Pallidotomy for medically refractory status dystonicus in childhood

CARLO EFISIO MARRAS | MICHELE RIZZI | LAURA CANTONETTI | ERIKA REBESSI | ALESSANDRO DE BENEDICTIS | FRANCESCO PORTALURI | FRANCO RANDI | ALESSANDRA SAVIOLI | ENRICO CASTELLI | FEDERICO VIGEVANO

1 Neurosurgery Unit, Department of Neuroscience and Neurorehabilitation, IRCCS Bambino Gesù Children's Hospital (BGCH), Rome; 2 Department of Neurosurgery, IRCCS Fondazione Istituto Neurologico 'Carlo Besta', Milan; 3 Neurorehabilitation Unit, Department of Neuroscience and Neurorehabilitation, IRCCS Bambino Gesù Children’s Hospital (BGCH), Rome; 4 Neurology Unit, Department of Neuroscience and Neurorehabilitation, IRCCS Bambino Gesù Children’s Hospital (BGCH), Rome; 5 Intensive Care Unit, Department of Emergency, IRCCS Bambino Gesù Children’s Hospital (BGCH), Rome, Italy.

Correspondence to Erika Rebessi, Department of Neuroscience and Neurorehabilitation, Neurosurgery Unit, Bambino Gesù Children’s Hospital, IRCCS 4, Piazza Sant’Onofrio, 00165 Rome, Italy. E-mail: erika.rebessi@opbg.net

This article is commented on by Lumsden on pages 607–608 of this issue.

AIM Status dystonicus is a rare and potentially fatal condition of continuous and generalized muscle contraction that can complicate dystonia. As status dystonicus is usually refractory to traditional pharmacological therapy, alternative and invasive strategies have been developed, but so far there are no guidelines on status dystonicus management. Pallidotomy has shown good results in status dystonicus treatment.

METHOD We report indications, surgical strategy, and outcome of bilateral pallidotomy in four pediatric patients (four males; mean age at surgery 11y 5mo) with secondary dystonia, who developed refractory status dystonicus. Pallidotomy was performed in the area corresponding to the mid portion of the globus pallidus internus.

RESULTS This procedure allowed patients to recover the pre-status dystonicus condition, controlling dystonic postures and movements of trunk and limbs. Moreover oromandibular dystonia, which is resistant to conservative approaches and deep brain stimulation, was significantly reduced. No postoperative complications were registered.

INTERPRETATION Our study suggests pallidotomy as a feasible treatment in patients with secondary dystonia complicated by status dystonicus.

Dystonia is a clinical syndrome characterized by sustained muscle contraction, frequently causing torsional and repetitive movements, or abnormal postures, which can be focal or generalized. Dystonia may sporadically evolve toward a life-threatening condition of severe generalized dystonia, called status dystonicus or dystonic storm. Status dystonicus is often triggered by events such as fever, infections, exposure to medications, or their abrupt cessation. This condition could rapidly lead to rhabdomyolysis, metabolic failure, and bulbar complications, requiring admission to the intensive care unit (ICU) for sedation and ventilation. Status dystonicus can be refractory to traditional pharmacological therapy. Alternative and invasive strategies have been developed but no guidelines for status dystonicus management have been unequivocally defined. Literature reports reveal that a gradual multistage approach, from enteral pharmacological therapy to ICU procedures and surgery (continuous infusion of intrathecal baclofen, thalamotomy, pallidotomy, or deep brain stimulation) may be required. A recent exhaustive literature report on status dystonicus showed that surgery appears to be an effective treatment. The aim of this study was to report indications, surgical strategy, and outcome of bilateral pallidotomy in a series of four children affected by secondary dystonia who developed status dystonicus.

METHOD From March 2011 to January 2012, four pediatric patients (four males), presenting with severe forms of secondary dystonia, were referred to the Department of Neuroscience and Neurorehabilitation of the Bambino Gesù Children’s Hospital in Rome, which receives tertiary patients for specialist consultations. All patients had an exacerbation of the disease with status dystonicus, refractory to medical treatment (Table I), and were prospectively studied. They fulfilled the diagnostic criteria for status dystonicus according to the definition by Manji. This includes the development of increasingly frequent and severe episodes of generalized dystonia necessitating urgent hospital
admission and accompanied by one or more of the following life-threatening complications: bulbar weakness compromising upper airway patency with the risk of progressive impairment of respiratory function leading to the development of respiratory failure; exhaustion and pain; and metabolic imbalances.

