

Short bowel syndrome

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Outline

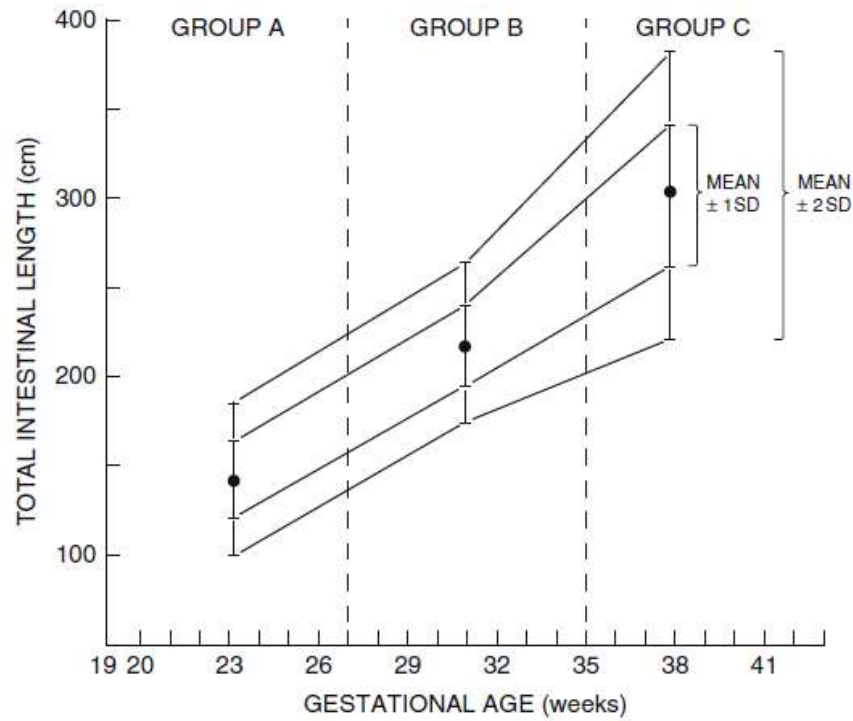
1. Definition
2. Epidemiology
3. Etiology
4. Pathology
5. Management
6. Outcome



1. Definition

- **Residual small bowel length is 25%-30% for given GA (~75 cm for term)**
 - Measurement should be with a thread laid along the antimesenteric but Intestinal loops shrink during manipulations making it difficult to measure real intestinal length.
 - Long term survival on enteral nutrition has been described with **11 cm (5% of bowel) with ileocecal valve** or with **25 cm(10% of total) without ICV.**
- **Requirement of TPN (> 6 wk)**
 - a functional description of intestinal failure (significant maldigestion and malabsorption)





- **Bowel length with gestational age. Reaches ~250 cm at term**



2. Epidemiology

- Most cases occur in **neonatal age group**
- prevalence increased due to the progress in **intensive care**
- 0.1% in all live births and 0.5% among ICU admissions



3. Etiology

- Prenatally acquired
 - **Intestinal atresia (21%)**
 - Gastroschisis (10%)
 - Hirschsprung disease (7%)
- Postnatal
 - **Necrotizing enterocolitis (36%)**
 - volvulus (19%)
- In older children
 - chron's disease
 - mesenteric ischemia



4. Pathophysiology: Bowel resection

- Resection of jejunum
 - **Transient reduction** in absorption b/c ileum has a much greater capacity for adaptation
- Resection of ileum
 - **vitamin B12** absorption
 - **bile acid** absorption - diarrhea, fat soluble vitamin malabsorption
 - fatty acid-Ca binding >> free oxalate (oxaluria) + inc enterohepatic circulation of bilirubin (sludge/stones)
 - **Water** secreted from UGI during hypertonic meal (Colon may not be able to compensate)



4. Pathophysiology: Adaptation

- Adaptation follows intensive intestinal resection
 - macroscopic - bowel dilatation, dysmotility, stasis and bacterial overgrowth
 - microscopic - enterocyte proliferation, muscle hypertrophy, inc villous height and crypt depth
 - Colon = inc Water absorption and bacteria ferment macronutrients into short chain fatty acids
- Stimulated by - ACE, gastrin, GH, IGF, enteroglucagon, glutamine, prostaglandin , vitamin D
- **90% of the patients can be weaned** after long term TPN.



