

Neonatal short bowel syndrome

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S U M M A R Y

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Neonatal short bowel syndrome is a disease with a high morbidity and mortality. The management of these patients is complex and requires a multidisciplinary approach. Recent advances in medical and surgical treatment options have improved outcomes. The following review highlights salient points in the management of this challenging patient population.

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1. Introduction

The management of neonatal intestinal failure has been transformed by recent medical and surgical innovations. These advances, in conjunction with the evolution of specialized multidisciplinary intestinal failure centers, have resulted in significant reductions in morbidity and mortality.¹ Survival now approaches 90%.¹

2. Definition

Neonatal intestinal failure is defined as intrinsic bowel disease resulting in an inability to sustain growth, hydration, or electrolyte homeostasis. Classically neonatal short bowel syndrome (SBS) is a subset of this disease process that is a consequence of actual small intestinal loss or resection. Mucosal enteropathies and motility disorders are the other causes of intestinal failure that do not involve bowel loss. At times these etiologies coexist in the same patient. For example, a neonate with surgical necrotizing enterocolitis (NEC) may have SBS secondary to small intestinal loss, compounded by malabsorption and disordered motility secondary to damaged but viable bowel that has remained in situ. In practice, the terms intestinal failure and SBS are often used interchangeably. Animal models of SBS are defined by a small intestinal resection of $\geq 80\%$.² No such convention exists in patients although the length of the remaining small intestine is highly correlated with neonatal parenteral nutrition (PN) dependence (Figure 1). In the absence of any surgical bowel lengthening and tapering procedure, 35 cm of neonatal small bowel is associated with a 50% probability of weaning from PN.³ However, a relatively wide variance is evident in Figure 1 and patients with

much longer bowel lengths sometimes do not wean from PN. Poor motility and/or ischemic injury incurred to the remaining bowel may contribute to this finding. Conversely, neonates with as little as 10 cm of residual small intestine can sometimes be weaned from PN (Figure 1). Some additional inaccuracies in neonatal small bowel length interpretation are introduced by differing measurement techniques (the convention is to record the antimesenteric length with no tension applied to the intestine) as well as variant gestational ages at the time of assessment. The more premature the neonate the more likely the intestine will grow in length subsequently. Generally premature neonates have a greater capacity for intestinal growth, and hence bowel adaptation, than full term infants. Nonetheless, residual neonatal small intestinal length remains the single major positive clinical predictor of ultimate enteral feeding tolerance.^{3,4}

Although studies are somewhat conflicting, the presence of an ileocecal valve has for many years been deemed a secondary favorable prognostic sign in SBS.⁵ Conceptually it is important to realize that the presence of an ileocecal valve is a marker for remaining ileum and this may in fact be the underlying important determinant for weaning from PN rather than the presence of the valve itself. The loss of colonic length has a relatively modest effect upon the necessity for long term PN. In general, the colon's prime function is fluid and electrolyte absorption.

An alternate method of assessing neonatal SBS is the serum citrulline concentration. Citrulline is a non-structural amino acid that is primarily synthesized in the intestinal mucosa and hence reflects mucosal mass. Serum citrulline is highly positively correlated with intestinal length and the ability to wean from PN. Intestinal failure patients with a serum citrulline level persistently $< 12 \mu\text{mol/L}$ are usually unable to wean from PN.⁶ A purely functional definition of neonatal SBS may also be used. The simplest and most frequently applied is PN dependence for more than three

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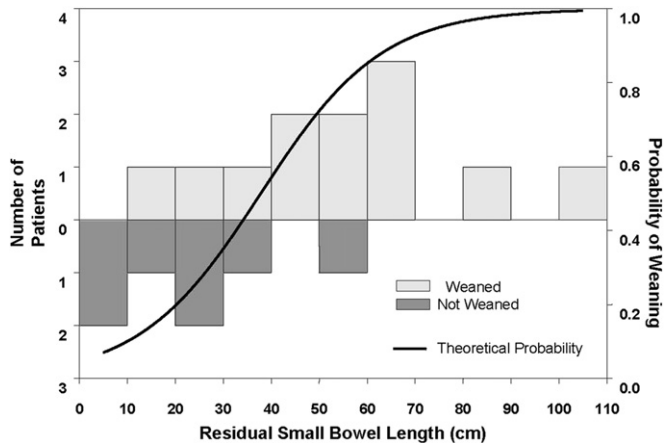


