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Motor outcome of dystonic camptocormia treated with pallidal neurostimulation

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ABSTRACT

Background: Deep brain stimulation of the internal pallidum (GPi-DBS) is effective for various types of drug-refractory primary dystonias. Rare clinical forms as dystonic camptocormia may profit but available data are scarce.
Methods: We here report on a retrospective clinical assessment of three patients with primary dystonic camptocormia treated with GPi-DBS.
Results: All three patients showed marked response to bilateral GPi-DBS within days to weeks after surgery which was preserved in the long-term (38–45 months after implantation: mean improvement 82% as rated on the Burke Fahn Marsden Dystonia Rating Scale, 89% in the subitem "trunk"). Two patients developed mild stimulation induced speech problems (stuttering or dysarthria) which resolved with reprogramming or were acceptable in return for the control of dystonic symptoms.
Conclusions: The diagnosis and treatment of camptocormia will continue to require expert knowledge in movement and neuromuscular disorders, but DBS may expand treatment options in this difficult patient population.

1. Introduction

Deep brain stimulation of the internal pallidum (GPi-DBS) has become first choice treatment in drug-refractory primary segmental and generalized dystonia [1,2]. There is also good evidence for primary focal [3,4], cranio-cervical [5,6], and tardive dystonia [7–9]. Overall, GPi-DBS is a comparatively safe surgical procedure and its clinical benefit is documented for more than 10 years [10].

We here present clinical data to the efficacy of GPi-DBS in a rare form of primary dystonia: primary dystonic camptocormia. The syndrome is clinically defined by phasic and tonic ventral flexion of the thoraco-lumbal spine due to dystonic activity in abdominal muscles [11]. It is typically aggravated by standing and walking, alleviated in reclined position and disappears during sleep. Severe malposition of the spine causes pain and orthopedic problems and relevantly affects mobility and other daily life activities. This is an entity which is distinct from camptocormia in Parkinson's disease or muscle disease [12,13]. Conservative treatment options include, similar to other forms of (primary) dystonia, anticholinergics, benzodiazepines and selective muscle denervation by botulinum toxin [11,14]. The success, however, is often limited. So far, a total of 13 cases of GPi-DBS with dystonic camptocormia have been reported in the literature, all showing marked symptomatic improvement (Table 1) [15–20].

We here add to the current literature clinical long-term follow-up (>3 years) in three patients with primary dystonic camptocormia treated with bilateral GPi-DBS.

2. Patients and methods

From our large group of >100 dystonic patients treated with GPi-DBS at the Departments of Neurology and Neurosurgery, Kiel University, we retrospectively selected three patients (1 male/2 female) with primary dystonic camptocormia. Two patients had only truncal dystonia, one patient suffered from generalized dystonia with prominent disability resulting from severe lumbar flexion of the spine. Ethical approval was waived and all patients

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Table 1
Summary of clinical data and outcome parameters of the Kiel patients and the other published cases of dystonic camptocormia treated with pallidal neurostimulation.

Study	Pat. No. (gender)	Clinical presentation/etiology	Age at onset/age at surgery	Last follow up (FU) [months]	Clinical outcome	
					BFMDRS pre/FU	Subitem trunk pre/FU
Nandi, D et al., 2002	1 (M)	Generalized dystonia/tardive	35/39	6	Severe truncal bending impairing stance and gait/unaided upright walk	
Fukaya, C et al., 2006	1 (M)	Inexplicitly described/primary	13/17	>6	50/0 (100%)	Not stated
	2 (M)	Inexplicitly described/primary	45/46	>48	32/4 (88%)	Not stated
	3 (M)	Inexplicitly described/primary	44/49	>6	48/32 (33%)	Not stated
O'Riordan, S et al., 2009	1 (M)	Pure camptocormia/primary	57/67	60	Severe truncal bending impairing stance and gait/unaided upright walk	
Sakas, DE et al., 2010	1 (F)	Pure camptocormia?/primary	23/26	44	Severe truncal bending impairing stance and gait/unaided upright walk	
	2 (M)	Pure camptocormia?/primary	15/21	42	Severe truncal bending impairing stance and gait/unaided upright walk	
Capelle, HH et al., 2011	1 (F)	Segmental dystonia/primary	47/75	18	21/12 (43%)	12/4 (67%)
	2 (M)	Segmental dystonia/primary	26/39	12	34/20 (41%)	12/4 (67%)
	3(M)	Generalized dystonia/primary	25/40	9	58/12 (79%)	12/4 (67%)
	4 (F)	Pure camptocormia (myopathy)/secondary?	64/69	18	12/6 (50%)	12/6 (50%)
Hagenacker, C et al., 2013	1 (M)	Pure camptocormia/primary	53/60	24	20/4 (80%)	16/4 (75%)
	2 (F)	Generalized dystonia/primary	?/?	12	81/26 (68%)	16/2 (88%)
Reese, R et al., 2013	1 (F)	Generalized dystonia/primary	44/62	39	19/10 (47%)	6/2 (67%)
	2 (M)	Pure camptocormia/primary	48/49	38	4/0 (100%)	4/0 (100%)
	3 (F)	Pure camptocormia/primary	52/54	45	6/0 (100%)	6/0 (100%)

