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OUTCOMES IN CHILDREN WITH INTESTINAL FAILURE FOLLOWING LISTING FOR INTESTINAL TRANSPLANT

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Abstract

Purpose—The purpose of this study was to describe the population of pediatric patients waiting for intestinal transplant and to evaluate the risk of death or transplant by specific disease states.

Methods—We studied the United Network for Organ Sharing (UNOS) database (1/1/1991–5/16/08) for patients ≤21years-old at first listing for intestinal transplant and examined their age, sex, weight, and diagnoses. Time to list removal was summarized with cumulative incidence curves. Multinomial logistic regression was used to compare relative risk ratios for removal from the list for transplant, death or other reasons.

Results—We identified 1,712 children listed for intestinal transplant (57% male, 51% < 1 yr, weight 8.1kg (IQR 6.1–14.1) at listing). Median age and weight at transplant (N=852) were 1yr (IQR 1–5) and 10kg (IQR 6.5–16.3). Regression analysis demonstrated significant differences in outcomes among disease conditions ($p < 0.001$). Compared to the gastroschisis group, the relative risk ratio for death versus transplant was higher in the NEC group ($p=0.015$), lower in the SGS group ($p=0.001$) and not different in the volvulus group ($p=0.94$) after adjustment for weight and sex.

Conclusions—We conclude that the relative risk of transplant versus death varies significantly by the disease condition of the patient.

Keywords

Intestinal Failure; Intestinal Transplant; Outcomes; Gastroschisis; Necrotizing Enterocolitis; Volvulus; Short Gut Syndrome

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The incidence of pediatric intestinal failure is increasing. There are a variety of disease conditions that lead to intestinal failure. The use of intestinal transplant to treat children with intestinal failure is becoming more common. The outcomes of children with intestinal failure while on the wait list for an intestinal transplant by disease state are unknown.

Introduction/Background

Intestinal transplant for the management of pediatric patients with intestinal failure is becoming more common. Since its conception in the 1960s, there has been considerable growth in the utilization of intestinal transplant; accelerated after the development of cyclosporine and tacrolimus in the 1980s. The Organ Procurement and Transplantation Network (OPTN, optn.org) database reported an increase in the total number of procedures for patients ≤17years-old, performed annually, from as few as 3 procedures in 1990 to as many as 96 procedures in 2006. The increasing role of transplant in the management of intestinal failure is largely due to the improved survival that has been reported comparable to lung transplantion[1] and approaching liver transplantion2.

The reported median wait time for intestinal transplant is 300 days, with the wait-list mortality as high as 300 to 600 per 1000 patient-years at risk[3]. Given the long wait time and high mortality, Medicare has mandated that intestinal transplant only be utilized in patients with intestinal failure after total parenteral nutrition (TPN) therapy has failed. The criteria for TPN failure include impending or overt liver failure, thrombosis of central veins (loss of access), frequent central line-related sepsis, and frequent, severe dehydration[4]. Yet, despite the long wait times for scarce organs, in some centers intestinal transplant has been accepted as a viable treatment for patients with intestinal failure.

Intestinal failure results from either an inability to absorb fluids or nutrients due to inadequate function or from the lack of absorptive surface area secondary to massive bowel resection or maldevelopment. The most common diagnoses responsible for intestinal failure include necrotizing enterocolitis (NEC), gastroschisis, volvulus and atresia. Despite the increasing use of intestinal transplant in children, and despite a decade of intestinal transplant experience, little is known about the impact of these underlying disease states on outcomes[5]. The objectives of our study are to describe the population of pediatric intestinal failure patients waiting for intestinal transplant, examine their outcomes, and evaluate the risk of death or transplant by specific disease states.

Methods

Database & Patients

The United Network of Organ Sharing (UNOS) is charged by the Human Resources and Services Administration (HRSA) to maintain the United States (US) transplantation network. A prospectively collected database of transplant patients called the Standard Analysis File (SAF) tracks pre-transplant, transplant and follow-up information on these patients and is maintained by the Scientific Registry of Transplant Recipients (SRTR). The database contains information on all intestinal transplants performed in the US and reported to the OPTN since October 1, 1987. This data is publicly available to any citizen requesting the information from UNOS. As the data in the SRTR file are de-identified, the University of Washington Human Subjects Division granted a Certificate of Exemption (HSD#34595) for our work.

