Should Developmentally Delayed Children Be Listed For Solid Organ Transplants?

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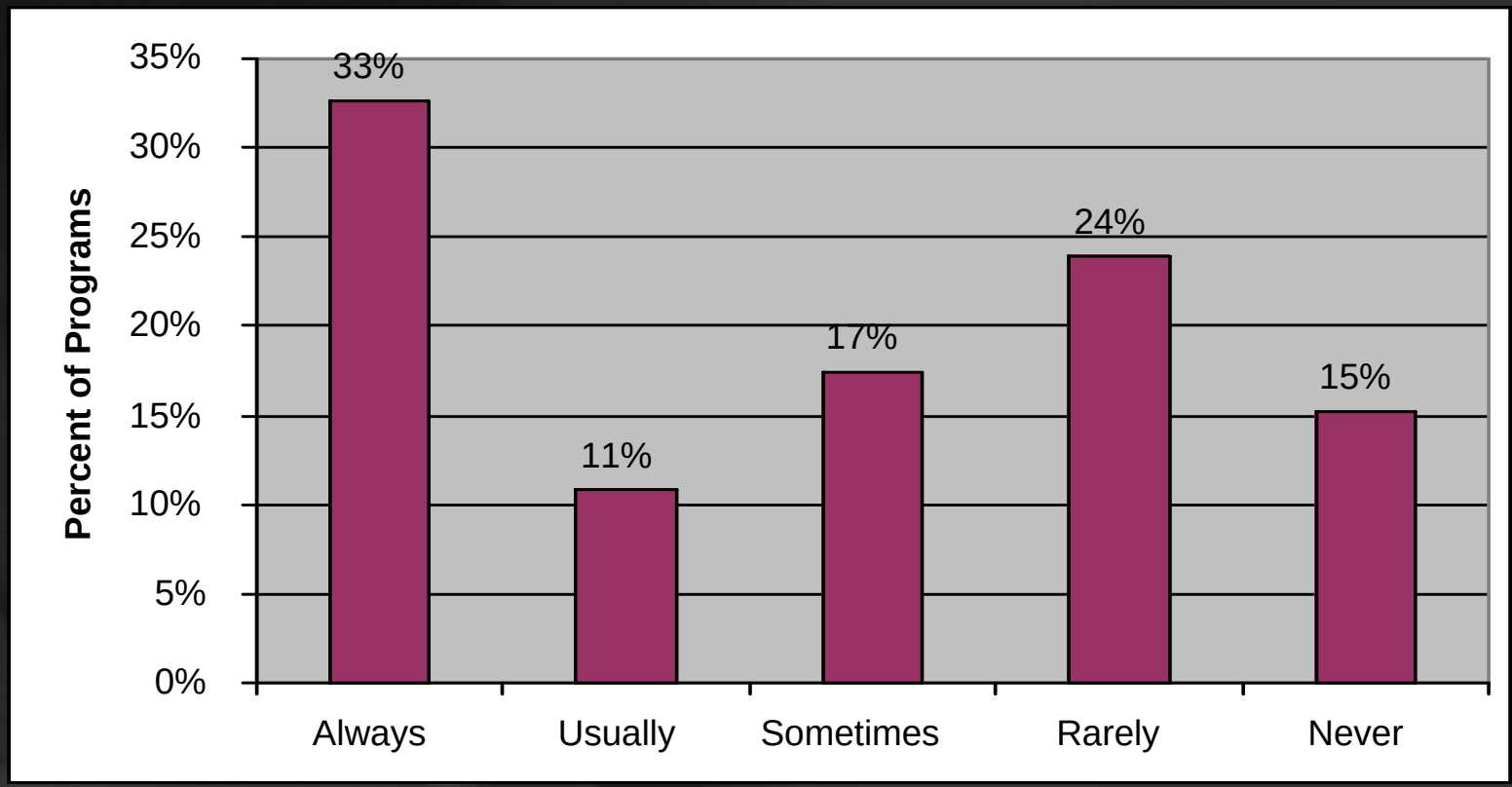
Wide Range Of Possible Cases

