ICD-10-CM: Short Bowel Syndrome and Intestinal Failure Proposal Overview September 8-9, 2020

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Disclosures

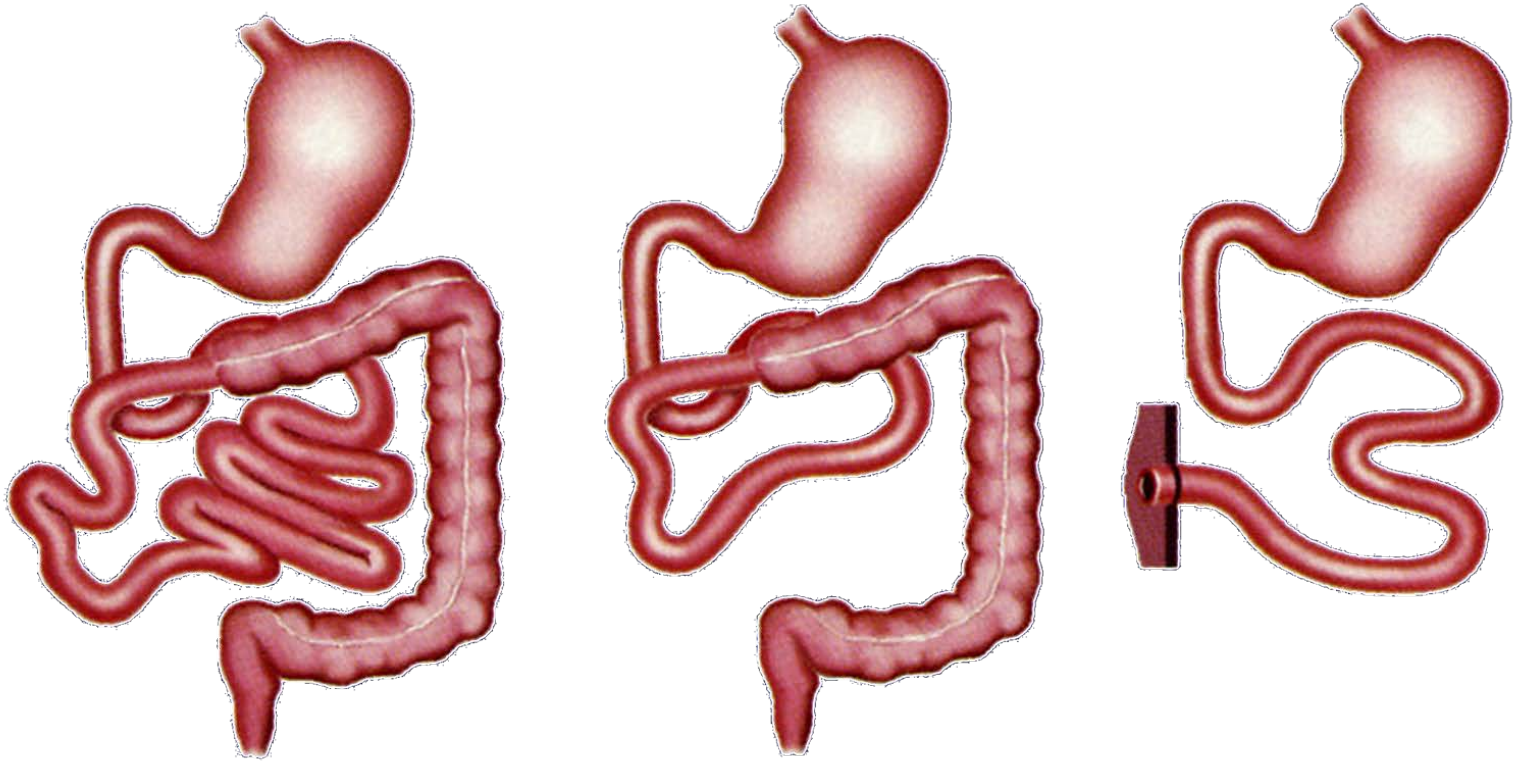
- Anthem Health – employee
- Takeda Pharmaceuticals – consultant

Agenda

- Overview of Short Bowel Syndrome (SBS) and Intestinal Failure (IF)
- Describe the relationship between SBS and IF
- Explain why new and appropriate codes for SBS and IF are critically important
- WHO MSAC recommends approval for ICD-11 codes for SBS & IF

How do patients typically develop ***Short Bowel Syndrome***?

