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# Management of Short Bowel Syndrome in Children

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SURGERY GRAND ROUNDS 3/21/2013

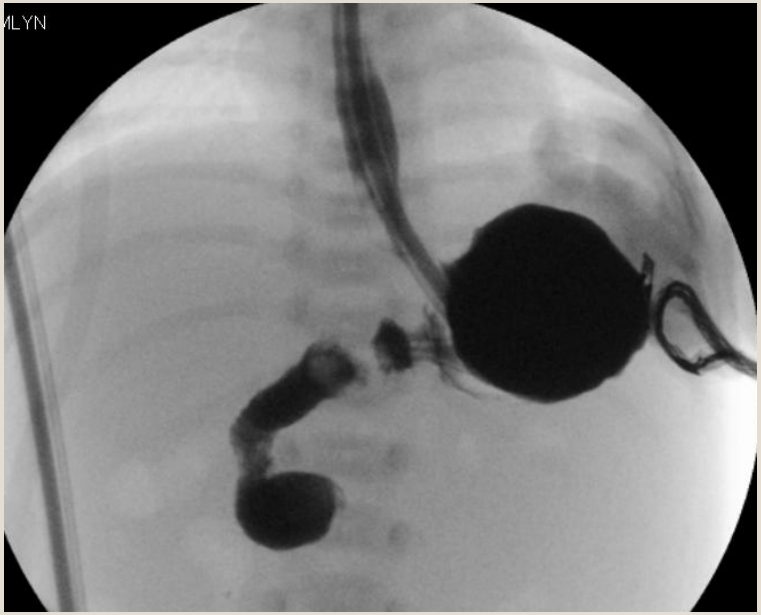
# Case Report

- Female newborn, born at 35+5 weeks gestation via c-section, birth weight 2780g, APGAR 9/9
- DOL#0-1:
  - admitted to NICU for respiratory distress requiring CPAP and r/o sepsis
- DOL#2:
  - no meconium, abdomen soft,
  - AXR: scattered gas filled loops of bowel and dilated transverse colon
  - drop in hct requiring transfusion of PRBC
- DOL#3:
  - abdominal distention and bilious drainage from OGT
  - AXR: remained unchanged
  - pediatric  surgery consulted

# Case Report

UGI:

- Obstruction at the second/ third portion of the duodenum




BE:

- Patent colon with cecum in the right upper quadrant adjacent to the obstructed duodenal loop

# Case Report

- DOL#3:
  - UGI/BE concerning for malrotation with volvulus
  - Urgent surgical exploration: malrotation with volvulus and necrosis of the small bowel from third portion of duodenum to terminal ileum
  - Detorsion of small bowel, closure of abdomen
  - Return to NICU for resuscitation
  - Planned second look in 24h
- DOL#4:
  - Second look: unchanged necrosis of small bowel
- DOL#5-11 (POD#1/2-6/7)
  - Awake, alert, on abx, TPN, NCPAP

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- DOL#12 (POD#8/9):
  - Exploratory laparotomy, resection of necrotic small bowel with anastomosis of mid duodenum to terminal ileum
  - Gastrostomy
  - Broviac placement

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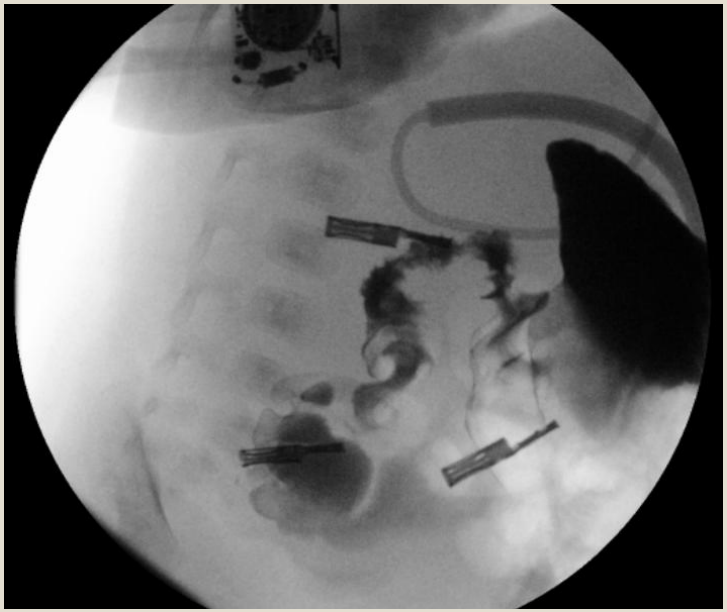
- DOL#21 (POD#9/17/18)
  - Off abx, on RA, TPN, starting feeds via gastrostomy
- DOL#32 (POD#20/28/29)
  - Tolerating feeds at 3cc/h via gastrostomy
  - TPN
  - UGI: esophageal reflux, delayed passage of contrast into ileum
- DOL#72
  - Tolerating feeds at 12 cc/h via gastrostomy
  - Continuous TPN
  - Weight: 3905g



