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Chronic Intestinal Failure and Short Bowel Syndrome in Adults: Principles and Perspectives for the Portuguese Health System

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Keywords
Intestinal failure · Home parenteral nutrition · Short bowel syndrome

Abstract

Background: Short bowel syndrome is a devastating malabsorptive condition and the most common cause of chronic intestinal failure (CIF). Patients need parenteral support for months or years. Ideally, it should be delivered at home, reducing limitations in everyday life activities. ***Summary:*** The Portuguese Health Directive 017/2020 was the first step in the regulation of home CIF management, and more patients are now being treated in an ambulatory setting. However, much work still needs to be performed in this area. Our country lacks a network of units capable of providing home parenteral nutrition (HPN), and only a few centers have expertise to take care of these complex patients: fluid support, oral, enteral, and parenteral nutrition; disease/HPN-related complications; pharmacologic treatment; and surgical prevention/treatment. Providing adequate transition from pediatric to adult care is a mandatory issue that should only be addressed by expert centers. ***Key Messages:*** Implementation of a national network, as well as the creation of an intestinal failure registry, with an initial focus on adult patients, will start a new era in the identification and management of these complex CIF patients.

Falência Intestinal Crónica e Síndrome do Intestino Curto em Adultos: Princípios e Perspetivas para o Serviço Nacional de Saúde

Palavras Chave
Falência intestinal · Nutrição parentérica domiciliária · Síndrome do intestino curto

Resumo

Contexto: A síndrome do intestino curto constitui uma condição clínica devastadora e mal-absortiva, sendo a causa mais comum de falência intestinal crónica (FIC).

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Estes doentes carecem de suporte parentérico durante meses ou anos, idealmente em ambulatório, reduzindo as suas limitações no seu dia-a-dia. ***Sumário:*** Em Portugal, a Norma 017/2020 constituiu um primeiro passo oficial na abordagem da FIC, abrangendo cada vez mais doentes em contexto de ambulatório. Contudo, em Portugal não existe rede de serviços/unidades que possam providenciar nutrição parentérica domiciliária e apenas alguns centros possuem competência no tratamento de doentes com FIC, nomeadamente no manejo da fluidoterapia, nutrição oral, entérica e parentérica, complicações associadas à doença e/ou à própria nutrição parentérica domiciliária, tratamento farmacológico e ainda na prevenção/tratamento cirúrgico. Proporcionar uma adequada transição da idade pediátrica para a idade adulta constitui um aspeto fundamental que apenas deve ser operacionalizado entre centros de referência. ***Mensagens-chave:*** A implementação de uma rede nacional de FIC, assim como a criação de um registo nacional de FIC, com foco inicial no doente adulto, iniciarão uma nova era na identificação e abordagem adequada destes doentes.

Introduction

Short bowel syndrome (SBS) is a malabsorptive condition that results from the loss of intestinal length due to disease or resection [1]. SBS is the most common cause of chronic intestinal failure (CIF) [2]. Patients need parenteral support during months or years, parenteral nutrition (PN), or hydration/electrolyte supplementation. Parenteral support should be delivered at home, reducing limitations to everyday life activities, including working for adult patients and studying for children and adolescents. Functional classification includes all chronic metabolically stable patients requiring intravenous supplementation (type III) and some less metabolically stable patients requiring intravenous supplementation during weeks or months (type II) that can be effectively managed in an ambulatory setting.

For decades, Portuguese patients with CIF faced multiple barriers in receiving home PN (HPN). PN was considered a pharmacological treatment to be used only in hospital ward. Only a few institutional teams with home hospitalizations or a similar organization can provide HPN to some citizens [3]. In 2020, a new regulation from Portuguese authorities (*Direção-Geral da Saúde*), "Norma 017/2020" [4], set the conditions for home CIF management. Regulation of HPN, procedures, and structure of the nutrition support team (NST) have been organized, and an increasing number of intestinal failure (IF) patients are being treated in an ambulatory setting.