Common types of resection/anastomosis occurring in patients with **Short Bowel Syndrome**



Ileocolonic Anastomosis

Jejunocolonic Anastomosis

End-Jejunostomy

Short bowel syndrome (SBS) is the result of physical loss and functional deficiency of portions of the intestine, primarily due to surgical resection

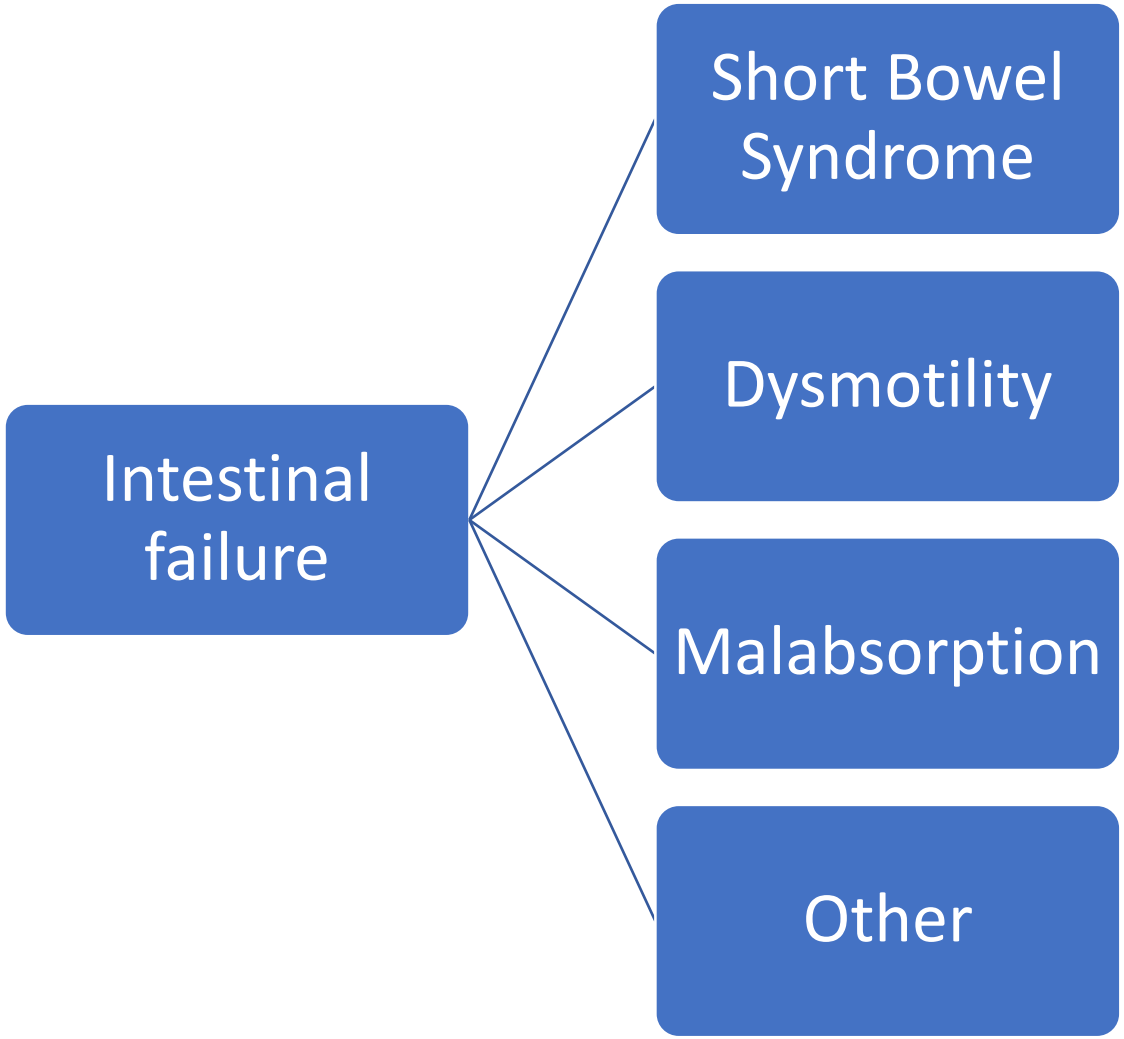
Malabsorptive spectrum of SBS is wide. Clinical features include:

- Malnutrition
- Dehydration
- Electrolyte disturbances
- Diarrhea/Increased outputs

Common causes of SBS include:

Adults	Children
Crohn's disease	Necrotizing enterocolitis
Vascular events	Midgut volvulus
Bariatric surgery complications	Intestinal atresia
Trauma	Gastroschisis
Volvulus	Malrotation
Malignancy	Trauma
Strangulated hernia	Hirschsprung's disease
Small bowel fistula	Vascular events