# Intestinal Failure

- Inability of the GI tract to provide sufficient absorption capacities to cover nutritional requirements for maintenance in adults and growth in children
- Functional and structural adaption for 1-2 years after resection
- Permanent intestinal failure is determined after 2 years
- Parenteral nutrition is first line treatment, allowing satisfactory growth and acceptable quality of life
- Intestinal transplant is indicated for failure of PN
- 5 year survival rate on PN is 75%

# Causes of Intestinal Failure in Children

- Short bowel syndrome 68%
  - Volvulus 15%
  - Gastroschisis 24%
  - Necrotizing enterocolitis 16%
  - Atresia 9%
  - Other 4%
- Motility Disorders 14%
  - Long segment Hirschsprung disease, chronic intestinal pseudoobstruction
- Mucosal Defects 10%
  - microvillus inclusion disease, tufting enteropathy
- Retransplantation 5%
- Others 3%
  - tumors

# SBS: Pathophysiologic changes

- Loss of intestinal absorptive surface
- More rapid intestinal transit
- Gastric hypersecretion
- Inactivation of pancreatic enzymes
- Loss of bile salts


- Malnutrition
- Weight loss
- Diarrhea
- Steatorrhea
- Vitamin deficiency
- Electrolyte imbalance
- Cholelithiasis
- Nephrolithiasis
- Liver Disease

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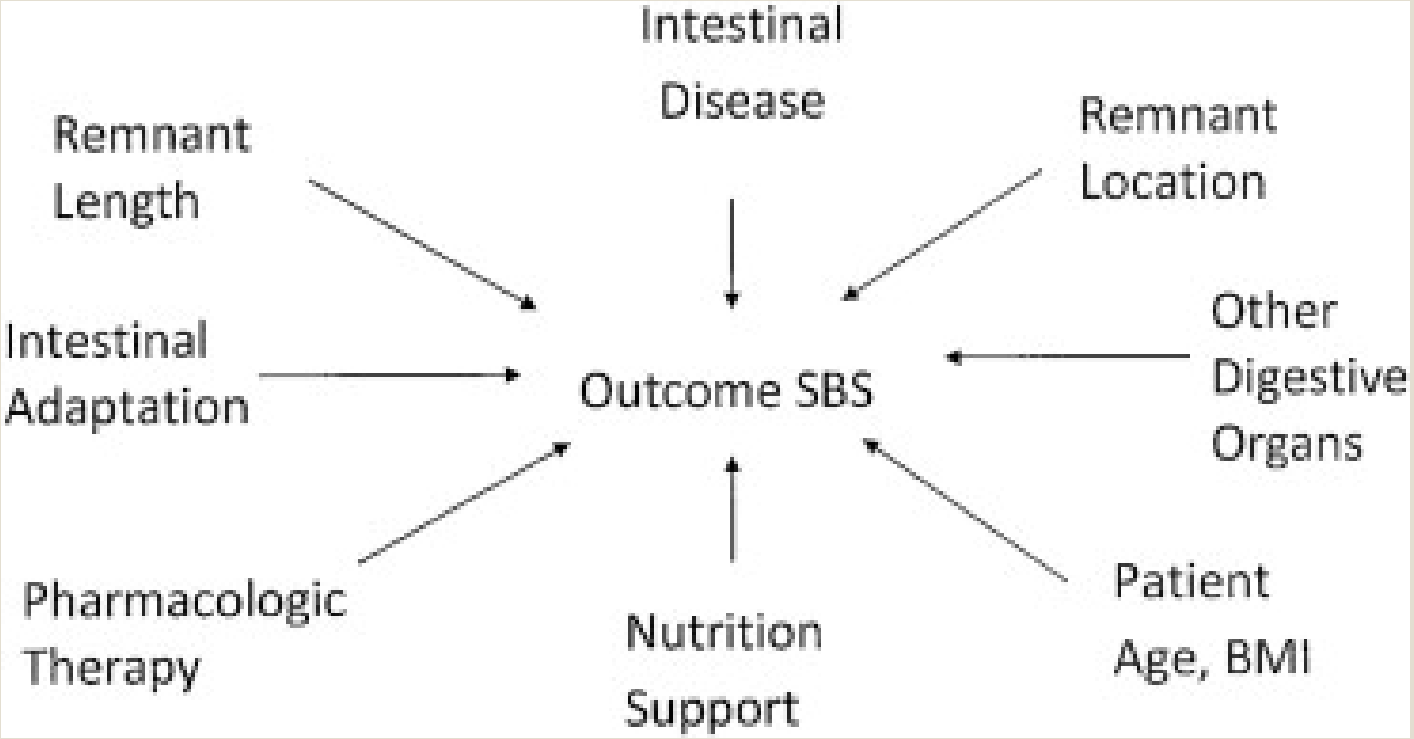
# Factors influencing Outcome