5. Management



5. Management approach

- Acute phase = Duration depends on underlying disease
 - insufficient absorption, dysmotility, diarrhea, and gastric hypersecretion and hypergastrinemia.
 - **restoring and maintaining fluid**, electrolyte, acid-base equilibrium and minimizing nutrient loss.
- Adaptation = slower and often takes **more than a year** to reach its peak (unrelated to length of bowel)
 - balanced parenteral nutrition and **stepwise increasing enteral feeding**
- Maintenance phase. = If constant malabsorption rate of > 30%
 - **surplus of enteral calories** has to be consumed, **supplemented** by vitamins, elements, minerals



5.1. Nutrition

- **TPN is the primary therapy** utilized in the **immediate postoperative** period
- **enteral nutrition** should be initiated as soon as possible (**best stimulus for adaptation**)
- **Continuous infusion preferred** over bolus (Avoids gastric distension and constant load to microvilli)
- When condition improves, oral feeding of **small amounts of breast milk**
 - Human milk is preferred b/c contain protective factors and support intestinal adaptation.
- **Stool volume** increment by >50% is an indication to reduce amount/ concentration
- Other = high-soluble fiber (if colon intact), vitamin and mineral supplement



5.2. Supplemental therapy

- **hormones stimulate adaptation** (not yet proved sufficiently.)
 - Human growth hormone
 - Glucagon-like peptide 2 / teduglutide
 - Glutamine
- **Other medications**
 - PPIs – dec high output diarrhea
 - Loperamide – slow transit
 - Octreotide – inhibit secretions
 - Cholestyramine – prevent choleric diarrhea
 - Oral calcium – for oxaluria
 - Probiotics – dec bacteria translocation and stimulate bowel regrowth



5.3. Surgery: tactics in extensive resection

- Restoration of continuity (stoma closure) as soon as possible
- ? *Distal stoma reinfusion* ?
- intestinal atresia = dilated bowel should be preserved instead of resected in the usual way
- In volvulus = second-look procedures to decide which parts of the intestine are definitely lost.
- extensive NEC = questionable bowel should be decompressed by an enterostomy, not resected





Distal stoma refeeding in children with ileostomy: A method for facilitating restitution of intestinal transit.

María Zornoza-Moreno,¹ José Alejandro Ruiz-Montañez¹



Journal of Pediatric Surgery
Volume 50, Issue 5, May 2015, Pages 779-782

CAPS Paper

Mucous fistula refeeding in neonates with enterostomies ☆☆☆

Candace A. Haddock^a, Jennifer D. Stanger^a, Susan G. Albersheim^b, Linda M. Casey^c, Sonia A. Butten^{a, d, e}

Intestinal Failure

Safety of mucous fistula refeeding in neonates with functional short bowel syndrome: A retrospective review



Clinical Research Reports

Reinfusion of Succus Entericus Into the Mucous Fistula Decreases Dependence on Parenteral Nutrition in Neonates

Douglas Drenckpohl, MS, RD, CNSD, LDN, Ravindra Vegurta, MD, Lisa Knaub, RN, Mark Holterman, MD, PhD, Huaping Wang, PhD, Kamlesh Macwan, MD, and Richard Pearl, MD

Original Article

Mucous fistula refeeding decreases parenteral nutrition exposure in postsurgical premature neonates ☆☆☆

Colin D. Gause^a, Madoka Hayashi^b, Courtney Haney^c, Daniel Rhee^a, Omar Karim^a, Brian W. Weir^{d, e}, Dylan Stewart^a, Jeffrey Lukish^a, Henry Lau^a, Fizan Abdullah^a, Estelle Gauda^b, Howard I. Pryor II^{a, f, g}



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The feasibility of routine use of distal stoma refeeding method in newborns with enterostomy

