1. Introduction

Camptocormia, which is also known as bent spine syndrome, is characterized by abnormal posture of the trunk with marked forward flexion of the thoracolumbar spine, which increases during standing and walking and abates in the recumbent position (Azher & Jankovic, 2005). Camptocormia is a disabling symptom that occurs during the course of Parkinson’s disease (PD), but the optimized medical and surgical therapy for PD-associated camptocormia remains to be established (Finsterer & Strobl, 2010; Doherty et al., 2011). PD-associated camptocormia is generally thought to be unresponsive to levodopa (Azher & Jankovic, 2005). In most patients with PD, the extreme anterior bending is not or poorly improved, or even worsened, in response to levodopa administration, and the severity of the bent spine is often unchanged during the medication-on and -off phases (Melamed & Djaldetti, 2006), although an exceptional case has been reported (Ho et al., 2007). Although some reports have shown that deep brain stimulation (DBS) in the subthalamic nucleus (STN) (Hellmann et al., 2006; Yamada et al., 2006; Sako et al., 2009; Umemura et al., 2010; Capelle et al., 2011; Asahi et al., 2011) and globus pallidus internus (GPi) (Micheli et al., 2005; Capelle et al., 2011; Thani et al., 2011) is effective in treating camptocormia, the overall efficacy of DBS in relieving PD-associated camptocormia has not been determined. This review introduces the use of DBS in the treatment of medically refractory camptocormia in patients with PD.
Figure 1. A patient having PD-associated camptocormia in the standing (A) and recumbent (B) positions.

2. Definition and diagnosis

Camptocormia was first described by Earle (1815) and Brodie (1818). Camptocormia, which is also referred to as “bent spine syndrome”, was initially considered, especially in war times, to be a psychogenic disorder and a conversion reaction to war stress (Karbowski, 1999). Camptocormia is associated with various etiologies, including parkinsonian syndromes, dystonia, vascular lenticular lesions, and muscular and rheumatologic disorders. It was first described in association with PD by Djaldetti et al. (1999).

The term camptocormia is now used to describe marked forward flexion of the thoracolumbar spine that varies between 30 and 90 degrees, presents in a sitting position and typically increases during walking, and completely disappears in the recumbent position, but there are no criteria with a clear consensus for diagnosing camptocormia. Most of the diagnoses are made by subjectively assessing the patient’s posture. Diagnosing camptocormia in PD patients is based on clinical examination alone. Nevertheless, some specific findings might suggest alternative diagnoses. For example, weakness of the truncal extension suggests concomitant myopathy or anterior horn cell disease. In addition, camptocormia can occur in other parkinsonian syndromes, such as multisystem atrophy, progressive supranuclear palsy, or corticobasal degeneration. Patients with psychogenic disorders sometimes develop movement disorders due to conversion syndromes or malingering. Bent
spine that is due to simple kyphosis that is associated with degenerative vertebral spinal changes is easily ruled out, as this phenomenon remains largely unchanged when the patient is in the recumbent position.

3. Epidemiology

Among the 16 patients with camptocormia that have been described by Azher & Jankovic 2005, the most frequent etiology is PD. The reported prevalence of camptocormia in PD varies widely. Four studies have described the prevalence rates of camptocormia in patients with PD as being between 3% and 6-17% (Ashour & Jankovic, 2006; Lepoutre et al., 2006; Tiple et al., 2009; Abe et al., 2010). This wide range probably reflects the different thresholds that have been used for diagnosing camptocormia, the lack of a clear definition, and the different populations that have been studied. Most epidemiological studies show a positive association between camptocormia and disease severity (Ashour & Jankovic, 2006; Bloch et al., 2006; Tiple et al., 2009; Margraf et al., 2010), the male gender, older age, longer disease duration, prominent axial involvement, motor fluctuations, and autonomic symptoms (Lepoutre et al., 2006). In addition, camptocormia is associated with a high prevalence of lumbar or thoracolumbar scoliosis (in 61% of the patients) and mild to moderate low-back pain (in 77% of the patients). On average, camptocormia presents 7–8 years after the onset of parkinsonism in PD (Azher & Jankovic, Djaldetti et al., 1999; Bloch et al., 2006; Lepoutre et al., 2006; Margraf et al., 2010; Spuler et al., 2010).

