Spinal deformity and Parkinson disease: a treatment algorithm

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Object. The authors review the literature on the treatment of spinal deformity in patients with Parkinson disease (PD) and formulate a treatment algorithm.

Methods. The authors provide representative cases of patients with PD and spinal deformity who underwent deep brain stimulation (DBS) or spinal surgery.

Results. In patients with PD and spinal deformity who undergo spinal surgery there is a high rate of acute and delayed complications. Patients who undergo DBS, while having significantly fewer complications, often do not regain sagittal balance.

Conclusions. Cases involving PD and camptocormia have a high rate of complications when spinal surgery is performed. The authors prefer to offer spinal surgery only to patients with coexisting spinal stenosis causing radiculopathy or myelopathy. Patients with PD and camptocormia without spinal stenosis may be considered for DBS, but the results are mixed. (DOI: 10.3171/2010.1.FOCUS09288)

Key Words • Parkinson disease • camptocormia • spinal deformity • Pisa syndrome

Parkinson disease is a neurodegenerative disorder that affects over 1 million people in the US.19 It is estimated that the lifetime risk of developing PD is 1.5%.7,10 With the aging of the US population, the prevalence of PD will likely continue to grow.5 A recent estimate of the prevalence of deformities (involuntary trunk flexion/camptocormia, anterocollis, scoliosis) in PD was 33.5%.2

The cardinal motor signs of PD are 4–6-Hz resting tremor, rigidity, bradykinesia, and gait disorder/postural instability. Other symptoms include stooped posture, hypophonia, and paucity of facial expression. However, it can be difficult to diagnose PD correctly, and the early signs of PD can often be subtle. This has particular relevance when trying to understand the impact of surgical interventions on this population.19 In the later stages of PD, the patients have a risk of developing dementia.1,22 The dementia may contribute to death due to PD.21

Patients with PD may present with postural deformities. Several factors can contribute to postural instability including axial rigidity, poor trunk coordination, orthostatic hypotension, and difficulty integrating various sensory inputs. The postural instability contributes to increasing difficulty with transfers, gait, ability to live independently at home, and falls.3

A number of spinal deformities have been described in association with PD. The stooped posture classically associated with PD is the most common abnormality. Other disorders include camptocormia, myopathy-induced postural deformity, Pisa syndrome, and degenerative scoliosis. Here, we review the literature on surgical treatment of spinal deformity in patients with PD, including effects of DBS and spinal instrumentation.

Camptocormia

Camptocormia, or “bent spine syndrome,” is an extreme forward flexion of the thoracolumbar spine, which often worsens during standing or walking, but completely resolves when supine. The term itself is derived from the Greek “kamptos” (to bend) and “kormos” (trunk). While the condition was described as early as 1818, the term camptocormia was first proposed in 1914 to describe the forward flexion posture of some soldiers in World War I who had to move through the trenches in a bent posture to avoid injury.8,18,28,31

Camptocormia is used to describe the extreme forward flexion of the spine associated with a number of causes including dystonia, Tourette syndrome, amyotrophic lateral sclerosis, myopathy, myositis, multiple system atrophy, PD, and conversion disorder. While initially thought to be a rare manifestation of PD, recent estimates of the prevalence of camptocormia in patients with PD vary from 3–12.9%.2,20,33 It is unclear if the
prevalence of camptocormia varies with the severity of the PD.3,6,11,20,33

Medical management of camptocormia in PD remains suboptimal. Azher and Jankovic14 have reviewed the cases of 16 patients with camptocormia associated with PD (11 patients), dystonia (4 patients), and Tourette syndrome (1 patient). Twelve patients received levodopa therapy with minimal or no improvement in camptocormia. In 9 patients botulinum toxin Type A was administered into the rectus abdominus muscle, with improvement in 4 patients. Ho et al.37,38 have described a single patient in whom camptocormia improved after adjustments in dopaminergic therapy. Von Coelln and colleagues37 reported on 4 patients with PD who received ultrasound-guided injections of botulinum toxin Type A into the iliopsoas muscle. While they found the technique to be safe, with only patients receiving the highest doses reporting mild weakness of hip flexion, they also found no significant postural improvement. Bloch et al.5 have reported a case control study (8 patients in each arm) and found that patients with PD and camptocormia responded poorly to levodopa treatment and had levodopa-unresponsive axial symptoms.