Intestinal Failure is the inability to absorb sufficient nutrients and/or fluid necessary to maintain nutritional autonomy



Short Bowel Syndrome (SBS) & **Intestinal failure:** A Relationship

Some SBS patients do not require parenteral support any longer, but have a history of parenteral support

Some SBS patients require parenteral support

Short Bowel Syndrome

Intestinal Failure

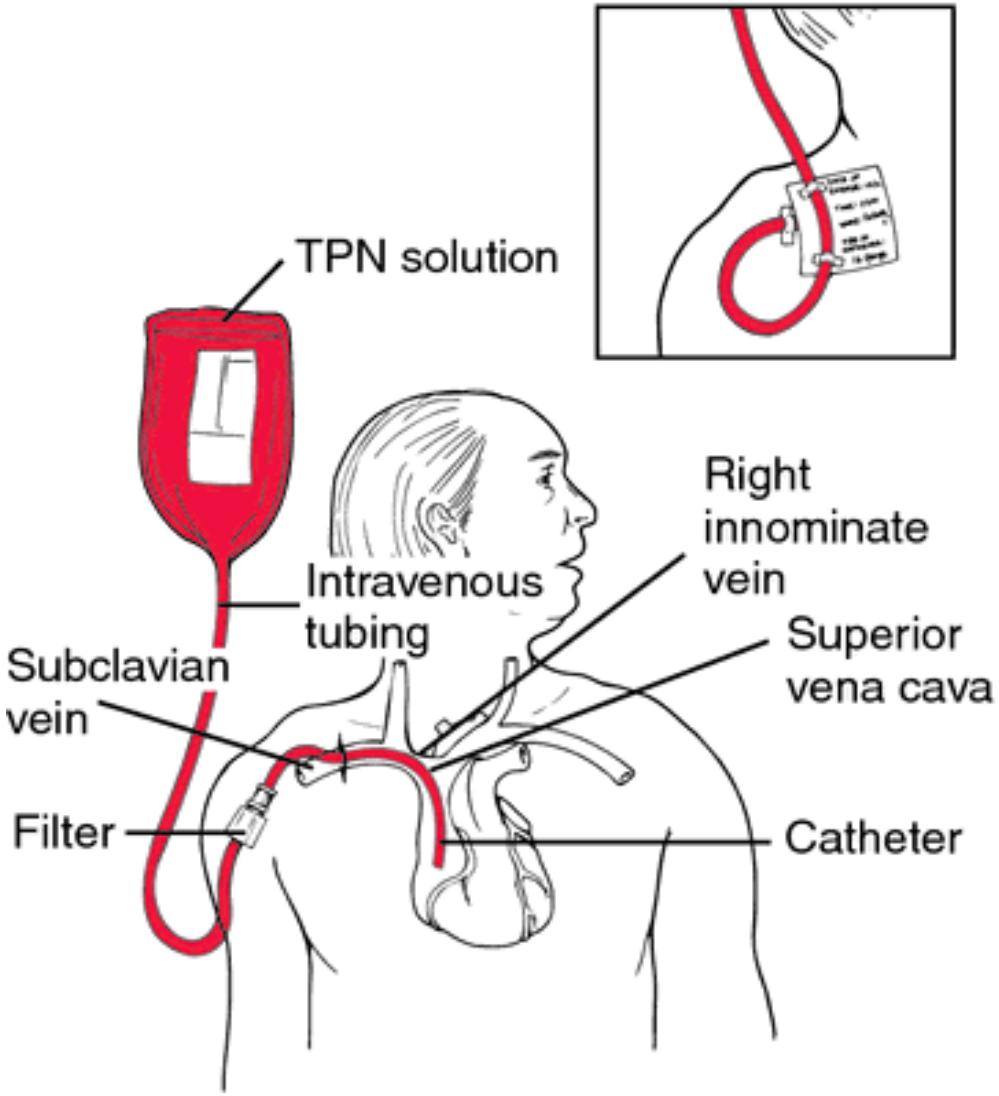
The reduction of gut function below the minimum necessary for the absorption of macronutrients and/or water and electrolytes, such that intravenous supplementation is required to maintain health and/or growth

Having less than 200 cm of residual small bowel with or without colon in an adult and for children (< 18 yrs), less than 25% of the normal length of intestine for their respective age

FYI: Some IF patients require parenteral support, but do not have SBS

What happens in **Intestinal Failure?**

Amount of physical loss and functional deficiency of intestine function will impact degree of parenteral support required



Management of **SBS** & **IF** are complex

- Parenteral support is a life-saving therapy (lasting months to years)
- Care involves prevention & treatment of:
  - Nutritional deficiencies
  - Dehydration
  - Central line infections
  - Complications impacting multiple organs (e.g. liver, kidney, bones, brain)

Duggan CP, Jaksic T. Pediatric Intestinal Failure. *N Engl J Med*. 2017;377(7):666-675.

