What Is the Role of Developmental Disability in Patient Selection for Pediatric Solid Organ Transplantation?


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The National Organ Transplant Act stipulates that deceased donor organs should be justly and wisely allocated based on sound medical criteria. Allocation schemes are consistent across the country, and specific policies are publicly vetted. Patient selection criteria are largely in the hands of individual organ transplant programs, and consistent standards are less evident. This has been particularly apparent for patients with developmental disabilities (DDs). In response to concerns regarding the fairness of transplant evaluations for patients with DDs, we developed a transplant centerwide policy using a multidisciplinary, community-based approach. This publication details the particular policy of our center. All patients should receive individualized assessments using consistent standards; disability should be neither a relative nor an absolute contraindication to transplantation. External review can increase trust in the selection process. Patients in persistent vegetative states should not be listed for transplantation.

Abbreviations: AASLD, American Association for the Study of Liver Diseases; ADA, Americans with Disabilities Act of 1990; AST, American Society of Transplantation; BCH, Boston Children’s Hospital; CEC, Community Ethics Committee; DD, developmental disability; ID, intellectual disability; ISHLT, International Society for Heart and Lung Transplantation; MCS, minimally conscious state; PTC, Pediatric Transplant Center; PVS, persistent vegetative state; TLAC, transplant listing advisory committee; UNOS, United Network for Organ Sharing

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National solid organ allocation policies in the United States, starting with the National Organ Transplant Act of 1984, stipulate that organs should be justly and wisely allocated based on sound medical criteria. Nevertheless, individual transplant programs are given wide latitude in determining patient selection criteria, and the processes of decision-making regarding transplant eligibility for individual transplant programs neither have been the subject of national policy nor have been evaluated in a rigorous manner. Consequently, it is not surprising that there is marked variation with respect to how developmental disability (DD) is factored into candidacy determinations at different pediatric transplant centers (1).

Federal law (2) established in 2000 that deceased donor organs should be equitably allocated with consideration to sound medical judgment, best use, minimized waste and broad patient access. National organizations and the United Network for Organ Sharing (UNOS) ethics committee have applied these balancing mandates to DD by making general statements within consensus guidelines. Table 1 summarizes the official statements, which generally claim that DD ought not to be an absolute contraindication to transplantation and that patients should not be categorically excluded based on DD status. The International Society for Heart and Lung Transplantation guideline is distinguished by the fact that it specifies that DD may be regarded as a relative contraindication based on comparative lack of benefit.

The Pediatric Transplant Center (PTC) at Boston Children’s Hospital (BCH) became interested in developing a policy about the use of neurodevelopmental criteria in patient selection decisions, believing that individual transplant programs under the PTC should be practicing consistently in this regard. Institutional experience with several ethically challenging cases combined with well-publicized reports (3)
of children with DDs being declined for solid organ transplantation further established the necessity of practice using an adequately vetted policy. We formed a neurodevelopmental task force to review this topic and to develop a hospitalwide policy. For this viewpoint, we have defined critical terms, described the process used to develop our policy, set forth our policy, addressed the federal Americans with Disabilities Act of 1990 (ADA) and offered a pathway forward to address this important issue.

**Definition of Terms**

The task force defined particular terms, drawing on federal and expert consensus guidelines as appropriate authorities in this regard.

1. **Developmental disability**: A severe, chronic disability of an individual person that is attributable to a mental or physical impairment or combination of mental and physical impairments. Furthermore, the condition (i) is manifested before the person turns 22 years of age, (ii) is likely to continue indefinitely, (iii) results in substantial functional limitations, and (iv) reflects the person’s need for interdisciplinary services that are anticipated to be needed for an extended period of time (4). DDs include conditions such as cerebral palsy, intellectual disability, autism, epilepsy, deafness and blindness as well as global developmental delay not otherwise specified.

- **Minimally conscious state (MCS)**: A condition of severely altered consciousness in which minimal but definite behavioral evidence of self- or environmental awareness is demonstrated. Behaviors can include following simple commands, gestural yes/no responses, intelligible verbalization and purposeful behavior (5).

- **Persistent vegetative state (PVS)**: A condition of complete unawareness of the self and the environment accompanied by sleep–wake cycles with either

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**Table 1: Excerpts from official organization policy statements pertaining to potential recipients with developmental disabilities**