4. Clinical features

Camptocormia occurs mostly in patients with PD in more advanced stages of disease progression, but, in a few cases, it appears even in the early stage (Melamed & Djaldetti, 2006). In some patients, the onset is subacute with the development of significant flexion over days to months (Lepoutre et al., 2006; Margraf et al., 2010; Spuler et al., 2010). In the majority of patients with camptocormia, the initial symptoms of PD are bradykinesia and rigidity and, less frequently, tremor. In almost all patients, the initial signs and symptoms are predominantly asymmetrical (Melamed & Djaldetti, 2006). The truncal forward flexion is more prominent when standing (see Fig. 1A) and walking, but complete straightening of the back in the recumbent position (see Fig. 1B). Normally, the strength in the abdominal and paravertebral muscles is normal. Some patients can, upon external command or strong self-will, straighten themselves up, but only for very short periods and at the expense of severe fatigue (Melamed & Djaldetti, 2006). In quite a number of cases, camptocormia is associated with lower back pain (Bloch et al., 2006; Lepoutre et al., 2006; Margraf et al., 2010), but, in others, it is painless (Melamed & Djaldetti, 2006). Some patients report a feeling of being pulled forward or a sensation of tightening in their abdomen (Azher & Jankovic, 2005). If the deformity is long established with secondary fixed changes, patients might complain of breathlessness due to restricted lung capacity or of difficulty lying flat in bed due to hip or knee contractures; the
latter can be accompanied by skin irritation in the flexed segment (Bloch et al., 2006). Neurological examination often reveals marked axial rigidity (Bloch et al., 2006; Lepoutre et al., 2006). The strength of the trunk and hip extension are normal unless testing is precluded by fixed posture or pain (Lepoutre et al., 2006). The paraspinal muscles can have a wooden consistency, and the rectus abdominis often feels tense (Azher & Jankovic, 2005). There might be compensatory hyperextension of the neck in order to obtain a normal visual field. There is often mixed deformity, with deviation also in the coronal plane.

5. Pathogenesis of camptocormia in PD

The pathophysiology of the axial postural abnormalities in PD is not well understood, and it seems to be heterogeneous. However, two possible causes for the camptocormia genesis in patients with PD have been proposed: dystonia and myopathy.

5.1. Dystonia

Axial or action dystonia is considered a possible etiology of camptocormia in patients with PD (Ponfick et al., 2011). The first report on camptocormia in PD patients has suggested that bent spine might represent an action dystonia resulting from dysfunction of the striatum (Djaldetti et al., 1999). Sławek et al. (2003) have described a PD patient who showed painful camptocormia that improved markedly after an unilateral pallidotomy, and the findings that DBS has a beneficial effect on camptocormia support a dystonic etiology (Micheli et al., 2005; Thani et al., 2011). Another study on camptocormia that was associated with lenticular lesions has also reported the critical role of the striatum and pallidum in the maintenance of axial posture (Nieves et al., 2001). Thus, it is possible that PD-associated camptocormia may serve as an axial dystonia, a result of dysfunction of the basal ganglia controlling the reticulospinal pathway that projects to the axial muscles (Djaldetti et al., 1999; Nieves et al., 2001; Azher & Jankovic, 2005; Bloch et al., 2006; Melamed & Djaldetti, 2006).

5.2. Myopathy

Recent studies have shown detailed evidence for camptocormia that is caused by a myopathy of the paraspinal muscles. Gdynia et al. (2009) have investigated paraspinal muscle biopsies in 19 patients with PD who presented with camptocormia or dropped head syndrome. Thirteen patients showed myopathic changes in electromyography recordings, and magnetic resonance imaging showed slight fatty degeneration of the erector spine musculature in three patients. Histopathological analyses of the patients with PD have demonstrated a wide spectrum of abnormalities in the skeletal muscles of those with camptocormia and dropped head syndrome. Spuler et al. (2010) have examined 17 patients, 13 of whom had a myopathic pattern in electromyographic recordings. The authors found apparently folded proteins in the muscle biopsies, suggesting the possibility of a myopathy that was induced by an accumulation of aggregated proteins. Margraf et al. (2010) have examined 15 patients with PD and campto-
carmia with electromyography, muscle magnetic resonance imaging, and biopsy of the paravertebral muscles. They showed increased levels of creatine kinase in 9/15 patients, myogenic electromyographic changes in 8/15 patients, and myopathic changes in the muscle biopsies in 12/15 patients. They claimed that the cause of camptocormia in idiopathic PD is a focal myopathy and that the myopathy has a progressive course, resulting in degeneration of the paravertebral muscles.

