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Intestinal transplantation



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A B S T R A C T

Intestinal transplantation has now emerged as a lifesaving therapeutic option and standard of care for patients with irreversible intestinal failure. Improvement in survival over the years has justified expansion of the indications for intestinal transplantation beyond the original indications approved by Center for Medicare and Medicaid services. Management of patients with intestinal failure is complex and requires a multidisciplinary approach to accurately select candidates who would benefit from rehabilitation versus transplantation. Significant strides have been made in patient and graft survival with several advancements in the perioperative management through timely referral, improved patient selection, refinement in the surgical techniques and better understanding of the immunopathology of intestinal transplantation. The therapeutic efficacy of the procedure is well evident from continuous improvements in functional status, quality of life and cost-effectiveness of the procedure. This current review summarizes various aspects including current practices and evidence based recommendations of intestinal transplantation.

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Indications and contraindications

Intestinal failure (IF) results from various conditions that cause anatomical short gut syndrome due to extensive small bowel resection, intestinal atresia, and volvulus or from functional short gut due to

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absorption or motility disorders like Hirschsprungs disease, necrotizing enterocolitis, pseudo obstruction. Loss of functioning gut mass leads to malabsorption of important micronutrients and fluid-electrolyte imbalance leading to malnutrition and progressive weight loss [1].

The advent of home parenteral nutrition (HPN) has revolutionized the prognosis in patients with IF from a near fatal outcome to reported 1 and 4 year survival rates of 90% and 84% respectively [2]. However, parenteral nutrition (PN) is disadvantaged with high cost and poor quality of life from treatment-specific morbidities like central line associated blood stream infections (CLABSI) and parenteral nutrition associated liver disease (PNALD) [3]. IF and PN dependency is believed to be permanent in adults with 60–80 cm of remnant bowel without ileocecal valve and with >2 yrs duration on PN. In pediatric population the chances of weaning PN become slim with <30 cm of remnant bowel and with 36–48 months of PN dependency [3–5]. Patients with permanent intestinal insufficiency are constantly faced with the threat of morbidity and mortality from effects of longstanding PN use.

Most patients with IF benefit from a multidisciplinary intestinal rehabilitation approach with integrated medical and/or surgical strategies geared to promote intestinal adaptation and restore enteral autonomy. This reserves the option of intestinal transplantation for 10–15% of patients with irreversible intestinal failure who progress to develop complications of long term PN [6].

An exciting milestone in small bowel transplantation was reached in October 1999, at the Sixth Annual International Small Bowel Transplantation Symposium in Omaha, when experts in the field proposed indications for intestinal transplantation. Following this in the year 2000, Center for Medicare & Medicaid Services (CMS) approved isolated small bowel intestinal (ISBTx), combined liver-intestinal(ILTx), and multivisceral transplantation(MVTx) as the standard of care for patients with irreversible intestinal and PN failure, adding a new dimension to the management of irreversible intestinal failure [7].

Subsequently in a report published to serve as a referral guide for care providers specific indications for intestinal transplantation included [8]: (1) overt or impending liver failure caused by PNALD; (2) multiple thromboses of central veins limiting central venous access; (in infants when 2 of the 4 available standard access sites i.e. bilateral subclavian and internal jugular veins, have been lost to thrombosis and in an older child or an adult patient, when 3 of the 6 available standard access sites—bilateral internal jugular, subclavian and femoral veins—have been lost to thrombosis) (3) more than two episodes of catheter-related infection requiring hospitalization in any year; (4) a single episode of fungal line infection; and/or (5) frequent and severe episodes of dehydration, despite IV fluid supplementation and TPN.

Over the years several other indications were approved including unresectable, benign or slow-growing mesenteric tumors like desmoid, congenital mucosal disorders such as microvillous inclusion disease, complete splanchnic venous thrombosis precluding liver transplant alone and ultra-short bowel syndrome with residual small intestine <10 cm in infants and <20 cm in adults [9–11]. Other potential indications which were perceived as important indications in current practice but not uniformly accepted

Underlying Indications for Intestinal Transplantation (ITx).

Pediatric	Incidence (%)	Adult	Incidence (%)
Gastroschisis	21	Ischemia	23
Volvulus	17	Crohn's disease	14
Necrotizing enterocolitis	12	Trauma	10
Pseudo-obstruction	9	Desmoid tumors	9
Intestinal atresia	8	Motility disorders	8
Retransplant	8	Volvulus	7
Hirschsprung's disease	7	Short gut, other	7
Microvillous inclusion disease	6	Retransplant	6
Malabsorption, other	4	Miscellaneous	5
Short gut, other	4	Other tumors	4
Other	4	Gardner's syndrome	3
Motility	2		
Tumor	1		

From Intestinal Transplant Association (ITA): Intestine transplant registry: 25 years of follow-up results (<http://intestinaltransplantassociation.com>).

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