# Prognosis

- 80% of children with neonatal onset SBS wean from PN between age 1-2
- 10% die from complications
- 10% remain on permanent PN
  - Remnant length <30-40 cm
  - Absent ileocecal valve
  - Colon resection
  - Tolerance of <25% of enteral calories at 3 months of age
- Multidisciplinary approach at specialized center improves outcome

Kaufman et al: Indications for pediatric intestinal transplantation: A position paper of the American Society of transplantation. Pediatr Transplantation. 2001;5:80-87

# Intestinal Rehabilitation

enhance intestinal absorption and function

eliminate long-term dependency on PN

Medical Rehabilitation

- Maintain nutritional status
- Maximize enteral nutrient absorption
- Prevent complications

Surgical Rehabilitation

- Preserve intestinal remnant length
- Improving intestinal motility
- Increasing absorptive area

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# Maintain nutritional status

- Replacement of fluid and electrolyte losses
- Parenteral Nutrition
  - Continuous then cyclic PN
  - Lipid minimization and alternate lipid preparations to reduce IFALD
- Enteral Nutrition
  - Start as early as possible postop with hypoosmolar feeds
  - Gradually increase as enteral adaption occurs
  - Enteral independence is goal

# Dietary recommendations for SBS

**Table 2**
**Dietary recommendations for short bowel syndrome**

| | Colon in Continuity | No Colon in Continuity |
|---|---|---|
| Fluids | Hypotonic and/or ORS | ORS |
| Carbohydrate | 50%–60% of caloric intake<br>Complex carbohydrates<br>Limit simple sugars<br>Low lactose | 40%–50% of caloric intake<br>Complex carbohydrates<br>Limit simple sugars |
| Fat | 20%–30% of caloric intake<br>Adequate essential fats<br>MCT/LCT | 30%–40% of caloric intake<br>Adequate essential fats<br>LCT |
| Protein | 20%–30% of caloric intake | 20%–30% of caloric intake |
| Fiber | 5–10 g/d soluble fiber for net secretors | 5–10 g/d soluble fiber for net secretors |
| Oxalate | Restriction | No restriction |

*Abbreviations:* LCT, long-chain triglycerides; MCT, medium-chain triglycerides; ORS, oral rehydration solution.