6. Treatment

6.1. Pharmacotherapy

In the majority of cases with advanced PD, camptocormia is thought to be unresponsive to levodopa (Azher & Jankovic, 2005; Margraf et al., 2010). Depending on the investigated cohort, up to 20% of the patients with PD and camptocormia profit from levodopa therapy (Bloch et al., 2006). The adjustment of dopaminergic therapy by carbidopa-levodopa and entacapone has been shown to result in improvements in camptocormia. Fujimoto (2006) has described the deterioration of camptocormia in patients who were treated with dopamine agonists, thus suggesting that the withdrawal of these agents might lead to an improvement of camptocormia in some cases. The poor responses to these medical agents can be explained by the fact that, when postural abnormalities and postural instabilities appear, patients are already at an advanced stage of the disease with severe axial symptoms, and all of their symptoms are known to respond poorly to levodopa, suggesting the involvement of non-dopaminergic pathways (Campbell et al., 2003). Furthermore, postural reactions that support surface perturbations are resistant to dopaminergic therapy (Carpenter et al., 2004).

In patients who seem to have a predominantly dystonic element, another treatment option might be botulinum toxin injections in the rectus abdominis muscles (Azher & Jankovic, 2005; Jankovic, 2009; Bonanni et al., 2007; Lenoir et al., 2010). Azher and Jankovic (2005) have found this to be successful in selected patients, but few have reproduced their positive results.

6.2. Spinal surgery

Spinal surgery has been used to attenuate postural abnormalities in patients with camptocormia associated with PD, whereas it has significant complications, and often requires revision surgery (Babat et al., 2004; Peek et al., 2009; Koller et al., 2010; Wadia et al., 2011).

6.3. DBS

Stereotactic neurosurgery of the basal ganglia is a therapeutic alternative for patients with advanced PD. As DBS of the STN or GPi produces a significant improvement in the motor symptoms in patients with severe PD, it also can be effective in treating camptocormia associated with PD (see Table 1.).
### Table 1. DBS for camptocormia: literature review

<table>
<thead>
<tr>
<th>DBS target</th>
<th>author/year (year)</th>
<th>No of patients</th>
<th>age</th>
<th>duration of PD (years)</th>
<th>Improvement</th>
<th>follow up (months)</th>
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<tr>
<td>STN</td>
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<td>1</td>
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<td>n.a.</td>
<td>0/1</td>
<td>n.a.</td>
</tr>
<tr>
<td></td>
<td>Hellman et al. (2006)</td>
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<td>53</td>
<td>25</td>
<td>1/1</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>Yamada et al. (2006)</td>
<td>1</td>
<td>71</td>
<td>11</td>
<td>1/1</td>
<td>20</td>
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<tr>
<td></td>
<td>Sako et al. (2009)</td>
<td>6</td>
<td>53-71</td>
<td>5-11</td>
<td>6/6</td>
<td>5-46</td>
</tr>
<tr>
<td></td>
<td>Capelle et al (2010)</td>
<td>2</td>
<td>73/65</td>
<td>12/15</td>
<td>1/2</td>
<td>12-16</td>
</tr>
<tr>
<td></td>
<td>Umemura et al. (2010)</td>
<td>8</td>
<td>59-79</td>
<td>8-20</td>
<td>6/8</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td>Upadhyaya et al (2010)</td>
<td>1</td>
<td>59</td>
<td>n.a.</td>
<td>0/1</td>
<td>2</td>
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<tr>
<td></td>
<td>Asahi et al. (2011)</td>
<td>4</td>
<td>60-69</td>
<td>7-13</td>
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<td></td>
<td>Lyons et al. (2012)</td>
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<td>63</td>
<td>19</td>
<td>1/1</td>
<td>3</td>
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<tr>
<td>GPi</td>
<td>Micheli et al. (2005)</td>
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<td>62</td>
<td>10</td>
<td>1/1</td>
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<tr>
<td></td>
<td>Upadhyaya et al. (2010)</td>
<td>1</td>
<td>59</td>
<td>n.a.</td>
<td>0/1</td>
<td>15</td>
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<td>13</td>
<td>1/1</td>
<td>14</td>
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</tbody>
</table>