Short Bowel Syndrome and **Intestinal Failure** are associated with high healthcare utilization

- Research studies constrained without specific codes
  - SBS is a leading indication for isolated small bowel transplant & combined small bowel/liver transplant[1]
  - Average hospitalization stay was approximately 11 days
  - Average re-admission for patients that receive home parenteral nutrition rate is 1-2 times per year, although those with SBS cannot be selected out (Oley Foundation data)

1. Organ Procurement and Transplantation Network. Reasons for Intestine Transplants. https://optn.transplant.hrsa.gov/data/organ-datasource/intestine/ [Accessed 30 January 2020]
2. Raphael BP et al. Analysis of Healthcare Institutional Costs of Pediatric Home Parenteral Nutrition Central Line Infections. J Pediatr Gastroenterol Nutr. 2018 Oct;67(4):e77-e81.

Professional society position statements and clinical guidelines in place for **Short Bowel Syndrome** and **Intestinal Failure,** yet No ICD-10-CM code exists

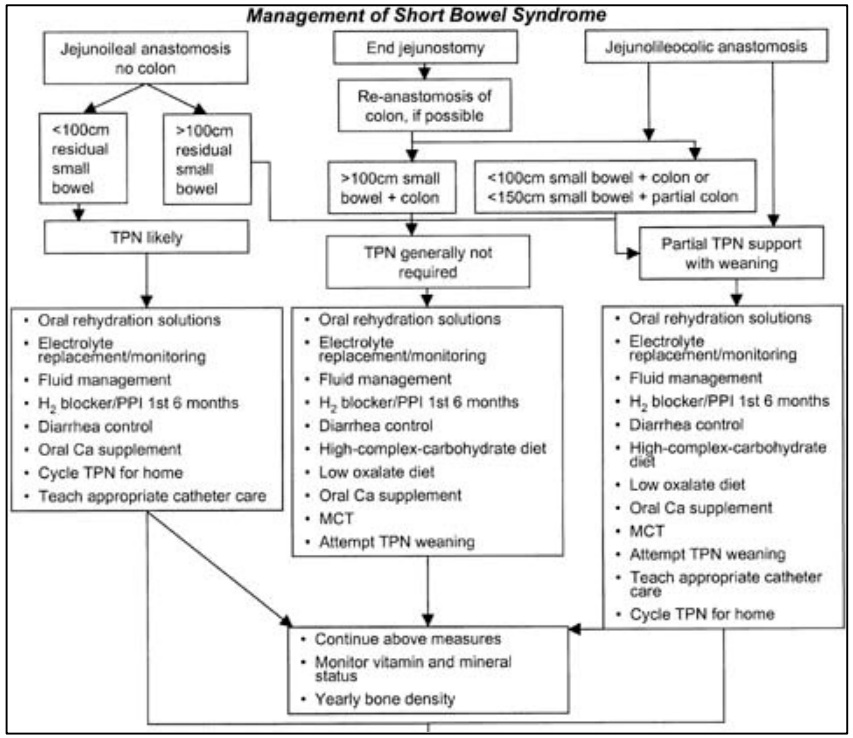


Table 6
Complete list of statements on chronic intestinal failure due to benign disease (absence of end stage cancer disease).

#	Statement	Grade of evidence	Strength of recommendation
	Management of home parenteral nutrition for benign chronic intestinal failure		
1	We recommend that the aims of an HPN programme include provision of evidence-based therapy, prevention of HPN–related complications such as catheter-related infections and metabolic complications and ensure quality of life is maximized.	Very low	Strong
2	We recommend regular audit of therapy and outcomes against standards to ensure safety and efficacy of an HPN programme.	Very low	Strong
3	We recommend that patients selected for an HPN programme have confirmed intestinal failure that	Very low	Strong

Table 1. Nutrition Support Clinical Guideline Recommendations in Pediatric Patients With Intestinal Failure.