Figure 1. Correlation between small intestinal length and parenteral nutrition (PN) dependence in neonates with short bowel syndrome. The lightly shaded bars represent those individuals weaned to full enteral nutrition; the darkly shaded bars reflect patients failing to attain enteral autonomy. The sigmoid-shaped dark line plots the theoretic probability of weaning from parenteral nutrition based upon remaining small bowel length. It may be seen that 35 cm of residual small intestine is associated with ~50% probability of weaning from PN. (Adapted with permission from Andorsky et al.³).

months.^{3,4} More complex amalgams of functional and anatomic considerations have also been suggested.⁷

3. Causes of neonatal short bowel syndrome

The causes of pediatric SBS vary according to the specific clinical setting surveyed. The Center for Advanced Intestinal Rehabilitation at Children's Hospital Boston manages >200 children with SBS, ~60 of whom are on home PN at any one time. The primary etiologies of SBS in this cohort are: NEC (35%), intestinal atresia (25%), gastroschisis (18%), malrotation with volvulus (14%), with the remainder comprising a compendium of rarer diagnoses, such as Hirschsprung's disease extending into the small bowel (2%). When comparing the causes of intestinal failure between different centers it is important to note that the incidence of NEC is highly dependent upon birth weight, with those neonates between 500 and 750 g having a 12% incidence of NEC.⁸ In extremely low birth weight neonates there is a symmetric 3% decrease in NEC incidence for each 250 g increment in birth weight >750 g.⁸ In NEC the prime predictor of eventual intestinal failure remains residual small bowel length.⁴ It follows that centers with large numbers of extremely low birth weight infants (especially at the lower ranges) will have a preponderance of patients with SBS secondary to NEC.

4. Incidence and mortality of short bowel syndrome

A large cohort of hospitalized neonates encompassing 16 tertiary care centers in the USA demonstrated an incidence of SBS between 0.7% and 1.1% depending upon birth weight.⁹ These estimates, however, did not include term infants. A study from the Province of Ontario, Canada, estimated the incidence of SBS to be 24.5 per 100,000 live births. The occurrence of SBS was much higher in infants born at <37 weeks' estimated gestational age as compared to term newborns (353.7/100,000 live births vs 3.5/100,000 live births respectively). The mortality in this study was three times higher for SBS patients than that of a control cohort matched by underlying diagnoses.¹⁰

The mortality associated with SBS has a bimodal distribution.^{10,11} Mortality is seen in the early postoperative period from

complications associated with the underlying disease process and attendant surgery. Patients surviving long term succumb from the delayed complications of intestinal failure-associated liver disease (IFALD) and sepsis. The major cause of sepsis in SBS is catheter-associated bloodstream infection (CABSI). Based upon a retrospective review, a large intestinal transplant center in the USA has reported a five-year survival of 95% in SBS patients weaned from PN as opposed to 52% in those individuals remaining on PN.¹²

5. Associated complications

The most frequent complication of SBS is an inability to grow and develop without supplemental PN. However, a common misconception is that bowel adaptation is time-limited. In fact the intestine continues to adapt over many years and in our program 90% of intestinal failure patients eventually wean from PN. In addition to continuing adaptation with time it must also be remembered that energy and protein requirements actually decrease significantly on a per kilogram basis as the child ages. Hence, it is not uncommon that neonates who were dependent upon PN for adequate growth will wean from PN as toddlers and even later.

Three specific complications are frequently associated with neonates who have significant SBS. These are IFALD, CABSI, and bacterial overgrowth.

5.1. Intestinal failure-associated liver disease

The cause of IFALD is multifactorial and includes prematurity, PN toxicity and recurrent sepsis. Prematurity with an attendant immature liver has been implicated in the increased incidence of IFALD in neonates compared to older children. Another very important factor contributing to PN toxicity appears to be lipid quantity and type. Appropriate feeding modifications are discussed later in the section on PN. Recurrent episodes of sepsis are also undoubtedly deleterious to liver function and considerable effort is warranted to minimize CABSI.

