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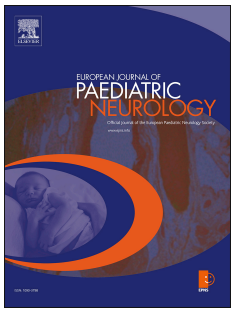

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What parents think and feel about Deep Brain Stimulation in paediatric secondary dystonia including cerebral palsy: A qualitative study of parental decision-making

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Highlights/ what this paper adds:

This is the first study to explore parental experiences in decision-making for deep brain stimulation consent in children with secondary dystonia illustrated with verbatim interview quotations

An over-riding desire to do the best for the child helped families come to terms with potential risks of DBS surgery

A lack of a child-specific prognosis for outcome of DBS was outweighed by a desire to avoid regret at failing to make a decision to go forward with DBS surgery

Parents of higher-functioning children had to overcome the fear of their child losing function as a consequence of DBS surgery

Remarkable parental resilience in the face of a life devoted to the care of their dystonic child was a constant feature in all cases

Parents strongly respect the opinions of the professionals advising them and there is therefore a burden of responsibility to provide better evidence of efficacy of DBS for secondary dystonias

This study will help families and health-care professionals understand how families take important decisions to help children with secondary dystonia disability

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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.

Key Words: secondary dystonia, deep brain stimulation, decision-making, informed consent, bioethics

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[7,8,9] 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 disorder service were identified and recruited directly from the clinic and invited to take part by the Clinical Psychologist within the team. The tertiary hospital is a national centre in the United Kingdom for the assessment and management of childhood movement disorders. Its intervention strategies are similar to those in use in the other centres in the country with the addition of over 10 year's experience of deep brain stimulation for children with dystonia.

Consecutive sampling was employed to select a homogenous sample that met the inclusion and exclusion criteria: patients had a diagnosis of secondary static dystonia that developed during infancy (birth to 2 years); had DBS surgery at less than 17 years of age and the surgery had occurred 12-24 months prior to the interview. Parents were excluded if they were unable to comprehend and speak English fluently to avoid biases in data interpretation. All of the eight families who were invited to take part consented to participate.

All eight participants identified themselves as a main carer: seven mothers and one father were interviewed. The children of the parents were between three and seventeen years of age at the time of surgery. Three of the children were male, and five were female. Despite fulfilling inclusion criteria of a diagnosis of secondary static dystonia, there was variability in dystonic aetiology. Of the eight children, six had a diagnosis of cerebral palsy (CP), one had an inherited genetic condition; and one diagnosis was unknown. All children fulfilled inclusion criteria since they were born with or developed dystonia during childbirth, or as a result of complications during birth or in the neonatal period. The children's motor and verbal capabilities varied. Motor ability was defined using the Gross Motor Function Classification System[21] and communication ability using the Communication Function Classification System[22]. Two children experienced complications with their DBS system in the year post surgery. This demographic information and surgery information is summarised in Table 1.

Table 1: Child Demographics

Child Pseudonym	Gender	Dystonia Subtype	Age at time of DBS surgery	CFCS Level*	GMFCS Level*
Imogen	Female	Secondary Dystonia	17 years old	I	IV
Megan	Female	Secondary Dystonia	16 years old	I	I
Wade	Male	Secondary Dystonia	16 years old	I	II
Philip	Male	Secondary Dystonia	14 years old	IV	IV
Charlotte	Female	Secondary Dystonia	12 years old	I	V
Ivy	Female	Secondary Dystonia	3 years old	IV	V
Billy	Male	Secondary Dystonia	9 years old	III	IV
Emily	Female	Secondary Dystonia	11 years old	III	IV

**Gross Motor Function Classification System (GMFCS)) – This is a five level physiotherapist rated classification system based on self-initiated movement and has an emphasis on sitting, walking and wheeled motor ability. Typically children at 'level I' can walk without restrictions, but have difficulties with more advance motor skills, whereas at 'level V' all areas of motor function is limited and children require assistive technology and physical assistance.*

