Intestinal Transplant in Children

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KEYWORDS

• Intestine • Short gut syndrome • Pediatric • Transplant

KEY POINTS

- Outcomes of intestinal transplantation have dramatically improved; pediatric recipient survival is up to 95% and 70% at 1 and 3 years, respectively.
- Aggressive and innovative intestinal rehabilitation efforts have improved outcomes of children suffering from short gut syndrome — intestinal transplantation is now an integrated treatment modality when rehabilitation efforts fail.
- Early referral to a transplant center is crucial to optimize transplantation option and outcomes.
- Long-term intestinal transplant outcomes depend on excellent compliance with follow-up and immunosuppressive regimen.
- Chronic rejection incidence is dramatically decreased with new immunosuppressive protocols but remains the Achilles heel of pediatric intestinal transplantation.

HISTORY

The first reported human intestinal transplant was performed by Lillihei and coworkers in 1968.[1] Many attempts followed over the next decades and were met with high failure rates due to graft rejection and infectious complications. The past decade has seen major advances in the field, ushering in a new era where intestinal transplantation is the treatment of choice for many with intestinal failure.

The international 1-year mortality from pediatric intestinal transplantation significantly declined over the past decade, from 30% to the current 10% to 15%. This has mainly been the fruit of a multidisciplinary approach adopted in transplant centers, spanning the spectrum of diagnosis and treatment of intestinal failure to short-term and long-term care of post-transplant patients. Pediatric age carries many additional special

Disclosure Statement: The authors have nothing to disclose.
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Pediatr Clin N Am 64 (2017) 613–619
http://dx.doi.org/10.1016/j.pcl.2017.02.002 **pediatric.theclinics.com**

considerations along the spectrum of care that continue to cause challenges but also offers growth opportunities. In particular, pediatric intestinal transplantation indications and timing are changing as a result of new developments in diagnostic and treatment tools.

This article reviews updates on pediatric intestinal transplantation and highlights future directions.

INTESTINAL FAILURE AND REHABILITATION

Intestinal failure can be broadly defined as the inability of the intestines to function, which can either be secondary to anatomically short intestine or lack of adequate absorptive intestinal function.

Intestinal Failure Causes

- Anatomic loss
  - Intestinal resection (volvulus, NEC, gastroschisis, intestinal atresia, Crohn disease)
  - Intestinal bypass
- Congenital mucosal disorders
  - Microvillous inclusion disease
  - Tufting enteropathy
- Intestinal motility disorders
  - Hirschsprung disease
  - Intestinal pseudo-obstruction
  - Megacystis microcolon hypoperistaltic syndrome

Every effort should be made for intestinal rehabilitation in patients who have some preserved ileal length.[2,3] In addition to nutritional and fluid rehabilitation, surgical procedures are commonly used to increase the intestinal absorptive surface and control bacterial overgrowth resulting from intestinal stasis. Such procedures include but are not limited to bowel lengthening, such as serial transverse enteroplasty and longitudinal intestinal lengthening and tailoring.[4] Trials are ongoing with enteral growth hormones and glutamine to improve the mucosal health. In addition, control of bacterial overgrowth, avoiding infections, and minimizing lipids exposure have greatly decreased early liver failure, allowing more time for rehabilitation efforts.[5] For some patients, however, these efforts remain temporary bridges that only help to reduce morbidity and mortality while affected patients await transplantation.

Complications of Intestinal Failure

Intestinal failure is a debilitating condition that can result in life-threatening complications. Intestinal transplantation has been a life-saving treatment of such patients. The most serious encountered complications are parenteral nutrition (PN)–associated liver disease, sepsis and loss of intravenous access.[2,3]

PN liver disease or intestinal failure–associated liver disease has been reported in as many as 50% of patients with intestinal failure who received PN for more than 5 years. It is more frequently seen in infants and toddlers versus adolescents and adults. Progressive liver disease can be identified by persistent elevations in serum bilirubin for more than 3 months to 4 months. These patients are also at risk of developing other complications, such as portal hypertension, variceal bleeding, and liver failure. All efforts should be made to perform intestinal transplant in these children while the liver disease is still reversible. Once liver failure is established, survival on the transplantation wait list is greatly affected and remains at less than 50%. Recently, intravenous lipids minimization and the use of polyunsaturated lipids have helped improve the incidence of (and