Subthalamic nucleus DBS has been reported to improve camptocormia associated with PD. Sako et al.27 have reported on 6 patients in whom they documented a mean improvement of 78 ± 9.1% in thoracolumbar angle after bilateral STN stimulation. Hellmann et al.,16 Yamada et al.,29 and Micheli et al.23 each reported on a single case of PD and camptocormia in which the patient improved after bilateral subthalamic DBS or GPi stimulation. Reports on the responsiveness of camptocormia in PD patients have been inconsistent. Of the 16 patients reported on by Azher and Jankovic,14 one underwent placement of bilateral STN electrodes with no improvement in camptocormia. Most recently, Umemura et al.34 reported on a retrospective analysis of 18 patients (8 with camptocormia, 10 with Pisa syndrome) who underwent subthalamic DBS placement. In 13 patients with a moderate postural abnormality, 11 patients ultimately improved. In the 5 patients with severe postural abnormality, 2 patients improved slightly. Deep brain stimulation has also been reported to improve camptocormia associated with other movement disorders. Fukaya et al.33 reported improvement of camptocormia in 3 patients with primary dystonia who underwent placement of bilateral GPi DBS. Nandi et al.24 published a case report of a patient with tardive dyskinesia and camptocormia who responded to the placement of bilateral GPi electrodes for DBS. However, it is unclear if the pathophysiology of camptocormia in PD is similar to that of camptocormia associated with primary dystonia.

Babat et al.4 reported on 14 patients with PD who underwent spinal surgery (mostly short-segment laminectomies/fusions; 1 patient underwent multiple-level cervical corpectomy, 1 underwent deformity correction, and 1 underwent L-1 transpedicular decompression for burst fracture). They noted that 11 patients underwent 22 additional operations at the same or adjacent levels for instability. Four of these patients had hardware failure or pullout, requiring 10 additional operations. Their conclusion was that the primary cause of failure was persistent kyphosis or segmental instability. Peek et al.29 recently described the case of a patient with PD and camptocormia who underwent posterior T7–ilium fixation. The patient required several surgical revisions, prolonged hospitalization, and rehabilitation. Although they were ultimately successful in restoring spinal balance, their conclusion was to consider surgical intervention only after subthalamic nucleus DBS has been performed and then only in patients who were highly motivated to walk.

Myopathy Associated Postural Deformity in PD

Inflammatory myopathy of the paraspinal muscles can mimic the appearance of camptocormia in PD. Wunderlich et al.38 have described a 63-year-old man with PD in whom a camptocormia-like deformity developed. They noted hyperintensity (consistent with edema) within the paraspinal muscles and histopathological features consistent with myositis. The patient was treated with steroids and they noted marked improvement in forward flexion.

Myopathy with nemaline rods, end-stage myopathy, with autophagic vacuoles, mitochondrial myopathy, and necrotizing myopathy have all been associated with camptocormia in patients with PD.15,25,30 Gydnia et al.15 have studied 19 consecutive muscle biopsies obtained in patients with PD and either camptocormia or dropped-head syndrome (anterocollis), finding abnormal muscle biopsies in all patients. Although MRI images were not abnormal in all patients, MR imaging generally showed fatty degeneration of the paravertebral musculature or neck extensor musculature in many of them. Electromyography was also generally consistent with myopathy changes.

Pisa Syndrome

Pisa syndrome is characterized by a lateral flexion of the trunk when sitting or standing.2 In addition, there is an associated backward axial rotation of the spine. The term itself is derived from the image of the patient leaning like the Leaning Tower of Pisa. The condition was first described by Ekbom et al.5 in 1972. It is generally associated with the use of neuroleptics, antiemetics, and/or cholinesterase inhibitors.12,36 Treatment generally consists of removing the offending medication or reducing the dosage. Gambarin et al.4 reported on one patient with PD in whom Pisa syndrome developed without having received neuroleptic drugs, antiemetics, antipsychotics, selective serotonin reuptake inhibitors, or benzodiazepines. Cannas et al.9 described 8 patients with PD and Pisa syndrome whose symptoms were brought on by the introduction of or increase in a dopaminergic medication. In a single case report Santamato et al.29 described a patient with PD and Pisa syndrome who benefited from a rehabilitation program and botulinum toxin Type A therapy after medication adjustments were made.