The database contains unique entries that represent a listing event. An individual patient may have more than one entry within the database. For example, individuals may be listed simultaneously at two centers to maximize their opportunity to receive a graft, and patients that have received an initial graft may suffer graft failure and require another listing. In both situations, patients may have more than one entry in the database, but all entries would contain the same, unique identification number.

Based on OPTN data of May 16, 2008, we identified 2,882 cases of listing for intestinal transplantation. Of these, 1,977 listings were of subjects ≤ 21 years-old at listing representing

1,744 individual patients. Patients listed 3 or more times (29 patients and 93 listings) were excluded from the study as an analysis of this small `outlier' population may skew our analysis. The remaining 1,712 patients listed once or twice represent the study population for our analysis of intestinal transplant. (Figure 1)

Entries where date data were chronologically improbable, such as cases when transplant date preceded the listing date, we designated the transplant date as the listing date (6 cases). Instances of multiple listing (as described above) were reconciled (198 groups). When a subject had multiple or overlapping listings at two or more centers, the earliest listing date was chosen as the representative listing date. When subjects required re-listing, follow-up data was updated to reflect re-transplants, deaths or those patients still waiting on the list.

Patients less than one year of age were categorized as <1 years-old. Weights used are in kilograms and are specific to listing or transplant. Patients with multi-organ transplants such as liver and pancreas were included in the study.

Diagnostic Definitions

The underlying disease condition leading to intestinal failure was determined using the most specific entry among all diagnosis categories (primary, secondary, and text) provided within the UNOS database. In certain cases where an `other/unknown' diagnosis was indicated under a primary diagnosis, or if a diagnosis was missing, either the secondary or text diagnosis was utilized as the transplant diagnosis, if reasonably applicable. In situations where both the secondary and text diagnosis was not compatible with any of the above categories, the diagnosis was left as `other'. Data for diagnosis was ascertained from `at discharge' if available; if not, then `at transplant', and if not, then `at registration'.

A comparison of the diagnoses provided in the OPTN database and our grouping of diagnostic categories for this study is provided (Table 1). In the OPTN database, short gut syndrome (SGS) is a large category heading encompassing gastroschisis, atresia, NEC, volvulus, as well as shortened gut due to resection, and other unspecified reasons. We were able to identify patients with NEC, gastroschisis and volvulus separately for our analysis. The study category of other short gut syndrome (SGS) therefore includes the remaining OPTN-defined patients; mass resection of bowel due to Crohn's disease, tumor, arterial or venous thrombosis, atresia and other, unspecified reasons. Similarly, the category of functional bowel problems comprises a large group including Hirschprung's disease, and the latter diagnosis was separated into its own category for analysis. The other functional bowel problems were grouped as one category including neuronal intestinal dysplasia, neuropathic or myopathic pseudo-obstruction, protein losing enteropathies, microvillous inclusion disease, and other or unspecified conditions.

Statistical Analysis

Categorical data are summarized with percentages. Numeric data are summarized with medians and interquartile range statistics. Comparisons between transplanted and non-transplanted patients were made using the two-group mean comparison t-test. Analyses were formed using STATA version 10.0 (STATA corporation, College Station, Texas). All p-values are two-sided and all confidence intervals presented have a confidence level of 95%.

Cumulative incidence curves

We utilized cumulative incidence curves to describe the time from listing for intestinal transplant to the time of removal from the list. Reasons for removal from the list were categorized into three groups. The first group represents death or poor condition and encompasses the database categories of; `medically unsuitable, died, condition worsened/too sick for transplant and death during the procedure'. The second group represents successful

intestinal transplantation and encompasses all database categories referring to successful transplantation. In addition, each member of the successful transplant group was required to have a date entry associated with a successful transplant procedure. Our third group contains subjects removed from the list for any other reason. This includes the database categories of; `improved, refused, transfer, error and other' as we did not feel confident combining any of these reasons for list removal with the definitive first two categories. At the time of analysis, there were also patients still waiting on the list for transplant, so the cumulative incidence of removal from the list for all of these three categories does not sum to 1.0.

