Pediatric Intestinal Transplantation

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KEYWORDS

- Pediatric intestinal transplantation • Intestinal failure • Immunosuppression
- Short gut syndrome

KEY POINTS

- Intestinal transplantation is a successful procedure for children with life-threatening complications of irreversible intestinal failure.
- Intestinal transplantation has shown improvement in survival rates owing to advances of surgical techniques and immunosuppressive therapies.
- Infection and chronic rejection are the most common causes of graft loss after intestinal transplantation.
- Close monitoring, early recognition, and prompt treatment of viral infections have improved survival.
- Efforts are directed toward the prevention and management of immunosuppressant-related morbidities to achieve ideal outcomes in pediatric intestinal transplantation.

INTRODUCTION

Intestinal transplantation (ITx) has continued to be an important treatment modality for pediatric intestinal failure despite development of increasingly successful intestinal rehabilitation outcomes. Owing to challenges with long-term graft function due to chronic rejection, ITx has been generally reserved for children with irreversible intestinal failure suffering from complications of total parenteral nutrition.[1] Complications of total parenteral nutrition such as frequent sepsis, multiple central line infections, lack of intravenous access, and parenteral nutrition–associated liver disease are major factors for consideration of ITx in an patient with intestinal failure.

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The initial trials for ITx developed by Lillehei in 1959 and Starzl in 1960 developed the experimental basis in dogs that established the technical cornerstones. In the cyclosporine era, experiences were mostly unsuccessful until the 1990s, when the long-term survival of patients with isolated small bowel transplantation occurred. The first human multivisceral transplant (MVT) was also performed by Starzl and colleagues[2–5] in 1983 and 1989 in 2 pediatric cases, but clinically reproducible results were not achieved until after the introduction of tacrolimus in 1989. The reasons for initial graft failure were mostly ascribed to unpredictable allograft rejection and recipient multiple organ disease severity as compared with isolated liver transplant recipients. The introduction of tacrolimus and then antibody conditioning with antithymocyte globulin or other antibodies significantly improved outcomes in pediatric ITx. However, chronic rejection and the prevention and management of immunosuppressant-related morbidities remain the most significant challenges to achieving ideal outcomes in this field.[6,7]

INDICATIONS

According to The Intestinal Transplant Registry (ITR) data, the main disease indications for ITx in the pediatric population are the presence of anatomic/surgical short gut and functional/motility disorders (**Box 1**). Between 1985 and February 2013, 1611 pediatric patients underwent ITx in 55 centers. Gastroschisis (22%), volvulus (16%), necrotizing enterocolitis (14%), and intestinal atresia (4%) were the main contributors of surgical short gut. Motility disorders (18%) were the major nonsurgical short gut indication with malabsorption (8%), with additional indications owing to tumors (1%) and re-ITx cases (8%).[8]

Box 1
Pediatric intestinal transplantation indications

Short gut syndrome
Volvulus
Gastroschisis
Necrotizing enterocolitis
Intestinal atresia
Ischemia
Trauma
Motility disorders
Primary pseudo-obstruction
Hollow visceral myopathy
Microvillus inclusion disease
Hirschsprung's disease
Other indications
Neuropathy with extensive involvement of gastrointestinal system with stomach
Gastrointestinal neoplastic disorders
Intestinal polyposis
Intestinal retransplantation