We aimed to address the current Portuguese reality for adult patients with SBS-CIF and the implications of the health directive 017/2020. We also reviewed the composition of multidisciplinary NST and the transition from pediatric to adult care. Finally, on behalf of Núcleo de Nutrição em Gastrenterologia (NNG), there is now a need to develop a national network of skilled centers and to organize SBS-CIF care, including patients requiring all types of parenteral support (PN and/or hydration/electrolyte supplementation). With standardized care, we will certainly contribute to improving the outcomes of these complex patients. Because SBS is the most important etiology of CIF, other causes of CIF will not be covered in this review.

The Current Portuguese Reality for Adult Patients with SBS-CIF

CIF is a rare organ failure worldwide [5], and the estimation of SBS-CIF prevalence is extremely variable. One of the reasons for this variability is the methodology used for the SBS-CIF estimation. Several studies have used HPN as a surrogate marker of SBS-CIF, which is frequent in older studies [6, 7]; however, it carries some setbacks: not all patients needing HPN have SBS-CIF, and not all SBS-CIF need PN. Other studies were based on SBS-CIF hospitalizations [8]. More recent studies are based on national SBS-CIF registries, which are easily accessible to researchers [9]. Another reason for the wide variation is the study or survey period. In Europe, the prevalence varies from 6 to 34 per million citizens in several countries, which is lower than the 2022 American study [10–15]. In contrast, data on CIF are scarce in Portugal. Antunes et al. [16] reported a prevalence of 27 cases per million inhabitants of the pediatric population in 2018. Furthermore, a Portuguese nationwide survey conducted in 2019 found only 20 children and 11 adults undergoing HPN (although 2 known patients were not included for logistic reasons), equivalent to 1 adult patient per million, the lowest European prevalence [17]. This likely reflects the lack of a national registry and the complete absence of a network of units capable of providing HPN and treating patients with SBS-CIF. In fact, the survey only identified five centers that took care of adult patients.

It is expected that the incidence and prevalence of CIF will increase in the coming years due to improvements in care, introduction of new drugs (e.g., teduglutide), and the associated increased life expectancy [5]. However, owing to

its low incidence and prevalence, as well as the complex medical and surgical issues associated with long-term PN, it is essential to establish a wider network of referral centers that are proficient in the management of SBS-CIF patients.

The Portuguese Turning Point – Health Directive 017/2020

As stated, patients with SBS-CIF had major difficulties in accessing PN at home since PN was only considered a hospital-based treatment. In addition, patients living in geographically isolated areas with limited health resources do not have access to HPN. "*Norma 017/2020: Implementação da Nutrição Entérica e Parentérica no Ambulatório e Domicílio em Idade adulta*" [4], a government initiative, was, indeed, a major step in clinical/artificial nutrition in Portugal. It starts with the creation of an NST named *Grupo de Nutrição Entérica e Parentérica* (GNEP), whose main objective is to manage the prescription of enteral nutrition (EN) or PN at the hospital level. For all patients previously identified as suffering from malnutrition or nutritional risk that would benefit from HPN, an individualized nutritional plan should be developed. It aims to achieve enteral autonomy by progressively reducing PN and increasing oral/enteral feeding, if tolerated.

According to Health Directive 017/2020, HPN should be prescribed when EN is contraindicated or when nutritional support is required, but needs cannot be met by the enteral route. Examples of disorders leading to HPN other than SBS in adults in Portugal (stated in the health directive) include the following:

- cancer-associated cachexia (e.g., after intestinal resection and Lisboaradiation/treatment-related enteritis);
- inflammatory bowel disease causing severe malnutrition;
- mesenteric ischemia with abdominal angina;
- proximal enteric fistula;
- chronic dysmotility diseases not controlled by therapy.

Table 1 summarizes the therapies, equipment, and medical devices essential for HPN according to health directive 017/2020, highlighting the complexity, resource intensiveness, and expense to be considered with HPN.