- Anencephalic
- Profoundly developmentally impaired (no mobility, no measurable IQ)
- High level of functioning, capable of communicating, attending school

Survey (Chris Richards, et al)

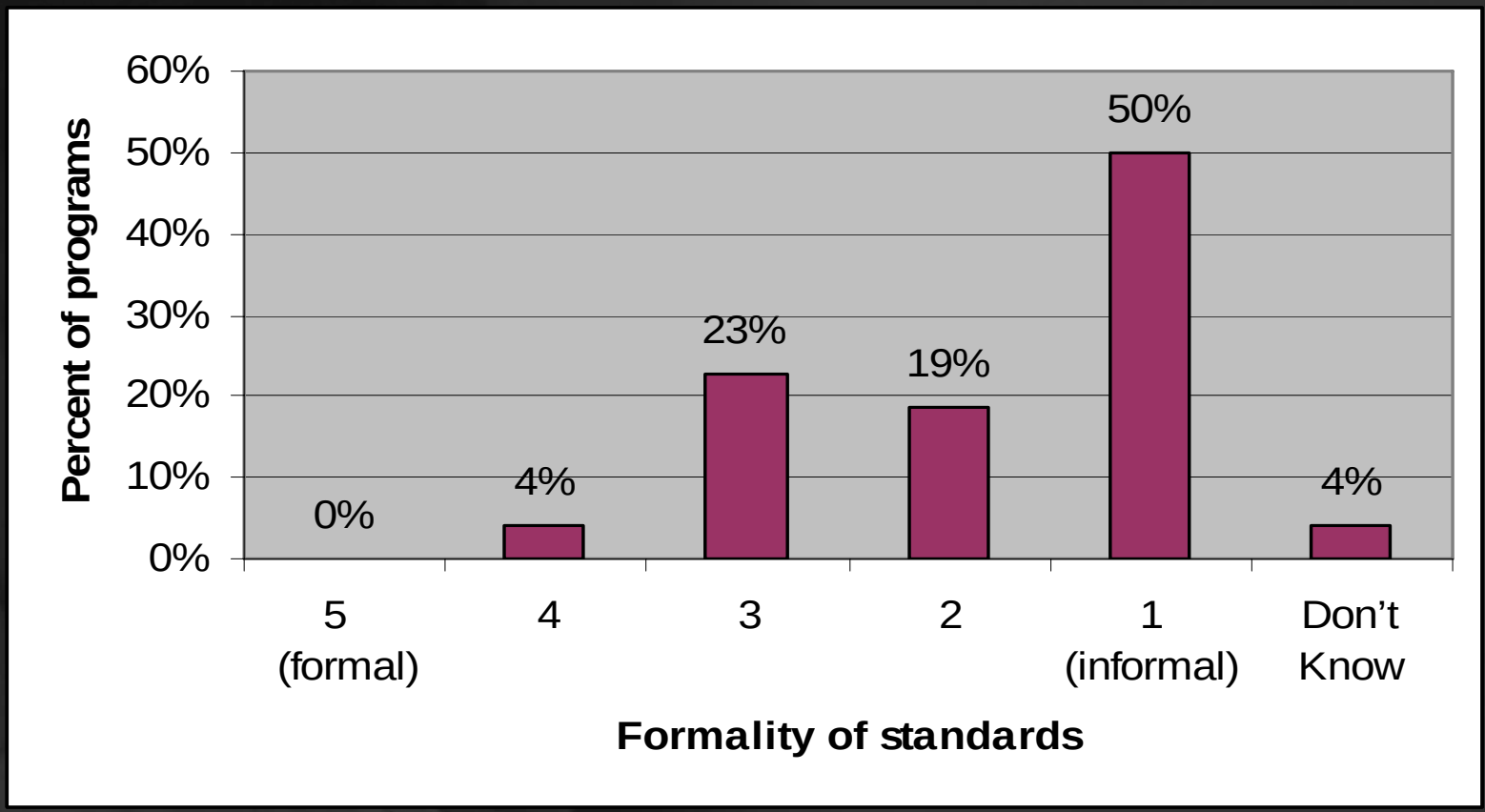
- Most active kidney, liver, and heart transplant programs in the U.S.
- Surveyed medical directors, surgical directors, or transplant coordinators (listed contact persons on web sites)
- 88 programs at 45 hospitals surveyed
- 50 responses (57% or programs)