<table>
<thead>
<tr>
<th>Professional society (year)</th>
<th>Specific language</th>
<th>Notes</th>
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</thead>
<tbody>
<tr>
<td>ISHLT, heart transplantation (2006) (15)</td>
<td>“Mental retardation or dementia may be regarded as a relative contraindication to transplantation.”</td>
<td>Acknowledges variation in practice across different programs; does not define “mental retardation”</td>
</tr>
<tr>
<td>ISHLT, lung transplantation (2006) (16)</td>
<td>No specific mention of developmental disability</td>
<td>Absence of consistent and reliable social support system listed as an absolute contraindication</td>
</tr>
<tr>
<td>AST, renal transplantation clinical practice guidelines (2001) (17)</td>
<td>“A cognitive deficit should not automatically exclude a patient from transplantation.”</td>
<td>Notes that caregivers’ assessment of patients’ quality of life is important</td>
</tr>
<tr>
<td>AASLD, liver transplantation in adults (2013) (18)</td>
<td>No specific mention of developmental disability</td>
<td>Lack of adequate social support system listed as contraindication to liver transplantation</td>
</tr>
<tr>
<td>AASLD, liver transplantation in children (2014) (19)</td>
<td>“The presence of a severe intellectual or developmental disability has raised concerns of candidacy for liver transplantation. Those concerns center upon compliance with a rigorous and lifelong post-transplant management schedule, potential for increased risk for malignant or infectious complications related to genetic or physical disabilities, and assessment of quality of life. Unfortunately, data to address these concerns are very limited.”</td>
<td>Neurocognitive testing recommended to identify areas warranting early intervention but no recommendation made with regard to candidacy</td>
</tr>
<tr>
<td>UNOS ethics committee (2007) (20)</td>
<td>“Patients with disabilities should not be excluded from consideration for transplant solely by virtue of their disability.”</td>
<td>Notes that evaluation for transplant is a “local issue” and that teams should focus on whether a patient will garner “significant/meaningful medical benefit” from the transplant; “significant/meaningful” is not defined</td>
</tr>
<tr>
<td>American Medical Association (1995) (21)</td>
<td>“A major concern in making quality of life decisions is the possibility of discrimination against the disabled population.”</td>
<td>Notes that priority to those whose needs are greatest to prevent a very bad outcome protects all patients, including the disabled</td>
</tr>
</tbody>
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AASLD, American Association for the Study of Liver Diseases; AST, American Society of Transplantation; ISHLT, International Society for Heart and Lung Transplantation; UNOS, United Network for Organ Sharing.
complete or partial preservation of hypothalamic and brain stem autonomic function. The vegetative state is considered to be persistent when the diagnosis of irreversibility is established to a high degree of certainty, based on available predictive models. The Multi-Society Task Force on PVS, for example, considers 3 months after a nontraumatic injury to be persistent, whereas 12 months is used as a criterion for traumatic injuries (6).

Process and Justification

In 2012, the PTC convened an ad hoc task force to examine the role of DD in transplant evaluation and/or listing decisions. The task force was composed of transplant professionals from medicine, surgery, nursing, social work and psychology and included representation from all of the solid organ transplant programs. The task force also included representatives from the BCH ethics advisory committee.

Our task force used multiple methods to develop our policy, including literature review, ethical analysis, consultation with the Boston, Massachusetts–area Community Ethics Committee (CEC), repeated multidisciplinary discussions and legal review.

The task force determined that for purposes of listing for transplant, differences in organ or patient survival between DD and non-DD recipients should be considered meaningful only if they were substantial. The literature cited in Table 2 suggests that they are not. Importantly, this literature has to be understood as being limited in several regards. First, Wightman et al reported in 2014 the first large study comparing 3-year renal transplant recipient outcomes using UNOS data and acknowledged that cognitive developmental categories are nonstandardized and could vary from center to center or even from reporter to reporter. Second, retrospective case series could be biased toward reporting successful or unsuccessful outcomes. Third, all of the reports missed patients with DD who were not referred to transplant centers, who were not listed, or who remained on the waitlist. The literature is summarized as follows:

- Some patients who have DDs may have other medical issues that preclude them from receiving a transplant.
- If no other medical issues exist that would contraindicate transplant, patients with DDs have no worse organ and patient survival outcomes (in some cases, they have better outcomes) than other transplant recipients.
- Patients who have significant DDs must have strong social support systems in place to help facilitate successful posttransplant experiences.
- The extent of an individual patient’s later DD is often difficult to confidently prognosticate, especially in infants.

Our ethical analysis centered on the defensibility of provider-based assessment of benefit. It is routine for programs to exclude patients when anticipated survival benefit is poor. A child with DD should be declined because of irremediable comorbid conditions that negatively affect the odds of posttransplant survival in the same way a child without DD and similarly serious comorbid illness should be declined. Moreover, no child should be listed if the

