

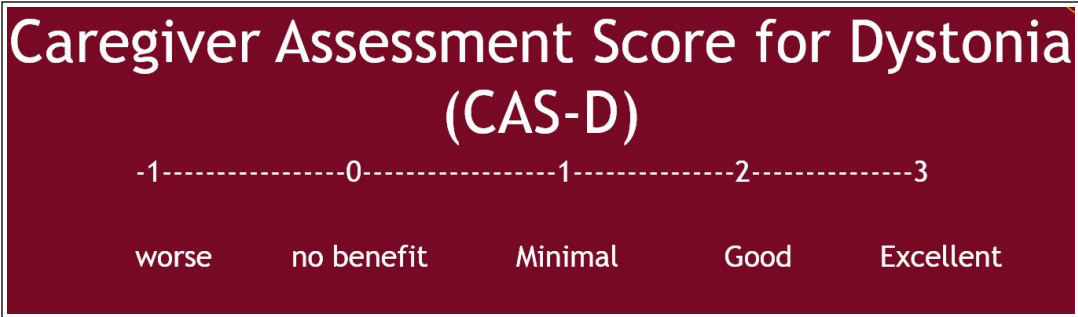


Introduction

Deep Brain Stimulation (DBS) of the Globus Pallidus Interna has been shown to be extremely effective in primary generalized dystonia. There is much less evidence for the use of DBS in patients with secondary dystonia. We present a series of 14 patients with secondary dystonia who underwent pallidal DBS at our institution.

Methods

Approval by the **Children's Hospital Los Angeles** Institutional Review Board was obtained prior to initiation of this study. A retrospective review of patients with secondary dystonia who received treatment with DBS between February 2011 – December 2014 was performed. Pre-operative and post-operative videos were scored using the **Barry Albright Dystonia (BAD) score**. In addition, a **Caregiver Assessment Score for Dystonia (CAS-D)** was completed using the following scale: -1(worse), 0(no benefit), 1(minimal benefit), 2(good benefit), 3(excellent benefit).



Results

BAD scores improved by 4% ($p=0.07$) with minimal improvement in patients with kernicterus (n=3, 0% improvement) and the most improvement in dystonia secondary to drug overdose (n=1, 22% improvement). However, **even with 0% BAD improvement, patients with kernicterus had life-impacting changes that were not reflected by BAD scores:** patient A had less opisthotonus with a significant decrease in respiratory complications, patient B was able to dress herself, and patient C was able to independently drive his wheelchair after DBS treatment. In addition, even in patients with minimal change on BAD scoring, caregivers reported decreased contractures and spasms leading to improved comfort levels. The mean CAS-D score was 1.4 with 2, 4, 6, and 2 caregivers reporting excellent, good, minimal and no benefit respectively. In addition, there were no complications such as infections or hematoma in our series.

Conclusions

DBS does provide benefit to a segment of the secondary dystonia population. Current dystonia scoring does not adequately reflect patient improvement after DBS. Larger studies for secondary dystonia focusing on age, etiology, and patterns of dystonia are needed to learn which patients will respond best to treatment.

References

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	Age at implant	CAS-D	Preop BADS	Postop BADS	Additional Observations
Kernicterus					
A	13	1	28	28	Less opisthotonus, significant decrease in respiratory complications
B	16	3	23	23	Increase in functional independence
C	18	1	26	26	Increase in functional independence
Cerebral Palsy					
D	20	0	25	24	
E	17	1	28	28	
F	20	2	27	19	
G	14	0	22	22	
H	19	1	28	28	
I	6	2	28	28	
J	13	1	27	27	
K	11	2	28	28	
L	16	3	28	28	
Drug Overdose					
M	20	1	27	21	
Glutaric Aciduria 1					
N	17	2	24	24	