Percent of programs reporting how often neurodevelopmental status is considered in the transplant listing decision.



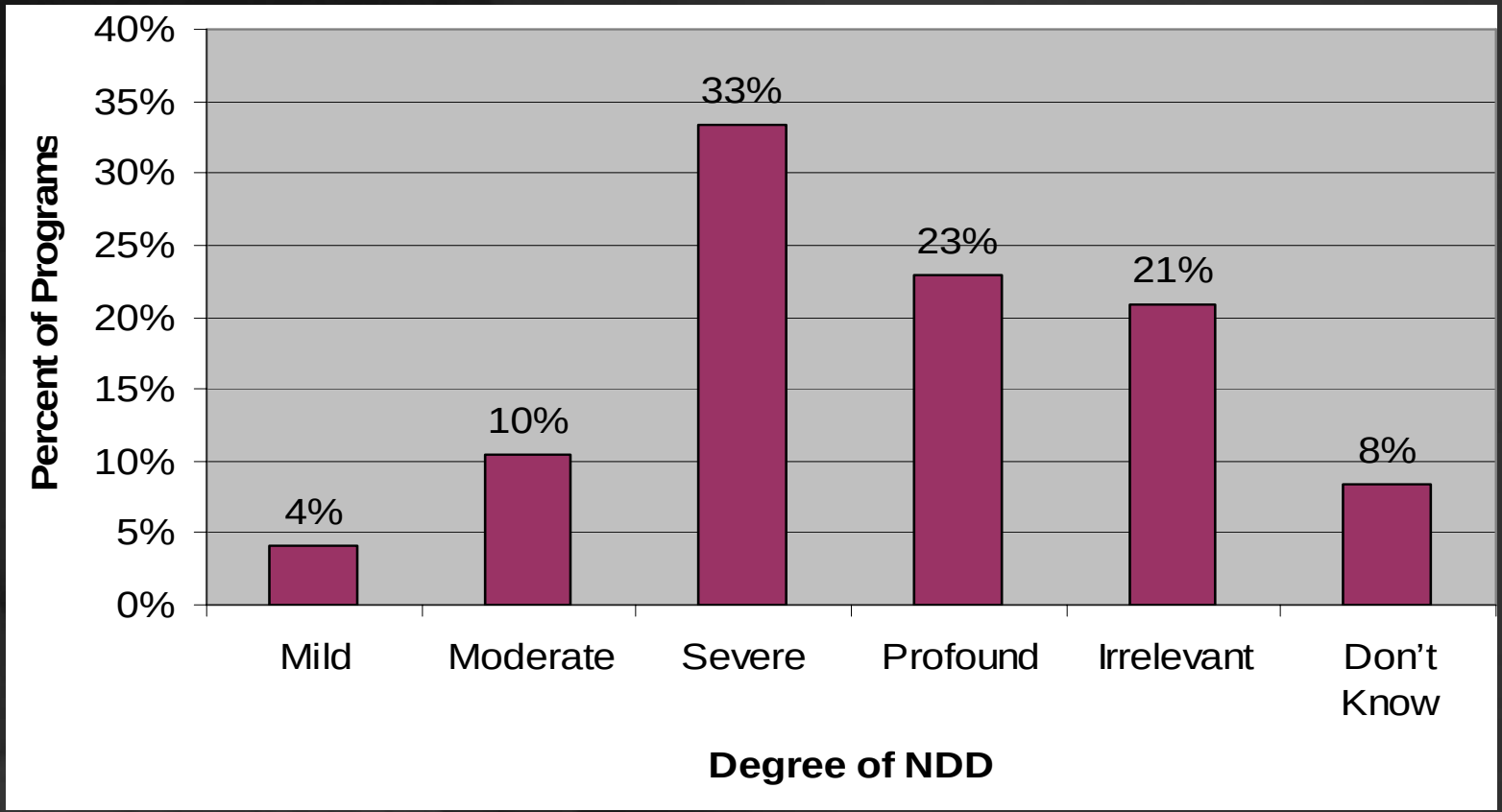
Percent of programs describing the formality of standards by which NDD is used in the listing decision.