Serpil Sancar, Murat Sanal, Oliver Renz & Paul Hechenleitner

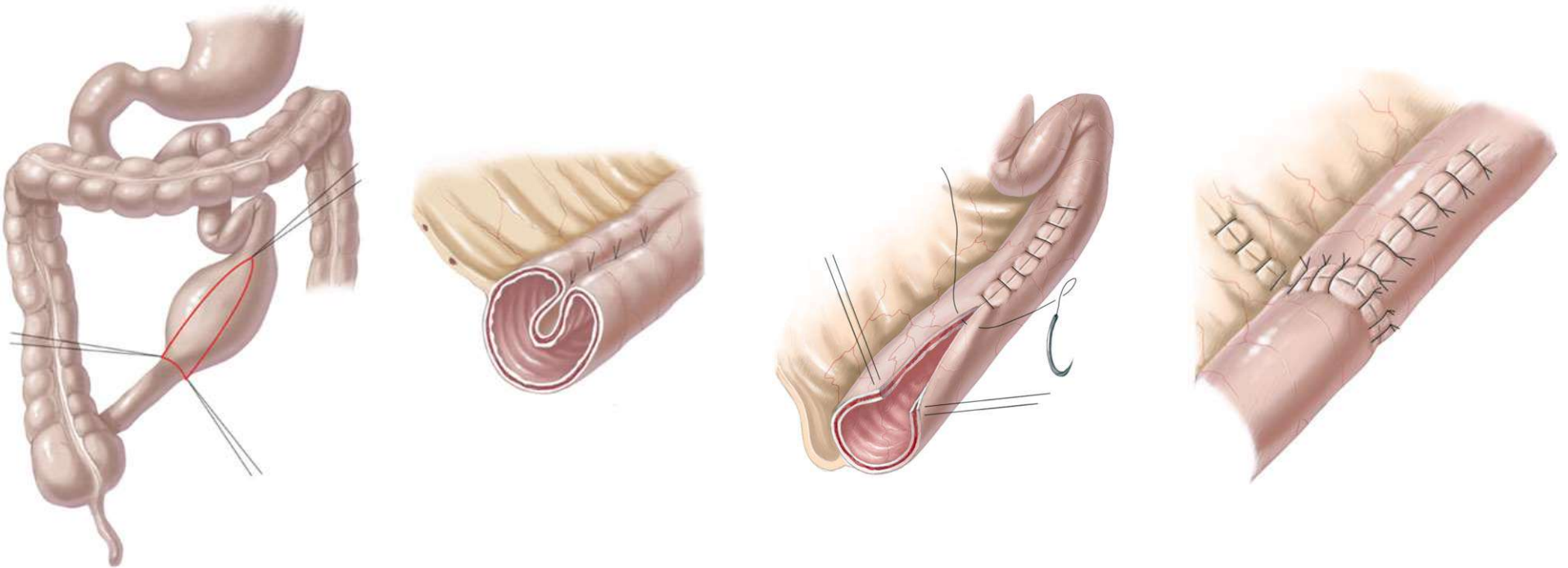
- **Mucus fistula Re-feeding of proximal stoma effluent:** a safe, easy and inexpensive technique to facilitate restitution of intestinal transit, promote absorption, prevent the atrophy of the distal segment and prepare bowel for anastomosis. Potential to contribute to weight gain and decrease PN dependence. Reported complications include mucus fistula perforation and bleeding.



