Short Bowel Syndrome in the NICU

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KEYWORDS

• Short gut • Intestinal adaptation • Intestinal rehabilitation • Short bowel syndrome • Necrotizing enterocolitis • Malabsorption • Small bowel transplant • Bowel conservation

KEY POINTS

- In neonatal intensive care units, the most common cause of intestinal failure is surgical short bowel syndrome, which is defined as a need for prolonged parenteral nutrition following bowel resection, usually for more than 3 months.
- The clinical course of short bowel syndrome patients can be described in the 3 following clinical stages: acute, recovery, and maintenance.
- A cohesive, multidisciplinary approach can allow most infants with SBS to transition to full enteral feeds and achieve normal growth and development.

DEFINITIONS: INTESTINAL FAILURE AND SHORT BOWEL SYNDROME

Intestinal failure is defined as a significant reduction in the functional gut mass below a critical threshold necessary to maintain growth, hydration, and/or electrolyte balance.[1,2] Intestinal failure can occur because of surgical resection of bowel, congenital anomalies, or functional/motility disorders. In neonatal intensive care units (NICUs), the most common cause of intestinal failure is the surgical short bowel syndrome (SBS), which is defined as a need for prolonged parenteral nutrition following bowel resection, usually for more than 3 months.[3]

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EPIDEMIOLOGY OF SBS IN NEONATES AND YOUNG INFANTS

Surgical SBS was recorded in 0.7% (89/12,316) of very low birth weight infants born during the period 2002 to 2005 at the National Institute of Child Health and Development neonatal research network centers.[4] The frequency of SBS increased in an inverse relationship with birth weight; the incidence of SBS in infants weighing 401 to 1000 g was 1.1% (61/5657), nearly twice that in infants with a birth weight of 1001 to 1500 g (28/6659; 0.4%).[4] In Canada, data from a large tertiary NICU show an overall incidence of SBS as 22.1 per 1000 admissions, and 24.5 per 100,000 live births.[5] The incidence was higher in infants born at less than 37 weeks' gestation (353.7 per 100,000 live births) than in full-term infants (3.5/100,000 live births). The SBS rate of case fatality was 37.5%.[5] Similarly, in a study from 7 tertiary neonatal units in Italy, intestinal failure was seen in 0.1% (26/30,353) of all live births and 0.5% (26/5088) among those admitted to the NICU.[6]

Attempts to estimate the incidence and prevalence of SBS have been constrained by the rarity of the condition, variation in nomenclature, difficulty in providing a clear definition of the study population at tertiary institutions because of complex referral patterns, and paucity of follow-up data.[3,5] Population-based studies are sparse. Although the need for home parenteral nutrition has been used as a surrogate for SBS in some studies, this approach has important limitations; some patients may require parenteral nutrition because of a diagnosis other than SBS, such as malignancy, whereas some infants with SBS who may have been weaned off parenteral nutrition before discharge from the hospital may not be included.

CAUSE OF SBS IN NEONATES AND YOUNG INFANTS

Necrotizing enterocolitis (NEC) is the most common cause of SBS (35%) in neonates, followed by intestinal atresia (25%), gastroschisis (18%), malrotation with volvulus (14%), followed by less common conditions, such as Hirschsprung disease with proximal extension of aganglionosis into the small bowel (2%) (**Fig. 1**).[2] In another study, infants with SBS who eventually required intestinal transplantation had the following primary diagnoses: gastroschisis (25%), intestinal volvulus (24%), NEC (12%), intestinal pseudo-obstruction (10%), jejunoileal atresia (9%), Hirschsprungs disease (7%), and other conditions (13%).[7] In very low birth weight infants, NEC remains the predominant cause of SBS. In the National Institute of Child Health and Development cohort, 96% of cases of SBS were due to NEC. Congenital defects (gastroschisis, intestinal atresia) accounted for 2% and volvulus accounted for the remaining 2%.[4]

Duro and colleagues[8] enrolled 473 patients with a diagnosis of NEC. Among the 129 patients who required surgery, 54 (42%) developed SBS, which was significantly more

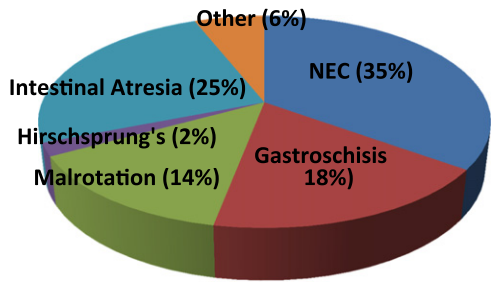

Fig. 1. Causes of short bowel syndrome in neonates and young infants.