<table>
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<tr>
<th>Study (year)</th>
<th>Type of delay/disability studied</th>
<th>Organ and patient group</th>
<th>Outcome</th>
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<tbody>
<tr>
<td>Wightman et al (2014) (23)</td>
<td>UNOS data set; defined categories “definite,” “probable,” and “no” DD</td>
<td>2292 pediatric kidney recipients between 2008 and 2011</td>
<td>332 patients definite DD, 117 probable DD; no significant difference in patient or organ survival compared with the non-DD group</td>
</tr>
<tr>
<td>Sharifian et al (2007) (24)</td>
<td>Bardet–Biedl syndrome</td>
<td>Five pediatric kidney recipients</td>
<td>No organ loss; one had poor renal function due to compliance</td>
</tr>
<tr>
<td>Samelson-Jones et al (2012) (25)</td>
<td>Mental retardation or comparable DDs</td>
<td>Five adult heart recipients</td>
<td>Four of five lived several years, one died due to declining health of caregiver</td>
</tr>
<tr>
<td>Benedetti et al (1998) (26)</td>
<td>Mental retardation (IQ &lt;70)</td>
<td>Eight kidney recipients (five adult, three teen)</td>
<td>1- and 5-year survival 100%; one died at 10 years from sepsis</td>
</tr>
<tr>
<td>Uemura (2005) (27)</td>
<td>Significant mental retardation</td>
<td>One combined liver/kidney</td>
<td>Excellent liver and kidney function 5 years after transplant</td>
</tr>
<tr>
<td>Galante et al (2010) (28)</td>
<td>DDs</td>
<td>16 kidney recipients (11 children, five adults)</td>
<td>Four patients died of infection; all survivors had functioning organs and 100% adherence</td>
</tr>
</tbody>
</table>

DD, developmental disability; UNOS, United Network for Organ Sharing.

procedure, overall, would cause him or her clear harm. This would be the case for patients with certain medical conditions (e.g., intractable seizures, acute intracranial bleed, severe combined immunodeficiency, severe clotting or bleeding disorder) that would be predicted to worsen because of the required surgery or immunosuppression.

Compared with the quantity-of-life benefit afforded by transplantation, the quality-of-life benefit is subjective; its value is substantially derived from what patients or parents report from their own experiences. There are well-documented discrepancies between clinician- and patient- or family-reported quality of life (7,8). On this basis, patients with DDs have historically encountered (9) and continue to encounter (1,10) unequal access to transplant. Given the documented vulnerability of patients with DDs to discrimination, it is morally troubling to compare the quality of different children’s lives for the purposes of excluding those whose lives are not considered good enough by some.

The task force concluded that a scarce resource should not be used to prolong the survival of people who very likely have no conscious experience. In its recommendations, it set an absolute threshold for quality-of-life benefit, and patients in a PVS fall below this threshold. Admittedly, it is often complicated to determine whether a patient is in a PVS (11); in general, disorders of consciousness have become more complex and interrelated (12). Careful collaboration with expert neurologists is a critical component of this work.

The task force identified a particular value judgment that drove the recommendation to set an absolute threshold, as noted above, namely, that biological survival in a PVS does not constitute an adequate benefit for solid organ transplantation. It also recognized that deceased donor organs are a community resource and that, in large part, community values ought to be reflected in the balancing of equity and utility that underlies the systems of organ allocation and patient selection.

For these reasons, the task force sought assistance from the Boston-area CEC, a volunteer-based organization with representation from diverse socioeconomic, cultural, religious and educational backgrounds. The CEC provides independent reports and opinions on topics brought forward by members of the Harvard Ethics Leadership Group—representatives of the various ethics services within the Harvard University teaching hospitals—and other groups. Task force representatives drafted detailed questions for the CEC and met with the CEC in 2013. The CEC report from February 2014 based on the committee’s deliberations is publicly available online (http://www.medicaethicsandme.org/p/publications-of-cec.html). The CEC used an absolute lack of benefit argument to claim that it is ethically justifiable, or even obligatory, to exclude patients in a PVS from receiving transplants.

The CEC found no justification for excluding children or adults from transplantation solely on the basis of DD. Moreover, in its review, a clear difference was noted between the transparency and publicly vetted organ allocation procedures for people on the waitlist compared with the more opaque processes used by individual transplant teams to determine whether patients are candidates for transplantation. Based on the CEC’s considered opinion, we adopted an approach that increases transparency and accountability with respect to candidacy decisions for patients with DDs. We recommend formation of a transplant listing advisory committee (TLAC) with the purpose of ensuring that institutional transplant selection criteria are fair and nondiscriminatory and that patients declined for evaluation or listing at BCH have been granted a fair and unbiased review, particularly with respect to characteristics that identify vulnerable or protected classes of people, such as people with disabilities who may be protected under the ADA. This will be accomplished through periodic review of selection criteria and formalized statistical comparison of patients accepted and declined for organ transplantation to protect against otherwise unintentional biases in transplant selection decisions.

Our task force met periodically over the course of ≈2 years. Core task force members presented preliminary recommendations to a multidisciplinary group representing all of the solid organ transplant teams and the PTC leadership. This iterative work had the purpose of building adequate consensus around a very difficult topic, and we recognized that unappreciated or unrecognized serious dissent could jeopardize adoption of our recommendations by each of the transplant programs.

