

Vascular Access in Children with Short Bowel Syndrome

Subjects: Surgery

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Short Bowel Syndrome and intestinal failure are chronic and severe conditions that may require life-long parenteral nutrition in children. Survival of these children relies on the correct functioning of central venous catheters; therefore, careful management, prevention, and treatment of complications is of paramount importance. Despite a growing awareness on preserving the vascular real estate, a certain number of patients still experience a progressive and life-threatening exhaustion of vascular access.

Keywords: children ; intestinal failure ; central venous catheter ; central vascular access ; short bowel syndrome

1. Introduction

Short Bowel Syndrome (SBS) is a rare and severe condition characterized by the loss of intestinal length leading to intestinal failure (IF) with malabsorption of nutrients and fluids ^[1]. Causes of SBS may include surgical resection due to inflammatory bowel disease, bowel ischemia/necrosis, trauma, and, in the pediatric population, necrotizing enterocolitis, motility disorders, complications of abdominal wall defects, volvulus, or bowel atresia. There is no consensus in the literature on a precise definition of SBS in terms of intestinal length in children, but intestinal length less than 25% than expected for the gestational age would probably lead to long-term or life-long intravenous supplementation ^[2]. Moreover, the definition of SBS should not rely on intestinal length alone, because clinical manifestations are mostly dependent on the ability of the remaining intestine to undergo adaptation ^{[3][4]}.

In SBS patients, the remaining intestine is not able to absorb sufficient amounts of nutrients and fluids to support the patient's survival and growth ^[5]. Following an extensive resection, the remaining intestine will undergo a process of adaptation: changes in bowel structure, length, and diameter occur to improve absorption and maximize the contact between the mucosa and nutrients ^[5]. Subsequently, changes will happen in villus height and crypts' depth, improving the bowel's ability to absorb nutrients and fluids ^[5]. The efficacy of this adaptation process depends on several factors, such as primary diagnosis, residual intestinal length, and function ^{[5][6][7]}. Therefore, parenteral nutrition (PN) plays a major role in the management of these patients to maintain an optimal nutritional status and growth rate and to provide them with enough macro- and micronutrients.

In recent years, the use of omega-3-lipid emulsions in PN formulas as well as the institution of Intestinal Rehabilitation Centers has determined great improvement in survival rates in SBS patients ^{[8][9]}. Therefore, the need for viable long-term central venous access in affected children has progressively increased, since PN is often required for several years and in some cases for a life-long period.

Given the above, preserving central vascular accesses (CVA) in these patients is essential. PN delivery requires a reliable and sustainable central venous access; in patients who may need PN for short periods of time (2–3 months), peripherally inserted central venous catheters (PICCs) can be used ^[10]. However, skin-tunneled or subcutaneously implanted central venous catheters (CVCs) in the internal jugular or subclavian vein are recommended for the delivery of long-term PN ^[11]. Specifically, following the statements of the latest NASPGHAN position paper ^[12], only tunneled, single lumen, cuffed silicone catheters should be used in children with IF.

When available, an upper extremity access should be the preferred location and the superior vena cava (SVC) should be the first location of CVC insertion, accessed through either the internal jugular veins, the brachial veins, or subclavian veins. If access to these sites is lost, the femoral or saphenous veins can be exploited to reach the inferior vena cava (IVC). Generally, preferred sites of access include the deep veins of the neck and chest (e.g., internal jugular or brachiocephalic veins) or the deep veins of the arm and shoulder (brachial, proximal basilic, and axillary) ^{[13][14]}. Other

conventional routes, including femoral, subclavian, and cephalic veins, are currently utilized but seem to be associated with higher rates of mechanical and infectious complications [15].

2. Children with Short Bowel Syndrome

Management of vascular access needs to be part of a comprehensive and multidisciplinary strategy aiming, as a first step, to preserve the vascular real estate of children requiring long-term PN. Therefore, a wise and careful planning of vascular accesses associated with a detailed knowledge of the patient-specific vascular anatomy since the beginning of the child's clinical history is mandatory.

Actually, in highly specialized multidisciplinary teams, up to 90% of catheters are placed by interventional radiologists [16]. All children with IF should benefit from the expertise of a hybrid, multidisciplinary, combined surgical and radiological team from the beginning of their clinical history. Furthermore, this approach should be mandatory for those children experiencing a progressive exhaustion of their CVAs despite all attempts.

Progressive loss of vascular access is the consequence of multiple, intertwined factors, mostly central line infections and vascular thrombosis.

Central Line Associated Bloodstream Infections (CLABSI) are a major cause of morbidity and mortality in PN-dependent patients [17]. The literature reports a highly variable incidence in CLABSI rates [10]: a systematic review by Dreesen et al. in 2013 reported a range of 0.38–4.58 per 1000 catheter days among the adult population, while Chu et al. reported an incidence of 8.6 per 1000 catheter days in children [17][18]. Prevention of CLABSI starts with family education, training, use of prevention bundles on CVC insertion sites, and catheter-care maintenance protocols [19]. In SBS patients, multiple studies demonstrate the efficacy of a taurolidine-citrate-heparin catheter lock in determining a clinically substantial and cost-beneficial reduction in CLABSI occurrence [20][21][22].

