Clinical Neurology and Neurosurgery 135 (2015) 62–68

Contents lists available at ScienceDirect

Clinical Neurology and Neurosurgery

journal homepage: www.elsevier.com/locate/clineuro

# Deep anterior cerebellar stimulation reduces symptoms of secondary dystonia in patients with cerebral palsy treated due to spasticity

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ARTICLE INFO

*Article history:*
Received 23 November 2014
Received in revised form 2 May 2015
Accepted 16 May 2015
Available online 23 May 2015

*Keywords:*
Cerebellar neurostimulation
Cerebral palsy
Spasticity
Dystonia

ABSTRACT

*Introduction:* Deep anterior cerebellar stimulation (DACS) is a neuromodulation therapy of spasticity. Bilateral DACS is applied in young patients with cerebral palsy (CP). In these patients symptoms of spasticity coexist with symptoms of focal or segmental dystonia, which can cause chronic pain. We performed the study to investigate the therapeutic effects of DACS in spasticity, secondary dystonia and pain.
*Methods:* We examined 10 from 13 patients with CP treated with DACS due to spasticity in years 2006–2012. We compared Ashworth scores of spasticity, VAS scale of pain and UDRS (Unified Dystonia Rating Scale) score before DACS and after it in follow-up lasting from 2 to 11 years it in these patients basing on clinical examination and evaluating forms given by the patients or parents.
*Results:* We received statistically significant reduction of spasticity in upper extremities (median: from 3 to 1,5 in Ashworth scale) in 8 patients ($p = 0{,}01$), in lower extremities in 7 patients (median: from 3 to 1,75) ($p = 0{,}02$). Symptoms of focal dystonia were reduced. Total score for the UDRS (median = 18,0 before surgery) after DACS decreased significantly (median = 10,3) ($p = 0{,}043$). Change in consecutive parts of UDRS before (median = 1,6) and after (median = 1,0) surgery in 7 patients had statistical significance ($p = 0{,}0179$). There were not significant changes in intensity of pain before and after surgery ($p = 0{,}108$).
*Discussion:* Chronic bilateral DACS aimed for spasticity treatment not only decreases muscular tone in quadriplegic or paraplegic patients with CP but also is associated with reduction of symptoms of focal or segmental, secondary dystonia.



## 1. Introduction

Cerebral palsy (CP) is a relatively rare condition (affecting approximately 2–3 in every 1,000 live births) [1,2]. Dystonic CP is caused by damage to the basal ganglia or cerebellum and causes symptoms such as involuntary movements and muscle spasms. It is estimated that dyskinetic CP has a prevalence of 0.27 per 1000 live births [3]. Dystonic forms with choreoathosis are met in up to 15% of patients with CP [1,4]. Patients with this type of CP have symptoms of spasticity which coexist with symptoms of focal or segmental dystonia. Pharmacological treatment of dystonic movements is unsatisfactory and is burden with side effects [5].

Surgical therapy for the secondary dystonias is generally perceived to be less effective than for primary disease [5–7]. In secondary dystonia effects of neurostimulation can be low [8–10]. Deep brain stimulation (DBS) of the globus pallidus internus (GPi) has been described to be an effective and safe treatment in patients with primary generalized dystonia [11]. GPi stimulation is currently the mainstay surgical treatment for patients with dystonia, particularly for generalized dystonia [12,13]. In general in patients with primary dystonia, the improvement in Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) has been reported to be of 40–70% after GPi DBS, but in patients with secondary dystonia, the improvement in the BFMDRS has been reported to be of 10.6–41% [8,11,12]. Patients with secondary dystonia due to previous perinatal insults showed a mean improvement in BFMDRS of 41.9%, with better results in purely dyskinetic patients (mean improvement of 61.2%) [14].

Bilateral pallidal stimulation can also be an effective treatment in adult patients with dystonia-choreathosis CP [15]. DBS of GPi can produce 23.6% mean improvement in movement component of BFMDRS and 9,2% in disability component of BFMDRS in patients with dyskinetic CP according to results of meta-analysis recently published [16].

In the 1972 the electrical stimulation of the cerebellar cortex was used for the first time to treat abnormal motor disorders,

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http://dx.doi.org/10.1016/j.clineuro.2015.05.017
0303-8467/

**Table 1**
Characteristics of patients and results of Ashworth and VAS scores. Follow-up in years, age in years.

| No. | Initials | Sex | Age | Follow-up | Ashworth before | | Ashworth after | | VAS | | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | | | | Upper | Lower | Upper | Lower | Before | After | Pain area |
| 1 | E.O | F | 18 | 10 | 3 | 2 | 1,5 | 2 | 5 | 4 | Knees |
| 2 | E.K | F | 22 | 10 | 2 | 2 | 1,5 | 1,5 | 2 | 2 | Knee hip |
| 3 | D.M | M | 30 | 2 | 3 | 3 | 1,5 | 1,5 | 3 | 1 | Knee |
| 4 | E.Ł | F | 27 | 6 | 3 | 3 | 2 | 2 | 0 | 0 | n/d |
| 5 | B.W | M | 26 | 4 | 1 | 2 | 1 | 1,5 | 0 | 0 | n/d |
| 6 | E.Z | F | 26 | 5 | 3 | 3 | 1 | 1 | 0 | 0 | n/d |
| 7 | J.M | F | 28 | 6 | 3 | 3 | 0 | 0 | 10 | 0 | Hands feet |
| 8 | S.K | M | 27 | 11 | 4 | 3 | 3 | 3 | n/d | n/d | n/d |
| 9 | A.O | F | 17 | 3 | 3 | 4 | 2 | 3 | n/d | 1 | Thighs |
| 10 | K.M | F | 26 | 3 | 2 | 2 | 2 | 2 | 8 | 8 | Back |

but with varying degrees of success. Cooper treated patients with muscular hypertonia in CP with the anterior lobe of cerebellum stimulation [17]. Several hundred patients with spasticity (mainly from CP) were operated in various centres by cortical cerebellar stimulation in the seventies [18–21]. Rose Davis, who has performed more than 600 operations, presented results of the cerebellar cortex stimulation and concluded that cerebellar stimulation reduces spasticity diminishing of hypertonus in muscles, co-contractions, spasms and athetoid movements in 85% of CP patients with various degree (marked 25%, moderate 34%) [20].