Thompson et al. Current Management of the Short Bowel Syndrome. Surg Clin N Am. 2011;91:493-510

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# Pharmacologic treatment

- Slow transit / diarrhea
  - Loperamide
  - Diphenoxylate
  - Narcotics
  - Cholestyramine
  - Pancreatic enzymes
- Reduce GI secretion
  - $H_2$ receptor antagonists
  - Proton pump inhibitors
  - Octreotide
  - Clonidine
- Treat bacterial overgrowth
  - Antibiotics
  - Probiotics
  - Prokinetics
- Glutamine, hormonal treatment

# Hormonal Treatments

- Growth hormone combined with glutamine
  - Effects short lived, results inconclusive in metaanalysis
- GLP-2 and its analog teduglutide
  - Hormone strongly associated with intestinal growth and post resection intestinal adaptation
  - Reduction in PN compared to placebo in RCT over 52 weeks
- EGF
  - transiently increased carbohydrate absorption and improved tolerance to enteral feeding in pediatric patients
  - Under investigation
- GLP-1 agonist exenatide
  - GLP-1 receptor agonist approved for type 2 DM
  - Improvement in intestinal transit in case series

Thompson et al.Current Management of Short Bowel Syndrome. Curr Prob Surg.2012:49(2);52-119
O'keefe et al. Safety and Efficacy of Teduglutide after 52 weeks of treatment in patients with shirtnBowel syndrome intestinal failure. Clin GastroenetrolHepatol.2013:13:87-6

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# Surgical Rehabilitation

- Preserve intestinal remnant length
  - Minimize resection (tapering, stricturoplasty)
  - Restore continuity
  - Recruit additional intestine
- Improve motility
  - Relieve obstruction
  - Taper dilated bowel
  - Prolong intestinal transit (reversed segments, artificial valve, colon interposition)
- Increase absorptive area
  - STEP
  - Bianchi

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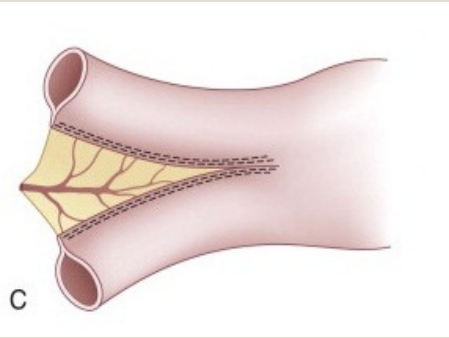
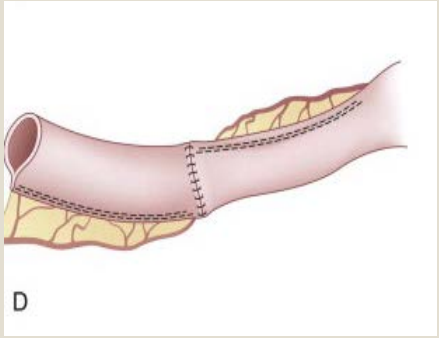
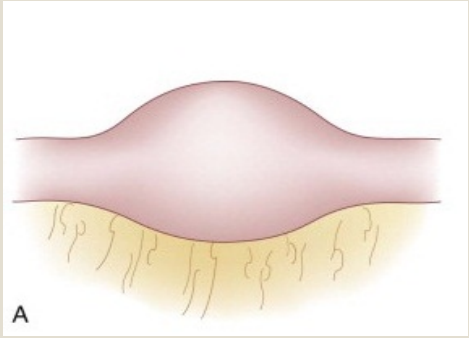
# Surgical Rehabilitation

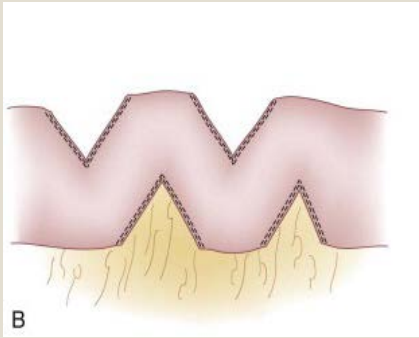
- Bianchi enteroplasty (1980)
  - Longitudinal division of loop of distended bowel
  - Each mesenteric leaf supplies one of the divided segments
  - Divided segments are anastomosed
  - Early complication rate 20%
  - 50-80% sustained clinical improvement




- STEP: Stepwise transverse enteroplasty (2003)
  - Alternating staple lines
  - Reducing diameter while increasing length
  - Mortality 8%
  - 80% clinical improvement
  - Can be done more than once on same segment