Even in correctly managed patients, life-long need for PN may lead to progressive loss of conventional access routes, such as the axillary, external jugular, internal jugular, subclavian, saphenous, brachio-cephalic, and femoral veins. These veins can become unavailable due to stenosis and/or thrombosis. Regardless of the underlying pathology requiring long-term central lines, the overall reported rate of thrombosis and stenosis in children varies from 26% to 75% [23][24][25]. Thrombotic occlusion of the superior and inferior vena cava may occasionally occur, thus compelling to find direct and challenging access to the heart.

Prevention and/or aggressive treatment of these complications are essential to ensure a long-term functioning of chronic vascular access in patients who totally depend on these devices. For this reason, over the decades, specific guidelines, such as the comprehensive ESPEN guidelines, have been produced to maximize the longevity of vascular access and to prevent the exhaustion of the vascular real estate [11].

Unfortunately, despite a growing awareness of the importance of preservation of vascular assets, loss of standard access sites is a common clinical challenge, especially in children with chronic IF, who are dependent on PN. Loss of one central vein can be documented in the majority of children with intestinal failure (57%), while loss of two or more central veins is reported in 40–46% [26][27]. Critical loss of vascular accesses with progressive exhaustion of the vascular real estate, is, per se, one of the indications to intestinal transplantation, according to the American Gastroenterology Association (AGA) [28] and ESPEN guidelines, respectively [29]. The latest ESPEN indications to intestinal transplantation are summarized in

Table 1.

Table 1. Indications for intestinal transplantation adapted from ESPEN guidelines [29].

Evidence of Advanced or Progressive Intestinal Failure—Associated Liver Disease
<p>Hyperbilirubinemia > 75 μmol/L (4.5 mg/dL) despite intravenous lipid modification for >2 months</p> <p>Elevated serum bilirubin and/or reduced synthetic function (subnormal albumin or elevated international normalized ratio), and/or laboratory evidence of portal hypertension and hypersplenism persisting for >1 month in the absence of confounding events</p> <p>In children thrombosis of 3 discrete upper body central veins (left subclavian and internal jugular, right subclavian and internal jugular) or occlusion of a brachiocephalic vein (in adults evaluate on a case-by-case basis)</p>

Evidence of Advanced or Progressive Intestinal Failure—Associated Liver Disease

Life-threatening morbidity in the setting of indefinite PN dependence, as suggested by:
In children, 2 admissions to an intensive care unit after diagnosis of intestinal failure because of cardiorespiratory failure (mechanical ventilation or inotrope infusion) due to sepsis or other complications of intestinal failure;
In adults, on a case-by-case basis

Invasive intra-abdominal desmoids in adolescents and adults

Acute diffuse intestinal infarction with hepatic failure

Failure of first intestinal transplant

First-line management of long-term catheter needs to focus on punctual protocols for prevention and treatment of complications: infection, dislocation, device deterioration, and obstruction.

Efforts should aim to salvage the actual catheter or, when the device needs to be changed, to reuse the same vessel by guide-wire substitution of the central line or by more aggressive techniques such as pharmacological (systemic and/or local) or mechanical thrombolysis, if required. Over the decades, multiple techniques for vessel reutilization emerged, mostly from the experience of adult endovascular radiologists. Therefore, children should benefit from these expertise and data should be collected to elaborate systematic guidelines.

To overcome life-threatening complications, alternative routes have been developed and further implemented, as detailed in **Table 2**. In the last decades, some helpful solutions have been learned from the vast armamentarium broadly used in the adult population.

Table 2. Types of central vascular access [30].

Conventional Accesses	Non-Conventional Accesses	Last-Resource Accesses
Jugular	Azygous	Transhepatic
Subclavian	Translumbar	Direct right atrial insertion
Femoral	Intercostal veins	Gonadal vein
	Mammary	
	Arteriovenous fistula	

Moreover, discussion by experienced and multidisciplinary teams with specific competences is mandatory when it comes to the use of alternative access sites. The decision to proceed to use alternative routes should come after implementation of strategies aiming to treat the venous obstruction or stenosis by all possible means (balloon dilatation, mechanical thrombolysis, stenting).

Alternative sites or salvage procedures should be proposed only by experienced teams in accordance with an intestinal transplant center.

Obviously, despite these attempts, alternative routes for central line access will always be necessary for a small number of children. Technical improvements allow safe access of unusual veins; therefore, surgeons should be aware of these techniques to deal with rare but very challenging anatomical conditions. When extensive thrombosis of the larger vessels prevents the placement of a new central line, intercostal, hepatic, gonadal, and inferior epigastric veins should be kept in mind as last-resource but safe and effective routes.

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