**Communication Function Classification System (CFCS) – CFCS scale ranges from 'level I' indicating minimal impact on communication, where as children at 'level V' struggle to communicate effectively and be understood by even familiar people.*

2.3 Measures

Semi-structured interviews were used to elicit rich and detailed accounts of individuals' lived experiences[23]. The researcher adopted an exploratory participant-led approach to explore what was meaningful for each participant. Additional 'prompt' questions could then be used to inquire about interesting and unexpected areas[24]. The researcher developed an interview schedule to guide the interviews through familiarisation with the literature, and consultation with the clinical team and service-users. The interview schedule was made up of open-ended questions to encourage unbiased narrative and reflection. Initial warm up questions were designed to help build rapport, and broad introductory questions were used to allow the participant to construct the parameters of the conversation and speak about what was personally meaningful for them. A pilot interview was completed to refine the interview schedule based on 'sensitivity', 'clarity' and 'content'. In line with a participant-led interview, the researcher was guided by how comfortable participants were in talking and how much they wanted to say. Consequently there was variation in interview length: Interviews ranged in length from 62 to 129 minutes.

2.4 Procedure

Patient and participant demographics were primarily gathered by the researcher from the interviews. However, the clinical psychologist collected diagnosis and surgery details from the hospital database. The researcher interviewed those who agreed to take part. Parents were offered a choice of interview location: Five parents were interviewed in a private clinic room at the hospital and three interviews were completed in participant homes.

A local NHS Research Ethics Committee granted approval and informed consent was obtained from all participants. Service user consultation took place throughout the design and data collection to ensure the study was grounded in parents concerns and had clinical utility and applications for service delivery. Consultation informed exploratory aims, study procedure decisions, the interview schedule, the information, consent and debrief sheets.

2.5 Data Analysis

Interviews were digitally recorded and transcribed verbatim by the researcher. Qualitative Interpretative Phenomenological analysis of each transcript was conducted. The interviews were analysed following the flexible IPA guidelines[23], in which the researcher followed an iterative process of reading and re-reading, initial line-by-line exploratory coding, developing emergent themes and then clustering and collapsing emergent themes for each individual transcript. The clusters were then compared across cases to develop super-ordinate theme labels which synthesised an overall representation of participant experience.

To maintain validity and provide a credibility check[25] the first transcript was independently coded by the clinical psychologist, and two authors (AA and TO) discussed, clarified and agreed themes emerging from the analysis to ensure that the themes were grounded in participant perspectives and were supported by verbatim quotes. Two families also provided a validity check and reported that the themes accurately represented their experiences. To achieve a rigorous IPA, as recommended by Smith[26] extracts from at least half of the participants were represented in each theme.

3. Results and Discussion

This study captured the decision-making process of families who had undergone DBS surgery for secondary dystonia, and it is the first study to explore DBS decision-making in any population. The superordinate theme identified was *'facing the uncertainty of decision-making'*, which was comprised of four sub-themes: the *context of disability & hope for a better life*, *parents' attitude to parenting a child with disability*, *uncertainty of outcome and potential risks and involving children* and *trusting professionals*. These themes are supported by quotations from participant interviews, either embedded within the narrative descriptions of each theme or as stand-alone quotations.

This study provides a new understanding of the psychological processes and complexity of factors influencing decision-making. For all parents the decision for their child to undergo DBS was viewed as significant, with life altering consequences for the child. This decision involved consideration of a number of factors, before deciding to go ahead with the surgery. This decision process was set in the context of their child being physically, functionally, psychologically and socially disadvantaged due to disability. The treatment context offered potential for long term benefit in reducing dystonic spasms, but had short term costs (hospitalisation, surgery), longer term costs (recovery, setting adjustments) and the consequences of their child being dependent on a technical device. Undoubtedly, the overall sense was that parents were trying to do what was best for their child.

