Clinical guidelines for the managment of intestinal failure secondary to pediatric short bowel syndrome. Executive summary

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Intestinal failure (IF) secondary to pediatric short bowel syndrome (SBS) is an uncommon disease with a high morbidity and mortality. An adequate management requires an interdisciplinary team formed by specialist in pediatric nutrition, surgeons, nurses, and social workers, among others. The development of such teams, now known as intestinal rehabilitation teams, helps to reduce morbidity and mortality and allows most patients to achieve intestinal autonomy.

This is an uncommon disease, so there is not enough scientific evidence regarding different approaches to it. Therefore, we believed it was necessary to develop these *Clinical management guidelines* based on the modified Delphi method. In the setting of the Argentine Association for Enteral and Parenteral Nutrition (*Asociación Argentina de Nutrición Enteral y Parenteral*, AANEP), 16 experts gathered to discuss and agree on the main aspects of clinical management and establish common criteria so that the results of different groups may be adequately disseminated.

Undoubtedly, this document will be of help to patients, institutions, and funders.

Below we describe the main aspects with a strong agreement among experts (level of agreement > 80 % + rate of abstention < 5 %).

Four different aspects were discussed.

1. Introduction, definitions, and epidemiology

- IF is defined as a reduction of functional intestinal mass below the minimum necessary to maintain nutrient digestion and absorption, electrolyte balance, nutritional status, and growth. The management of IF requires parenteral nutrition (PN).
- The most common cause of IF is SBS, which mainly results from a massive small intestine resection, leaving a residual bowel length of less than 25 % of that expected for gestational and/or chronological age. In addition to bowel length, the functionality of the residual segment should be taken into consideration.
- The conditions leading to surgical resection include congenital anatomical abnormalities, intestinal atresia and/or gastroschisis, neonatal ischemia or necrosis (secondary to enterocolitis) or secondary to volvulus or vascular thrombosis. Two or more conditions in a patient may lead to SBS.
- The main causes may vary among the different reporting referral facilities. Most SBS cases in the

pediatric population are observed in the neonatal period. The greater survival of preterm infants accounts for the higher prevalence of necrotizing enterocolitis (NEC) in some facilities. According to experts, NEC is not the main cause of SBS in Argentina.

- Several factors influence the clinical course of SBS patients:
  a. Positive variables include a longer residual bowel segment; preservation of the ileum, ileocecal valve, and colon; a younger age at the time of resection.
  b. Unfavorable variables include a long-term enterostomy; more surgical procedures; residual bowel dysfunction; cholestasis; sepsis episodes; and the presence of underlying diseases.
  c. In relation to causative diagnoses, complicated gastroschisis and intestinal atresia usually have a less favorable course than other diagnoses.
- The management of pediatric patients with IF/SBS requires an interdisciplinary team, formed by health care providers with experience and/or knowledge about these conditions, who may be consulted or work as part of a network of specialty facilities. The team assigned to patient follow-up should also include their caregivers.

2. Enteral nutrition

- Enteral nutrition (EN) is defined as nutrient supply directly to the stomach or small intestine, regardless of the route (oral, nasogastric tube, stoma) or the type of feeding.
- EN should be started as soon as possible for patients receiving PN because it promotes physiological responses that may improve the bowel adaptation process.
- Breast milk (BM) is the best feeding option for infants with IF/SBS receiving EN.
- For patients with IF/SBS who cannot receive BM, there is no consensus about which infant formula to use. Polymeric formulas (containing whole proteins and long-chain triglycerides) may favor bowel adaptation. Formulas containing protein hydrolysates and medium-chain triglycerides (MCTs) may favor absorption in the early stages.
- For patients with IF/SBS, oral feeding should be indicated whenever possible, as long as it is not formally contraindicated.
- Then feeding should change to EN up to the maximum volume tolerated by the patient. Tolerance will be based on volume and digestive loss characteristics (fecal output), abdominal distension, pain or discomfort, and vomiting.

3. Parenteral nutrition

- Electrolyte and energy intake in patients receiving PN should be individually adapted based on their clinical condition and in accordance with the general recommendations for pediatric patients established in the ESPGHAN/ESPEN 2018 guidelines.
- The recommendations about protein intake for patients with IF/SBS are similar to those for other pediatric patients receiving PN. Protein intake should be adequate to ensure growth and a positive nitrogen balance.
- To estimate the resting energy expenditure (REE) in pediatric patients with IF/SBS, the recommendation is to use the Schofield predictive equation, which takes age, sex, weight, and height into consideration.
- To estimate energy intake, non-protein calories provided as glucose and lipids are used. The recommendation is a 60-70 % carbohydrate (glucose) proportion and a 30-40 % lipid proportion of non-protein kilocalories.
- Also, it is recommended to use mixed lipid emulsions with a lower omega-6 long-chain polyunsaturated fatty acid content, a lower omega-6/omega-3 ratio, a lower phytosterol content, and a higher alpha-tocopherol and MCT content.
- A cyclic infusion of PN is recommended for stable patients with IF/SBS, prolonged hospitalization with PN or receiving PN at home.
- PN in hospitalized patients should be administered by trained nursing staff following management guidelines or standards that have been validated and endorsed by scientific societies and agreed upon by the health care facility. At home, PN should be administered by trained nurses or parents/caregivers who have been trained in the technique and following strict rules.
- Intestinal failure-associated liver disease (IFALD) is defined as liver and/or biliary dysfunction resulting from intrinsic factors of IF and others inherent to its medical or surgical management. It may progress to end-stage liver disease or stabilize or resolve through bowel adaptation stimulation.
- For PN at home, semi-implantable, tunneled,

single-lumen, central venous catheters (Hickman or Broviac type) with the smallest caliber possible should be used.
- Non-tunneled (PICC type) catheters or percutaneous central lines should be used in newborn infants. In all cases, the use of silicone or polyurethane central venous lines (CVLs) with the smallest caliber possible to infuse the necessary volume to the patient is encouraged.

4. Pharmacological and surgical treatments: Referral criteria
- Severe diarrhea in children receiving EN may be managed with opioids if accelerated bowel transit is suspected or with exchange resins if the cause is suspected to be bile acid malabsorption.
- There is not enough evidence to recommend the use of probiotics for the prevention or treatment of bowel overgrowth in patients with SBS.
- There is no consensus about absolute and relative indications for the drug treatment with teduglutide in pediatric patients with IF/SBS.
- Those requiring a simple surgical procedure (stoma closure, reanastomosis) may undergo reconstruction at a local level. A complex autologous gastrointestinal reconstruction (including stoma closure with stenosis resection, reduction-tapering of the proximal dilated segments, intestinal lengthening, fistula treatment) is an indication for referral to an intestinal rehabilitation facility. An intestinal lengthening surgery should be considered for patients with an adequate bowel dilatation (3-4 cm) and a residual segment > 40 cm, which prevents the advance of EN and/or leads to translocation infections and/or is considered a limitation for weaning from PN.

Final comment

The full version of this document can be accessed online and includes 42 items accepted by experts with a high level of agreement. We believe that this document will help to provide a better care to these patients, to whom we dedicate this article.

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