n.a. = not applicable

6.3.1. **STN-DBS**

STN-DBS is thought to be superior to GPi-DBS in improving the cardinal motor symptoms of PD and in reducing the dosage of dopaminergic medications in PD patients (Moro et al., 1999; Deep-Brain Stimulation for Parkinson’s Disease Study Group, 2001; Krack et al., 2003). As far as we are aware, 25 patients in 9 reports have been reported as having undergone STN-DBS for PD with camptocormia, and 19 of the patients (76%) showed improvements of the camptocormia. Sako et al. (2009) have described 6 PD patients with severe camptocormia who underwent bilateral STN-DBS. In this series, improvements of the camptocormia were reported in all of the patients after STN-DBS (see Fig. 2). Improvement was defined as a change in the thoracolumbar angle that was sustained for a mean follow-up period of 17 months. Umemura et al. (2010) and Capelle et al. (2011) have shown a positive effect of STN-DBS on PD-associated camptocormia, but not in all of the patients. In their studies, only the severity of the postural abnormality was shown to be related to improvements in the postural abnormalities after STN-DBS; moderate, rather than severe, postural abnormalities tended to improve after surgery (Umemura et al., 2010; Capelle et al., 2011). Asahi et al. (2011) have described 4 cases with advanced PD with camptocormia that were treated with STN-DBS, with improvements of the camptocormia in 3 patients but no improvement in one patient. In their computed tomography study, the single patient who did not show improvement of the camptocormia exhibited marked muscle atrophy and degeneration of the fatty tissue in her
paraspinal muscle (Asahi et al., 2011). Thus, STN-DBS is a potential treatment for camptocormia in patients with PD, but the outcomes have varied from excellent improvement to only mild improvement or no benefit. Camptocormia usually does not respond to levodopa treatment and STN-DBS is thought to be effective mainly for the dopa-responsive PD symptoms. The mechanisms by which STN-DBS improves camptocormia in some patients and not in others remain unclear. As the recent reports that have shown the beneficial effects of STN-DBS in the treatment of primary dystonias (Kleiner-Fisman et al., 2007; Novak et al., 2008; Ostrem et al., 2011; Fonoff et al., 2012), axial posturing could be controlled by the STN as a function of the basal ganglia.

6.3.2. GPi-DBS

The GPi is an alternative target that can be considered in the treatment of camptocormia. Previous research on patients with PD and camptocormia that was treated with GPi-DBS is limited to only four cases, with reports of no improvement in posture in one and improvement in posture in three (Micheli et al., 2005; Upadhyaya et al., 2010; Capelle et al., 2011; Thani et al., 2011). If camptocormia is considered a form of dystonia, then it is not surprising that high-frequency DBS of the GPi results in improvements because there is a large body of evidence of a dramatic effect of pallidal surgery on various forms of dystonia (for a review see Vidailhet et al., 2013). Micheli et al. (2005) have described a PD patient in whom bilateral GPi-DBS showed beneficial and sustained improvement of the PD symptoms and improvement of the camptocormia six months after surgery. In addition, Thani et al. (2011) have reported slow but steady improvement of camptocormia by six months, which was sustained at 14 months, after GPi-DBS in patients with PD. This progressive resolution of symptoms is reminiscent of the response that is expected in other forms of dystonia, with optimal improvement often requiring months.
7. Conclusion

Reports of success in controlling axial posturing in patients with camptocormia with both STN and GPi stimulation support the notion that the basal ganglia plays an important role in the maintenance of posture. Although both STN-DBS and GPi-DBS can be potential surgical means for treating camptocormia in patients with PD, further studies need to be performed in order to confirm this conclusion and to select PD patients with medically refractory camptocormia who are optimal candidates for STN or GPi DBS.

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