Selection criteria for bilateral pallidotomy included the presence of status dystonicus, which was not responsive to pharmacological treatment, and associated with a rapid and severe decline of clinical conditions (Table I).

The method used to target the area for pallidotomy was based on the 3D built volume routinely used in neuronavigation. The technique needs the AC-PC definition to build the stereotactic volume with the X, Y, and Z coordinates; the definition of the globus pallidus internus (GPI) is not based on the use of the stereotactic coordinates [18–21mm laterally, 6mm vertically, and 2mm anteriorly to the intercommisural point to target the posteroverentral (VPL) GPI], but on visual direct targeting on magnetic resonance imaging (MRI) inversion recovery sequences. Afterwards, the relationship to the midcommisural point was obtained by the direct targeting that was usually lateral (X) 14 to 18mm, anteroposterior (Y) 3 to 5mm, and vertical (Z) 2mm. This target fits with the coordinates used for pallidotomy and with those proposed by other authors for DBS.4

Before the onset of status dystonicus, and before surgery, all patients underwent a dystonia assessment including neurological evaluation, videotape recording, and the administration of the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS), brain MRI and cranial-computed tomography (CT). These clinical and imaging evaluations were repeated 1, 3, and 6 months after surgery, and every 6 months thereafter.

Table I: Aetiology and clinical features of the status dystonicus series

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex, age (y)</th>
<th>Diagnosis</th>
<th>Age at dystonia onset</th>
<th>Age at status dystonicus onset (y)</th>
<th>Precipitating factors</th>
<th>Status dystonicus symptoms</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M, 15</td>
<td>Chromosomopathy</td>
<td>3y</td>
<td>1st status dystonicus: 12</td>
<td>Continuous oromandibular, trunk, limbs movements Pain</td>
<td>Fever</td>
<td>Partial amputation of the tongue Pneumonia</td>
</tr>
<tr>
<td>2</td>
<td>M, 19</td>
<td>Epileptic encephalopathy</td>
<td>1mo</td>
<td>17</td>
<td>Generalized dystonic movements, opisthotonus, psychomotor agitation Pain</td>
<td>Eyelids clonus, opisthotonus, upper limbs movements, fixed dystonic posture of the left upper limb</td>
<td>Respiratory distress</td>
</tr>
<tr>
<td>3</td>
<td>M, 6.5</td>
<td>Idiopathic bilateral striatal necrosis</td>
<td>4mo</td>
<td>5</td>
<td>Enteritis/ileo-ileal Invagination</td>
<td>1st status dystonicus: H1N1 infection</td>
<td>Respiratory distress</td>
</tr>
<tr>
<td>4</td>
<td>M, 12</td>
<td>Severe neonatal cerebral hypoxia</td>
<td>8mo</td>
<td>1st status dystonicus: 8</td>
<td>1st status dystonicus: unknown</td>
<td>Generalized dystonic movement of eyes, mouth, head and limbs, opisthotonus</td>
<td>Severe dysphagia, sepsis, deep vein thrombosis</td>
</tr>
</tbody>
</table>

Informed consent was given by parents of all four patients. Data collection, recording and analysis was performed according to our institutional review board and waiver of consent was obtained for data collection, deidentification and analysis.

**Surgical treatment**

Pallidotomy was performed, under intravenous general anesthesia, by a frameless stereotactic neuronavigation robotic system (ROSA: RObotized Stereotactic Assistant; Medtech, Newark, NY, USA). The targeted area corresponding to the mid portion of the GPi, was preoperatively defined by a direct targeting on brain MRI (fast spin echo inversion recovery sequence), and merged with brain CT. Microrerecording and macrostimulation were performed under electromyography (EMG) monitoring to detect the nucleus and to exclude the involvement of the pyramidal fibers of the internal capsule by the lesioning. The radiofrequency lesion (Cosman RFG-1; Cosman Medical Inc., Burlington, MA USA) at 40V for 60 second was performed at the target, 3mm above, and 2mm below it. A second trajectory (2mm posterior, 2mm laterally to the first target) was used to perform lesioning.

**Case 1**

The first patient was 15 years old and had started to experience from hypertonia and severe psychomotor delay.
in the first year of life. Genetic testing displayed a chromosomalopathy (deletion of the long arm of chromosome 22).

