

Short Bowel Syndrome (SBS):

"A journey through the age spectrum using evidence-based practice"

Toolkit for Healthcare Professionals



This toolkit was developed by **Skymedcare** in collaboration with **John DiBaise, MD** (Mayo Clinic, Phoenix, AZ), **Stephanie Oliveira, MD, CNSC** (Cincinnati Children's Hospital, Cincinnati, OH), and **Carol Rees Parrish, MS, RDN** (retired from UVA Health, Charlottesville, VA).

This activity was supported by an independent medical grant by **Takeda**. You can access this activity's four patient vignettes and accompanying roundtable discussion at **My-lme.com**.

FAST FACTS ABOUT SBS¹⁻⁷

- SBS is a rare malabsorptive condition that occurs due to physical and functional loss of portions of the small intestine, affecting <50,000 people in the US.
- Clinical manifestations include diarrhea, dehydration, nutrient imbalances, and malnutrition, often requiring specialized nutritional therapy.
- Although SBS can occur congenitally, it is more often acquired due to surgical resections:

CAUSES OF SBS^{1,5-7}

Extensive small bowel resection:

CHILDREN

ADULTS

Congenital/perinatal disease (~80%)

Necrotizing enterocolitis
Malrotation with midgut volvulus
Jejunal/ileal atresia
Gastroschisis
Extensive aganglionosis
Long-segment Hirschsprung's disease

Non-natal (~20%) Children – Young adult

Trauma
Surgical complications
Volvulus
Crohn's disease
Vascular thrombosis
Malignancies

Surgical complications
Crohn's disease
Mesenteric ischemia
Volvulus
Adhesive obstruction
Trauma
Malignancies

HOW IS SBS DIAGNOSED?^{2,4,12}

- Diagnosing SBS is challenging due to its variable symptoms.
- SBS is diagnosed clinically in patients usually manifesting fluid, electrolyte, micronutrient imbalance/abnormalities, growth failure in children, or bone disease, and generally in the context of:

1 Specific vitamin and micronutrient deficiencies may give a clue to the diagnosis.²

CHILDREN

"An intestinal length of <25% of normally expected by age¹³ or the need for PN for >60 days after intestinal resection"¹⁴

ADULTS

"A small intestinal length of <200 cm in continuity with or without colon"^{3,13}

CLINICAL TESTS AND WORK UP^{1,2,4,8,11,12,15,16}

- Remnant bowel anatomy (length and region).
- Residual function (absorptive capacity).

This will help:

- ✓ Refining diagnosis.
- ✓ Establish nutritional prognosis.
- ✓ Design a plan for intestinal **rehabilitation** and need for surgical/pharmacological management.

Tests and investigations may include:

- Medical history (including intraoperative reports) and physical exam.
- Abdominal CT scan, MRI, ultrasound, or plain X-rays (i.e., for obstructions).
- Stool and fecal fat tests (stool frequency, stool/ostomy volume, 24-h steatorrhea).
- CBC, vitamins and minerals as indicated.

SBS can be **classified** by pathophysiological and anatomical criteria:^{2,3,8,9-11}