We present representative cases from our experience of treating camptocormia in patients with PD. We then propose a treatment algorithm incorporating a multimodality treatment strategy.
Spinal deformity and Parkinson disease: a treatment algorithm

Case Reports

Case 1: Bilateral STN DBS

This patient was a 59-year-old man with advanced idiopathic PD and motor fluctuations. Preoperatively it was noted that he had severe camptocormia (Fig. 1 left). He underwent placement of bilateral STN deep brain stimulators and a right chest dual channel pulse generator. He was discharged to an acute rehabilitation facility on postoperative Day 6 in good condition for several weeks after his hospitalization. At 2-year follow-up, his gait and ease of ambulation had improved, but he had no significant improvement in his camptocormia posture (Fig. 1 right).

Case 2: Bilateral GPi DBS

This patient was a 59-year-old man with PD and severe camptocormia. Parkinson disease was diagnosed based on the initial presenting symptoms of a severe stooped posture, decreased fine finger movements, and bilateral hand tremor. The patient’s camptocormia only minimally responded to dopaminergic medications, and he would fall several times a day. He had begun wearing kneepads to prevent further injury from his frequent falls. He underwent placement of bilateral GPi DBS electrodes to alleviate his parkinsonian symptoms as well as treat his camptocormia. He was discharged to an acute rehabilitation facility on postoperative Day 5 in good condition. At 15-month follow-up, while having some improvement in his parkinsonian symptoms, he continued to suffer from severe camptocormia.

Case 3: Short-Segment Spinal Fusion

This patient was a 68-year-old man with PD, severe camptocormia, chronic low-back pain, bilateral lower-extremity pain, and severe spinal stenosis (Fig. 2 left). He had previously undergone a left L4–5 hemilaminotomy that gave him 6 weeks of relief. Lumbar MR imaging demonstrated multilevel high-grade stenosis at L2–3, L3–4, and L4–5. He also had mobile Grade 1 L4–5 anterolisthesis. He underwent an L1–5 posterior spinal fusion with Smith-Petersen osteotomies at L2–3, L3–4, and L4–5. In addition, a transformaminal lumbar interbody fusion was performed at L2–3, L3–4, and L4–5. Postoperatively, a marked bradykinesia and rigidity developed, which ultimately improved over the following several days. Furthermore, he remained confused during much of his acute inpatient course and was treated for diarrhea due to Clostridium difficile. He was ultimately discharged to rehabilitation in good condition with a thoracolumbosacral orthosis brace on postoperative Day 17. At 6-week follow-up he was in a skilled nursing facility. Imaging studies caused concern regarding the progression of postural kyphosis above his construct (Fig. 2 right). At 3 months, CT scanning of the lumbar spine demonstrated hollowing of the L-1, L-2, and L-5 screws. At 6 months, imaging studies suggested pseudarthrosis at L3–4 and L4–5, and we were concerned by the possibility of discitis at these levels. The patient underwent CT-guided biopsy of these levels and the cultures grew Enterobacter cloacae. He was started on long-term antibiotic therapy.

Case 4: Long-Segment Spinal Fusion

This patient was a 65-year-old woman with PD who complained of back pain and worsening capacity to ambulate due to her sagittal imbalance (Fig. 3). She required a wheelchair when outside of her home. She had no significant spinal stenosis. She underwent a T3–4 posterior spinal fusion. After 7 days of hospitalization, she was discharged to rehabilitation. Initially she was ambulatory with a walker and was weaned off oral narcotics. She returned to clinic 4 months postoperatively with recur-
rent back pain and was found to have pseudarthrosis due to partial screw pullout at L3–4 as well as a new lateral fixed listhesis at L4–5 (below her fusion) (Fig. 4 left). She underwent posterior spinal revision surgery in which the fusion extended to the sacrum and pelvis (Fig. 4 right). Again she was discharged to rehabilitation after 7 days of hospitalization and regained her ability to ambulate with a walker. She returned to clinic 2 years later in a wheelchair. She reported that she ambulated short distances in her home, but she was not motivated to ambulate outside her home and preferred to be in a wheelchair. She reported her back pain was improved and her capacity to sit upright was improved. A radiographic fusion was achieved.