# Bianchi vs STEP

- Single Center review of lengthening procedures over 24 years
- Comparing survival, TPN weaning, complications
- 64 patients (14 adults)
- Complications 10%

| | Bianchi | STEP |
|---|---|---|
| Procedures (#) | 43 | 34 |
| Remnant length before lengthening | 44 cm | 45 cm |
| Remnant length after lengthening | 68 cm | 65 cm |
| Overall survival (3.8y) | 88% | 95% |
| Time to discontinuation of TPN (months) | 8.4 | 4.8 |
| TPN weaning | 55% | 60% |

Sudan et al. Comparison of intestinal lengthening procedures for patients with short bowel syndrome. Ann Surg.2007;246(6):593-604

# Complications of Short Bowel Syndrome

| Catheter related | Hepatobiliary | Metabolic | Renal | Gastrointestinal |
|---|---|---|---|---|
| **Infection** | **Intestinal failure associated liver disease** | Fluid and electrolyte abnormalities | Chronic renal failure | Gastric hypersecretion |
| **Loss of vascular access** | Cholelithiasis | D-Lactic acidosis | Nephrolithiasis | Small bowel bacterial overgrowth |
| | | Micronutrient deficiency | | Changes in colonic flora |
| | | Metabolic bone disease | | |
| | | Osteoporosis and osteomalacea | | |

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# Intestinal Transplantation

- 1989:  first successful intestinal transplant
- 2011: 2067 intestinal transplants and 629 liver-pancreas-intestine transplants performed in the USA (75% in children)
- Cost-effective compared to PN at 2 years
- Early referral and listing are important to ensure greatest opportunity to obtain a transplant

# Indications for Intestinal Transplantation in children

- Failure of TPN (impending life-threatening complications)
  - Intestinal failure-associated liver failure
  - Thrombosis of ≥2 central veins
  - ≥2 episodes/yr severe sepsis especially fungemia
- Intestinal failure that virtually always results in early death despite optimal parenteral nutrition
  - Extreme short bowel syndrome
  - Congenital intractable epithelial disorders
- Controversial indications
  - Intestinal failure with frequent hospitalizations, high enteric fluid losses or pseudoobstruction
  - Patient unwillingness to accept long-term parenteral nutrition (PN)

Kaufman et al: Indications for pediatric intestinal transplantation: A position paper of the American Society of transplantation. Pediatr Transplantation. 2001;5:80-87