Multinomial logistic regression

We performed a multinomial logistic regression analysis to compare the relative risk ratios (RRR) for different outcomes across diagnosis groups. The primary interest in this multinomial model was to evaluate the risk of removal from the list for death/poor condition versus removal from the list for successful transplant. The relative risk of death/imminent death versus successful intestinal transplantation was compared among the four most common diagnostic groups: gastroschisis, other short gut syndrome, necrotizing enterocolitis and volvulus. Gastroschisis was chosen as the reference group, as it was the most common diagnostic category. In this formulation of the model, a relative risk ratio above 1.0 for a particular diagnostic group indicated poorer outcomes for that group compared to the reference group (i.e. increased mortality and decreased rates of successful transplantation). Other factors initially considered in the regression analysis include the subject's sex, age and weight. Due to collinearity, the age variable was removed and only weight was utilized in the final model.

Results

Overall Population

We identified 1,712 children initially listed for intestinal transplant (57% male). Fifty-one percent were less than 1 year old at initial listing, and 31% were 1–5years of age. The median weight and height at listing were 8.1kg (IQR 6.1–14.1kg) and 66cm (IQR 60–86.4cm), respectively. The most common conditions at listing were gastroschisis (23%), other short gut syndrome (21%), necrotizing enterocolitis (19%), volvulus (12%), and functional bowel syndrome (12%). (Table 2)

Patients Transplanted & Not Transplanted

Of the 1,712 initially listed for intestinal transplant, during the study period, 852 (50%) patients were transplanted, 694 (40%) were not transplanted, and 166 (10%) patients were awaiting intestinal transplant at the time of our analysis (Table 2). Fifty-four percent of those transplanted were female. Twenty-five percent of patients were less than one year-old at operation while 51% were between the ages of 1 and 5 years. Among those transplanted, the median height and weight were 74cm (IQR 65–109cm) and 10.7kg (IQR 7.8–21.7kg), respectively. Sixty-nine percent of patients also received a liver transplant. (Table 3)

A comparison of mean ages and weights at listing between those that were transplanted and those that were not transplanted revealed a statistically significant difference in both variables. Compared to patients that were removed from the list, transplanted patients were older (3.58 vs. 1.75 years of age, $p<0.0001$) and heavier (15.9kg vs. 10.9kg, $p<0.0001$) at the time of listing.

Outcomes Following First Listing for Intestinal Transplant

The cumulative incidences of removal from the transplant list for death/poor condition, removal for successful transplantation, and removal for any other reason are summarized over time in Figure 2 for the four most common diagnostic categories. This graphic depicts the

cumulative incidence estimates for the three outcomes simultaneously (stacked vertically), so that the fraction of the total cohort experiencing each type of outcome can be evaluated in relation to the fraction experiencing the other outcomes at any given point in time. The relative risk of removal for death/poor condition versus removal for a transplant is most favorable in the SGS group (14% death: 75% transplant) and least favorable in the NEC group (37% death: 42% transplant). The significance of the differences in this relative risk parameter by diagnostic group was further evaluated via multinomial logistic regression modeling. Overall, diagnostic grouping was a highly statistically significant predictor of the relative risk, even after adjusting for patient sex and weight ($p<0.001$). Taking the largest diagnostic category, gastroschisis, as the reference group, the relative risk ratio for death versus successful transplant was 1.55 for NEC (95% CI: 1.09–2.20), 0.98 for volvulus (95% CI: 0.64–1.50) and 0.46 for SGS (95% CI: 0.29–0.73). (Table 4) These results indicate that while volvulus and gastroschisis have similar outcomes, NEC has significantly poorer outcomes (increased mortality, decreased transplantation), and SGS has significantly better outcomes (decreased mortality, increased transplantation).

Discussion

While nutrition and growth is possible with parenteral nutrition, the majority of patients with intestinal failure ultimately require intestinal transplant for long-term survival. Our finding of a discrepancy in the relative risks of death or progression to transplant among different disease conditions is striking. In our analysis of the four most common conditions leading to intestinal failure and listing for intestinal transplant, we demonstrated the relative risk of death was significantly higher in the NEC group, similar for those diagnosed with volvulus, and improved for those within the broad category of short gut syndrome, compared to the gastroschisis group. Since no study to date has described such differences in outcomes, the definitive explanation for these findings is not yet clear. Many possibilities exist. First, there are as of yet no standardized criteria for listing for intestinal transplant, much less an agreed upon definition of intestinal failure. Although the Medicare criteria outlined above must be met, the specific weight, age and number of comorbidities allowed at a certain center may be different than those at another center. Additionally, centers may have different qualifying criteria for patients with NEC compared to those with gastroschisis. Standardizing the listing criteria in terms of demographic and clinical variables may eliminate the differences between populations. Second, there may be as of yet unknown differences in treatment and referral patterns among different diagnoses leading to intestinal failure. Patients with NEC may be treated more commonly in tertiary centers with transplant programs, leading to earlier referral than patients with gastroschisis or volvulus. This study evaluated patients that were listed, and therefore there may be a selection bias towards survival among patients referred from non-transplant centers. Lastly, there are clearly natural differences in these populations. Patients with NEC have an ongoing systemic inflammatory insult and are generally of low birth weight whereas patients with gastroschisis and volvulus have more acute issues and are much more often full or near full-term. The survival benefit of a complete in utero gestation has been demonstrated previously[6] and previous work has noted the negative relationship between small body size and survival to transplant at a single center[7].