Schvarcz et al. reported positive effects of cerebellar dentate nucleus stimulation on spasticity, improvement in balance and posture of the patient [22].

High-frequency stimulation of the anterior lobe of the cerebellum was introduced by Galanda and has been designated as a neuromodulative method of treatment of spasticity. Instead of paddle type cortical electrodes, cylindrical ones for deep brain stimulation were placed in the region of the superior cerebellar peduncle. Intermittent stimulation was performed to decrease muscle tonus and general relaxation. As a result improvement in choreoathosis, in speech and mood was also observed [23,24]. Experience of Galanda et al. in treatment of CP with stereotactic stimulation of the anterior lobe of the cerebellum prompted us to conduct the study on the clinical effects of this method in diminishing symptoms of spasticity, secondary dystonia and pain. We introduced this method in 2003 in patients with CP. The study was performed to investigate the effects of DACS, in particular therapeutic effects in dystonic symptoms in these patients [25].

## 2. Material and methods

We examined 10 from 13 patients with CP treated with DACS in whom symptoms of focal or segmental dystonia occured. Three patients were excluded due to early postoperative infection in site of implantation. Retrospectively we scored them basing on the examination and interview of patients or their parents. A total 10 patients (3 men) were enrolled into the study with median age 26 (range 18–30) at the time of inclusion. Observational period lasted from 1 to 6 years (median 5,5) (Table 1). After ethics committee approval procedures of implantation of two electrodes into the anterior lobe of the cerebellum were conducted under general anaesthesia. Stereotactic frame was fixed to the head of patient and two burr holes were made in occipital area. Electrodes for DBS were implanted according to plan executed on MRI scans. Intraoperative CT was performed in an operating room before the surgery. CT scans were fused with MRI plan, then coordinates for stereotactic procedure were calculated. The planning of the operation was performed using a fusion CT/MRI (BrainLAB stereotactic frame and software) (Fig. 1). The quadripolar electrodes were implanted from the both sides. The enter points were placed in the suboccipital area below the transversus sinuses. Pulse generator (Activa Kinetra, Medtronic, USA) or (Libra XP, St.Jude Medical, USA) was usually localized in subclavian area where extentions connecting to electrodes were inserted. Evaluation of the localization of the electrodes was performed intraoperatively with CT imaging in an operating room during the same procedure or after implantation the electrodes in the radiological department. We used two electrodes and bipolar intermittent (on 30 min, off 3,5 h) stimulation: the initial parameters of the stimulation were usually as follows: amplitude: 1,4–2,4 V; pulse width: 150–180 μs; rate: 130 Hz; polarisation: 0 and 4 minus, 3 and 7 plus.

We measured the results of muscle tone in Ashworth scale, involuntary movements in Unified Dystonia Rating Scale UDRS, and pain in VAS score before surgery and at the end of follow-up. UDRS consists of 11 components: subscales with scores from 0 (lack of involuntary movements) to 4 (the highest intensity of movements) on each. These components are duration of symptoms, intensity of symptoms in face, eyes, jaw, larynx, neck, shoulder, hand, trunk, pelvis and foot. We asked parents or caregivers to assess the effects of this treatment and fill out the forms with these scales.

### 2.1. Statistical analysis

All statistical analyses were performed using Statistica version 10 (StatSoft) and Microsoft Excel 2007. The distribution of results differed from the normal distribution, the correlation analysis of the data was performed using the nonparametric *Spearman's* test, and the comparative studies were analysed statistically using the nonparametric *Mann–Whitney U* test. To confirm the significance of differences between related variables the nonparametric *Wilcoxon* test was used. The $p$ value <0,05 was considered statistically significant.

## 3. Results

Spasticity in upper extremities measured in Ashworth scale decreased significantly ($p = 0{,}01$) in 8 patients (from median 3,0 to 1,5) (Fig. 2), in lower extremities spasticity was reduced in 7 patients ($p = 0{,}01$) (from median 3,0 to 1,75) (Fig. 3). We also noticed that reduction of spasticity was more expressed in elder patients over 26 years: in upper and lower extremities was 30% in comparison to patients younger than 26 years where the percentage of reduction was respectively 20 and 10%. There were not significant changes in intensity of pain before and after surgery ($p = 0{,}108$).

Total score for the UDRS (median = 18,0 before surgery) after DACS decreased significantly (median = 10,3) ($p = 0{,}043$) (Fig. 4a). The most dominant and significant change in UDRS was observed in reduction of involuntary movements in neck, in hand, in pelvis and in duration of these symptoms (Table 2). Change in consecutive parts of UDRS before (median = 1,6) and after (median = 1,0) surgery in 7 patients had statistical significance in Wilcoxon test ($p = 0{,}018$) (Fig. 4b). In 3 patients we did not observed reduction of