3.1 The Context of Disability & Hope for a Better Life

The driving motivator to consider DBS Surgery was the parental desire to give their child a better life. This decision was therefore set in the context of the child's physical difficulties and wider social and emotional experiences of dystonia:

Obviously severely affected by cerebral palsy from a mobility point of view. She has athetoid and dystonic CP. Obviously urr, quadriplegic so basically full body.

Because of his speech impediment he (pause)…urm because he's embarrassed about his speech, anybody who tries to be friendly he feels that they're pitying him, he, he doesn't feel that he's accepted for who he is.

Difficulties with fine motor skills, and hand-writing and all that sort of thing, which obviously (pause) has affected his education, over the years

Parents had different hopes for surgery, some were physical (more control, reduction in spasm), others were functional (improve participation in activities) and some were about QoL (pain reduction).

Think it was maybe to do with her arms. Because that's a lot of impact, all that dislocating and spasm. You know she's trying to do stuff at school, or doing anything, and it keeps getting in the way.

So urm (pause), giving her the best possible outcome long term, was my most important aim and obviously to alleviate discomfort and pain, because as a mum its awful to see and you know you want to help

These hopes were consistent with the functional priorities of parents identified in the literature and the different priorities for higher and lower ability children[14]. Beyond functional concerns, parents of more able children were also motivated by a desire for their child to have a more 'normal life' through participation in age-appropriate activities, independence and looking visibly 'more normal'.

Our hopes and really I think, the reason we went ahead with it was you know…the same as for any parent, for Wade to be able to live independently, you know on his own, without needing any help from anyone else to do

This was the first study to focus on the visible aspect of dystonia and how feeling different, could motivate families to undertake DBS to try and achieve a sense of normality.

3.2 Parents' Attitude to Parenting a Child with Disability

This decision was located in the context of parent's belief system and experiences. Parents believed they must do everything to help their child achieve their full potential and provide the best opportunity in life:

I don't want her to be stopped doing things because of her disability. I want her to be able to experience everything that everyone else can experience

The DBS decision was therefore in keeping with how parents have reacted and coped with disability throughout the child's life. Parents struggled with their inability to 'fix' the child and, as such, appeared to go to any lengths to overcome the barriers and restrictions caused disability:

I want to solve everything, and as a mum you want to wave a magic wand and make it all go away and you can't, so the next best thing is to try and do, give her a whole range of experiences

There was a sense of parental responsibility throughout the accounts. And what was most striking was the lengths parents went to provide for their children and the strength of parental love and devotion that shines through:

My kids have always been the most important thing in my life, I'd do anything for them

Research has previously identified parents' concern about ensuring everything possible is done for their child[27,28,29]. Notably, all hopes were about the child and parents never voiced their difficulties as a motivator to undergo surgery. This speaks to the parents' unwavering commitment to give their child a better life. This study offers new insight into the broader relational and social context in which DBS decision-making takes place, and is in line with previous studies of elective surgery where social, emotional and psychological factors were important in decision-making[30, 31, 32].

3.3 Uncertainty of Outcome and Potential Risks

Deciding to have DBS was a very 'big decision' and a difficult process for all parents. For all parents this process was made up of different stages. All parents faced the dilemma of deciding whether the child should have surgery with its associated risks and no certainty of what impact DBS would have. Parents experienced 'fear' that their children would end up 'more damaged':

> *I thought that she, you know 'cause its brain surgery at the end of the day isn't it. I thought she might come out and she wouldn't be able to speak, she wouldn't be able to see, you know. I think my main fear was that they would do something else to make her more disabled*

Many parents described overwhelming fear because of the meaning of brain surgery:

> *Because it was, you know, we don't wanna, something as big as surgery, brain surgery,… It was a big decision to make*

The meaning of neurosurgery was clearly significant and has been under-researched in the literature. In this study neurosurgery was perceived to be more risky than other types of surgeries, and resulted in greater decision-making burden for families.