IFALD is reflected biochemically by elevations of serum transaminases and direct bilirubin followed later by increases in the prothrombin time (PT) and international normalized ratio (INR). Once splenic enlargement occurs, thrombocytopenia may evolve. Hypoalbuminemia is usually a later finding. Clinically one may note persistent jaundice, scleral icterus and an enlarged liver and spleen. Routine liver function assessment is mandated in PN-dependent patients with SBS. Any signs consistent with portal hypertension require further Doppler ultrasound evaluation of the liver, spleen, hepatic arteries, portal vein and hepatic veins. The improvement of IFALD is associated with a normalization of the direct bilirubin followed by a delayed resolution in elevated serum alanine aminotransferase.¹³ Despite normalization of bilirubin levels, significant residual liver damage and even cirrhosis may be present on subsequent liver biopsy.¹⁴ In general, liver damage of a degree less than cirrhosis may be fully reversible. A non-invasive intravenous ¹³C methionine breath test has recently been reported that differentiates cirrhotic from non-cirrhotic infants with IFALD.¹⁵

5.2. Catheter-associated bloodstream infections

SBS patients often require a central venous catheter, which serves as a conduit for the administration of PN. This type of catheter may be placed at the bedside, in the operating room or in the interventional radiology suite. The positioning of these catheters into the central venous circulation should always be confirmed by radiologic evaluation. The key technical elements of insertion are the maintenance of aseptic technique and an effort to avoid

ligating the vessel used for insertion (particularly if a large vessel that can be reutilized is cannulated). Over the last 10 years no patient in our intestinal failure program has lost all central access and no patient has been referred for intestinal transplantation for this indication.

One of the major complications of central venous lines is CABS. Various protocols have been implemented for line care¹⁶ and salutary results have been obtained in many intensive care units. Data from the Vermont–Oxford Neonatal Network, a consortium that entrains data regarding ~80% of all very low birth weight neonates born in the USA, has shown a significant year-over-year reduction in central line infections. These are highly encouraging trends but CABS remains a major issue in SBS neonates.

Any suspicion for CABS needs to be thoroughly assessed. A child with SBS and symptoms of fever, lethargy, irritability, or ileus (usually manifesting as abdominal distension) may have CABS. The most important investigation to confirm CABS is a blood culture through the central line. If the patient's clinical presentation is congruent with CABS, broad spectrum intravenous antibiotics are started through the central line. Neonates with SBS have a very high incidence of CABS due to enteric organisms.¹⁷ Once the organism's antibiotic sensitivities are available the antimicrobial therapy is narrowed. Fugal infections and hemodynamic instability require urgent central line removal.

A promising therapy in the prevention of CABS is the use of ethanol locks. Ethanol has been demonstrated to have the ability to penetrate biofilms that form on central lines, and no bacteria or fungi have been reported to be resistant to ethanol.^{17,18} A recent review of infants with SBS, who are on home PN, has shown a four-fold reduction in CABS in the cohort-administered ethanol locks.¹⁷ The rate of line loss was significantly decreased.¹⁷

5.3. Bacterial overgrowth

Bacterial overgrowth can occur in up to 60% of children with SBS.¹⁹ Areas of disordered motility and bowel dilation offer an ideal environment for abnormal bacterial propagation. The adverse effects of bacterial overgrowth include: abdominal pain, worsening motility, mucosal ulceration with bleeding, deconjugation of bile acids, and the generation of toxic byproducts such as D-lactic acid. It is also thought that bacterial overgrowth potentiates translocation and hence septicemia.

The treatment for suspected bacterial overgrowth is largely empiric. A typically used strategy is the administration of enteral antibiotics for seven days followed by an interval of no antibiotic administration. Antibiotics that are commonly used include those effective against anaerobes or Gram-negative organisms. The use of quantitative duodenal cultures to guide therapy in refractory cases has had success.²⁰ Bowel-tapering procedures may be necessary in those patients where the complications of bacterial overgrowth cannot be managed medically.²¹

D-Lactic acidosis is a classic problem that may be associated with SBS patients who have bacterial overgrowth.²² In the course of normal anaerobic mammalian metabolism L-lactate is generated. L-Lactate is easily converted to pyruvate and used in intermediary metabolism. However, bacteria produce D-lactic acid. This moiety is not well metabolized by humans and hence toxic levels can accumulate. Signs of D-lactic acidosis in neonates include an anion gap acidosis and seizures; older children may additionally demonstrate confusion, slurred speech and delayed cognition. The diagnosis is confirmed by laboratory assay of serum D-lactate. The treatment for this condition is rehydration and eradication of the underlying bacterial overgrowth using enteral antibiotics.