5.3. Surgery: for fast transit and dysmotility

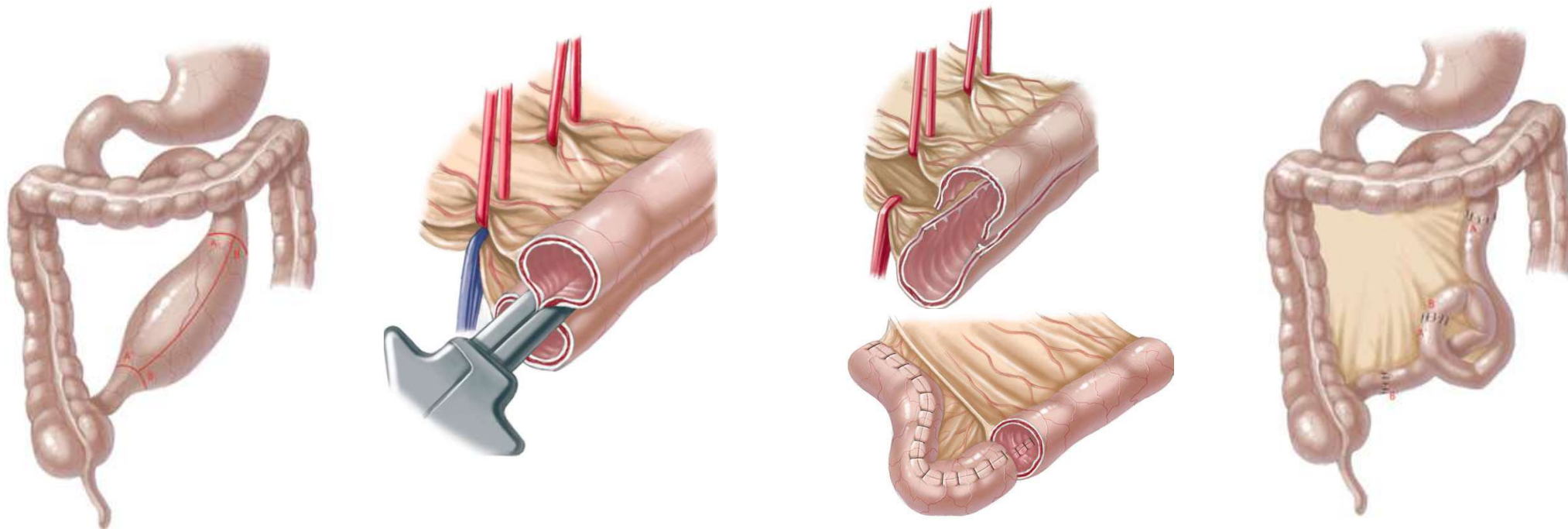
- **10% will benefit from surgery** (never recover bowel function in order to wean off PN)
- **Indication** = unable to obtain 10–50% of caloric intake by 6 months on PN
- To improve peristalsis
 - Tapering
 - Tapering Lengthening
- To increase transit
 - Antiperistaltic (reversed) small bowel segment
 - Colonic interposition
 - Intestinal valves and pouches





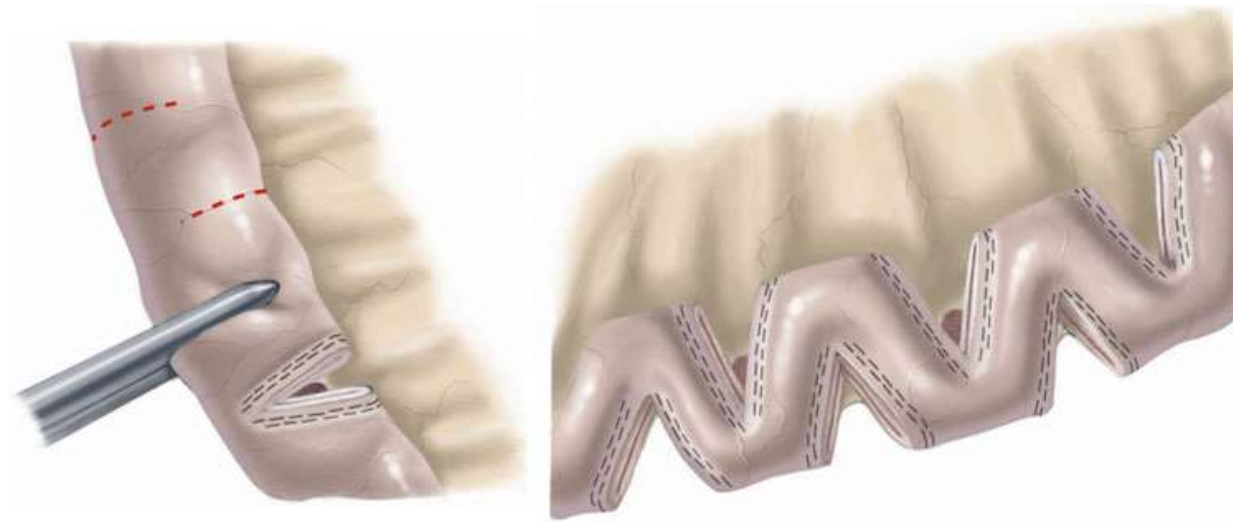
- **Intestinal refashioning (tapering):** can be achieved surgically by tailoring the antimesenteric side of a dilated loop, either by resection of triangular antimesenteric segment (if enough absorptive area remains available and stasis is the only problem) or by infolding the excessive part of the intestinal circumference in a longitudinal way (without losing absorptive surface area).