At the age of 3 years, dystonic movements of trunk and limbs, not responsive to any drugs, associated with spastic tetraplegia appeared. Subsequently, the patient had frequent respiratory tract infections, and a percutaneous enteral gastrostomy (PEG) became necessary because of malnutrition. At the age of 12 years, after severe pneumonia, a marked worsening of dystonia occurred, evolving within a week to status dystonicus. The patient was then admitted to ICU for sedation and intubation followed by a tracheostomy.

Brain MRI showed a diffuse atrophy, bilaterally involving the cortex and subcortical structures (caudate, putamen, and thalamus). Administration of benzodiazepine, thiopentone, and phenobarbital, resulted in control of the status dystonicus after a period of 7 months. The patient was then discharged requiring assisted ventilation during sleep, and daily intake of several drugs (clonazepam, tetrabenazine, baclofen, tizanidine, and phenobarbital). His neurological condition became stable for 1 year but, after mononucleosis, a second episode of status dystonicus occurred with severe continuous oromandibular movements causing amputation of the tongue. The patient was readmitted to the ICU with the same management performed during the first status dystonicus. The medical management became ineffective and, 2 months after the onset of status dystonicus, bilateral pallidotomy was performed.

Case 2

The patient, aged 19 years at the time of writing (May 2013), had a diagnosis of epileptic encephalopathy of unknown etiology in the first year of life. At the age of 18 months, rapid dyskinetic eye movements, frequent startles, episodes of restlessness, and upper limbs dystonic movements were observed. Rectal diazepam administration allowed control of dystonia. Because of malnutrition, at the age of 14 years, he underwent PEG. Two years later, the patient had a severe worsening of the dystonic condition; after an episode of hyperpyrexia, he developed a status dystonicus, refractory to enteral therapy with phenobarbital, baclofen, tetrabenazine, and diazepam. The clinical condition was complicated by renal failure with hypermyoglobinemia, rhabdomyolysis, and respiratory distress. He was admitted to the ICU of our hospital, but deep sedation with midazolam and propofol proved ineffective and the intubation was followed by a tracheostomy. At that stage, a bilateral pallidotomy was performed.

Case 3

The patient, aged 6 years had a diagnosis of bilateral striatal necrosis of unknown aetiology. In the first months of life, generalized hypertonia was observed, followed by cognitive delay. Cerebrospinal fluid examination revealed a low level of homovanillic acid and he started oral therapy with levodopa, which was partially successful. At the age of 4 years, after a febrile episode, the patient presented with upper limb tremor, eyelid clonus, opisthotonus, and a fixed dystonic posture of the left upper limb. Two months later, during severe enteritis complicated by ileo-ileal invagination, a rapid worsening of movement disorder associated with loss of head control and environmental contact was noticed. He was admitted to the ICU and sedated; a PEG was also performed. Fifty days from status dystonicus onset, a bilateral pallidotomy was carried out.

Case 4

This 12-year-old patient had a severe hypoxic-ischemic event at birth, followed by a spastic-dystonic tetraplegia with mental retardation. At the age of 8 months, generalized dystonia occurred with axially fixed dystonic postures and dystonic movements of the four limbs. Brain MR showed diffuse atrophy involving the basal ganglia. At the age of 8 years, after an acute respiratory infection caused by the H1N1 flu virus, the patient had status dystonicus, which caused pain and oxygen desaturation episodes. The patient was admitted to the ICU; invasive mechanical ventilation and sedation with midazolam, propofol, and thiopentone were performed. He underwent an intrathecal baclofen (ITB) pump positioning, which halted the status dystonicus. In addition he underwent PEG positioning. Three months after surgery, the ITB system became infected and was removed. The patient continued to take phenobarbital, baclofen, tetrabenazine, and lorazepam, resulting in status dystonicus control. At the age of 10 years, without any apparent provoking factor, he had a status dystonicus relapse with clinical features similar to the first episode. A novel ITB pump implantation was performed, but it was removed soon after because the hardware became infected. A tracheostomy was then performed and several drugs were administrated without efficacy. The patient developed sepsis and deep vein thrombosis, and 2 months from status dystonicus onset, a bilateral pallidotomy was performed.

Medical and surgical management are summarized in Table II.

RESULTS

In the series the mean age of dystonia onset was 16.5 months (range 4-36). The mean age at onset of the first episode of status dystonicus was 10.8 years (range 5-17); the mean period between status dystonicus onset and surgery was 46 days (range 14-60).