Decision-making was influenced by the severity of child disability, and there was a contrast in decision-making between parents: parents whose children were more physically able with high cognitive functioning perceived their children as having more to lose than parents of children who were severely impaired.

> *Wade had a reasonable quality of life before… And urm the fact that before DBS he could walk, you know he had his intelligence and that sort of thing, and had reasonable speech so the idea that any of those could be affected in a bad way was probably one reason why we took a while to decide*

> *So ultimately in my mind set what have I got to lose… from the kind of physical point of view even if the surgery went wrong, Ivy wasn't going to lose anything, because she couldn't do anything*

This process influenced the ease of decision-making, and consequently the length of time it took to make the decision. Parents of more able children appeared to agonise over this decision, and displayed ambivalence as they often changed their minds, whereas parents of less able children were not tormented by the uncertainty of if they had made the right decision. This decision was also experienced as *difficult* because parents could be offered no certainty of DBS outcome. The lack of guarantee for positive outcome or certainty of how the DBS would change the child's dystonia made the decision more difficult for every parent:

This lack of certainty of what could be achieved was compounded because each child's disability was completely different, and because of the lack of a thorough understanding of how DBS affects children with secondary dystonia:

> *Because it's secondary, there's a lot more questions, is it worth doing? You can't give me any definite answers… I don't know about anyone else, but for*

me that was the biggest thing ever, I'm doing this but is it actually going to work?

This uncertainty represented the main struggle for parents, and Mishel's Uncertainty in Illness theory[33] conceptualises how unfamiliar procedures and potential change in health status, lead to increased uncertainty and distress.

Ultimately, all parents made the final decision by privileging the hope for a better life over all perceived risks, and that surgery was 'worth a chance' if it 'would give a glimmer of making life easier' for the child:

Hope of a positive outcome, was, overweighed any other objections I think

Parents believed the surgery was worth the chance. In terms of models of decision-making regret theory[34] proposed that in conditions involving risk people often make decisions, by weighing up consequences of a possible action with consequences of different decisions. It proposes that people are motivated to take action to avoid future regret. In this context, parents described feeling lucky to be offered DBS, and there was a sense of parents wanting to try all options, and find out conclusively if DBS could help their child to avoid regret:

but we moved on and we decided it was worth trying because I think in life if you try something and either you don't like it or it doesn't do what you anticipate, you've tried, but if you don't try you never know, and I think regret or looking back on things and saying 'I wish I had' is far more painful, more difficult than not trying them at all.

These extracts clearly demonstrated how parents struggled with the uncertainty of decision-making. Some parents continued to be affected by the burden of responsibility and difficulty accepting they have made the right decision.

3.4 Involving Children and Trusting Professionals

A key feature was listening to the views of the child and involving them throughout the decision-making process. Because DBS is an elective surgery which has the long-term impact of being dependent on a technical device, parents sought to involve children as much as possible considering their age and cognitive abilities:

Its her brain, its us making that decision for her, she needs to have some say in it, as best she can, at the age that she was

Children attended all the appointments and were involved in discussions from the beginning. However, parents also held a protective role in keeping positive and minimising risks to try alleviate children's worries and concerns. However, ultimately responsibility for the decision fell to the parents:

and I think the concerns that we had instantly were are we gonna go ahead with this without her full adult consent, she's still young, can we expect to make this decision for her, because she's still a child, this is us, deciding what to do, and it was, that was the concern that we had

Another important factor was the trust and value parents placed in professional opinion. Professionals seemed to hold a position of power in influencing parents to go ahead with the surgery.

Healthcare professional power and competency has previously been shown to influence parents' decision-making[29]. It seems that for elective surgery, when professionals can't guarantee positive outcome because of the heterogeneity of secondary dystonia, parents engaged in a long process of weighing up perceived benefits and costs as a family, and were very reliant on professionals in the face of this uncertainty.

