Deep brain stimulation (DBS) is increasingly considered for the treatment of refractory neurodevelopmental conditions affecting children. In 1999, Coubes and colleagues were among the first to report on the use of pediatric DBS for primary dystonia. Since then, pediatric DBS has gained considerable traction, with several case series reported. Although dystonia remains the most common indication for pediatric DBS, the technology has also been applied to treat epilepsy and Tourette syndrome. More recently, DBS of the nucleus accumbens and amygdala have been reported for self-injurious behaviors.

The majority of the evidence for DBS in children has been derived from the adult literature, in which the procedure has been shown to be safe and effective for an increasing number of indications. However, pediatric DBS has many substantial differences, with important bioethical, social, and legal considerations. First, patients and their caregivers represent a uniquely vulnerable population. Second, given biological differences between adults and children, the procedure in pediatric populations can be considered a surgical innovation and is still investigational for all indications. Third, the natural history of a disease within the context of a child’s neuromuscular and cognitive development must be factored into treatment decisions. Furthermore, the surgical risks and technical nuances of DBS differ from those in adults. Thus, evidence from the adult DBS literature may not be readily translatable to pediatric populations.

With the exception of DYT1 dystonia (for which there is level II-2 evidence), all indications for DBS in pediatric patients are in the early stages of investigation; thus, the procedure represents a surgical innovation, i.e., a procedure that “departs in a significant way from standard or accepted practice.” Surgical innovations are characterized by evolving techniques, outcome measures, and patient selection. Three systematic reviews and meta-analyses (two manuscripts currently under review and one paper in press) have described the conduct of DBS in pediatric populations (Table 1), all emphasizing significant heterogeneity and the need for prospective studies to guide patient selection. The framework we describe here is intended to be applied to DBS in children in all settings, most of which are presently experimental or under the realm of surgical innovation. Ultimately, the most basic ethical principles protecting research subjects flow from the Nuremberg Code (1948), the Declaration of Helsinki (1964), and the Belmont Report (1979); however, in children with a medically complex condition who may benefit from novel investigational treatments, a more targeted framework is needed.

Herein we expand on differences in the conduct of DBS between adults and children and the unique ethical challenges they pose. First, we describe ethical issues raised by the vulnerability of the patient population, the novelty of the application of DBS for expanding indications in children, and the conduct of the procedure in children despite limited evidence. We then propose a bioethical framework for the evaluation of children for DBS. Although ethical issues may figure more or less prominently depending on the individual patient and the indication, the framework we put forth may be generalized to shape personal, institutional, and social policies regarding DBS in children. It is not our intention to prescribe the steps and thought process for carrying out pediatric DBS, but rather to advance dialogue regarding the ethical underpinnings of this promising technique in children.

The Population: Ethical Challenges in Vulnerable Patients

The pediatric population being considered for DBS is...
participate in decision-making. In the case of DBS, there are often asked to consider rather than a specific age. Beginning at age 7 years, competence is more often associated with prior experience rather than a specific age. Such decision-making spans not only the perioperative period, but also subsequent decision points such as device programming, further treatment, and potential revisions. While many of these principles apply to the conduct of DBS in children, several unique pediatric considerations should be recognized. Children may not be able to articulate their wishes due to intellectual immaturity or neurocognitive disability, leaving caregivers to make difficult decisions. In cases in which the child has not yet declared their interests, decisions are based on the child's presumed best interest rather than their right to autonomy and self-determination. Such decision-making spans not only the perioperative period, but also subsequent decision points such as device programming, further treatment, and potential revisions. Depending on the child's age and abilities at initial intervention, he or she may begin assuming a greater role in subsequent decision-making, an evolution that all stakeholders within the circle of care must anticipate. There is no accepted age at which children are deemed competent and capable of medical decision-making. Competence is often more associated with prior experience rather than a specific age. Beginning at age 7 years, children with a prior surgical history are often asked to participate in decision-making. In the case of DBS, there is usually no prior experience, and few such cases have been performed; therefore, advice and experience from family support groups with related experience are limited. Children with prior non-DBS surgical experience might participate better in consent discussions regarding general surgical risks, but would be unlikely to have added insight into the specifics of undergoing DBS. In some cases, there is a dilemma between early versus delayed intervention to allow the child to better participate in decision-making. A delay in treatment may result in undue harm from disease sequelae, such as musculoskeletal deformity from spastic conditions, worsening epilepsy, and so forth. Early intervention is most warranted for conditions with strong evidence such as primary DYT1 dystonia, but is less indicated where evidence is heterogeneous such as in secondary dystonia. The concept of “double jeopardy” may apply: First, these children suffer as a result of their disabilities, and second, lower priority is given to treatments that may improve their quality of life. A “relational” view of decision-making holds that treatment (and evidence for and against intervention) should be considered within the context of the child's subjective experience with the illness. Consultation with medical bioethicists may be prudent to maintain the strictest standards for patient care. The Procedure: Known and Unknown Risks of Surgical Innovation Although DBS for movement disorders in adults is an accepted treatment option, in children it still represents a surgical innovation, even for the most common indication, dystonia. As mentioned above, an innovation is defined as a procedure that significantly departs from standard or accepted practice and is characterized by evolving techniques, outcome measures, and patient selection. The extent to which a surgical innovation deviates from standard practice is directly related to the extent to which it requires ethical oversight and regulation. Before proposing DBS for a pediatric patient, the health team should recognize that the risks, and often the benefits, of the procedure are not well understood. Available data are limited to small case series with specific institutional protocols that are poorly generalizable to other centers. Risks of the procedure may be higher in children when that procedure is performed using protocols derived from adult DBS. For example, early case series on pediatric DBS have reported higher rates of infection ranging from 5% to 33%, as compared to approximately 5% with adult DBS. Since then, two series in which the pulse generator was implanted at a later date both reported an infection rate of 0%. Furthermore, DBS leads and batteries are sized for use in adults, and the risks of skin erosion and hardware fracture seem to be higher in children. An appreciation of the ways in which a standard procedure in adults (i.e., DBS) may be an innovative intervention in children is critical to determining the extent of ethical oversight required for its use in clinical and experimental indications.

Surgical innovations are also associated with evolving outcome measures. Current instruments used to measure the success of DBS treatment may not be ideal, even for the most widely studied indication, dystonia. Originally created to assess primary torsion dystonia in adults, the Burke-Fahn-Marsden Dystonia Rating Scale (BFMDRS) has been adopted for pediatric use and is now the most commonly reported measure of dystonia impairment. Intended for use in secondary dystonia, the Barry-Albright Dystonia Scale (BADS) is also frequently cited. These scales often fail to capture a child’s experience with illness (i.e., dystonia). For example, a recent study highlighting this showed that DBS improved individualized functional goals in the absence of significant changes in BFMDRS scores in children with secondary dystonia.

Despite the uncertainties introduced by evolving techniques, patient selection, and outcome measures, clinicians have an obligation to apply DBS in appropriately selected cases. Children with dystonia may benefit from surgical intervention, but the decision to proceed must be based on a thorough evaluation of the potential benefits and risks of the procedure.