

Intellectual Disability and Pediatric Lung Transplantation

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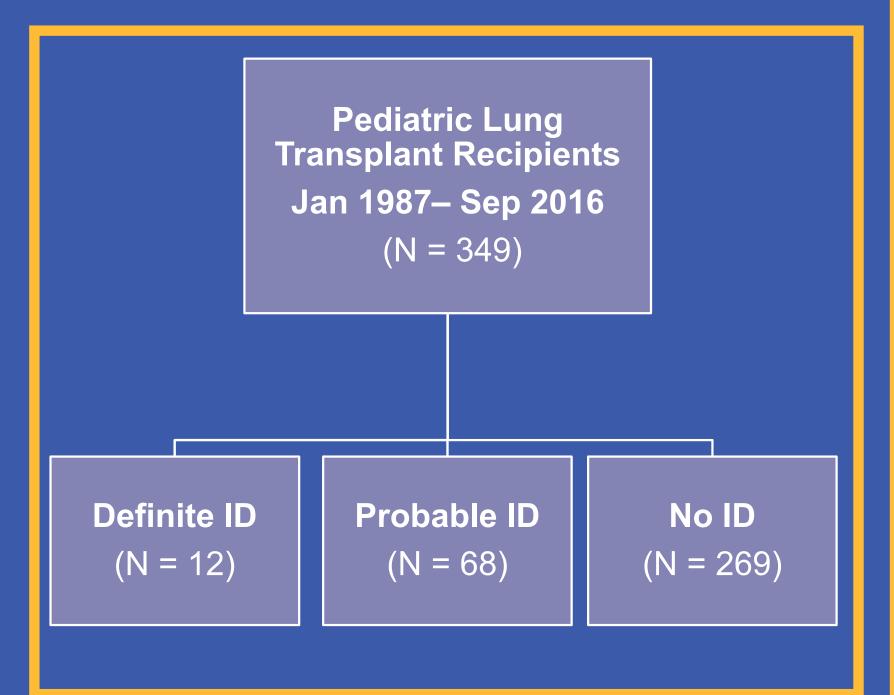


Background

- Pediatric lung transplantation is a rare procedure, limited by a shortage of available donor lungs.
- Transplantation in children with intellectual disability (ID) purports an ethical dilemma due to risk of medication non-compliance and potentially worse outcomes.
- We sought to evaluate outcomes of lung transplantation in the pediatric population, stratified by presence of intellectual disability.

Methods

- Retrospective analysis of pediatric lung transplant recipients performed from the UNOS database between Jan 1987 – Sept 2016.
- Patients stratified into definite, probable and no intellectual disability based on academic progress and standing.
- Survival analysis was performed via Kaplan-Meier method & censored at 5y.
- Multivariable Cox hazard models were adjusted for age, diagnosis, procedure, ischemic time, waiting list time, preoperative life support, donor age, and lung allocation score.