Neither intraoperative problems nor postoperative complications occurred. Mean follow-up after surgery was 18.2 months (range 15-22). All the patients showed a progressive improvement of dystonia without any recurrence of status dystonicus. Resolution of status dystonicus was obtained within 21 to 60 days after surgery (mean 38). After pallidotomy, dystonic oromandibular, axial and limb postures and movements improved. All the patients recovered to their pre-status dystonicus condition and, at the time of writing, they are not restricted to lying in bed and can spend time in a wheelchair. In patients 1, 2, and...
postoperative dystonia improved compared with the pre-status dystonicus dystonia, as shown by reductions in the severity scores of the BFMDRS (Table III). At the latest follow-up, all the patients maintained a pharmacological politherapy.

Case 1
Status dystonicus disappeared with a strong reduction of dystonic movements 40 days after surgery. Mild oromandibular dystonic movements, and sporadic spasms of right arm and legs, induced by discomfort and pain, remained. Currently, the patient requires mechanical ventilation only during the night. At 22 months of follow-up, the postoperative results are stable (BFMDRS: severity 16, disability 30).

Case 2
A progressive improvement of status dystonicus was observed within 1 month of surgery. Three months after surgery the patient regained independent respiratory function and showed mild dystonic movements of the four limbs. He also recovered environmental contact and the ability to sit in a wheelchair. The BFMDRS severity/disability scores at the last follow-up were 4/30.

Case 3
Status dystonicus was controlled 3 weeks after surgery and dystonic symptoms dramatically improved 30 days later (Fig. 1). A further progressive neurological improvement was observed during the 3 months after surgery. Up to now (15mo after surgery), trunk hyperextension and limbs spasms have disappeared; the patient is able to sit in a wheelchair. After an emotional trigger, smooth dystonic facial and upper limbs spasms can appear (BFMDRS severity/disability scores: 57/30).

Case 4
Resolution of status dystonicus happened 2 months after surgery. At the last follow-up (15mo), mild sporadic dystonic movements of the trunk, limbs, and of the oromandibular district, usually aroused by emotional triggers, are observed. The patient recovered the capacity for oral feeding and the ability to stay in a wheelchair (BFMDRS severity/disability scores: 44/26) (Table III).
At time of induction of general anesthesia, all patients monitored by Bispectral index (BIS) showed a low baseline value of 60 to 70 because they were still sedated. During anesthesia and surgery, BIS was maintained between 35 and 50. The requested time to increase the BIS value to 60 to 70 was about 10 minutes.

Micro-electrode recording was performed in all four patients. We analyzed four recording tracks because four out of eight were affected by external electrical fields. The recordings showed Globus pallidus externus (GPe) firing activity 7 to 8mm above the target (frequency discharges 10 (SD 5Hz), usually followed by a relative decrease in

![Figure 1: Globus pallidus complex activity for various depths (8 to 1mm above the MRI-based target). The top of the figure shows a selected GPifiring recording frame (1s). Gpi, globus pallidus internus; GPe global pallidus externus.](image)

<table>
<thead>
<tr>
<th>Case</th>
<th>Status dystonicus outcome after pallidotomy</th>
<th>Follow-up (mo)</th>
<th>BFMDRS before status dystonicus</th>
<th>BFMDRS at latest follow-up</th>
<th>Drugs at latest follow-up (per d)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Resolution after 40d</td>
<td>22</td>
<td>Severity: 101/120, Disability: 30/30</td>
<td>Severity: 16/120, Disability: 30/30</td>
<td>Phenobarbital 75mg, Tetrabenazine 12mg, Tizanidine 8mg, Baclofen 26mg, Nitrazepam 10mg, Clonazepam 0.8mg, Phenobarbital 100mg, Tetrabenazine 50mg, Baclofen 75mg, Lorazepam 3mg, Lamotrigine 63mg, Valproic acid 1200mg, Rufinamide 900mg</td>
</tr>
<tr>
<td>2</td>
<td>Resolution after 30d</td>
<td>21</td>
<td>Severity: 84/120, Disability: 30/30</td>
<td>Severity: 4/120, Disability: 30/30</td>
<td>Phenobarbital 75mg, Tetrabenazine 12mg, Baclofen 10mg, Clonazepam 2mg</td>
</tr>
<tr>
<td>3</td>
<td>Resolution after 21d</td>
<td>15</td>
<td>Severity: 60/120, Disability: 30/30</td>
<td>Severity: 57/120, Disability: 30/30</td>
<td>Tizanidine 12mg, Baclofen 10mg, Lorazepam 3mg, Valproic acid 1200mg, Rufinamide 900mg, Baclofen 50mg</td>
</tr>
<tr>
<td>4</td>
<td>Resolution after 60d</td>
<td>15</td>
<td>Severity: 77/120, Disability: 26/30</td>
<td>Severity: 44/120, Disability: 26/30</td>
<td>Phenobarbital 150mg, Clonazepam 2.8mg, Lorazepam 4mg, Baclofen 50mg</td>
</tr>
</tbody>
</table>