The Interdisciplinary Nutrition Team for SBS-CIF Adult Patients

The most important resource for IF management is multidisciplinary NST [2]. It includes health care professionals with different backgrounds and clinical nutrition training. The NST should include the following:

- Medical doctors, including gastroenterologists and gastrointestinal surgeons [18], are trained in providing the necessary care for digestive diseases. Gastroenterologists should be experienced in the management of medical disorders that cause CIF, such as Crohn's disease, dysmotility, and malabsorption. The experience and training of surgeons should be considered a subspecialty of digestive surgery according to the European Society of Coloproctology [19], and this is of special importance for patients with type II IF. Whenever needed, other specialists, such as radiologists, endocrinologists, and/or pediatricians, should be included in the NST.
- Nurses: They assist patients in a therapeutic manner. A major objective of nurses' interventions is to promote the autonomy and self-care of patients and caregivers in a safe and responsible manner. An individualized educational plan should be made to meet the needs of each patient/caregiver and should address principles of infection control and prevention, central venous catheter (CVC) precautions, training aseptic care of the CVC, and technique of management PN [20]. Nursing intervention is essential in preventing risks and complications associated with CVC, including monitoring inflammatory signs at the CVC insertion site and the integrity of the CVC, assessing the effectiveness and tolerance of PN, and evaluating the skills achieved. CIF nurses are essential to engage patients and caregivers in the management of their illness, promoting a feeling of independence, security, control, responsibility, normality, and trust.
- Dietitians with experience and training in CIF [21]. Most patients with CIF suffer from SBS, with some bowel extension capable of digestion and absorption, and the same scenario may be present in type II IF. Although virtually all patients with SBS-CIF will require PN, more than 50% will be able to be weaned completely from PN within 5 years. Therefore, rehabilitation should be initiated as soon as possible, and dietitians play a major role in this process. Oral feeding plays an important role in global nutrition and is a major contributor to the quality of life and well-being. It provides gut feeding, helps intestinal integrity and function by providing nutrition to enterocytes, and may contribute to the prevention of IF-associated liver disease. Dietitians must provide optimal oral intake, considering the PN, morphology of the remaining bowel, patient preferences, and lifestyle. EN should also be considered, especially in those with low PN dependence who are expected to be weaned off. Furthermore, the importance of restricting the intake of

Table 1. Therapies, equipment, and medical devices essential for HPN

PN equipment/therapies
Standard PN admixtures (two or three compartments) or personalized PN admixtures if the former does not reach nutritional needs
Injectable hydrosoluble and liposoluble vitamins
Bidistilled water
Sodium chloride 0.9% (100 mL and 500 mL)
Heparin
Hypertonic glucose (20 or 30%)
Rapid-onset insulin (if necessary)
Antiseptic (chlorhexidine or iodopovidone)
Disinfectant (alcohol)
Catheter lock with antimicrobial and antifungal properties (taurolidine 2% or equivalent)
Medical devices
Perfusion pump
Filter in line, 0.22 micron (two-chamber PN or admixtures without lipids) or 1.2 micron (three-chamber PN or admixtures with lipids)
Three-way stopcock
Sterile catheter cap
Surgical mask
Scrub cap
Surgical gloves
Sterile fields
Sterile syringe (2, 5, 10, and 20 mL)
Hypodermal needle (12 and 18G)
Sterile dressings (10×5 cm and 7.5 × 7.5 cm)
Film dressings (ex. IV 3000; op-site 3000 10×14 cm)
Container for sharp/cutting medical waste
Disposable medical gown
Other materials
Disinfectant for hand hygiene
Fridge space for storage of PN bags
Metallic support for PN admixture (if necessary)
Support for the perfusion bomb (if necessary)

PN, parenteral nutrition.

low-sodium fluids such as hypotonic fluids (e.g., water, tea, alcohol, and coffee) and hypertonic fluids (e.g., regular soda and fruit juices) should be emphasized.

- Pharmacists' main role is to collaborate and supervise the design, composition of macro- and micronutrients, implementation and monitoring of PN, and prevention of metabolic or catheter-related complications [22]. In addition, they play a very important role in preventing the possible interactions of PN with other medications, as well as the absorption of oral/enteral drugs in the context of intestinal dysfunction. Pharmacists' roles have been expanding beyond the supervision of compounding/dispensing PN, including direct care, consultations of PN patients, and education of patients/caregivers, which results in more adequate nutrition with fewer metabolic complications.
- Besides this core team, other healthcare professionals are also frequently needed. Physiotherapists and exercise professionals are necessary as these patients present with malnutrition and sarcopenia with impaired movement aptitudes. Several cases present with dysphagia or insecure swallowing; evaluation/rehabilitation may require a trained speech therapist. Finally, patients may present with comorbidities that may require a wide range of health professionals [23].