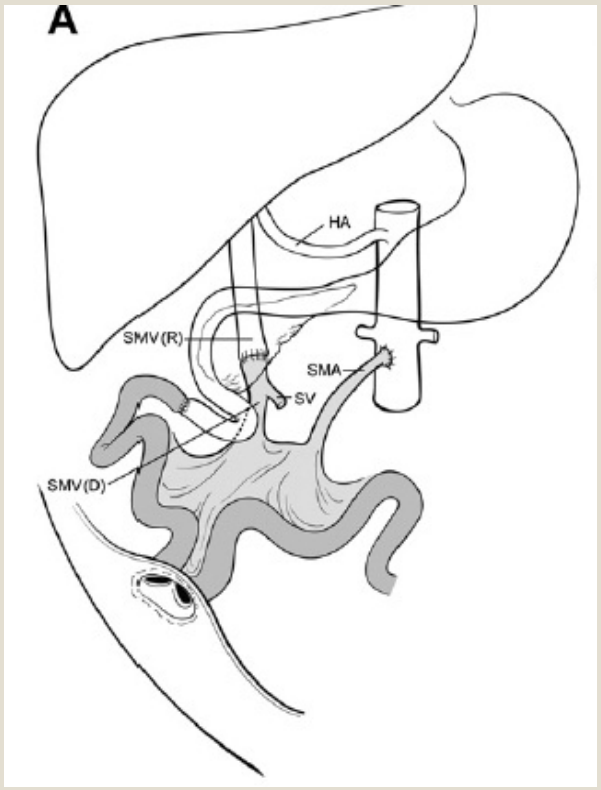
# Referral and Listing Criteria for IT

**Table 1**
**Recommended criteria for consultation or referral for IT assessment and for listing for IT**

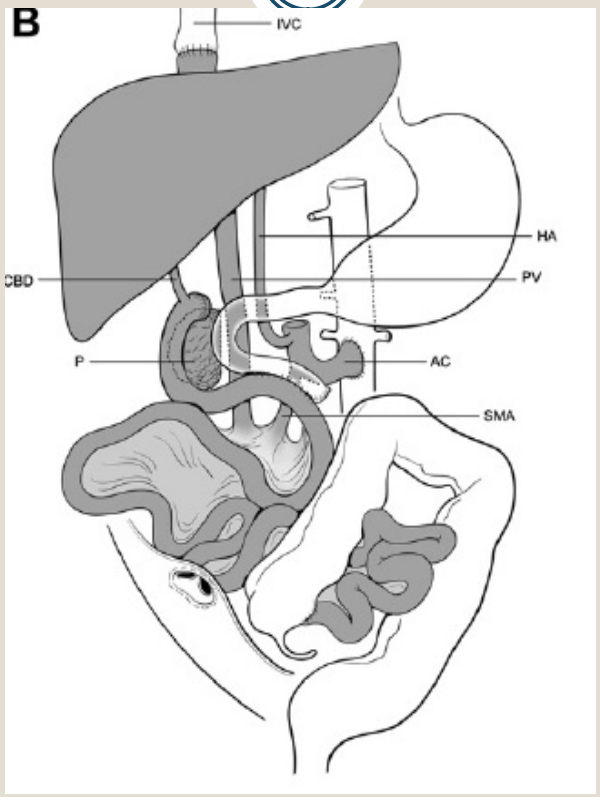
| Referral Criteria | Listing Criteria |
|---|---|
| Children with massive intestine resection | Small bowel length of <25 cm without an ileocecal valve |
| Children with severely diseased bowel and unacceptable morbidity | Intestinal failure with high morbidity and poor QOL |
| Microvillous inclusion disease or intestinal epithelial dysplasia | Congenital intractable mucosal disorder such as microvillous inclusion disease or tufting enteropathy |
| Persistent hyperbilirubinemia (>6 mg/dL) | Persistent hyperbilirubinemia (>3–6 mg/dL) and signs of portal hypertension, or synthetic liver dysfunction with coagulopathy |
| Thrombosis of 2 of 4 upper body central veins | Loss of more than 50% of the standard central venous access sites |
| Continuing prognostic or diagnostic uncertainty | Recurrent life-threatening episodes of sepsis resulting in multiorgan failure, metastatic infectious foci, or acquisition of flora with limited antibiotic sensitivities |
| Request of the patient or family | |

Avitzur et al. Intestine Transplantation in Children: Update 2010. Pediatr Clin N Am.2010;57:415-431

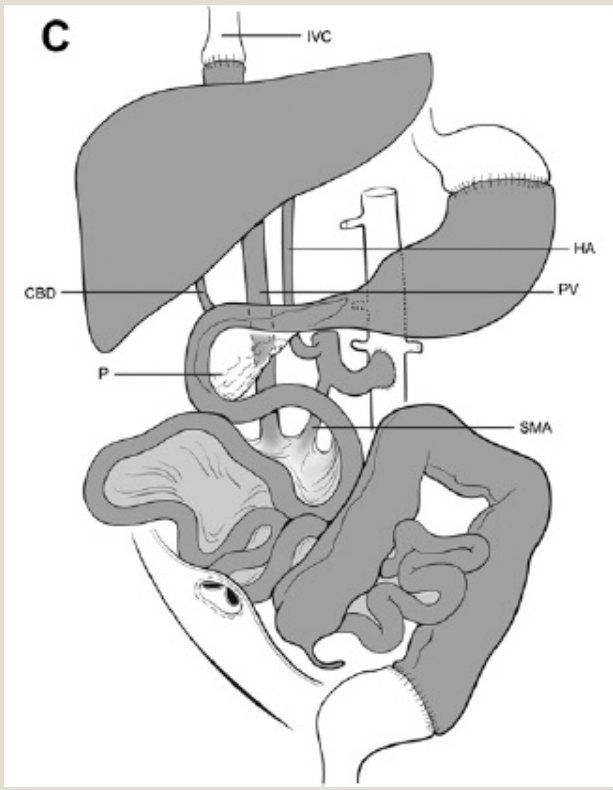
# Types of Intestinal Transplant




Isolated intestinal transplant




Intestine plus liver transplant




Multivisceral transplant

# Complications after Transplant

- Acute rejection
  - Breakdown of gut barrier function leading to bacterial translocation and infection
  - Main cause of graft loss and death
- Infections
  - Sepsis one of the main cause of death post-transplant
  - Immune suppression, breakdown of gut barrier and ischemia reperfusion injury contribute to high indidence
  - Viral infections can lead to severe and prolonged diarrhea
- Chronic renal failure
- PTLD
  - More common in ITx than in other solid organ transplant
- GVHD 5%

