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Management of Pediatric Intestinal Failure

Jacqueline Wessel, Med, RDN, CNSC, SCP, CLE[a], Meera Kotagal, MD, MPH[b], Michael A. Helmrath, MD[b,*]

[a]Division of Nutrition Therapy, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA; [b]Division of Pediatric Surgery, Cincinnati Children's Hospital Medical Center, Cincinnati, OH, USA

Keywords

• Intestinal failure • Enteral feeding • Pediatric • Intestinal reconstruction

Key points

- The management approach to children with intestinal failure is complex.
- It requires diligent management of nutrition with maximal effort given to achieving as much as possible via the enteral route, involving enteral tubes if necessary.
- Optimization of enteral feeding requires thoughtful surgical planning for access and should be done in a manner that minimizes starting and stopping feeds frequently.
- In these challenging patients, timely management and preferably prevention of complications can result in significant clinical benefit.

INTRODUCTION

Formal intestinal failure multidisciplinary programs greatly improve the care provided to patients by reducing the morbidity and mortality associated with this complex condition [1]. In general, these programs consist of gastroenterologists, pediatric surgeons, advanced care providers dedicated to intestinal rehabilitation, intestinal failure nutritionists, pharmacists with advanced training in parenteral nutrition (PN), and social workers. However, because more than 80% of all patients with intestinal failure present during the neonatal period, it is the general pediatric surgeon and neonatologist who make the early decisions that often greatly affect the outcome of these complex patients. This article focuses on the important role of surgical management of damaged

*Corresponding author. 3333 Burnet Avenue, MLC 2023, Cincinnati, OH 45229-3039.
E-mail address: Michael.Helmrath@cchmc.org

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intestine, effective means, and strategies to provide nutrition required for healthy growth and development. Attention to these key areas may attenuate the significant physical, emotional, and fiscal burdens that result in an immeasurable amount of distress to patients and their families [2].

Intestinal failure occurs in these patients because of an inability of their bowel to meet fluid and/or nutritional needs required to sustain normal physiology and growth without PN support. In children, short bowel syndrome (SBS) is the major cause of intestinal failure and results from congenital disorders and extensive surgical resection. The common causes of SBS include intestinal atresia, abdominal wall defects (primarily gastroschisis), intestinal volvulus, long segment Hirschsprung disease, complicated meconium ileus, and necrotizing enterocolitis (30% of cases, the most common cause). Data on the incidence and mortality related to SBS are sparse. A recent cohort of very low and extremely low birth weight neonates at 16 tertiary centers in the United States demonstrated incidences of SBS at 0.7% and 1.1%, respectively, although this excluded cases in term infants [3]. Given the increase in the overall number and survival of these at-risk patients it is logical to assume that the overall number of pediatric SBS will also continue to increase. Mortality in this population of patients occurs in a bimodal fashion, with the first peak corresponding to infants who undergo massive initial bowel resections and a later peak corresponding to complications of SBS, namely central venous catheter sepsis and intestinal failure–associated liver disease (IFALD). Survival rates for children with SBS have been quoted as 73% to 89%, with lower rates in patients requiring chronic total PN (TPN) [4–7].

The care of patients at risk for intestinal failure begins at birth for those with congenital problems and in the operating room for patients undergoing massive intestinal resection. Although the time course of intestinal adaptation predicts that many of these patients will eventually be able to obtain enteral autonomy, waiting for the few patients to declare themselves as having chronic intestinal failure misses an opportunity for prevention. Because enteral nutrition (EN) is the most effective therapy to augment adaptation, early treatment paradigms designed to minimize morbidity and establish early controlled feedings will likely benefit all patients at risk by reducing the overall need for PN support. Decisions made at this time should take into account several key points. The first is that rapid intestinal growth occurs during the third trimester and continues into the first year of life *when exposed to EN*, therefore short segments of intestine in infants have the greatest potential for significant increased growth [8,9].

Second, the intestine has a remarkable ability to heal after ischemic injury and it is often impossible to predict the fate of the bowel without the benefit of a significant period of time, often several weeks (not days) despite the appearance of "dead bowel" [10]. Early second-look operations should occur before committing to initial resection, and in the otherwise clinically stable infant should occur when definitive procedures can be performed. Proximal control of healthy bowel may be required to allowed distal diseased bowel the time required to heal. In such conditions as necrotizing enterocolitis, procedures