**Discussion**

The surgical management of patients with PD and spinal deformity is difficult for a number of reasons. First, a number of conditions that can be treated nonsurgically must be considered. For example, patients with severe motor fluctuations and transient abnormal truncal postures associated with wearing-on or wearing-off dystonia could respond to changes in antiparkinsonian medications. Patients with myopathy of the paraspinal muscles should also be excluded from surgical consideration. Patients with severe anterocollis or “dropped head” may suffer from a form of atypical parkinsonism that does not respond to DBS. Second, our experience, as well as the limited published surgical experience of spinal surgery in patients with PD, suggests that there is a high postoperative complication rate. Furthermore, the symptoms of postural instability, depression, and cognitive impairment, which are common features of later-stage PD, can make postoperative rehabilitation a challenge. Table 1 summarizes the current literature regarding camptocormia and either spinal surgery or DBS.

The role of DBS in the management of severe spinal deformity associated with PD is not yet known. Given that there is limited evidence that some patients’ spinal deformities may improve after DBS, the presence of spinal deformity should not be considered a contraindication for DBS as long as other standard criteria for DBS surgery exist. The standard criterion for DBS in PD is the presence of motor fluctuations in the setting of optimal medical management by a movement disorders neurologist (in patients who do not have dementia).

Our proposed algorithm attempts to classify the range of spinal deformity in patients with PD (Fig. 5). In any patient with PD and a spinal deformity, an appropriate history and physical examination, along with imaging studies, should be obtained. Furthermore, consultation with a movement disorders neurologist is essential in ensuring an accurate diagnosis of idiopathic PD and in ensuring that the patient’s medical regimen is optimized. Imaging studies should be acquired to evaluate for the possibility of myopathy, noted in particular with high T2 signal on MR imaging. If there is doubt, then a muscle biopsy can also be considered. Myopathy does not respond well to spinal surgery or DBS.

In patients in whom symptoms meet standard criteria for DBS (debilitating motor fluctuations in the setting of optimal medical management), DBS can be considered. However, it should be noted that in the experience of one of the authors (P.A.S.), at most 50% of the patients experience persistent benefit in their postural deformity following DBS of either the STN or GPi. Thus, if all parkinsonian motor symptoms other than abnormal posture are
Spinal deformity and Parkinson disease: a treatment algorithm

TABLE 1: Summary of reports on the treatment of camptocormia with medical management, DBS, or spinal surgery*

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>No. of PD Patients w/ Deformity</th>
<th>Type of Study</th>
<th>Intervention</th>
<th>Camptocormia Outcomes</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Azher &amp; Jankovic, 2005</td>
<td>11 with PD, 1 w/ generalized dystonia</td>
<td>case series</td>
<td>12 treated w/ dopaminergic medication adjustment; 2 received intrathecal baclofen infusion; 1 patient received bilat STN DBS</td>
<td>patients treated w/ medication adjustment only had minimal to no effect on camptocormia; 4 treated w/ botox noted moderate to marked improvement; no benefit to intrathecal baclofen infusion; no improvement in single patient who received DBS</td>
<td>none reported</td>
</tr>
<tr>
<td>Ho et al., 2007</td>
<td>1</td>
<td>case report</td>
<td>medical management</td>
<td>reported improvement w/ adjustments to dopaminergic therapy</td>
<td>none reported</td>
</tr>
<tr>
<td>von Coelln et al., 2008</td>
<td>4</td>
<td>case series</td>
<td>botox injection into iliopsoas</td>
<td>no improvement</td>
<td>transient &amp; mild itching at injection site in 1 patient</td>
</tr>
<tr>
<td>Bloch et al., 2006</td>
<td>16</td>
<td>case control</td>
<td>dopaminergic treatment</td>
<td>poor response</td>
<td>no significant difference between groups</td>
</tr>
<tr>
<td>Sako et al., 2009</td>
<td>6</td>
<td>case series</td>
<td>bilat STN stimulation</td>
<td>reported improvement</td>
<td>none reported</td>
</tr>
<tr>
<td>Hellmann et al., 2006</td>
<td>1</td>
<td>case report</td>
<td>bilat STN stimulation</td>
<td>reported improvement</td>
<td>none reported</td>
</tr>
<tr>
<td>Yamada et al., 2006</td>
<td>1</td>
<td>case report</td>
<td>bilat STN stimulation</td>
<td>reported improvement</td>
<td>none reported</td>
</tr>
<tr>
<td>Micheli et al., 2005</td>
<td>1</td>
<td>case report</td>
<td>bilat GPI stimulation</td>
<td>reported improvement</td>
<td>none reported</td>
</tr>
<tr>
<td>Umemura et al., 2009</td>
<td>8 w/ camp-tocormia, 10 w/ Pisa syndrome</td>
<td>case series</td>
<td>bilat STN stimulation early: 4/8 patients reported improvement; late: 5/8 patients reported improvement</td>
<td>none reported</td>
<td>1 had severe skin erosion at site of internal pulse generator requiring op repair; 1 suffered from prolonged depression</td>
</tr>
<tr>
<td>Peek et al., 2009</td>
<td>1</td>
<td>case report</td>
<td>spinal surgery</td>
<td>ultimately improved</td>
<td>multiple revision ops, percutaneous gallbladder drainage, prolonged hospital &amp; rehabilitation course</td>
</tr>
<tr>
<td>Babat et al., 2004†</td>
<td>14</td>
<td>case series</td>
<td>spinal surgery</td>
<td>NA</td>
<td>12 (86%) had reops; 4 (29%) had hardware pullout, requiring 10 additional ops</td>
</tr>
</tbody>
</table>