Question	Recommendation	Grade
1. Is ethanol lock effective in preventing bloodstream infection and catheter removal in children at risk of parenteral nutrition–associated liver disease (PNALD)?	A suggestion is made to use ethanol bloodstream infection (CLABSI) in children at risk of PNALD. Th and catheter removal is low and v effect of both decreased infection interpreted in light of the unknow formation and disruption of cathe	
2. What fat emulsion strategies can be used in pediatric patients with intestinal failure to reduce	Since the only fat emulsion in the U (SOE), a suggestion is made to re to treat cholestasis in children wi supporting this recommendation	

Professionals	Role and services
Pediatric surgeons	Gastrointestinal surgery, central venous catheter procedures. Inpatient and outpatient surgical management
Transplant surgeons	Assessment, surgery, immunosuppression
Pediatric gastroenterologists	Inpatient and outpatient medical management
Neonatologists	Initial inpatient management of premature and critically ill infants
Interventional radiologists	Central venous line management
Gastroenterology/parenteral nutrition nurses	Line and ostomy care, education
Pharmacists	Supervision, preparation of parenteral nutrition, drug-nutrient interactions
Registered dietitians	Nutritional monitoring and counseling, drug-nutrient interactions
Social workers	Access available resources; support
Physical/occupational/speech Therapists	Feeding, mobility and development
Child-life specialists	Child and family support, education
Psychologists	Individual treatment and family support
Medical educators	Instruction on self-care

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1. American Gastroenterological Association. American Gastroenterological Association medical position statement: short bowel syndrome and intestinal transplantation. Gastroenterology. 2003 Apr;124(4):1105-10
2. Pironi L. Home Artificial Nutrition & Chronic Intestinal Failure Special Interest Group of ESPEN. ESPEN guidelines on chronic intestinal failure in adults. Clin Nutr. 2016 Apr;35(2):247-307.
3. Wales PW. American Society for Parenteral and Enteral Nutrition, Teitelbaum D. A.S.P.E.N. clinical guidelines: support of pediatric patients with intestinal failure at risk of parenteral nutrition-associated liver disease. JPEN J Parenter Enteral Nutr. 2014 Jul;38(5):538-57.
4. Merritt RW, Cohran V, Raphael BP, Sentongo T, Volpert D, Warner BW, Goday PS, Nutrition Committee of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Intestinal Rehabilitation Programs in the Management of Pediatric Intestinal Failure and Short Bowel Syndrome. J Pediatr Gastroenterol Nutr. 2017 Nov;65(5):588-596.

What are the benefits of adding ICD-10-CM codes for **Short Bowel Syndrome** and **Intestinal Failure?**

- Improve data collection for epidemiological and outcomes research
- Measurement of outcomes and tracking care provided to patients
- Evaluation of frequency and severity of disease complications
- Quality improvement initiatives
- Identification of fraud and abuse

International Classification of Diseases, (ICD-10-CM/PCS) Transition – Background. https://www.cdc.gov/nchs/icd/icd10cm_pcs_background.htm Accessed 2/10/2020.

Summary

- **Short Bowel Syndrome** and **Intestinal Failure** are unique diseases with complex management, associated with high healthcare utilization
- Creation of Unique ICD-10-CM codes for Short Bowel Syndrome and Intestinal Failure will facilitate improved patient care and long-term health outcomes

THANK YOU

Appendix for Questions

Commonly used codes in lieu of Codes for **Short Bowel Syndrome** & **Intestinal Failure**

Codes	*Condition*	*Codes*	*Condition*
K90	Intestinal Absorption	**63.3**	Feeding difficulties
K90.0	Celiac disease	**K56.609**	Intestinal Obstruction
K90.1	Tropical sprue	**R63.0**	Anorexia
K90.2	Blind loop syndrome, not elsewhere classified	**E40**	Kwashiorkor
K90.3	Pancreatic steatorrhea	**E41**	Nutritional Marasmus
K90.4	Other malabsorption due to intolerance	**E43**	Unspecified protein-calorie malnutrition
K90.41	Non-celiac gluten sensitivity	**E44.0**	Moderate protein-calorie malnutrition
K90.49	Malabsorption due to intolerance, not elsewhere classified	**E44.1**	Mild protein-calorie malnutrition
K90.8	Other intestinal malabsorption	**E45**	Retarded development following protein-calorie malnutrition
K90.81	Whipple's disease	**E53.9**	Vitamin B deficiency, unspecified
K90.89	Other intestinal malabsorption	**E61.1**	Iron deficiency
K90.9	Intestinal malabsorption, unspecified	**E63.8**	Other specified nutritional deficiencies
K91	Intraoperative and postprocedural complications and disorders of digestive system, not elsewhere classified	**71.6**	Toxic liver disease with fibrosis and cirrhosis of liver
K91.2	Postsurgical malabsorption, not elsewhere specified	**76.9**	Liver disease unspecified