signed informed consent to be filmed for publication. Dystonic camptocormia was diagnosed according to the clinical presentation by GD and JV. Secondary causes were excluded by standard cranial MRI scans. Patients suggested for surgery had no biography of neuroleptic treatment and were refractory to various drugs as well as local botulinum toxin muscle injections. The patients suffered from typically patterned dystonic movements and had no inconsistent neurological findings or unusual disease course suggestive of a psychogenic movement disorder.

Details of the surgical procedures have already been extensively described [21]. Initial stereotactic coordinates (22 mm lateral to, 3 mm anterior, 4 mm below the midcommissural point) were adjusted to the individual stereotactic MRI after placement of a stereotactic frame. All surgical procedures were done under general anesthesia with propofol and remifentanil.

Microelectrode multichannel recordings (FHC, Bowdoinham, USA) combined with macrostimulation to check for capsular motor responses guided the bilateral implantation of quadripolar electrodes (model 3389, Medtronic) into the GPi. Pulse generators (Kinetra, Medtronic) were placed at the same surgical session. Postoperative MRI-scans verified electrode position and excluded asymptomatic cerebral hemorrhage.

Postoperative programming of the neurostimulator was done 4–5 days after surgery. Each of the four contacts was reviewed for both hemispheres (case served as anode; constant voltage, 60–90 μs pulse width, 130 Hz frequency). Acute clinical improvement and voltage-limiting side effects were assessed. For chronic stimulation, the lowest contact within the GPi with acceptable thresholds for side effects (typically ~3–4 V) was chosen for monopolar stimulation and programmed with a voltage at 10–20% below the threshold for side effects. Follow-up visits were routinely done around 4–6 weeks, 3 and 6 months after surgery, and whenever requested by the patient.

Severity of dystonia was retrospectively scored by RR on the global Burke Fahn Marsden Dystonia Rating Scale (BFMDRS) [22] as based on the blinded video documentation of the pre- and postoperative neurological assessments. Preoperative scores and scores of follow-up visits were not statistically compared owing to the small sample size. Data are shown as median (range) (Prism5 software, Version 5.04, GraphPad Software, Inc., La Jolla, CA, USA).

3. Results

Mean improvement in BFMDRS was 82% (preoperative median 6 with range 4–19 compared to 0 with range 0–10) and 89% in the subitem "trunk" (preoperative median 6 with range 4–6 compared to 0 with range 0–2) at the last follow up visit (38–45 months after implantation) (video; Fig. 1; Table 1). Symptomatic improvement was gradual over the first few weeks after surgery and nearly complete at 3 months follow-up. No surgery or device related

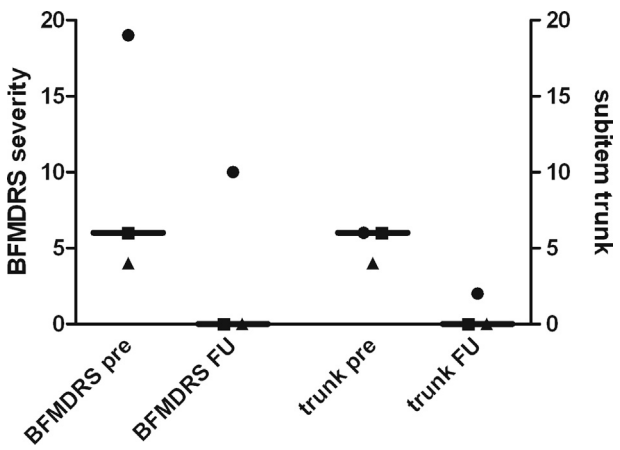

Fig. 1. Dystonia severity rated on the Burke Fahn Marsden Dystonia Rating Scale (BFMDRS) and its subitem "trunk" of our three cases with dystonic camptocormia. Horizontal bars represent the median. Individual scores: ●patient 1, ▲patient 2, ■patient 3 (see also Tables 1 and 2). Pre = prior to surgery, FU = last follow-up.

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