PATHO- PHYSIOLOGICAL CLASSIFICATION	WITHOUT colon in continuity	WITH colon in continuity																						
	End-jejunostomy (Type 1)	Jejuno-colonic (Type 2)	Jejuno-ileo-colonic (Type 3)																					
Resection	 ILEUM (ALL) + COLON	 ILEUM (MOST)	 JEJUNUM + ILEUM (retaining ≥10 cm of terminal ileum)																					
Colon	NO	PARTIAL	YES																					
Ileocecal valve	NO	NO	YES																					
Nutritional deficiency & Clinical features	<table border="1"> <tr> <th colspan="3">Absorption</th> </tr> <tr> <td colspan="3">Vitamin B12 Fat-soluble vitamins (e.g., A, D, E, K) Bile acids and SCFAs Water Electrolytes Amino acids</td> </tr> <tr> <td colspan="3">Fluid/electrolyte/nutrient imbalance</td> </tr> <tr> <th colspan="3">GI hormone secretion</th> </tr> <tr> <td colspan="3">PYY GLP-1 GLP-2</td> </tr> <tr> <th colspan="3">Gastric emptying</th> </tr> <tr> <td>Immediate dehydration post-surgery, hypotension, hypomagnesemia, renal failure risk.</td> <td>Weight loss, progressive severe malnutrition, diarrhea, steatorrhea, SIBO.</td> <td>Malnutrition rare.</td> </tr> </table>			Absorption			Vitamin B12 Fat-soluble vitamins (e.g., A, D, E, K) Bile acids and SCFAs Water Electrolytes Amino acids			Fluid/electrolyte/nutrient imbalance			GI hormone secretion			PYY GLP-1 GLP-2			Gastric emptying			Immediate dehydration post-surgery, hypotension, hypomagnesemia, renal failure risk.	Weight loss, progressive severe malnutrition, diarrhea, steatorrhea, SIBO.	Malnutrition rare.
	Absorption																							
Vitamin B12 Fat-soluble vitamins (e.g., A, D, E, K) Bile acids and SCFAs Water Electrolytes Amino acids																								
Fluid/electrolyte/nutrient imbalance																								
GI hormone secretion																								
PYY GLP-1 GLP-2																								
Gastric emptying																								
Immediate dehydration post-surgery, hypotension, hypomagnesemia, renal failure risk.	Weight loss, progressive severe malnutrition, diarrhea, steatorrhea, SIBO.	Malnutrition rare.																						
Use of PN	High risk of permanent dependence.	Mostly supplemental to oral/enteral feeding.	Usually not needed.																					

SBS COMPLICATIONS^{3,4,8,15,17}

- SBS may lead to severe and life-threatening complications.
- SBS is the leading pathophysiological cause of chronic IF (SBS-IF), accounting for ~2/3 of adults and ~1/2 of children of those with chronic IF.

"Persistent reduction of gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes such that IVS to maintain health and/or growth", in a patient who is metabolically stable.^{3,18}

- PN is a life-saving treatment in patients with SBS-IF, but it is **associated with morbidity, mortality, and a reduced QoL**.^{5,10,15}

ASSOCIATED COMPLICATIONS	CVAD	PN	Psychosocial problems
	Sepsis Occlusion Breakage Deep vein thrombosis	IFALD (i.e., steatosis, cholestasis, cirrhosis) Biliary complications (e.g., gallstones) Metabolic bone disease (e.g., low bone calcium uptake)	Impaired body image Reduced school attendance Reduced QoL of patients and caregivers

SBS MANAGEMENT GOALS^{6,16,19}



Optimize/maintain essential hydration and nutrition and, in children, promote normal growth and development.

Promote intestinal adaptation (restore absorptive capacity of the remnant bowel).



ACHIEVE FULL ENTERAL AUTONOMY

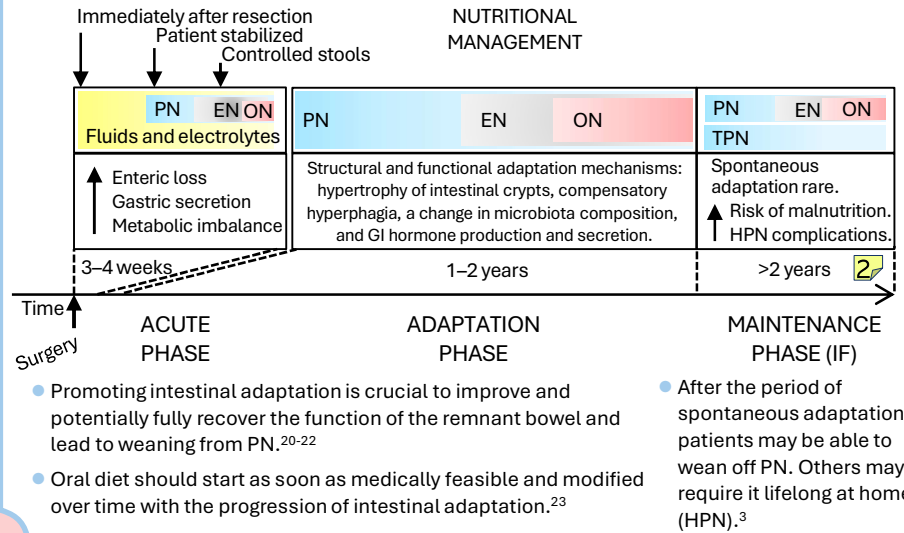
Reduce or eliminate the long-term dependence on PN and its associated complications.