# Outcomes

- Patient survival:
  - 1y: 80-94% (worse if <1y of age)
  - 3y: 65-84%
  - 5y: 50-70%
- Graft survival:
  - 1y: 88%
  - 3y: 74%
  - 5y: 58%
- 80-100% of pediatric recipients develop enteral autonomy within 1-3 months

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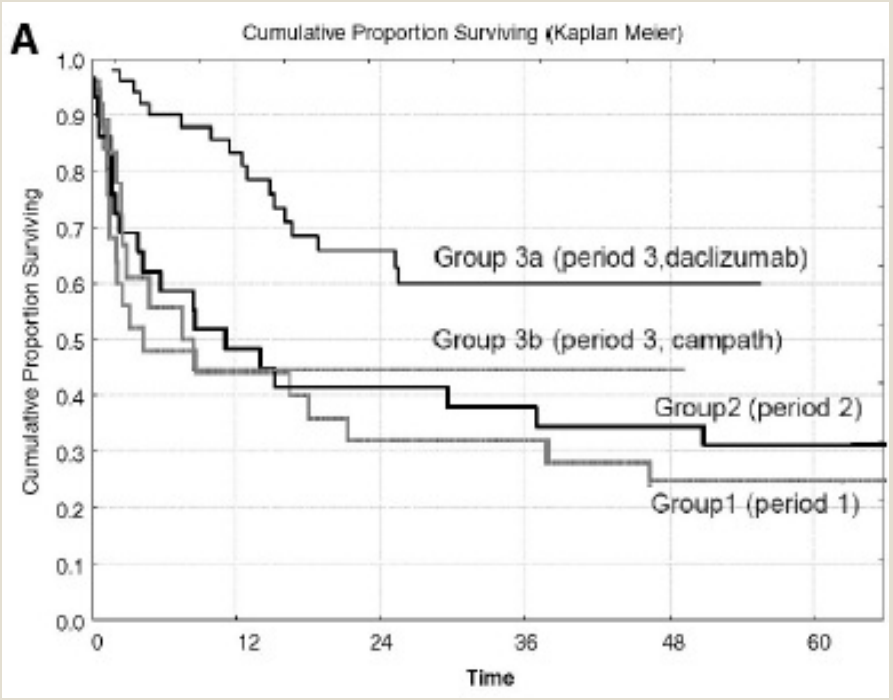
# Outcomes

- Single center retrospective review of 123 patients from 1994-2006
- Median age 16m (4m-17y)
- Median weight 9.5kg (4.4-67kg)
- Irreversible intestinal failure and complications of TPN
- Multivisceral or isolated intestine




Kato et al. Intestinal and Multivisceral Transplantation in Children. Ann Surg: 2006;243(6):756-766