The use of enteral probiotics to restore more normal flora to the gastrointestinal tract of neonates with SBS is theoretically

appealing. Unfortunately, PN-dependent patients with SBS-administered probiotics have developed CABS with the organism administered.²³ It is hypothesized that the probiotic translocates and infects the central line. Hence, the use of probiotics is not recommended.

6. Medical management

The current approach to managing patients with SBS is a multidisciplinary effort focused on nutritional, pharmacologic, and surgical interventions that achieve full enteral nutrition while minimizing the complications of PN therapy.

6.1. Parenteral nutrition

PN is a life-saving therapy in neonates with intestinal failure and has been clinically available since 1968.²⁴ The goal of PN is to provide adequate caloric intake, macronutrients and micronutrients to optimize growth and development. PN formulations are tailored to meet specific individual requirements. A major macronutrient modification for patients with SBS is the utilization of lipid-limited and/or fish oil formulas to obviate the deleterious consequences of IFALD. Soy lipid-based PN formulas administered at <0.5 g/kg/day have been shown to be effective in delaying or preventing cholestasis.²⁵ Soy formulas contain ω -6 lipids that tend to be proinflammatory, thus decreasing their allotment may be of importance in limiting hepatic toxicity. If lipids are reduced, caloric requirements must be met by glucose.

Fish oil is rich in ω -3 fatty acids that have potential anti-inflammatory effects upon the liver. The investigational use of a commercially available, but not US Food and Drug Administration (FDA)-approved, fish oil formula (Omegaven, Fresenius Kabi, Bad Homburg, Germany) at 1 g/kg/day has been associated with the reversal of hyperbilirubinemia and few apparent side-effects in neonates with intestinal failure.²⁶ Reversal of cholestasis was achieved in a mean time of 81 days. This is roughly equivalent to the interval for normalization in direct bilirubin seen in enterally fed children with IFALD.²⁷ It is intriguing that the provision of fish oil emulsions has not been generally associated with essential fatty acid deficiency in neonates.²⁸ A randomized controlled trial of a fish oil formula at 1 g/kg/day vs an equivalent reduced quantity of soy-based lipid has been initiated.

Low serum bicarbonate and sodium levels, both consequent to a reduced intestinal absorptive capacity, are common in patients with SBS. The former may be managed by increasing acetate in the PN. The latter is addressed by monitoring sodium balance and supplementing appropriate quantities. Prolonged sodium losses have adverse consequences in neonates and lead to growth failure.²⁹ As a rule, well-hydrated SBS patients with a urinary sodium of <10 mEq/L tend to be sodium deficient. The measurement of electrolytes from stomas can also provide valuable data to aid with accurate sodium repletion.

6.2. Enteral nutrition

Prompt transition to enteral nutrition is the most important intervention in neonates with SBS as it obviates IFALD and CABS. The provision of continuous feeds through a nasogastric (or if significant reflux is present a nasojejunal tube) may facilitate full enteral feeds in neonates with SBS. In long term patients the surgical (or endoscopic) placement of a gastric tube is indicated. The ideal enteral nutrition formulation for neonates with SBS remains controversial. It has been shown that both breast milk and commercially available elemental formulas are associated with a reduction in the time of PN dependence in neonates with severe SBS.³

Patients with SBS often have an absent or compromised terminal ileum. The loss of terminal ileal function is associated with a depletion of fat-soluble vitamins (A, D, E, K), vitamin B₁₂ and zinc. Since vitamins and trace elements are provided in PN, these shortages are usually not evident until several months after weaning to full enteral nutrition. A deficiency state may be imminent or present even in a child with normal somatic growth.³⁰ Fully enterally fed SBS neonates should have an assessment of serum vitamin A, 25-OH vitamin D₃, vitamin E, Zn, and vitamin B₁₂ at one and three months after the cessation of PN. The 25-OH form of vitamin D₃ most accurately reflects hepatic stores that are the main repository for this essential nutrient. The patient's PT, partial thromboplastin time (PTT), and INR may indirectly gauge vitamin K status. A failure of the PT, PTT and INR to respond to vitamin K injection may reflect underlying IFALD.

