Short Bowel Syndrome

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INTESTINAL FAILURE

• Designates a state of intestinal tract function that is inadequate to fulfill the nutrient and fluid requirements of the body.

• Implicit to the concept of IF is prolonged and potentially permanent requirement for parenteral nutrition (PN)

• Outcomes: recovery (ending PN) vs survival (on PN)

• If you need PN for >3 to 5 years will need it for life
Intestinal Failure

- Short bowel syndrome
- Persistent motility disorders
  - Long segment Hirschsprung’s disease
  - Intestinal pseudo-obstruction
  - Mitochondrial disorders
- Transport defects
  - Congenital chloride diarrhoea
  - Glucose-galactose malabsorption
- Epithelial dysplasias
  - Microvillus inclusion disease
  - Tufting enteropathy
Short Bowel Syndrome (SBS)

- Definition
- Etiology
- Physiologic Abnormalities
- Intestinal Adaptation
- Clinical Management
- Chronic Complications
- Surgical Options
- Transplant
Malabsorption in the presence of shortened small intestine.

Malabsorption includes nutrients, fluid and electrolytes (* nutrients).

Definition

Numerous attempts to anatomically define SBS.

*Canadian Assoc. of Paediatrics: SBS as the need for TPN greater than 42 days after bowel resection or a residual small bowel length of less than 25% expected for gestational age*

Functional definition is generally accepted.

- Malabsorption in the presence of shortened small intestine.
- Malabsorption includes nutrients, fluid and electrolytes (* nutrients)
Etiology

A syndrome with multiple etiologies.

- Gastroschisis
- Bowel atresia
- Tumors
- Volvulus
- NEC
- Hirschprung
- Radiation
- Crohn's
- SBS
Necrotising enterocolitis
NEONATES

Gastroschisis

Abdominal wall defects

Omphalocele
Physiologic/Anatomical differences

Functions of the small bowel vary by segment

Proximal (jejunum)

- Long villi and large absorptive surface
- [ ] of enzymes and transport proteins
- Large tight junctions
  ie. Epith. more porous
- Site of greatest nutrient absorption in the SI, leaky allows free and rapid flow of H2O and electrolytes
- Fluid and electrolytes flow plasma—lumen to dilute [ ] nutrient delivered from duodenum
- Mixing, digestion and subsequent carrier mediated transport of monosaccharidases, aa and dipeptides occur in the jejunum.

Distal (ileum)

- Shorter villi
- Lymphoid tissue
- Tighter junctions
- Efficient for absorption of fluid & electrolytes
- Absorption of Vit B12 and bile salts through site specific receptors
- Patient with a jejunostomy (ie ileal resection) susceptible to fluid losses from osmotic diarrhoea
- Ileum site of synthesis of GIT hormones affecting SI motility such as enteroglucagon and peptide YY.
- Resection of ileum impairs regulation of gut motility by nutrient ie. Fat-ileal brake
Physiologic/Anatomical differences

Functions of the small bowel vary by segment

- Resection of ileocaecal valve absences of 2 functions
  - barrier for reflux of colonic bacteria
  - regulates exit of fluid nutrient from the SI

Summary and Other Function

- Major consequence of resection of SI:- malabsorption due to reduction in absorptive area + loss of digestive enzymes and transporters
- Malabsorption of rapidly digested CHO produces tremendous osmotic diarrhoea. Proteins: larger molecule ingested in smaller quantities- less osmotic effect. Fats extremely large molecules less well absorbed little osmotic effect. Fat soluble vitamins are malabsorbed in SBS
- Extensive ileal resection cause impairment reabsorption of bile salts. Drops below critical micelar [ ]-preventing solubilazation of fat and fat soluble vitamins.
- Abnormalities in motility following resection ie Following ileal resection transit time faster thru jejunum and gastric emptying is more rapid
Intestinal Adaptation

Adaptation is the compensatory overexpression of the process that maintains normal mucosal integrity and function in response to enteric feeding.

Successful medical management of SBS is ultimately dependent on stimulation of the process of ADAPTATION.

Proliferative status of the intestinal epithelium is an important determinant of adaptation. Starvation decreases cell proliferation rate and increases duration of the cell cycle (concept of trophic feeding).
Changes that occur with adaptation

Hyperplasia of mucosal epithelium

Hyperplasia is preceded by increase in crypt cell production, increase in crypt depth and subsequent lengthening of the villi.

Dilatation increasing surface area

Duration: Months to years
Intestinal adaptation

A

Normal mucosa

B

Villus and crypt hyperplasia
Factors in adaptation

- Enteral nutrition: hydrolysed casein > trophic. LCT>MCT, SCFA, Fiber, Omega 3 FA
- Hormonal regulation eg enteroglucagon, gastrin, secretin, CCK. EGF, ILGF-1 Peptide YY
- Prostaglandins eg. PGE2 analogs in rat studies
- Polyamines- polycationic compounds eg. Putrescine formed from decarboxylation of ornithine by enzyme ODC which rises in proliferating tissue ie small intestinal epithelium
Synthesis and degradation of Polyamines

FIGURE 40.1-3 A diagram showing the synthesis and degradation of polyamines. DAO = diamine oxidase; ODC = ornithine decarboxylase.
Clinical Management
Chronic complications