BFMDRS, Burke-Fahn-Marsden Dystonia Rating Scale.
overall activity corresponding to internal medullary lamina, that lies between GPe and GPi. GPi activity was recorded at 3 to 2mm to the MRI-based target, with a mean firing discharge rate 35 (SD 10Hz; Fig. 2).

**DISCUSSION**

Status dystonicus is a rare clinical condition, occurring in some dystonic patients. As a result of its infrequency, it is difficult to define its incidence. It is more commonly reported in children with a mean age at onset of 14 years (SD 9.5).³

Status dystonicus causes pain and exhaustion, can lead to dehydration, rhabdomyolysis, acute renal failure, respiratory complications, and is associated with significant mortality. Despite the clinical relevance and the potential disabling nature of this disease, neither well-defined management guidelines nor standardized therapeutic approaches are still available.

This condition is not easily controlled by pharmacological therapy alone.²,³ Sedation and tracheal intubation are the only ways to reach a rapid resolution of spasms or dystonic movements, thereby avoiding bulbar, respiratory, and metabolic complications. Further therapeutic strategies, including surgery, are sometimes required. In refractory cases, thalamotomy, pallidotomy, and DBS of the GPi have been performed with encouraging results.⁵

In our four pediatric patients, non-invasive procedures, including ICU deep sedation, were ineffective in status dystonicus control. In patient 4, ITB permitted control of the first episode of status dystonicus, but it proved ineffective in the status dystonicus relapse. In both circumstances the hardware was removed because of infection. ITB usefulness in the treatment of status dystonicus is questionable, and the rate of complications of ITB, such as catheter site leakage and pump infection, is high.⁶

Literature data show good results of DBS also in the treatment of medical refractory status dystonicus, sometimes associated with clinical improvement of the baseline dystonic conditions.⁷,⁸ Despite the good results, DBS is associated with a complication rate, so-called hardware related complications, including infection of the DBS system (6.1%), skin erosion (1.3%), lead fracture (5%) or migration (5%), pulse generator malfunction, are more frequently observed in patients with dystonia.⁹

Hardware related complications lead to dystonia worsening and increase the risk of status dystonicus onset or recurrence.⁷,¹⁰–¹² The incidence of DBS complications in pediatric patients with status dystonicus may be even higher, considering aspects of soft tissue coverage, suboptimal nutritional status and prolonged mechanical ventilation with risk of status dystonicus recurrence and complaint for the patients and their family. Unlike patients with dystonia, those affected by status dystonicus more frequently could have tracheostomy and PEG with an increased risk of hardware related complications.

---

**Figure 2:** Status dystonicus in bilateral striatal necrosis of unknown aetiology (Case 3). On the left, the surgical planning (T1-weighted images with gadolinium: axial, coronal, and sagittal planes); on the right, postoperative MR (T2-weighted images: axial and coronal planes) showing the mid pallidotomy (I: entry point; B: target).
Lesional procedures, including pallidotomy, represent a viable alternative with several advantages, including a decreased need for access to specialists and clinical follow-up, and a lower risk of infection. Previous reports reveal that mere unilateral pallidotomy or even unilateral pallidotomy with contralateral GPi-DBS are well tolerated in the treatment of dystonia. Our group previously described a case of a young patient affected by primary dystonia, who benefitted from bilateral DBS but required device removal after a severe skin erosion. Before the lead removal, radiofrequency ablation of the GPi was performed through the DBS electrode resulting in long-term stable control of dystonia. Similar results are reached both with combination of GPi-DBS and contralateral pallidotomy and bilateral DBS. Moreover, pallidotomy reveals its efficacy in patients affected by status dystonicus, both as an independent option. A report by Hooper et al. shows different clinical conditions on which lesional procedures are preferred over neuromodulation. Whereas sham-controlled randomized clinical trials are available for bilateral GPi-DBS only, a similar outcome of bilateral pallidotomy in an open-label retrospective series suggested that pallidotomy is similar to GPi-DBS in effectiveness and safety.