Given this uncertainty, parents' spoke of preparing themselves emotionally for the procedure by carrying out extensive research, relying on the information and photographs provided by the medical team, and through conversations with other families who had been through the surgery. Having post-surgery photographs seemed to help families prepare and develop realistic expectations, In the face of perceived risks and uncertainties, knowledge of what the wound would look like and how the scar heals provided families an element of predictability and certainty otherwise lacking in this procedure. Furthermore, for some families knowledge of the visibility of the battery-pack was integral to their decision because their DBS surgery hopes involved looking more 'visibly normal'.

3.5 Conclusion and Clinical Implications

This study described for the first time, to our knowledge, the experience and perceptions of parents during decision-making for DBS surgery, which has implications for the clinical support offered to parents and families during the DBS process, and has led to a wider understanding of the factors that influence decision-making and how parents manage the process.

The decision to undertake DBS was a difficult and significant decision for all parents and regarded as having life altering consequences for the child and family. Parents were motivated by their hope for a better quality of life and sense of parental responsibility to help children achieve their full potential. Parent's balanced their hopes against perceived risks, the uncertainty of DBS outcome, and personal fears and reactions to neurosurgery. The decision-making burden appeared greater in parents whose children were less impaired, who perceived there to be more to lose. Parent's sought to involve children throughout the process, especially due to the long-term impact of being dependent on a technical device. Decisions were also influenced by the trust and value parents placed on professional opinion and recommendations.

Managing uncertainty was the prominent struggle for parents, and clinicians have a responsibility to ensure parents can make an informed decision with all the relevant information:

- In the face of uncertainty of DBS outcome, parents need to be provided with the latest outcome evidence for secondary dystonia to ensure informed decision-making.
- Clinicians need to be clear and informative about likelihood of DBS changes and support families to develop realistic expectations of change.
- Clear information and recent photographs should be provided of the location of DBS implant, stitching in the head, scaring after surgery, recovery process and the visibility of the battery pack under the skin.
- This study suggested parents of more able children (lower GMFCS scores) and children where there was a disparity between cognitive and physical functioning, struggled more with uncertainty and the responsibility of decision-making, perceiving there to be more to lose. Clinicians' should be aware that parents who were more ambivalent in their decision may be more vulnerable to experience distress during and after the surgery.
- Decision-making ambivalence could make parents vulnerable, and place professionals in a powerful position. Professionals should be mindful of this, and ensure parents are provided the time and information to reach their own decision. Parents should also be encouraged to speak with other families to help develop realistic expectations and fully consider DBS implications to ensure informed decision-making.

This study tentatively suggests that decision-making can be a stressful experience for parents and longer term follow up of families is required. More research is necessary to clarify this. Furthermore, a main struggle for parents was the lack of certainty of DBS outcome in secondary dystonia, calling for further research to fully understand the efficacy of DBS for secondary dystonia to allow families to make informed decisions[35, 36,37, 38,39].

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3.6 Strengths and Limitations

This study has three main key strengths. First, its qualitative IPA approach enabled collection of rich narratives that have yielded many insights into the lived experience of decision-making from the perspective of parents. An important strength was the use of credibility checks and reflexivity to maintain quality and validity of final themes. Finally, every parent approached agreed to take part, this reduces recruitment bias of people volunteering to share overly positive or negative experiences.

The main limitation was the heterogeneity of the sample. There was variability in parent characteristics as only one father was interviewed, and although not selected for, all parents identified as white British. It is therefore likely that the themes are representative of mother's experiences from one cultural group. Given heterogeneity of child characteristics it was difficult to ascertain what experiences were unique to secondary dystonia, and themes are therefore representative of children who have secondary dystonia and another diagnosis e.g. CP. This heterogeneity and small sample size clearly create a challenge in terms of being able to make reliable generalisations. There is a need for further research to explore these initial findings, and broaden our understanding of decision-making in DBS within a paediatric dystonia population. An outstanding question is how children experience the decision, which could contribute to an overall understanding of family decision-making.

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Keyoumars Ashkan: Review and Critique

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