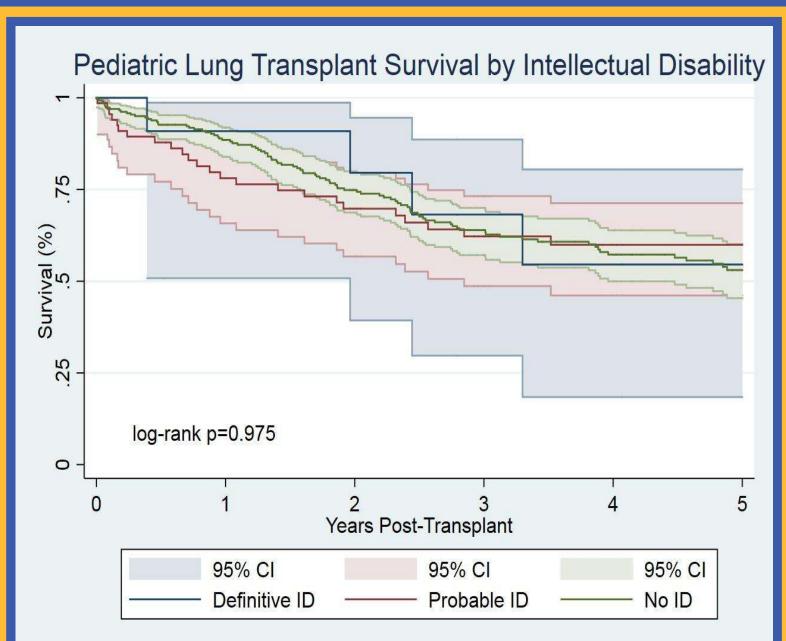


Results

Variables	Definite ID (N=12)	Probable ID (N=68)	No ID (N=269)	p-value
Recipient Age (yrs)	4.7 ± 6.3	8.3 ± 6.3	12.9 ± 4.4	<0.001
Male Gender	41.7%	45.6%	40.9%	0.782
Caucasian Race	66.7%	54.4%	74.0%	0.028
Lung Allocation Score	37 ± 18	37 ± 17	43 ± 19	0.002
Diagnosis Category				<0.001
A - COPD/Obstructive	16.7%	0.0%	1.5%	
B – Primary Pulmonary HTN	25.0%	20.6%	12.3%	
C – Cystic Fibrosis	8.3%	38.2%	57.6%	
D – IPF/Restrictive	16.7%	16.2%	8.2%	
Other	33.3%	25.0%	20.5%	
Diabetes Mellitus	8.3%	13.2%	26.8%	0.028
Cytomegalovirus Status	8.3%	1.5%	3.0%	0.412
Preoperative Dialysis	0.0%	1.5%	0.0%	0.126
Mean PA pressure (mmHg)	48 ± 30	41 ± 17	38 ± 21	0.446
PCW pressure (mmHg)	10 ± 3	10 ± 4	11 ± 4	0.920
Waiting Time (days)	112 ± 126	222 ± 334	208 ± 386	0.517
Single Lung Transplantation				
Donor Age (yrs)	8 ± 15	9 ± 10	16 ± 12	<0.001
Ischemic Time (hrs)	5.8 ± 1.3	5.7 ± 1.7	5.6 ± 1.4	0.523
Preoperative Ventilation	50.0%	26.5%	13.0%	<0.001
Preoperative ECMO	8.3%	2.9%	5.6%	0.598

Values presented as mean ± standard deviation or percent of population.

COPD = chronic obstructive pulmonary disease; PAP = pulmonary artery pressure; PCWP = pulmonary capillary wedge pressure; VAD = ventricular assist device; UNOS = United Network for Organ Sharing.



5-year Survival Curves (p=0.975)

- Definite ID: 54.6%
- Probable ID: 59.9%
- No ID: 53.0%

Cox Regression:

- Definitive ID: 1.50 [0.52-4.30], p=0.455
- Probable ID: 1.10 [0.67-1.81], p=0.712

Variables	Definite ID (N=12)	Probable ID (N=68)	No ID (N=269)
30-day Mortality	0 (0%)	1 (1.5%)	10 (3.7%)
Acute Rejection at 1 year	28.6%	20.8%	20.4%
Causes of Death			
Graft Failure	0.0%	10.7%	7.7%
Rejection	33.3%	21.4%	28.9%
Infection	0.0%	28.6%	7.7%
CV	0.0%	3.6%	5.8%
Malignancy	0.0%	0.0%	1.0%
Other	66.6%	35.7%	49.0%

Post-Transplant Outcomes/Complications

Thirty-Day Mortality: p=0.521
Acute Rejection: p=0.870
COD: p=0.254

Discussion

- Patients with evidence of intellectual disability were younger, had lower LAS, less diabetes, less cystic fibrosis diagnosis, and more preoperative life support.
- Despite the low incidence of pediatric lung transplantation, short term outcomes and in-hospital mortality is low across all ID groups.
- 5-year survival rates between ID groups were non-different with no difference in the adjusted mortality risk.
- Overall, results suggest that with appropriate patient selection, ID is not a contraindication to transplantation and compliance rates are likely similar.

Limitations:

- Retrospective design.
- Small number of pediatric lung transplants limits statistical power of analyses.
- Variation in treatment practices over time period may confound outcomes.

Conclusions

Pediatric lung transplantation can be safely performed in highly-selected patients with intellectual disability with equivalent short- and long-term outcomes. Intellectual disability should not be considered an absolute contraindication to transplantation.

Further studies are warranted to identify appropriate candidates for pediatric lung transplantation.

Disclosures

No authors for this presentation have relevant financial interests to disclose.