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What parents think and feel about deep brain stimulation in paediatric secondary dystonia including cerebral palsy: A qualitative study of parental decision-making

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ABSTRACT

Background: Dystonia is characterised by involuntary movements and postures. Deep Brain Stimulation (DBS) is effective in reducing dystonic symptoms in primary dystonia in childhood and to lesser extent in secondary dystonia. How families and children decide to choose DBS surgery has never been explored.
Aims: To explore parental decision-making for DBS in paediatric secondary dystonia.
Methods: Data was gathered using semi-structured interviews with eight parents of children with secondary dystonia who had undergone DBS. Interviews were analysed using Interpretative Phenomenological Analysis.
Results: For all parents the decision was viewed as significant, with life altering consequences for the child. These results suggested that parents were motivated by a hope for a better life and parental duty. This was weighed against consideration of risks, what the child had to lose, and uncertainty of DBS outcome. Decisions were also influenced by the perspectives of their child and professionals.
Conclusions: The decision to undergo DBS was an ongoing process for parents, who ultimately were struggling in the face of uncertainty whilst trying to do their best as parents for their children. These findings have important clinical implications given the growing referrals for consideration of DBS childhood dystonia, and highlights the importance of further quantitative research to fully establish the efficacy of DBS in secondary dystonia to enhance informed decision-making.

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1. Introduction

Dystonia refers to a heterogeneous group of movement disorders. The most recent consensus agreement on the definition of dystonia states that: Dystonia is defined as a movement disorder characterized by sustained or intermittent muscle contractions causing abnormal, often repetitive, movements, postures, or both. Dystonic movements are typically patterned and twisting, and may be tremulous. Dystonia is often initiated or worsened by voluntary action and associated with overflow muscle activation'.[1]

In childhood, dystonia is a heterogeneous disorder, with a wide range of causes and clinical features, varying severity and response to medical managements.[2] Dystonia has historically been classified by aetiology, either as primary or secondary. Primary dystonia is a movement disorder of unknown but proven or suspected monogenetic cause, where dystonia is the only neurological feature.[3] In secondary or acquired dystonia, the dystonia develops secondary to other conditions or identified disease processes such as cerebral palsy (the commonest cause of dystonia in childhood), neurometabolic, autoimmune, genetic and neurodegenerative conditions.[4] Children with secondary dystonia have been shown to spend a higher proportion of life living with dystonia, experience a greater severity of disability and have lower functioning capacity.[5] Dystonia impairs intentional movement, causing physical disability, functional impairment, and often pain and communication difficulties which prevent children from participating in activities of daily living, education, and age-appropriate social activities, and can lead to dependence on family members. This dependence places additional physical and emotional demands on parents, who often assume roles beyond the normative activities of parenting.

Management options for dystonia while increasing dramatically in choice[6] have little class I supporting evidence and most options are therefore applied 'off label' as agreed between the family/carers and the treating physician.[7] Although pharmacological management is commonly ineffective in generalised and multifocal dystonia[7–9] and is often accompanied by unwanted and adverse side effects.[10] There has been increased focus on emergent neurosurgical interventions for the management of dystonia, and childhood dystonia is now being routinely managed with Deep Brain Stimulation (DBS), a reversible 'non-lesioning' neurosurgical treatment[7] but usually only after demonstrating that dystonia has proven refractory to accepted pharmacological management options.[7]

Increasing evidence suggests DBS is successful in reducing childhood dystonia, demonstrating significant improvement on impairment focussed measures, such as the Burke Fahn Marsden Disability Rating Scale.[11,12] However, secondary dystonias appear to be less responsive to DBS compared with primary dystonia,[11] and improvements in motor scores have been shown to be more subtle and not as durable.[11] Studies have shown that impairment measures have failed to capture the subjective meaning of post DBS changes, or the functional priorities and concerns of parents.[13,14] The importance of duration of the dystonia has also been highlighted; with the response to DBS declining with increasing proportion of life lived with dystonia[11] and recommendations that surgery should be offered at a young age to minimise proportion of life lived with dystonia and maximise responsiveness and minimise or prevent inevitable fixed musculoskeletal deformities.[15]

DBS is now the management of choice for dystonia in certain specialised centres. In order to help ensure that DBS is used responsibly, it is necessary that professionals are attentive to the perspectives of patients.[16,17] Given the gap between professional experience of DBS and public understanding of the advantages and limitations of DBS functional neurosurgery it is perhaps surprising that to date, the exploration of decision-making in DBS surgical options has been ignored. Given the variability of outcomes in secondary dystonia, and growing evidence that impairment measures are not sensitive enough to detect small but significant changes[14] a greater understanding how parents experience and manage DBS decision-making would be valuable. The decision to undertake DBS for families with secondary dystonia comprises a combination of unique factors: children with variable cognitive and communication abilities (see Owen EJPN This edition), a lack of outcome certainty, a long term commitment to regular hospital follow up appointments and a daily commitment to battery charging.[18] Little is known about how these factors influence the decision to undergo DBS surgery. Understanding the DBS decision-making process of parents, and factors that are important to families, would help clinicians improve family preparation and support, and enhance the informed consent process. Greater support could also potentially reduce decision-making times, which have in certain cases taken many years as families opt to wait until the child is old enough 'to make their own mind' which is important because shorter dystonia duration and younger age at surgery have been associated with better outcomes after DBS.[11] Additionally, this paper by providing important insights on decision-making and thus informed consent can also contribute to and inform more general discussions on the ethical challenges of DBS.[19,20]

Our objective was to explore parents' decision-making processes and the factors that impact on their decision in a group of children with secondary dystonia who have undergone DBS.

2. Methods

2.1. Design

This cross-sectional qualitative study was conducted between July 2014 and January 2015. Semi-structured interviews were completed with eight parents of children with secondary dystonia who had undergone bilateral pallidal DBS to retrospectively explore parents' experiences of DBS decision-making.

2.2. Participants

Parents/main carers of patients with secondary dystonia attending a tertiary hospital specialist complex movement