1 = Informal, implicit, or unstated standards; 5 = Formal, explicit, or uniform protocols.



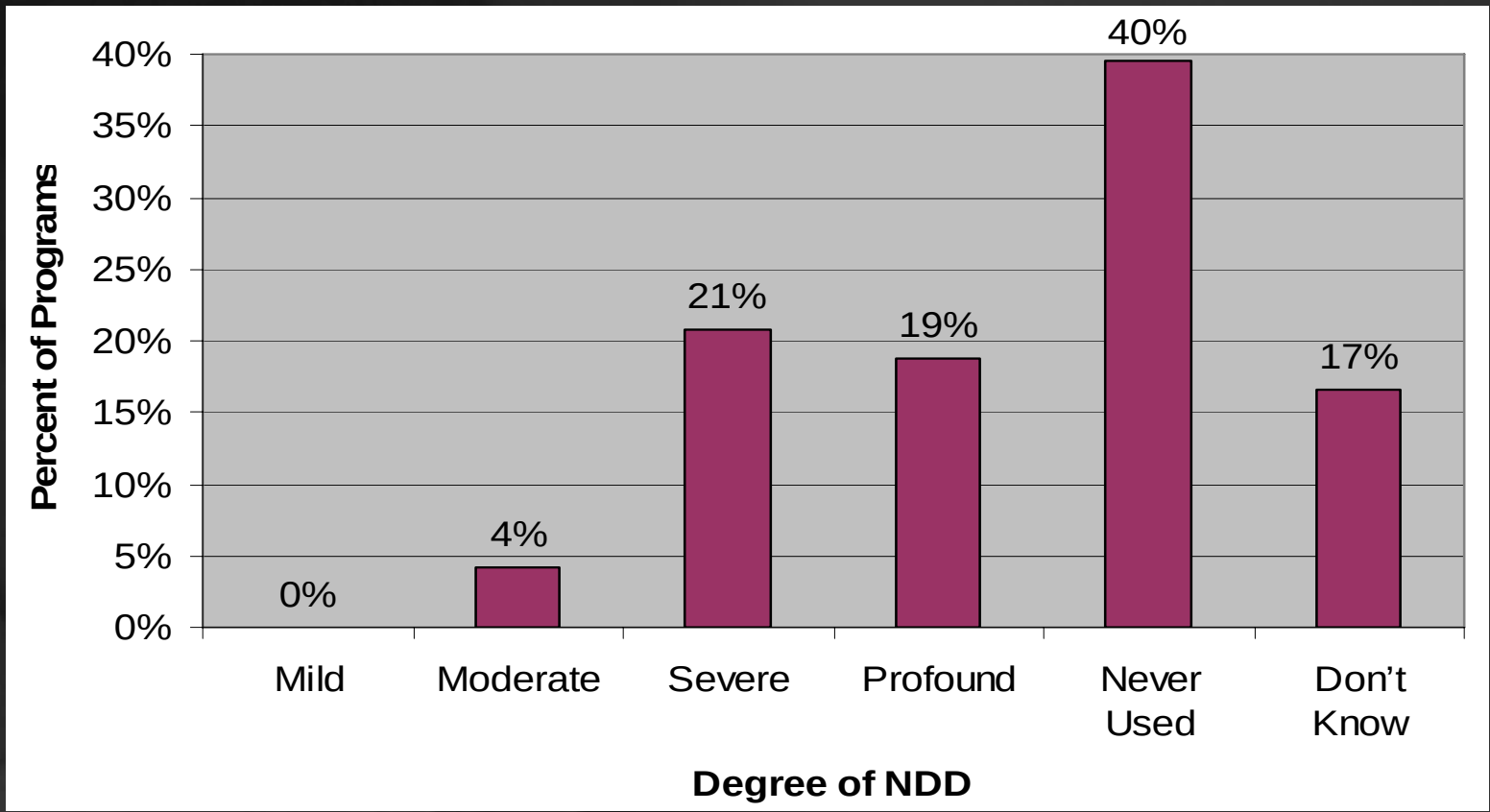
Percent of programs that consider NDD by itself as a relative contraindication to listing and, if so, at what degree of NDD.