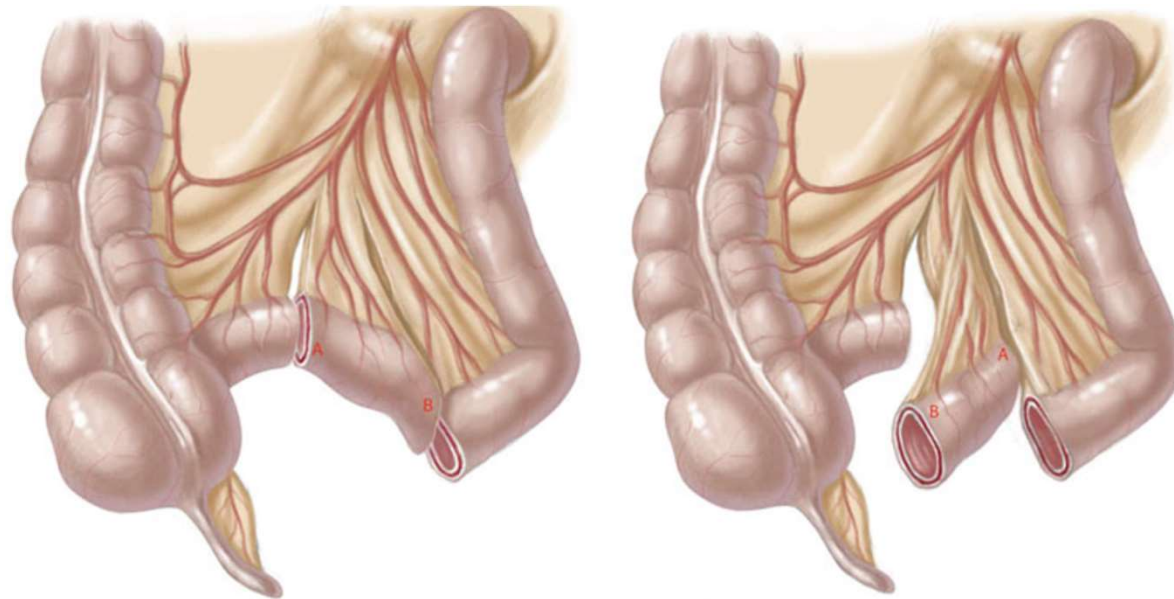
- **Longitudinal Intestinal Lengthening and Tailoring (LILT)/ Bianchi's** : longitudinal division of the dilated part into two separate segments. Both segments remain viable because the mesenteric vessels divide extramurally into branches supplying either side of the bowel separately. A Penrose drain facilitates the division of the segments. The two halves are then refashioned to tubes of normal intestinal diameter which are lined up in the isoperistaltic direction and anastomosed. The helixlike arrangement of the two separated parts allows the anastomoses to be performed with minimal traction on the vessels