*Botox = botulinum toxin Type A; NA = not applicable.
† This study did not specifically address camptocormia.

adequately treated medically, there is limited evidence to support DBS for camptocormia as the primary surgical indication.

In patients with camptocormia, one must also evaluate for the presence of myelopathy or radiculopathy due to spinal stenosis (Fig. 6). Patients with spinal stenosis and camptocormia who are candidates for DBS may first undergo DBS placement. In these patients, if the camptocormia does improve following DBS, a short-segment spinal decompression can be considered to treat spinal stenosis. If the patient does not meet standard criteria for DBS implantation (and does not undergo DBS), then spine surgeons may consider short-segment decompression and fusion alone. Long-segment spinal deformity correction and decompression should only be considered in patients who have minimal other comorbidities and are very well motivated to walk. It should be noted that long-segment spinal fixation in camptocormic patients is associated with a very high complication rate (essentially 100%).

In patients with camptocormia who do not have spinal stenosis, treatment follows a similar algorithm. If they have typical indications for DBS surgery in addition to their spinal deformity, then DBS can be offered. If they do not have indications for DBS, then long-segment deformity correction should probably not be offered because of the very high complication and revision surgery rate.

In patients who have a camptocormic posture with coexistent rigid degenerative scoliosis our recommendations are to consider short-segment decompression/fusion in patients with symptomatic stenosis, and to reserve long-segment deformity correction (with osteotomies) only for those patients who are motivated to walk, given
our experience with a high complication rate (> 50% major complications) (Fig. 7).

Conclusions

We present our algorithm for the management of patients with PD and spinal deformity. In our experience, patients with PD who undergo spinal surgery have a high rate of both acute and delayed complications. Nonsurgical management is preferred in this patient population. In addition, DBS can be considered in the correctly selected patient as an option for the treatment of some spinal deformities associated with PD. However, DBS is not universally effective in treating camptocormia.

Short-segment spinal decompression and fusion may be considered in patients with coexistent camptocormia and spinal stenosis with myelopathy or radiculopathy. Long-segment spinal fixation procedures should be performed sparingly due to the very high complication rates reported in the literature.

Disclosure

Author contributions to the study and manuscript preparation include the following. Conception and design: PV Mummaneni. Acquisition of data: PV Mummaneni, CD Upadhyaya, PA Starr. Analysis and interpretation of data: PV Mummaneni, CD Upadhyaya, PA Starr. Drafting the article: CD Upadhyaya. Critically revising the article: PV Mummaneni, PA Starr. Reviewed final version of the manuscript and approved it for submission: PV Mummaneni, PA Starr.

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