Zinc losses are accentuated in SBS by diarrhea or excessive stomal output. Severe zinc deficiency results in acrodermatitis enteropathica, characterized by a rash of the face, hands, feet, and genitalia. In patients with SBS and associated bacterial overgrowth, vitamin B₁₂ levels may be spuriously normal as bacteria can produce a biologically inactive analogue. In order to more accurately assess for vitamin B₁₂ deficiency, serum methylmalonic acid and homocysteine levels are also obtained.³¹

Convenient commercially available enteral preparations of the fat-soluble vitamins (usually including zinc) are available. Vitamin B₁₂ must be administered by injection or in older children via an intranasal route.

6.3. Prokinetic agents

In SBS where bowel motility disorders are present, the use of prokinetic agents may be indicated. This is a relatively frequent finding in SBS neonates with gastroschisis. Erythromycin administered orally or directly into the stomach increases gastric emptying and improves antroduodenal coordination. Studies in normal individuals demonstrate induction of phase III of the migrating motor complex.³² Azithromycin, a longer-acting analogue, may be utilized for the same purpose. However, tachyphylaxis to erythromycin and related drugs is common.

In neonates, the use of other agents that may promote motility is problematic. Octreotide may accentuate bowel ischemia, and metoclopramide can induce tardive dyskinesia. The latter complication has resulted in the FDA issuing a 'black box' warning regarding the protracted use of metoclopramide. Domperidone is available for the treatment of gastroparesis in the USA only by an investigational new drug application through the FDA because of concerns regarding cardiac arrhythmias. In 2000, cisapride was also withdrawn from the market in the USA due to concern for the induction of 'torsades de pointes' in susceptible individuals. It may be obtained from the manufacturer for selected patients who meet specific selection criteria, and follow a defined monitoring and dosage protocol. The reason that cisapride is still occasionally used, despite its potential for serious adverse-effects, is that it promotes motility in the small intestine as well as the stomach.³³

Fortuitously, aside from cases of true congenital intestinal pseudo-obstruction, motility issues tend to improve with time. It should be stressed that it is important to attempt oral feeding in neonates who are not aspiration risks as oral aversion is a risk for children with SBS. Clinically stable patients are often managed by continuous overnight g-tube drips and intermittent oral feeding during the day.

6.4. Controlling stool output

Neonates with SBS may have significant stool or stomal output that precludes transition to enteral feeds. Our practice is to advance

enteral feeds as long as stool or stomal output is <2 mL/kg/h. Continuous enteral feeds tend to cause less diarrhea in patients with SBS. If no mechanical or infectious issues are evident, loperamide may be used to decrease stool or stomal output. Stomal refeeding is also an effective strategy in patients with a long mucous fistula.

6.5. Hormonal therapy

Bowel adaptation in SBS is characterized by an increase in the mucosal surface area and often bowel dilation. The length of the bowel increases only in proportion to somatic growth and is not accentuated by SBS. The usual manner in which bowel adaptation is optimized is the administration of enteral nutrients.

Experimentally hormonal manipulation has also been attempted. The most promising of the hormonal therapies, at present, is the use of glucagon-like peptide 2 (GLP-2). This hormone acts on the bowel itself and its administration is associated with a marked increase in bowel adaptation, particularly in porcine models. The use of a long-acting GLP-2 analogue (teduglutide) significantly improved water and to a lesser degree nutrient absorption in adults with SBS.³⁴ Trials of teduglutide are ongoing, but no data regarding pediatric patients are yet available. A detracting aspect of GLP-2 therapy is that its effects are predicated upon continued administration of the agent. Theoretic concerns regarding the induction of gastrointestinal malignancy also exist.

7. Surgical treatment

The surgical treatment options for neonatal SBS include bowel conservation at the time of initial presentation, bowel lengthening operations and intestinal transplantation.

7.1. Bowel conservation

The initial surgical operation is aimed at limiting as much bowel loss as possible. In cases where the viability of the bowel is in question the use of a 'second look' operation within ~24 h may be employed. With this approach marginally viable intestine is left in situ, the patient is resuscitated and the bowel is then reassessed at a second operation. Only the frankly necrotic intestine is then excised. The use of a temporary transparent plastic 'silo' to cover the bowel is sometimes also helpful to avoid the risk of inducing abdominal compartment syndrome. Once bowel edema has subsided, delayed abdominal closure can be accomplished.

In the case of neonatal intestinal atresia and limited distal small intestine (usually <35 cm) a primary serial transverse enteroplasty (STEP) operation may be done. The dilated proximal segment is subject to the STEP procedure and then anastomosed to the distal bowel.³⁵ Details of this operation are provided in the section related to intestinal lengthening and tapering operations.