Literature data on the frequency of complications in patients who underwent bilateral pallidotomy are discordant: each group referring a different percentage of complications, including speech deterioration and disturbances of swallowing and drooling.

Anatomic observations show that the GPi is involved in both limbic system and extrapyramidal motor control. The motor part is represented by the posterior half of the nucleus, the limbic part corresponds to the anterior quarter, while an associative part is interposed. The anterior GPi has been targeted in patients affected by Tourette syndrome, characterized by motor and vocal tics, and also in Lesch-Nyan syndrome, on which self-mutilation is coupled with spasticity, dystonia, and other symptoms.

The VPL sensorimotor GPi, the so-called Laitinen and Leksell target, has been recognized as the optimal site for lesion surgery and DBS in Parkinson disease and in patients with dystonia. Choice of target position for electrode insertion in DBS for children has been described, at the junction of the anterior two-thirds and posterior-third of the GPi. In adults the target region is most commonly documented as the posteroventral GPi. Neuromodulation of a more anterior part of GPi proved effective in DYT1 positive patients, who received a bilateral GPI-DBS with two electrodes implanted in each nucleus after efficacy reduction of the lead on VPL.

In our series pallidotomy resulted in the mid portion of the GPi which represents the motor part of the GPi. This targeting allows for obtaining benefit on dystonic symptoms, without preventing patients from receiving a further gain from subsequent neuromodulation of the most posterior part (VPL) of the GPi. Considering the staged implantation of multiple electrodes at the level of GPi, it is otherwise possible to consider pallidotomy at the VPL level and a subsequent more anterior DBS. The relationship to the mid-commissural point collected in our series by the direct targeting (lateral X: 14-18mm; anteroposterior Y: 3-5mm; vertical Z: 2mm) was equivalent to other reported DBS series (anteroposterior 2-6mm; lateral 14-18mm; vertical 2mm). The utility of coordinates with reference to the mid-commissural point is limited, as has been previously reported by the Montpellier group. The target definition that we performed was not based on the use of the stereotactic coordinates, but on the visual direct targeting on MRI inversion recovery sequences. Moreover our small series is composed of secondary dystonia cases, which sometimes show distortion of pallidal boundaries (Fig. 1).

Although microelectrode recording (MER) could increase the risk of hemorrhage, it could also improve selection of the target and recognition of the surrounding structures, so optimizing the clinical response, while minimizing adverse effects coming from the involvement of internal capsule or optic tract by the lesion.

Microelectrode recording and stimulation techniques are well established tools to obtain direct measures of the activity of individual neurons within the basal ganglia, providing relevant insights into the pathophysiology of the movement disorders, which is not completely clear. In a recent study, Kojovic et al. demonstrated that primary and secondary dystonia have different pathophysiological mechanisms. Pathophysiological deficits in primary dystonia include reduced inhibition at many levels of the motor system and an enhanced cortical plasticity, while a role of cerebellar deficits has been suggested, but has yet to be clarified. Pathological mechanisms of secondary dystonia are not clear: a reduced intracortical inhibition has been observed, but this is not a specific characteristic of dystonia as a similar abnormality is found in other basal ganglia diseases. Maybe, the coexistence of reduced intracortical inhibition and an injury to basal ganglia, which can lead to a decreased inhibition of thalamic activity and consequently to an increased excitability of the motor cortex, can cause clinical expression of dystonia. Data recorded from GPi in our four patients show a mean firing rate of 35Hz, similar to those found in other series.

Vitek et al. have postulated that pallidotomy may work by interrupting an unregulated and disruptive circuit, allowing the remaining structures and circuits to function more normally.

STUDY LIMITATIONS

In this work, sample size is relatively small; this limitation is because of the low incidence of status dystonicus. Further multicentric investigations will be required to confirm the efficacy of the described technique.

Another source of weakness of this study should be the relatively short follow-up; however, we consider our outcome as a good result if we compare it with literature data on outcome of status dystonicus after pallidotomy and other surgical approaches.
CONCLUSION
Our results confirm the efficacy of the bilateral mid lesioning of GPi in treating pediatric patients affected by secondary dystonia complicated by status dystonicus.

REFERENCES