Our Policy

1. Patients should not be excluded from consideration for solid organ transplant solely on the basis of DD.
2. Organ and patient survival are the most legitimate outcomes to consider when determining whether a patient should be evaluated or listed for transplantation. If minimum thresholds are established in particular programs, these thresholds should not be different for patients with DD.
3. Biological survival is not a sufficient goal for transplantation. Survival for the purpose of having continued conscious experience is a minimum goal of transplantation.
4. Children in an MCS represent a challenging group to evaluate because it can be difficult to characterize the significance of the particular behaviors consistent with awareness.
5. A patient suspected of being in an MCS or a PVS should be evaluated by a neurologist with expertise in disorders of consciousness. The evaluation should
include an opinion about the nature and stability of the condition and an explanation for the meaning of any signs of consciousness that can be understood by any competent parent or guardian. This will help the transplant team and the family determine how beneficial transplant might be.

6. If the patient is found to be in a PVS by the neurologist, solid organ transplantation should not be offered.

7. Parents or guardians who desire transplant for their children or dependents with DD should be offered in-person transplant evaluations (as opposed to medical record reviews that do not involve a face-to-face meeting).

8. Evaluation for organ transplantation is a collaborative process between the organ transplant team and the patient, parents and/or legal guardians.

9. Provided that there are no medically significant comorbidities that unduly limit posttransplant survival, broad deference should be given to patients, parents and guardians when balancing the overall benefits and burdens of transplantation.

10. Deference to patients, parents and guardians is not complete. It is impermissible for transplantation to be offered to patients that will, on balance, be clearly harmed by the procedure, whether or not they have DD.

11. Families and legal guardians may decide that their children or dependents may not garner enough benefit to warrant the immediate and lifelong burdens of transplantation. Transplant teams have the profound responsibility of carefully educating families about the risks and benefits of transplantation and of collaboratively making the best decision possible, even when this means not pursuing transplantation.

12. Families and guardians deserve to be told, in person and in writing, the reasons for not being listed for transplantation.

13. Alternatives to transplantation ought to be offered in a caring and collaborative manner, including ongoing supportive, spiritual, and palliative care.

14. A TLAC will be established with representation from within and outside the hospital with the purpose of ensuring that listing criteria are fair and nondiscriminatory and that patients declined for evaluation or listing have been considered fairly and without bias against vulnerable and protected classes of people. The TLAC membership should include community members; representatives from nursing, social work, psychology, ethics, legal, surgery and medicine; and a statistician expert in the analysis of demographic variables in medicine. PTC leadership should appoint a chairperson of the TLAC, whose responsibility will include reporting findings to PTC leadership on a regular basis. Bylaws governing membership, authority, data gathering and analysis, documentation, and confidentiality ought to be drafted by the TLAC chairperson and approved by PTC leadership.

15. If the transplant team and parents or guardians cannot come to a mutually satisfactory decision, it is the responsibility of the transplant team to help interested families identify organ transplant teams at other institutions that could offer a second opinion or evaluation.

The ADA prohibits discrimination against patients with disabilities. As a transplant center that participates in the federal Medicare and Medicaid programs, we are bound by the ADA, and thus our policy must comport well with the requirements and complex interpretation of the law. In conjunction with the above policy, our review with hospital counsel helped us generate the following statement:

No patient will be considered ineligible to receive an organ transplant solely because of mental or physical disability. Each patient will undergo an individualized evaluation to determine whether he or she meets criteria for listing. These medical and non-medical criteria are used to select patients who are least likely to have limited post-transplant survival. For instance, patients must have adequate non-transplant organ function to safely maintain vital functions and properly metabolize medications post-operatively. So too, patients must have an adequate support system to ensure compliance with the post-transplant regimen necessary for organ survival. If patients’ medical or non-medical circumstances cannot be reasonably remediated and predict poor organ survival, they may be found ineligible for listing.

Going Forward

Through this process, our transplant programs have developed a consistent approach to evaluation and selection of patients with DDs; however, it is arguably unjust that candidacy with respect to DD varies substantially from center to center. Legal cases brought forward on violations of the ADA may not further the important project of increasing transparency, consistency and honest public dialogue with respect to formal and informal listing criteria. States have responded to high-profile cases by passing legislation barring exclusion from transplantation on the basis of DD (13,14). It is not clear that these laws will improve the transplant experience of patients with DDs. We hope to continue a national conversation about this topic in a forum that fairly considers the perspectives of key stakeholders while comporting with the modern ethical (and legal) imperative to equally value the lives of those with disabilities. We encourage the transplantation professional societies and the UNOS ethics committee to sustain the conversation within the transplant community and solicit opinions from the public.
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