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Definitions of intestinal failure and the short bowel syndrome



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The European Society for Clinical Nutrition and Metabolism defined intestinal failure (IF) as “the reduction of gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes, such that intravenous supplementation is required to maintain health and/or growth”. IF is classified as type 1-acute, type 2-prolonged acute and type 3-chronic IF. A short bowel syndrome (SBS) due to the intestinal malabsorption associated with a functional small intestine length of less than 200 cm is the most frequent mechanism of IF. SBS is a difficult and multifaced disease. Complications due to SBS itself and to treatments, such as long term home parenteral nutrition, can adversely affect the patient outcome. The care of SBS requires complex technologies and multidisciplinary and multiprofessional activity and expertise. Patient outcome is strongly dependent on care and support from an expert specialist team. This paper focuses on the aspects of the pathophysiology and on the complications of SBS, which are most relevant in the clinical practice, such as intestinal failure associated liver disease, renal failure, biliary and renal stones, dehydration and electrolyte depletion, magnesium deficiency and D-lactic acidosis.

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The European Society for Clinical Nutrition and Metabolism (ESPEN) has recently devised the definition and classification of intestinal failure (IF) [1] and the guidelines for chronic intestinal failure (CIF) management [2]. Furthermore, several updates and reviews on short bowel syndrome (SBS) have been published [3], prompted by the recent marketing approval in Europe and the US of an intestinal growth factor (teduglutide, a glucagon-like peptide two analogue) [4], as well as by new surgical techniques to improve bowel length and function in SBS patients [5]. The aim of this paper is to focus on the aspects of the pathophysiology and on those complications of SBS that are most relevant in the clinical practice.

Definition and classification of intestinal failure

Intestinal failure (IF) was firstly defined in 1981 by Fleming and Remington as “a reduction in the functioning gut mass below the minimal amount necessary for adequate digestion and absorption of food” [6]. In the following years, other experts further characterized this organ failure by including in the definition “the need of nutritional supplementation to maintain health and growth”. Jeejeebhoy considered IF “any gut dysfunction requiring either oral or intravenous supplementation (IVS)” [7]. Jeppesen and Mortensen proposed to define IF as “only those conditions requiring intravenous supplementation”, to call “intestinal insufficiency” those intestinal dysfunctions that need only enteral supplementation and define “oral failure” as the involuntary reduction of the ingestion of food [8]. Nightingale and Woodward categorized IF as “severe” when parenteral, “moderate” when enteral, and “mild” when oral nutrient/fluid supplements are needed [9]. Some Authors also included in the definition the need of micronutrients in association or alone [10]. Irving first described four mechanisms of IF, SBS, motility disorders of the bowel, SB parenchymal disease and intestinal fistula [11]. Shaffer devised a classification of IF, according to the onset modality, the metabolic characteristics and the expected outcome [12].

ESPEN recommendations, include a functional and a pathophysiological classification for both acute and chronic IF and a clinical classification of CIF [1]. IF has been defined as “the reduction of gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes, such that IVS is required to maintain health and/or growth”. The panel recognized that the diagnosis and quantification of IF would be optimally done by balance study techniques comparing nutrient requirement with nutrient absorption. However, as only few centres may have the facilities for these assessments, the need of IVS was considered an indirect criterion. The ESPEN definition highlights that two criteria must be simultaneously present to diagnose IF: a “decreased absorption of macronutrients and/or water and electrolytes due to a loss of gut function” and the “need for IVS”. This precludes patients receiving an IVS notwithstanding a normal intestinal absorptive function be considered as having an IF (e.g., disease-related hypophagia, anorexia nervosa, impaired swallowing or dysphagia, refusal of an otherwise effective enteral nutrition). To define the reduction of gut absorptive function that doesn't require IVS to maintain health and/or growth, the term “intestinal insufficiency” (or “intestinal deficiency” for those languages where “insufficiency” and “failure” have the same meaning) was proposed.

The “functional classification” was based on that originally proposed by Shaffer [12,13]: **type I IF** an acute, short-term and usually self limiting condition, commonly occurring in the peri-operative setting and/or in association with critical illnesses, and requiring IVS for a few days or a few weeks; **type II IF** a prolonged acute condition, often in metabolically unstable patients, such as those with an intra-abdominal catastrophe, enterocutaneous fistulae, or acute mesenteric ischemia, requiring complex multi-disciplinary care and IVS over periods of weeks or months; **type III IF** a chronic condition, in metabolically stable patients, who require IVS over months or years.

The “pathophysiological classification” of IF identified five major conditions: short bowel, intestinal fistula, intestinal dysmotility, mechanical obstruction, extensive small bowel (SB) mucosal disease. In the case of a short bowel, an enterocutaneous fistula or an extensive SB disease, the primary mechanism of IF is the malabsorption of the ingested food. When an intestinal dysmotility or an intestinal mechanical obstruction occurs, IF is primarily due to the restriction of oral/enteral nutrition or total oral fasting, in order to limit or avoid the feeding-related exacerbation of digestive symptoms.

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