallidus in the adoption, restoration, and maintenance of upright posture and bipedal walking.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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