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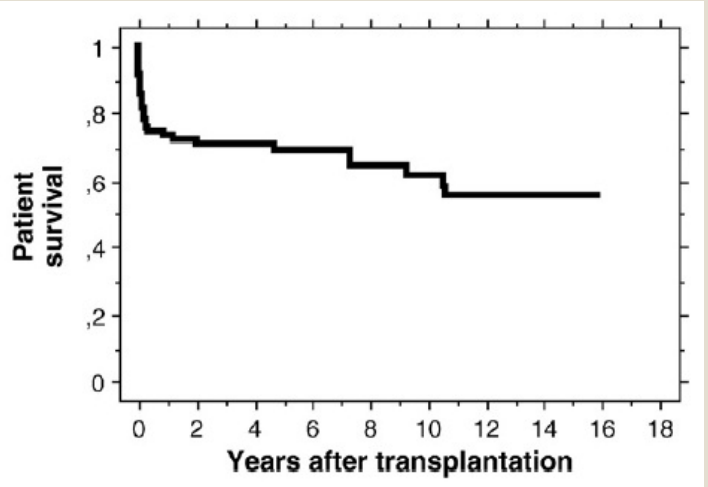
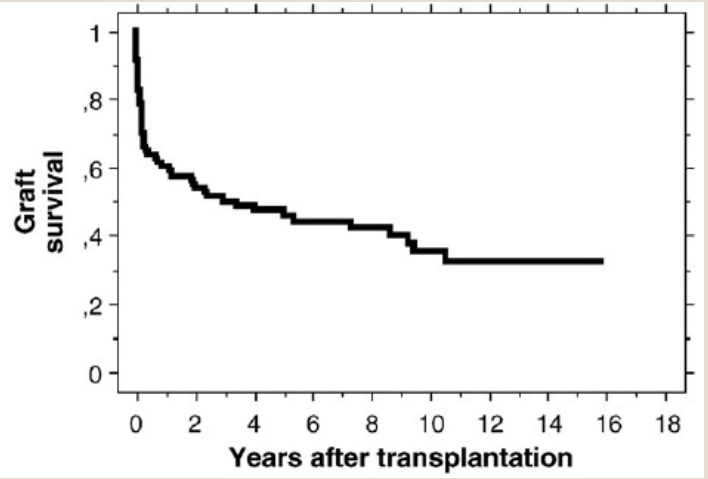
# Outcomes

- Single Center retrospective review of 87 children (93 transplants) from 1994-2010
- Patient survival (1/5/10y) 73%/69%/61%
- Graft survival (1/5/10y) 59%/46%/34%




Revillon et al. Indications and strategies for intestinal transplantation. J Ped Surg. 2011;46:280-283

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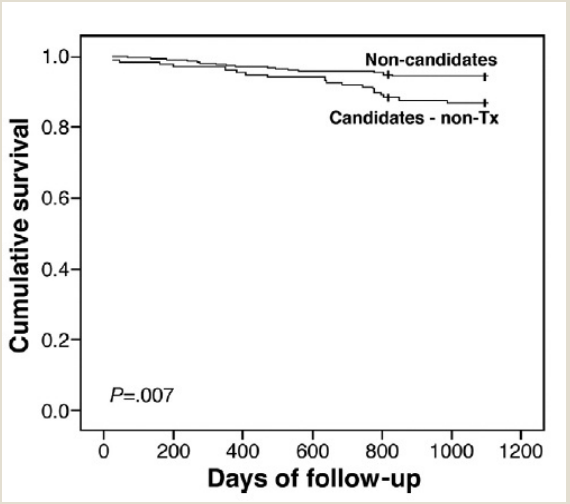
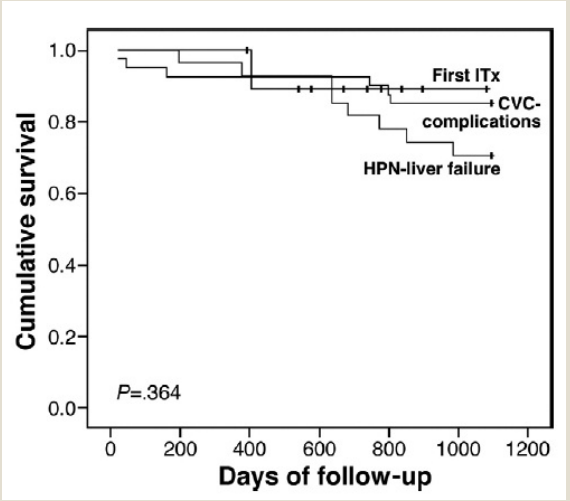
# Outcomes

- Multicenter cross-sectional survey of candidates and non-candidates for ITx (adult and pediatric)
- Evaluation of validity of AST listing criteria (failure of home PN/ high risk of death/ high morbidity)




Pironi et al. Survival of patients identified as candidates for intestinal transplantation: a 3 year prospective follow-up. Gatroenterol.2008;135:61-71

# Summary

- Multidisciplinary approach to SBS and progress in all aspects of management have greatly improved outcomes over the past 20 years
- Home PN is first line treatment
- Medical intestinal rehabilitation is initial treatment focus
- Surgical intestinal rehabilitation (lengthening procedures) may further improve function
- Short- and long-term survival for intestinal transplantation are improving
- Intestinal transplant is accepted treatment for patients with failure of PN

# References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