Short Bowel Syndrome

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.