The prompt establishment of bowel continuity through stomal closure is associated with more rapid weaning from PN.³ Usually the surgeon will wait approximately six weeks between operations in an effort to minimize the vascularity of any adhesions that may have formed. In cases where the neonate is unstable, the time of surgery may be further delayed. In practical terms, the closure of stomas is often governed by the anticipated stress that the operation would place upon the ventilatory capacity of the neonate. A baby with severe underlying lung disease, in whom intra-operative evaporative water loss and attendant fluid replacement are anticipated to cause an exacerbation of pulmonary problems, is likely to have surgery deferred. Careful discussions between the surgeon, neonatologist and anesthesiologist are mandatory when considering the timing of surgery in ill SBS patients.

7.2. Intestinal lengthening operations

The longitudinal intestinal lengthening and tailoring operation (LILT) was introduced in 1980.^{36,37} This ingenious but technically challenging operation utilized the concept that dilated small intestine is supplied by two separate leaves of mesentery. By carefully splitting the mesentery and then severing and anastomosing the intestine in an isoperistaltic manner the bowel could then be lengthened and tapered. Other clever but even more complex operations were also suggested.^{38,39} In 2003, a conceptually simple and reproducible bowel-lengthening and -tapering operation called the serial transverse enteroplasty operation (STEP) was introduced.^{35,40} This procedure relies upon the alternate application of surgical stapling devices (used routinely in other operations) to dilated bowel using a transmesenteric transverse approach. A zig-zag lengthening and tapering of the intestine ensues (Figure 2). The salient advantages of this operation are that it is technically straightforward, it results in a uniform bowel channel regardless of variable underlying bowel dilation, and it can be repeated if the bowel subsequently redilates.⁴¹ Animal studies have shown that the STEP is associated with improved nutrient absorption, enhanced growth and an overall increase in bowel surface area as reflected by elevated serum citrulline levels.² The latter effect is felt to be secondary to bowel adaptation that follows a STEP. Interestingly, a rodent model of the STEP procedure has shown that the operation results in elevations of postprandial GLP-2 levels and increased GLP-2 receptor expression.⁴²

The STEP operation has been performed successfully and safely in many centers around the world.⁴³ The most recent compilation of >100 STEP patients was presented by the International STEP

Registry at the 2010 Annual Meeting of the American Academy of Pediatrics. In this review, ~50% of all patients with intestinal failure refractory to maximal medical management were successfully converted to full enteral tolerance after the operation. The median time to attaining full enteral feeding approached two years. In selected neonates with intestinal failure, the STEP bowel-lengthening and -tapering operation has become a useful surgical option.

7.3. Intestinal transplantation

The first successful combined liver–intestine transplant in a patient with SBS was reported in 1990.⁴⁴ Intestinal transplantation has become a technically feasible option for babies with SBS. However, in the neonatal setting its role is limited to infants with concomitant irreversible life-threatening hepatic and intestinal failure. As techniques to preserve hepatic function in PN-dependent neonates have improved, the necessity for this type of transplant has decreased. The 10-year patient and graft survival for small bowel transplants that include the liver are 42% and 39% respectively.⁴⁵ The major problems associated with these transplants remain infection, chronic rejection and post-transplant lymphoproliferative disease (PTLD). Hence, small bowel–liver transplant or its related procedure – the multivisceral transplant – remains an option of last resort.

Neonates with marked liver disease and severe intestinal failure should be referred to centers of excellence in both bowel rehabilitation and transplantation capability. The decision for transplant is a difficult and dynamic one. Once IFALD has been ameliorated there is no emergent reason for transplant. For the older PN-dependent individual with intestinal failure the decision to have an isolated

