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## Preface

Intestinal failure in children carries a significant morbidity and mortality as well as social and economical burden. Intestinal failure can be defined as the reduction of gut function below the minimum necessary for the absorption of nutrients, such that intravenous supplementation is required to maintain health and growth. Intestinal failure in children is usually caused by three major conditions: short bowel, intestinal dysmotility, or extensive small bowel mucosal disease. The prevalence of intestinal failure remains unclear because the definitions used in different populations are variable. The estimated prevalences range between 2 and 13 per million children.

In children the intestinal length is related to the age of the child; therefore, a definition of a short bowel in absolute terms cannot be devised. The need for intravenous supplementation of nutrients and a residual small bowel length of less than 25 % expected for gestational age are suggested definitions of a short bowel in children. The main causes of short bowel in children are gastroschisis, midgut volvulus, bowel atresias, and necrotizing enterocolitis.

In addition to short bowel, intestinal failure may be due to congenital enteropathy, such as microvillus inclusion disease, and intestinal neuromuscular disorder, such as total bowel Hirschsprung's disease or intestinal pseudo-obstruction, which are associated with diminished effective absorptive small bowel.

The management of intestinal failure in children has evolved enormously during the last decades. The morbidity has decreased and the survival increased markedly. Since the 1980s the development of parenteral nutrition technology and the use of home parenteral nutrition have increased the safety of long-term parenteral administration of nutrients. The composition of parenteral nutrition formulas has evolved decreasing the metabolic complications that were very common especially in the newborn and infant population of intestinal failure patients. Intestinal failure-associated liver disease that along with septic complications of parenteral nutrition was the major complication threatening the life of intestinal failure patients has become less common and treatable by adjusting the intake and composition of parenteral fat. The central lines used for parenteral nutrition incorporate today chemical locks such as ethanol lock that protect the catheters from bacterial colonization and decrease the occurrence of catheter-related septic episodes.

Until recently, there has been less evolution in the medical treatment of intestinal failure. There are very few medications that significantly improve intestinal motility in the long term; moreover, their effects are commonly

unpredictable. Most patients require antibiotics regularly to control bacterial overgrowth episodes, but their use is associated with development of bacterial resistance. Teduglutide, a recombinant analog of glucagon-like peptide-2, is the first and a promising targeted therapeutic agent to treat short bowel-associated intestinal failure. It has been shown to reduce the requirement of parenteral nutrition significantly.

Surgical management of intestinal failure has been revolutionized by the development of autologous intestinal reconstruction techniques. Longitudinal intestinal lengthening procedure and serial transverse enteroplasty both increase the possibility of weaning off parenteral nutrition. In general, PN requirement decreases in the great majority of patients, while around half of children achieve enteral autonomy during the first two postoperative years.

Intestinal transplantation is an acceptable therapeutic option for patients with intestinal failure, but it is still reserved for patients who develop severe and life-threatening complications despite standard therapies or those who are not able to maintain a good quality of life. There has been a marked improvement in graft survival rates, especially for intestine alone grafts, over the past two decades.

Intestinal failure in children is a multifaceted condition that requires contributions of several medical and allied health professionals for hospital care and support at home. Therefore, the formation of a multidisciplinary intestinal rehabilitation team is vital to achieve optimal results. The team should include competent professionals in surgery, gastroenterology, and nutrition, a dietician, and staff experienced in handling central venous catheters and nutrient infusions. Special consideration should be given to the psychosocial support of the family during the whole treatment period.

This book has been compiled to summarize current knowledge on intestinal failure. The chapters are written by international experts in the field. The size of the book is kept compact to allow easy reading and timely and rapid updating of the content.

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