Improve QoL of patients and their caregivers.



Post-operatively, SBS evolves through **three phases**, each requiring a specific nutritional approach.^{4,5,20,21}



HOW ARE THESE GOALS ACHIEVED?

The management of SBS requires a **multidisciplinary approach** consisting of:

1 **A multidisciplinary team** familiar with the management of SBS^{12,15,17}
Including adult and pediatric specialists, dietitians, nurses, gastroenterologists, and surgeons.

2 **A variety of treatments to achieve enteral independence:**^{15,16,20}

Nutrition and hydration ^{8,20,22,25}	Pharmacological ^{2,8,17,20,24}	Surgical ^{20,22}
Nutrient, fluids, and electrolyte supplementation by intravenous, oral, subcutaneous, or intramuscular route, and based on remnant bowel and colon in continuity status. Calcium carbonate, magnesium oxide, potassium citrate, ferrous sulfate, ORS, soluble fiber, fat with high EFA content, vitamins (A, B12, C, D, K), etc.	Symptomatic treatments for complications. Antidiarrheal/antimotility agents: e.g., loperamide, codeine. Antisecretory agents: e.g., clonidine, ranitidine, omeprazole, lansoprazole. Bile salt binders: e.g., cholestyramine. Antibiotics: e.g., rifaximin. SBS-specific disease-modifying treatments after the adaptive period and PN weaning not achieved. rhGH*: somatropin. GLP-2 analog: teduglutide.	Reconstructive (to preserve/maximize function or remnant bowel in adults or increase length in children). Intestinal transplant for irreversible chronic IF.

*Somatropin; only registered for SBS in the US, but largely discontinued due to side effects.

2 **Intestinal growth factors** should be considered in SBS-IF patients requiring PN continuation if they are **stable after a period of post-surgery intestinal adaptation** (i.e., >1-2 years post surgery).^{8,18,24}

3 **Patient/caregiver education**^{8,22}
E.g., about the disease, HPN care, dietary management, regular monitoring, benefits/risks.

Intestintrophic therapy with the GLP-2 analog (teduglutide)

- Teduglutide is largely the only medication*, and the first GLP-2 analog, approved for the treatment of SBS (first approved by the US FDA and the EMA in 2012).^{20,24}
- Teduglutide is indicated for the treatment of adults and pediatric patients ≥ 1 year of age with SBS who are dependent on PN support.²⁶

Guidance	Recommendations
AGA clinical practice on SBS management - expert review (2022) ⁸	A series of best practice advice that included the use of teduglutide as a first choice in carefully-selected adult patients with SBS-IF with an unsuccessful or incomplete attempt to wean PN support.
ESPEN guidelines on chronic IF (2023) ¹⁸	Teduglutide as the first-choice in carefully-selected adult SBS patients who are candidates for intestinal growth factor treatment [grade of recommendation A].

WEANING OFF PN²³



- **Incremental, step-wise PN reduction to the minimum necessary** to maintain hydration and health (e.g., by reducing weekly infusion days or the daily volume and nutrient concentration).

Based on the ability to maintain appropriate hydration i.e., urinary volume output 1-2 L/day (adults) or until ≥75% of EN is tolerated (children) and to adhere to enteral/oral nutritional supplementation.^{15,23,27}

WHEN?

THE FUTURE OF SBS^{17,24}

Under development:

- Longer-acting GLP-2 analogues (glepaglutide, apraglutide).
- Long-acting GLP-1 analogue (vurolenatide).