Our data demonstrating the age and size difference between those that were transplanted and those that eventually died while waiting may point to the importance of early referral of such pediatric patients to a center where their intestinal failure therapy can be maximized[8–11]; if in fact, these centers can demonstrate improvement in nutritional status and weight gain. The difference in ages and weights between patients that were transplanted and those that were not may be accounted for in several ways. Transplanted patients may have been older because they suffered their intestinal insult at an older age and were therefore able to tolerate parenteral nutrition and intestinal failure better than younger patients. Alternatively, transplant surgeons

may prefer older patients who weigh more to younger patients who may have more physiologic compromise and malnutrition. Lastly, the supply of donor organs, or intestines, may be more available for older patients with larger body sizes. Regardless of the specific reason for differences noted among pediatric transplant populations, efforts to prolong intestinal viability, delay intestinal failure, lessen the dependence upon parenteral nutrition, improve nutritional status and promote weight gain should be bolstered in order to maximize the possibility of successful transplant.

Although transplanted patients were, on average, older and larger at the time of listing, it is not clear how long they dealt with intestinal failure prior to being listed. There have been several studies calling for `early referral' to specialized centers for the early management of intestinal failure[9, 10, 12]. There is little data available on the population of patients with intestinal failure prior to being listed for transplant. The time period from dependence upon parenteral nutrition or a clear definition of intestinal failure is critical to understanding the outcomes of this unique population. A clear and unified definition of intestinal failure is greatly needed to promote generalized understanding among clinicians and researchers. Additionally, further research into this unique population is necessary in order to keep these patients with intestinal failure alive as operative and intensive care therapies improve.

The majority (69%) of the transplants analyzed for this study had an associated liver transplant. Several studies have examined the relationship between a liver or multivsiceral transplant along with the intestinal transplant and outcomes. While some have demonstrated the feasibility of these procedures without additional morbidity or mortality[13,]14, others contend that the need for a liver in addition to an intestine transplant may lead to the higher wait-list mortality demonstrated among this population12.

There are several limitations to our study. The primary limitation is our dependence upon a clinical database; reliant upon individuals and centers to reliably diagnose, code and record diseases, conditions, and events. Misclassification and human error is possible. There were dates and entries within the database that were logically incorrect requiring identification and reconciliation. In addition, anatomical information such as the length of intestine, presence or absence of an ileo-cecal valve or the large intestine was not available. The UNOS database is the largest database for intestinal transplant in North American and provides information on patients awaiting this procedure from 1987. Although our data span more than two decades, improvements in care and technology specifically related to the management of intestinal failure patients and the use of parenteral nutrition may affect outcomes. Wait-times, donor availability, immunosuppression, surgical transplantation techniques, medical diagnostic and management techniques and computer technology have all changed during this period. There was opportunity for in- or out-migration from the database, however, these patients would likely have been captured by one center affiliated with the database. Despite these limitations, the use of such a comprehensive database over such a long time period has allowed us to study the largest pediatric cohort of intestinal failure and transplant patients to date.

Future studies regarding intestinal failure and intestinal transplant should first focus on developing uniform definitions of the conditions that predispose children to intestinal failure and most importantly, a uniform definition of intestinal failure, itself. Second, the criteria to qualify for an intestinal transplant must be developed, agreed upon and shared broadly to ensure similarity in these populations for outcome comparison purposes. Demographic information on age, gestational age, sex and weight is just as important as clinical factors such as specific disease condition, amount and functionality of viable intestine, presence of an ileo-cecal valve, amount of enteral and parenteral nutrition that is tolerated and the numbers and types of comorbidities in a given patient. The number and type of bowel-lengthening procedures must also be taken into account in order to compare patients equally. The preceding variables are

by no means comprehensive. A better understanding of the characteristics of these patients with intestinal failure awaiting intestinal transplant, their outcomes while on the waiting list, and the possible association of their outcomes with their initial disease state will not only help clinicians with the management of these patients, it will also aid policy-makers with resource allocation and families with expectations and family planning. Additionally, clear definitions and improved patient selection for transplant will have a significant impact on public health through improved resource utilization both in caring for the sick child with intestinal failure and in allocating the scarce resource of donor organs to appropriate recipients.