Whenever a department has a patient in a ward who may need HPN, the staff should contact the NST with experience in HPN, whether from the same institution or from another hospital. Together, the department staff and the NST should evaluate the need for HPN, stabilize the patient, and determine the composition of the PN mixture before discharge. After discharge, the patient

should be evaluated in an outpatient clinic on a weekly basis, with clinical and laboratory evaluations to assess the fluid, nutritional, and electrolyte status. When difficulties are recognized during the hospital-home transition, appointments may be more frequent, e.g., twice a week. Gradually, appointments may become less frequent according to HPN stabilization and patient autonomy and empowerment.

The NST should conduct regular audits and prepare regular reports of activities regarding the services provided in the field of HPN in outpatient/home settings. In this regard, the quality of care should be measured to evaluate HPN-related complications, hospital readmissions, weight change, and regular assessments of the patient's quality of life. The NST should promote and facilitate the continuous training of its members, with participation in courses and scientific meetings dedicated to clinical nutrition, such as those promoted by *Associação Portuguesa de Nutrição Entérica e Parentérica*, NNG, and the European Society for Clinical Nutrition and Metabolism.

Transition from Pediatric to Adult Care in SBS-CIF

In the CIF, the transition of care from pediatric to adult health services remains a delicate process, and a planned transition of care is essential. This transition is a planned and purposeful movement of adolescents and young adults with complex medical conditions from a child-centered to an adult-centered healthcare system [24].

The transition period is a time with a potential risk of morbimortality [25, 26], which reinforces the need for an organized, multidisciplinary, and individualized plan of care involving primary healthcare services. The NST must be prepared to provide care to these patients and to maintain, at least, the level of the previous care, which is crucial to maintain clinical stability and quality of life [27].

Children and adolescents with CIF have limited autonomy and dependence on the care provided by caregivers and healthcare teams. Adolescents are a distinct group of children and adults from physical, emotional, and psychological standpoints, making this transition even more complex.

There are different models of transitional care for chronic illnesses [24]. The most suitable model for CIF might be the transition model focused on the illness and their professionals [28].

In Portugal, most pediatricians select the age of 15–19 years as the most appropriate age to initiate the transition, which encompasses the recommended age by the American Academy of Pediatrics from 18 to 21 years [29, 30]. The key principles sustaining a successful transition are (1) information, (2) communication, and (3) planning/coordination [31]. According to the Italian Society of Pediatric Gastroenterology, Hepatology, and Nutrition and the Italian Society of Artificial Nutrition and Metabolism, the aims of this process, known as "acronym of 5 M" are (1) motivate independent choices, (2) move toward adult goals, (3) maintain previous care, (4) minimize the difficulties involved in the transition, and (5) modulate the length of transition [28].

In the first transitional appointment, the adolescent should attend simultaneous consultations with the pediatrician and the adult physician. Subsequent consultations should be performed according to a previously established plan. Initially, the place for consultations should be the usual pediatric environment and afterward, in the adult's environment. Most adolescents and young adults wish to be seen by the adult physician in their pediatric environment, but it is desirable to be familiar with adult facilities [28].

A Portuguese National Network for Adult Patients with SBS-CIF

In complex and rare diseases, it is essential to have dedicated professionals with medical and surgical resources, organized in multidisciplinary teams, and with knowledge and expertise in the management of these patients. In Europe, there are inequalities in accessing HPN, and there are no standardized models of organization or reimbursement. However, some countries have national centers for CIF, such as Denmark, France, and England [32]. In Portugal, most hospitals can provide PN in the setting of hospitalized patients (mainly type I and II IF) but not HPN, and there are only a few specialized centers to manage complex SBS-CIF patients.

Portugal, a country with 10.4 million inhabitants [33], has very few patients under nutritional support at home (related to several reasons including low expertise in the field and the absence of legal support for delivering PN at home, until recently). Owing to its rarity, a total of 50–150 adult CIF patients are expected to be managed with HPN a few years after the implementation of an organized network. To prevent the allocation of a very small number of SBS-CIF patients *per* center, insufficient to provide enough clinical experience to each center, it would be critical to constitute a maximum of 3–5 referral centers for adult CIF patients. These referral centers could work in close relation to support local NST. In geographically isolated areas, telehealth services, outreach clinics, and