"Mild" delay - IQ 50-70, "moderate" delay - IQ 35-50, "severe" delay – IQ 20-34, "profound" - IQ < 20. Key: NDD – Neurodevelopmental delay. "Irrelevant" – NDD is irrelevant to listing decision



Percent of programs that consider NDD by itself as an absolute contraindication to listing and, if so, at what degree of NDD.

"Mild" delay - IQ 50-70, "moderate" delay - IQ 35-50, "severe" delay – IQ 20-34, "profound" - IQ < 20. Key: NDD – Neurodevelopmental delay.



Other Findings

- 38% of programs said they had rejected a patient that would have been listed for transplantation had it not been for pts neurodevelopmental status
- Response to hypothetical scenarios further reflected fact that teams often would refuse to list patients based on cognitive status (even in cases of moderate delay) and there is no consistency in the field
- Consistent with other surveys of adults or mixed adult and pediatric populations

Summary Of Outcome: Renal Transplantation Of Pts With MR

Martens, Jones, and Reiss, "Organ transplantation, organ donation and mental retardation, Pediatric Transplantation, 2006: 10: 658-664

Table 2. One- and three-yr patient survival rates following renal transplantation in individuals with mental retardation

Transplant center	Age at transplant (yr)	1-yr patient survival*	3-yr patient survival†,‡
University of Illinois, Chicago, IL (24); E. Benedetti (personal communication, October 10, 2005)	16, 17, 17, 22, 23, 26, 31, 45	8/8	8/8
Children's Hospital, Stockholm, Sweden (25); M. Englund (personal communication, October 18, 2005; January 30, 2006)	1, 1, 5, 5, 7, 7, 9, 11, 12, 15 (three cases unknown)	13/13	12/12
North American Pediatric Renal Transplant Cooperative Study (26)	6–12 (8 cases), 13–17 (5 cases) >17 (1 case)	Information not available	11/14
The Hospital for Sick Children, Toronto, Canada (27)	7, 10	2/2	1/1
Massachusetts General Hospital, Boston, MA (28); O.S. Surman (personal communication, September 6, 2005)	36, adult (age not specified)	2/2	2/2
St Christopher's Hospital for Children, Philadelphia, PA (29); J.H. Baluarte (personal communication, September 6, 2005; January 12, 2006)	14, 20	2/2	0/1
Children's Hospital of Philadelphia, Philadelphia, PA; J.H. Baluarte (personal communication, September 6, 2005)	10, 11, 13, 17, 19, 34	6/6	4/4
Total across all studies		33/33	38/42

*Numerator = number of patients who survived 1 yr; denominator = number of patients transplanted.
†Numerator = number of patients who survived 3 yr; denominator = number of patients transplanted.
‡The number in the denominator may differ from the denominator in the 1-yr patient survival column because 3 yr have not elapsed since all cases were transplanted.