We have described the population of pediatric patients with intestinal failure awaiting intestinal transplant, and analyzed their outcomes on the waiting list. We have demonstrated that there are differences in the relative risk of death or transplant depending upon the specific disease condition, even after adjusting for sex and weight. This is the first study to perform an analysis of the national population of pediatric patients with intestinal failure eligible for and awaiting intestinal transplant. Further work is necessary to identify, define and follow factors among children with intestinal failure that affect outcomes.

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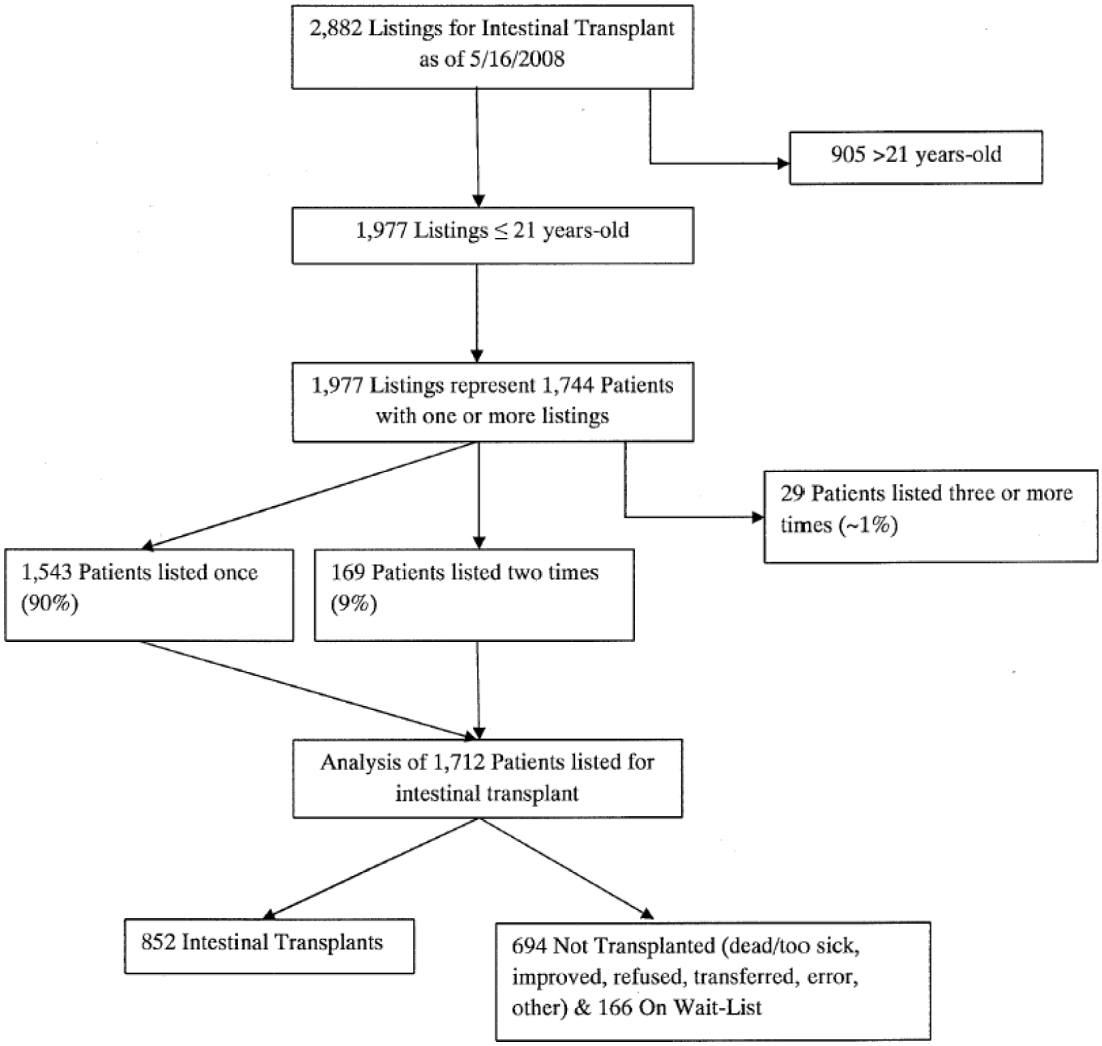