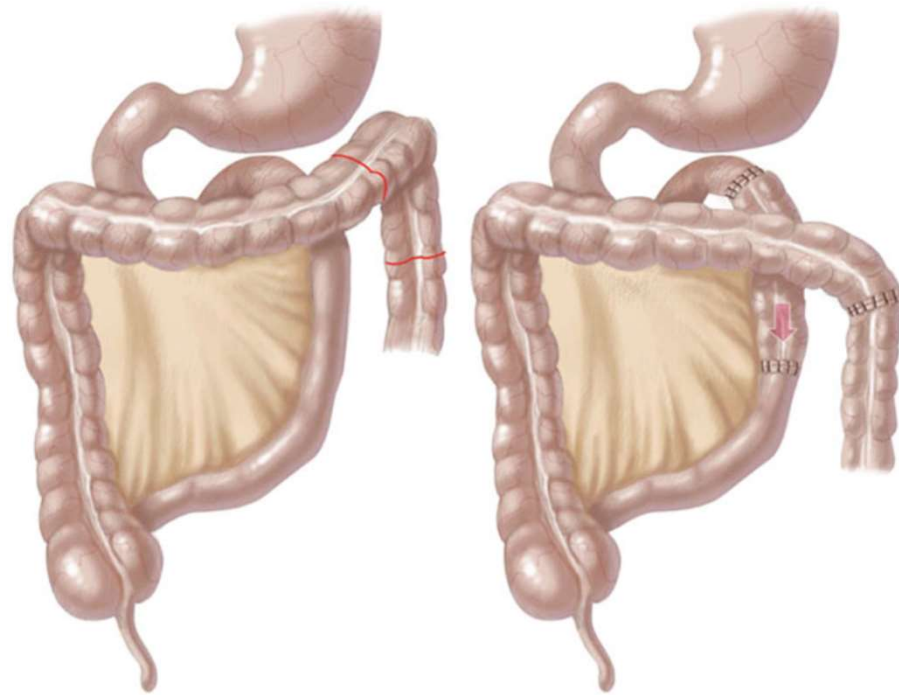
- **Serial transverse enteroplasty (STEP):** semicircular alternating incisions with the GIA stapler resulting in a zigzag-like elongation of the small bowel. Technically much easier when compared with the Bianchi method. It's a new procedure that gained significant attention worldwide





- **Segmental reversal of small bowel (SRSP):** The antiperistaltic intestinal segment should be interposed close to the ileocecal valve or at the end of the small bowel. The optimal length in newborns is around 3.0 cm.





- **Colon interposition:** can be used isoperistaltic to increase absorption or antiperistaltic to decrease transit. The isoperistaltic interposition should be proximal while the reversed interposition should be used distally. The length of the isoperistaltic is within 10–20 cm.





- **Nipple-like ileocecal valve.** The optimal length in newborns is not defined but will be around 1–2 cm. In a 15-year-old boy, a 4-cm-long valve worked well. The intestine is transected at an appropriate level and the last 2–4 cm of the end of the proximal bowel are everted and firmly fixed by 4/0 or 5/0 seromuscular interrupted sutures onto the underlying seromuscular bowel wall. The distal intestinal segment is then pulled over the everted bowel and finally anastomosed to the everted segment and to the proximal intestine by seromuscular interrupted stitches.



5.3. Surgery: Intestinal transplantation

- most effective method to increase intestinal absorptive area immediately
- Alone or together with liver transplant
- **Indications**
 - little or no small bowel remaining (last resort for refractory disease)
 - irreversible liver failure due to TPN
- High rejection rate in the past but current progress with new immunosuppressive agents
- Benefit some patients although 5-year survival rate does not lie much above 50%.



6. Outcome: Complications

- Malabsorption
 - Diarrhea, dehydration , electrolyte issues
 - Vitamin deficiencies, growth problems
 - Gall stones, kidney stones
- TPN
 - Central line – occlusion, infection
 - Liver failure
 - Bacterial translocation
- Bowel lengthening procedures
 - re-dilatation
 - bowel obstruction



6. Outcome: survival

- **Long-term survival between 72% and 90%**
- Depends on
 - length of the remaining bowel
 - quality of the motility of the remaining bowel
 - incidence of complications (IFALD, recurrent septicemia, central line infection)





That's all Folks!