

Case Series of Deep Brain Stimulation of Globus Pallidus Internus as Therapy for Dystonic Storm in a Single Center in Colombia

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Introduction

Dystonic Storm (DS) is a life-threatening-complication of dystonia characterized by a sudden and persistent condition in which episodes of dystonic movements become increasingly frequent and severe, requiring urgent hospital admission, that can lead to respiratory, metabolic and bulbar complications1,2. Pharmacologic treatment has been the mainstay management for this complication, however, many refractory patients will still require further treatments. Deep brain stimulation (DBS) of bilateral globus pallidus internus (GPi) is an interesting therapeutic strategy that has been used for dystonia and now it has been proposed to be used for DS.

Methods

We describe 5 cases with DS, that required emergent placement of a bilateral GPi DBS to control their symptoms. All five patients coursed with dystonic movements of different body regions including neck, extremities and trunk that interfered with ambulation. Severity was measured with the UDRS scale. Precipitating factors were identified in two cases (superior airway infection and electrode dislocation). Pharmacologic therapy wasn’t effective in any of the cases and ICU admission was necessary before surgery. Decrease of dystonic movements and resolution of abnormal postures was achieved in all five patients in a mean period of 6,6 days (1,6 days adults, 14 days pediatric).

Results

DS treatment is challenging. As reported by Fassano et al. in 89 cases of DS, pharmacological therapy was used as first line treatment in 82.4% of cases, ceasing DS only in 9 cases (10.1%), while surgery [either DBS (13 cases) or ablations] was used in 30.2% of cases being effective in the 33.7% of these events1. In our case series, pharmacologic therapy wasn’t sufficient to abort DS and DBS placement in bilateral GPi was effective for symptomatic control.

Learning Objectives

- 1) GPi DBS can be a suitable, versatile, reversible and adequate therapy of DS.
- 2) In pediatric population is important to consider the possibility of electrode dislocation and migration as a trigger for DS in a patient with a previous DBS placement for the treatment of dystonia.

References

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