Figure 1.
Study subject selection outline from UNOS database. (≤21 years-old at listing)

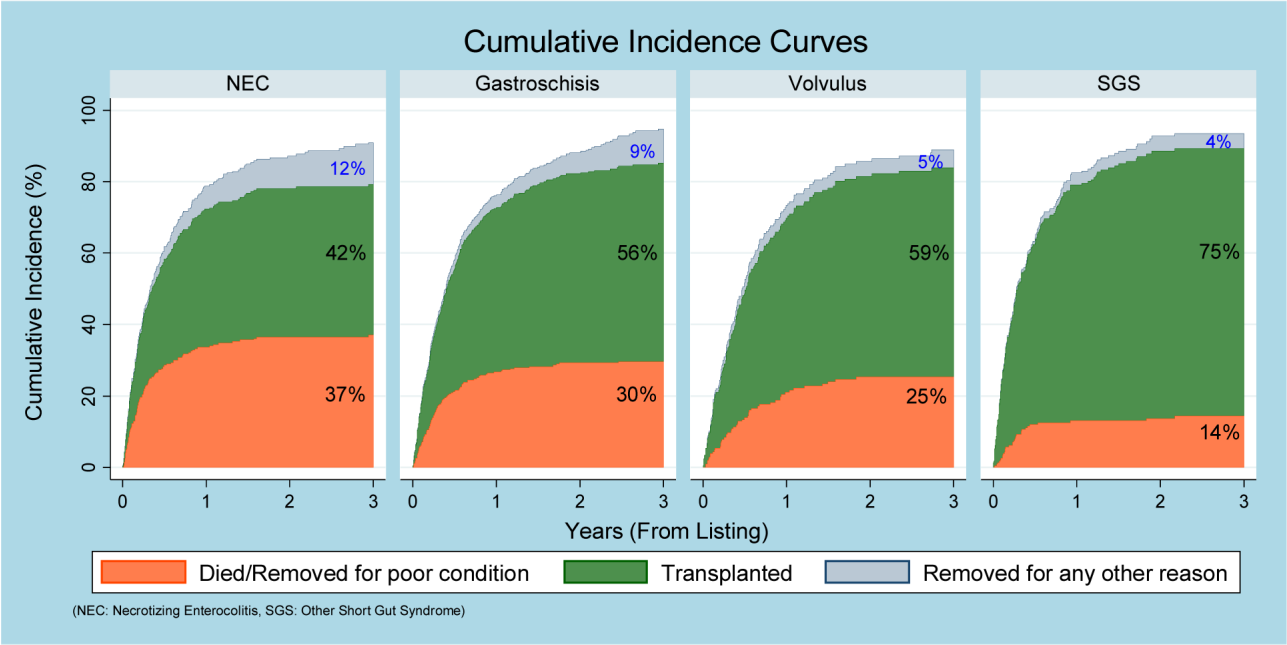

Figure 2.
Cumulative incidence curves from time of listing for intestinal transplant to removal from list for the four most common diagnostic categories.

Table 1

OPTN diagnostic categories and study diagnostic categories.

OPTN Diagnosis Categories	Study Diagnosis Categories
Short Gut Syndrome	
SGS: Gastroschisis	Gastroschisis
SGS: Intestinal Volvulus due to Adhesion, Malrotation, Persistent Obstruction	Volvulus
SGS: Necrotizing Enterocolitis	Necrotizing Enterocolitis
SGS: Intestinal Atresia, Mass Resection due to (Inflammatory bowel/ Crohn's disease, Mesenteric Arterial or Venous Thrombus, or Tumor), Other, Unspecified	Other Short Gut Syndrome
Functional Bowel Problem	
FBP: Hirschprung's	Hirschprung's
FBP: Microvillus Inclusion Disease, Neuronal Intestinal Dysplasia, Protein Losing Enteropathy, Myopathic Pseudo-Obstruction, Neuropathic Pseudo-Obstruction, other, unspecified	Functional Bowel Problem
Graft Failure	Other/Unknown/Missing/Graft Failure
Intestinal Disease Other Specify	
Retransplant/Graft Failure	
Other	
Not Reported	

