

# Short Bowel Syndrome

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## Pathophysiology

- Functional disorder caused by alterations of normal intestinal anatomy and physiology.
- Shortened bowel combined with malabsorption; dependent on parenteral nutrition >3 months.
- May result from: necrotizing enterocolitis, congenital bowel atresia, volvulus, gastroschisis, Hirschsprung's disease.
- After resection, the residual small bowel undergoes intestinal adaptation, stimulated by hormones and oral nutrients:
  - Mucosal hyperplasia.
  - Villus lengthening and increased crypt depth.
  - Bowel dilatation.
- Jejunum: long villi and large absorptive surface with high concentration of enzymes; site of greatest nutrient absorption.
  - If resected, will have transient or permanent nutrient losses.
  - Ileum can develop the absorptive capacity of the jejunum for various nutrients.
- Ileum: shorter villi, more lymphoid tissue, tighter epithelium; effective absorption of fluid and electrolytes; responsible for absorption of vitamin B12 and bile salts through receptors.
  - Other bowel will never develop ability to absorb B12 and bile salts.
  - Resection may impair bowel motility (many GI hormones produced in ileum).
  - Intestinal adaptation after massive ileal resection is more difficult than after jejunal.
- Malabsorption of rapidly digested carbohydrates produces osmotic diarrhea.
- Fat soluble vitamins may also be inadequately absorbed.
- No absolute number can be placed on the length of remaining bowel necessary for successful enteral nutrition; remaining bowel may be damaged and act dysfunctional.
- Best prognosis over time is for infants in whom the duodenum, distal ileum, and ileocecal valve can be preserved.



## Complications

- Long term TPN use
  - Hepatobiliary disease.
  - Catheter-associated sepsis.
  - Fluid and electrolyte imbalance.
  - Bacterial intestinal overgrowth.
- Malnutrition.
- Failure to thrive.
- Dumping syndrome: post-prandial tachycardia, diaphoresis, lethargy, watery diarrhea.
  - Marked increase in stools may indicate poor absorption, and enteral feedings should not be advanced.
  - If significant volume loss, rehydration therapy and electrolyte replacement will be needed.
- Mortality of 30-40% from sepsis or liver failure.

## Management/Feedings

- Recommend managing with consulting services in pediatric surgery, GI (and possibly the liver team), and neonatal dietitian.
- Attempt to wean off TPN as soon as possible.
- Enteral nutrition should be started promptly to promote intestinal adaptation.
- Usually started on elemental/semi-elemental diets containing free amino acids or small peptides, or breast milk.
  - There are no elemental preterm formulas.
  - Once they tolerate feeds, can gradually introduce a portion of preterm formula, fortifier, or supplements to improve the mineral and protein content of a term formula.
- These infants may have “leaky gut” with high rates of sensitization to cow’s milk or soy protein.
- High proportion of fat in long-chain fatty acids promotes more mucosal adaptation; but those with ileal resection may not be able to absorb these and may need more medium-chain fats.

- Feedings are started slowly and continuously to maximally saturate carrier proteins.
- Higher concentrated formulas may cause osmotic diarrhea.
- Oral feedings should be initiated promptly; may have solids at 4 months CGA (high in protein and fat and low in carbohydrate is preferred).
- For high stomas, may be beneficial to re-feed the proximal stoma effluent through the mucous fistula for additional nutrient and fluid absorption and to stimulate distal gut.
- For bacterial overgrowth, may need intermittent dosing of antibiotics that affect anaerobic bacteria.
- Infants with ileal resection are at risk for B12 deficiency; may need parenteral B12 every 1-3 months.
- Children with enterostomies or diarrhea are at risk for zinc deficiency and may need supplementation:
  - Poor growth, diarrhea, impaired wound healing, perianal and perioral skin rash, alopecia.
- Gastric acid hypersecretion:
  - More common in larger bowel resections and initiation of enteral feedings.
  - Can cause secretory diarrhea.
  - Ranitidine may help.

## **Surgical Treatments**

- Goals of surgery:
  - Slow intestinal transit.
  - Increase mucosal surface area.
  - Improve peristaltic function.
  - Increase intestinal length.
- May eventually require intestinal +/- liver transplant.