Table 2. Main topics in SBS-CIF in Portugal

What is the current Portuguese reality for adult patients with SBS-CIF?
Extremely low prevalence compared with other European countries, reflecting the complete absence of a national registry and a network of reference centers
The Portuguese turning point – what have changed with the health directive 017/2020?
Portuguese SBS-CIF patients had major difficulties in accessing PN at home (no legal support for it)
The health directive 017/2020 was a major step in clinical/artificial nutrition in Portugal
Regulation of HPN, procedures, and structure of the NST were organized
How should an NST be organized?
The NST should be composed of healthcare professionals with different backgrounds and clinical nutrition formation/training
The NST should include medical doctors (e.g., gastroenterologists/digestive surgeons), nurses, dietitians, and pharmacists
Other healthcare professionals may be needed (e.g., physiotherapists, speech therapist)
How to promote a stable transition from pediatric to adult care in SBS-CIF?
Most pediatricians select the age 15–19 years as the most appropriate to initiate the transition
In the first transitional appointment, the adolescent should attend simultaneous consultations with the pediatrician and the adult physician
Pediatric and adult centers may be located in different hospitals, providing that transition is effectively organized
What will change in Portugal regarding SBS-CIF care?
It is suggested to create 3–5 referral centers for adult SBS-CIF patients in relation with the local NST. Also, 2–3 pediatric referral centers should be organized
The organization of a Portuguese CIF Registry is ongoing and will focus initially on SBS-CIF adult patients
Although the pediatric register has not yet started, it may be unified with the adult register in the future

SBS-CIF, short bowel syndrome-chronic intestinal failure; PN, parenteral nutrition; NST, nutrition support team.

shared care models may overcome some challenges in managing these patients. Close collaboration with primary healthcare services, including blood collection and evaluation, may help patients living far from the hospital, although this is still not implemented, and SBS-CIF patients still need to come to the hospital. Sometimes, a "ready-to-use service" provided by an external corporation may be used to deliver several items at the patient's home, including PN admixture, systems, and perfusion bombs for PN delivery. Using such external partners may be useful; however, clinical follow-up must remain with the NST, and regular appointments should not be neglected [34]. Patients were required to have an available telephone number, ideally always available, to answer any emergent problems. Also, 2–3 pediatric referral centers should be organized. Pediatric and adult centers may be located in different hospitals, providing that the transition is effectively organized. These patients may live in different geographical areas, and it is important to offer healthcare in proximity. It is agreed that patients and GNEPs benefit from a large specialist center for discussion of more difficult cases and the promotion of a network model of care. It is crucial that the National Health System promotes this network of care and allocates funds to maintain their clinical practice, ongoing learning, and research with the aim of providing the best care for these patients according to international standards, such as those provided by the European Society for Clinical Nutrition and Metabolism [5].

A Portuguese IF Registry

The organization of the Portuguese SBS-CIF Registry is ongoing under the supervision of NNG, a special interest group in nutrition of the Portuguese Society of Gastroenterology (SPG). This IF Registry will be held on the Cerega platform (Centro Nacional de Registo de Dados em Gastrenterologia) and will use a data collection form similar to other collecting forms of European countries. As a first step, the register focuses on adult patients with CIF. Although the pediatric register has not yet started, it may be unified with the adult register in the future. Table 2 summarizes the main conclusions of this review.

Conclusion

CIF requires a multidisciplinary team composed of healthcare providers with different backgrounds, aiming for the successful treatment of these patients. In our

country, a great deal of work remains to be done in this area, and we hope that the implementation of a nationwide network as well as the creation of a CIF registry will start a new era in the identification and management of SBS-CIF patients.

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Statement of Ethics

Due to the nature of the article, ethical approval was not required.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

Francisco Vara-Luiz, Luísa Glória, Ivo Mendes, Sandra Carlos, Paula Guerra, Gonçalo Nunes, Cátia Sofia Oliveira, Andreia Ferreira, Ana Paula Santos, and Jorge Fonseca performed literature review, selection of studies, and writing. Luísa Glória and Jorge Fonseca conceived the study design, structured the content, and critically reviewed the manuscript. All the authors approved the final version of the manuscript.

Data Availability Statement

All the data analyzed during this review are included in this article. Further inquiries can be directed to the corresponding authors.

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