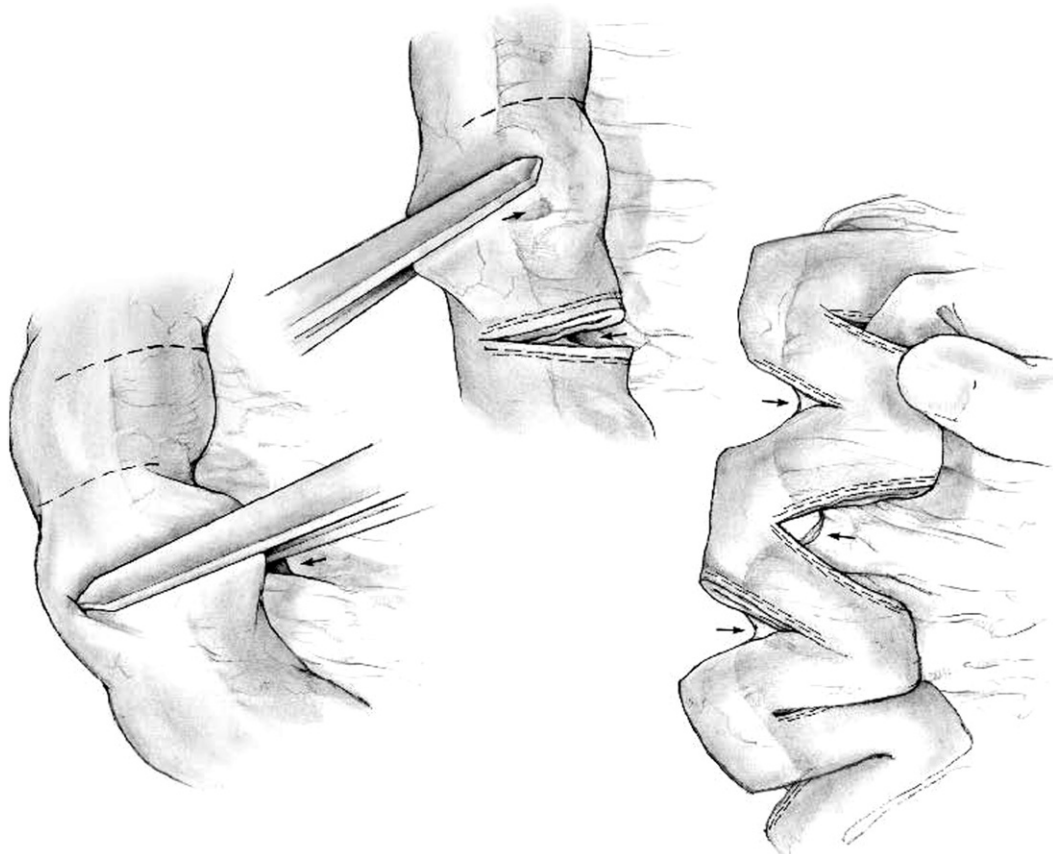


Figure 2. The serial transverse enteroplasty procedure involves the alternating application of a surgical stapling device to dilated bowel. The end result is a zig-zag lengthening and tapering of the bowel.

intestinal transplant is also not easy. The choice of an isolated intestinal transplant currently means accepting a reduction in survival for a potential improvement in quality of life. It is our current practice to offer such an option only to mature individuals and families who can accurately assess the risks and benefits of such an approach.

8. Conclusion

Neonatal SBS is a disease with comparatively high morbidity and mortality. The mainstay of management remains intestinal rehabilitation and, with the evolution of specialized intestinal failure centers, long term survival rates of about 90% are expected. A large majority of surviving neonates will transition to full enteral nutrition with appropriate management although the process may take years. Recent advances in hepatoprotective PN regimens and a reduction in CABSII has had a further beneficial effect upon outcomes. The use of hormonal modulation to facilitate intestinal adaptation remains an area of active investigation. In selected patients newer surgical bowel-lengthening and -tapering procedures, such as the STEP operation, appear to be safe and beneficial. Intestinal transplantation is reserved as a life-saving salvage option for a small group of ill neonates with irreversible hepatic and intestinal failure.

Practice points

- Neonatal intestinal failure is defined as intrinsic bowel disease resulting in an inability to sustain growth, hydration, or electrolyte homeostasis.
- Mortality associated with SBS has a bimodal distribution. Mortality is seen in the early postoperative period from complications associated with the underlying disease process. Long term survivors experience delayed complications from IFALD and sepsis.
- The current management of patients with intestinal failure focuses on nutritional, pharmacologic, and surgical interventions to achieve full enteral nutrition while minimizing the complications of PN therapy.
- Residual small bowel length, presence of ileum, and serum citrulline can be used as predictors for enteral feeding autonomy.
- Bowel-lengthening and -tapering procedures may be beneficial in selected patients.

Conflict of interest statement

The authors declare no conflict of interest related to this manuscript.

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