Case #1

- 10 year old boy with Alagille syndrome
- Liver failure, other organ system problems
- Abnormal growth factors
- Fall when young resulted in intracranial hemorrhage--profound neurodevelopmental delay

Case #1

- Pts mother is knowledgeable and has sought to have her son listed for transplant at three centers (each declined)
- Mother is hoping both to prolong pts life and to address puritis and thereby improve quality of life
- Prior development reports: wheelchair bound quadriplegic, with severe MR--no assignable IQ

Case #1--Assessment

- Parent Interview--Child recognizes family, requests action through eye gaze, verbalizes
- Child observation over time--6 week period both in and outpatient--videotape of home made by mother that captures requests for action, greeting, etc.

Case #1--Assessment

- Findings of child observation--no differentiation re communicative intent, no responsive facial expressions, no change in behavior, no recognition, no greeting, no negation
- Report findings to parent--went through video with mother to show that pt was unresponsive

Case #1

- Multiple organ system problems would complicate course for transplant, but
- Pts with Alagille are often listed for transplant
- Would have listed if not for IH and secondary developmental delay
- Should they list?

Case #2

- 10 year old girl with Wolf Hirschhorm syndrome and focal glomerulonephritis
- Profound retardation (no measurable IQ), seizures, poor muscle development
- Wheel chair bound patient had end stage renal disease
- Parents want dialysis and a transplant

Case #2--Assessment

- Parent interview--child recognizes family and request for action and objects through expressions, gestures and vocalizations; infrequent direct eye contact, but contact through mirrored reflection
- Child Observation--3 month period initially in patient and then clinic visit

Case #2--Assessment

- Findings--demonstrated differentiation of communicative intention, including greeting, negation, preference for parents, request for action; reciprocal activities
- Report findings to parents
- Should we offer dialysis to this patient?
- Should we list for a kidney transplant?

Case #3

- 13 year old boy with alpha-1 antitrypsin deficiency
- the child shows scleral icterus and varicosities over the umbilicus with lab values revealing an INR of 2.1 and bilirubin of 5.0 mg/dL
- The patient has no other complications (no infections, no renal disease, and no malnutrition)

Case #3

- The patient also has Fragile X Syndrome and has an IQ of 50
- The patient lives at a group home and, although attending school part-time, is at a second grade level and will likely not progress beyond that point.
- The child's legal guardians now are looking to the physicians to make decisions about continuing medical treatment and perhaps listing this patient as a transplant candidate

Consensus

- Split on Case #1
- (Largely) Yes on Case #2
- Clearly Yes on Case #3
- Clearly No on Anencephalic

Clash Of Values

- Justice--obligation to treat pts fairly, not to discriminate on basis of morally irrelevant characteristics
- Stewardship--obligation to make good use of a very scarce resource--for each organ, more than one pt. who would benefit from it, therefore obligation to maximize benefit

Consensus Conference

- Peds ethicists
- Transplant ethicists
- Parent of child with NDD
- Reps from Stanford liver, kidney, heart teams
- Transplant surgeons
- LPCH ethics committee representatives

Preliminary Consensus

- Well established (data driven) medical indicators associated with poor outcomes (including adherence) should be taken into account in listing decisions
- It is appropriate to consider the benefits and burdens of transplantation in deciding whether to list a child
- Parental perspective on benefit should be privileged

Preliminary Consensus

- All children entitled to consideration for transplantation (evaluation) if parents want it
- All decisions made on cases by case basis
- Developmental assessment by a skilled specialist should always be part of evaluation process
- Developmental specialist should participate in listing decisions with team

Preliminary Consensus

- Permanently unconscious patients should be listed for transplant
- Mild to Moderate NDD is not by itself a contraindication for transplant
- Paliative care should always be offered as an option where there is some question of the benefit/burden ratio of transplant

Preliminary Consensus

- There should be transparency in process of decision making by the program
- Better data on outcomes for NDD pts should be kept
- Appeal process should be set up to address conflicts between parents and listing team (e.g., to a board that includes relevant experts, ethicists, parental representatives with NDD)

Contributors

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LPCH ethics committee