Table 2

Demographics at Listing

N=1,712	All # (%)	Transplanted # (%)	Removed & Not Transplanted # (%)
Sex	1,712	852	694
Male	968 (57)	392 (46)	404 (58)
Female	744 (43)	460 (54)	290 (42)
Age at Listing			
<1yr	881 (51)	372 (44)	430 (62)
1–5yrs	536 (31)	285 (33)	193 (28)
6–11yrs	133 (8)	90 (11)	31 (4)
12–17yrs	99 (6)	59 (7)	29 (4)
18–21yrs	63 (4)	46 (5)	11 (2)
Median (IQR)	0 (0–21)	1 (0–5)	0 (0–1)
Missing	0	0	0
Weight @ Listing		837	678
1.00–2.49kg (LBW)	6 (<1)	2 (<1)	3 (<1)
2.50–4.99kg	216 (13)	82 (10)	107 (15)
5.00–9.99kg	840 (50)	371 (44)	398 (57)
10.00–19.99kg	324 (19)	193 (23)	96 (14)
20.00–39.99kg	173 (10)	109 (13)	46 (7)
>40kg	122 (7)	80 (10)	28 (4)
Median (IQR)	8 (6–14)	9 (7–18)	7 (6–10)
Missing	31 (2)	15 (2)	16 (2)
Height at Listing		784	605
<50cm	79 (5)	26 (3)	43 (7)
50–74.9cm	956 (62)	429 (55)	438 (72)
75–99.9cm	217 (14)	130 (17)	56 (9)
100–149.9cm	199 (13)	129 (16)	47 (8)
150–174.9cm	84 (5)	54 (7)	19 (3)
>175cm	19 (1)	16 (2)	2 (<1)
Median (IQR)	66 (60–86)	70 (61–101)	64 (58–74)
Missing	158 (10)	68 (8)	0 (0)
Diagnoses			
Necrotizing Enterocolitis	320 (19)	127 (15)	164 (24)
Volvulus	205 (12)	115 (14)	70 (10)
Gastroschisis	389 (23)	206 (24)	158 (23)
Other Short Gut Syndrome	362 (21)	172 (20)	148 (21)
Hirschprung's Disease	92 (5)	60 (7)	26 (4)
Functional Bowel Syndrome	212 (12)	134 (16)	55 (8)

N=1,712	All # (%)	Transplanted # (%)	Removed & Not Transplanted # (%)
Other*/Unknown/Missing/GF	132 (8)	38 (5)	73 (11)

* pseudo-obstruction, intractable diarrhea, trauma, microvillus inclusion disease, multiple polyposis, mesenteric venous thrombosis, desmoids tumor, crohn's

Table 3

Transplant Operation Demographics

N=852	# (%)
Age at Operation	
<1yr	210 (25)
1–5yrs	434 (51)
6–11yrs	97 (11)
12–17yrs	60 (7)
18–21yrs	45 (5)
Median (IQR)	1 (1–5)
Weight at Operation (N=710)	
1.00–2.49kg (LBW)	2 (<1)
2.50–4.99kg	26 (4)
5.00–9.99kg	295 (42)
10.00–19.99kg	197 (28)
20.00–39.99kg	108 (15)
>40kg	82 (12)
Median (IQR)	10.7 (7.8–21.7)
Missing	142 (17)
Height at Operation (N=618)	
<50cm	13 (2)
50–74.9cm	302 (49)
75–99.9cm	124 (20)
100–149.9cm	115 (19)
150–174.9cm	47 (8)
>175cm	17 (3)
Median (IQR)	74 (65–109)
Missing	234 (27)
Type of Operation (N=852)	
Small Intestine	813 (95)
Large Intestine	78 (9)
Simultaneous Liver	591 (69)
Simultaneous Pancreas	289 (34)
Whole:Segmental	838:13

Table 4

Relative risk of death vs. successful transplant by diagnostic categories

Diagnostic Group	Relative Risk Ratio (Death vs. Successful Transplant)*	95% Confidence Interval
Necrotizing Enterocolitis	1.55	1.09–2.20
Volvulus	0.98	0.64–1.50
Other Short Gut Syndrome	0.46	0.29–0.73

*Reference is Gastroschisis