Further research required regarding:

- Early treatment with GI hormones.
- Combinations (GLP-1/GLP-2 analogues, or other GI hormones).

Abbreviations: AGA, American Gastroenterological Association; CBC, complete blood count; CT, computed tomography; CVAD, central venous access device; EFA, essential fatty acids; EMA, European Medicines Agency; EN, enteral nutrition; ESPEN, European Society for Clinical Nutrition and Metabolism; FDA, Food and Drug Administration; GI, gastrointestinal; GLP, glucagon-like peptide; HPN, home parenteral nutrition; IF, intestinal failure; IFALD, intestinal failure-associated liver disease; MRI, magnetic resonance imaging; ON, oral nutrition; ORS, Oral rehydration solution; PN, parenteral nutrition; PYY, peptide YY; QoL, quality of life; rhGH, recombinant human growth hormone; SBS, Short Bowel Syndrome; SCFA, short-chain fatty acid; SIBO, small intestinal bacterial overgrowth; US, United States.

References: 1) Flahive CB, et al. *Curr Treat Options Peds*. 2015;7(1):1-16. 2) Short bowel syndrome. National Organization for Rare Disorders. Updated Feb 01, 2021. Available from: <https://rarediseases.org/rare-diseases/short-bowel-syndrome/>. 3) Pironi L. *Nutr Clin Pract*. 2023;38 Suppl 1:S9-S16. 4) Guillen B, Atherton NS. Short Bowel Syndrome. *StatPearls*. Updated Jul 17, 2023. 5) Massironi S, et al. *Dig Liver Dis*. 2020;52(3):253-61. 6) Muff JL, et al. *Children (Basel)*. 2022;9(7):1024. 7) Thompson JS. *Viszeralmedizin*. 2014;30(3):174-8. 8) Iyer K, et al. *Clin Gastroenterol Hepatol*. 2022;20(10):2185-94.e2. 9) Ber Y. *Nutrients*. 2021;13(7):2325. 10) Bering J, DiBaise JK. *Nutr Clin Pract*. 2023;38 Suppl 1:S46-58. 11) Aksan A. *World J Gastroenterol*. 2021;27(24):3440-65. 12) Fuglestad MA, Thompson JS. *Surg Clin North Am*. 2019;99(6):1209-21. 13) DA96.04 Short bowel syndrome. WHO ICD-11 MMS. 14) Merritt RJ, et al. *J Pediatr Gastroenterol Nutr*. 2017;65(5):588-96. 15) Parrish CR, DiBaise JK. *Gastroenterol Hepatol (N Y)*. 2017;13(10):600-8. 16) Short bowel syndrome (SBS) - AGA GI Patient Center. Available from: <https://patient.gastro.org/short-bowel-syndrome-sbs/>. 17) DiBaise JK. *Nutr Clin Pract*. 2023;38 Suppl 1:S4-8. 18) Pironi L, et al. *Clin Nutr*. 2023;42(10):1940-2021. 19) Managing Short bowel Syndrome - Diet & oral rehydration solution. Available from: <https://www.shortbowelsyndrome.com/sbs-management>. 20) Pironi L, et al. *Expert Opin Drug Saf*. 2021;20(12):1501-13. 21) de Dreuille B, Joly F. *Curr Opin Pharmacol*. 2022;65:102240. 22) Pironi L, et al. *Nutr Clin Pract*. 2024;39(1):141-53. 23) Roberts K, et al. *Nutr Clin Pract*. 2023;38 Suppl 1:S59-75. 24) Wauters L, Joly F. *Nutr Clin Pract*. 2023;38 Suppl 1:S76-87. 25) Ukleja A. *Gastroenterol Clin North Am*. 2019;48(4):525-50. 26) US Food and Drug Administration (FDA). GATTEX (teduglutide) for injection, for subcutaneous use - Prescribing information. Updated Feb 21, 2024. 27) Guideline on the use of parenteral nutrition in neonatal and paediatric units (2023). Royal College of Physicians of Ireland. Available from: <https://www.rcpi.ie/Faculties-Institutes/Faculty-of-Paediatrics>.