- Bacterial Overgrowth
- Watery diarrhoea
- Nutritional deficiency state
- TPN induced liver disease
- Catheter Related Complication
**Chronic Complications: Bacterial Overgrowth**

- Least recognized most treatable.
- N bacterial no.s in SI $10^3$ proximally to increasing in ileum.
- SBBO bacterial content $>10^5$ proximally:
- Predisposing factors: Slow motility, dilated bowel and absent ileocaecal valve
- These bacteria deconjugate bile salts impairing micellar solubilization resulting in steatorrhoea and malabsorption of fat soluble vitamins.
- Can also cause mucosal inflammation: worsening nutrient absorption
- Competes with host for B 12

**CONSIDER WHEN....**

- Bloating
- Cramps
- Diarrhoea or
- GI blood loss

Common cause of clinical deterioration in previously stable patients with SBS.
### Chronic Complications - Bacterial Overgrowth

**DIAGNOSIS**
- Demonstrate increased bacterial content in SI aspiration
- Breath hydrogen test: Glucose or lactulose: high fasting breath H2 level or rapid rise provided transit time thru SI not to rapid
- Urine indican
- Small bowel biopsy showing inflammatory changes
- *Other complications* D-lactic acidosis and small bowel colitis

**TREATMENT**
- Broad spectrum antibiotics
- Oral flagyl
- Oral gentamicin
- Bactrim
Chronic Complications:
Watery Diarrhoea

Results from excessive fluid secretion secondary to:
- Osmotic load when fed CHO
- Elevated gastrin levels hence a role for somatostatin analogs.
- Increase bile acid in the colon following resection of the ileum: Px cholestyramine. But in extensive ileal resection patients may have a deficiency and hence cholestyramine worsened steatorrhoea
Chronic Complications- TPN induced liver disease

- Major cause of death in children with SBS.

- Mech is unknown. Multifactorial?
  1) Toxicity of AA, lipid ???
  2) Toxin in unused bowel
  3) Lack of biliary secretions due to poor stimulus.

- Aim to have at least 30% enteral, prevent SBBO and decrease catheter related sepsis.
Catheter-Related Complication

- Infection/ Sepsis eg. Gram positive, negative, fungal
- Thrombosis- most patients on warfarin
- Loss of access
Surgical Options

- Repair of anastomotic strictures eg stricturoplasty
- Repair of dilated loops of bowel eg plication, tapering enteroplasty
- Procedure to decrease transit eg. reverse segment of bowel
- Bowel lengthening eg Bianchi procedure or STEP
FIGURE 40.3-2 Intestinal plication. A portion of the dilated bowel wall is inverted into the bowel lumen and secured by seromuscular sutures.

FIGURE 40.3-4 Taping enteroplasty. The antimesenteric portion of the dilated segment of bowel is removed. A portion of the absorptive surface is lost.

FIGURE 40.3-7 Serial transverse enteroplasty. Zigzag pattern of bowel created after sequential and opposite firings of a surgical stapler across the dilated bowel loop. The channel size is determined by the surgeon. The normal loops of bowel can be used as an internal guide. A gastrointestinal contrast study is performed on the seventh postoperative day prior to initiation of enteral feeds.
FIGURE 40.3-4 A. The mesenteric blood vessels within the two leaves of the mesentery are separated under the dilated bowel segment. 

B. The dilated bowel is divided longitudinally within the mesenteric leaves to create two parallel loops of bowel half the diameter of the original loop. 

C. The two bowel segments are anastomosed end to end to create a bowel segment double the length of the original. The bowel mesentery needs to be amenable to mobilization for this to be feasible. The bowel at “a” is the most proximal portion and the bowel at “c” is the most distal. The end of the reduced loop at “b1” is anastomosed to the proximal end of the other loop at “b2” to create an isoperistaltic segment of tapered and lengthened bowel.
### TABLE 40.2-1 INDICATIONS FOR INTESTINAL TRANSPLANT

<table>
<thead>
<tr>
<th>LIFE-THREATENING COMPLICATIONS ARISING FROM PARENTERAL NUTRITION THERAPY</th>
<th>TYPE OF TRANSPLANT</th>
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<tbody>
<tr>
<td>Impending loss of venous access, ie, when 2 of the 4 available sites have been lost in infants or 3 of 6 in older children</td>
<td>Isolated bowel transplant</td>
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<tr>
<td>Recurrent sepsis (especially if metastatic, eg, brain abscess or infective endocarditis, if unusually severe, resulting in multiorgan failure)</td>
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<td>Erratic fluid balance requiring hospitalization</td>
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<tr>
<td>Congenital intractable epithelial disorder, eg, microvillous inclusion disease and tufting enteropathy</td>
<td>Isolated bowel transplant or combined liver and bowel transplant depending on severity of hepatic complications of parenteral nutrition</td>
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<tr>
<td>Some cases of short-bowel syndrome</td>
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<tr>
<td>Irreversible liver disease: hyperbilirubinemia persisting beyond 3-4 mo of age and features of portal hypertension, splenomegaly, prominent superficial abdominal veins</td>
<td>Combined liver and intestinal transplant</td>
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</tbody>
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Adapted from Kaufman SS et al.\(^{13}\)
Bowel Transplant

FIGURE 40.2-2 Combined liver-bowel en bloc diagram. A = aorta; HA = hepatic artery; IVC = inferior vena cava; PV = portal vein; S = stoma.