

GUT SHORTY

or medical management of intestinal failure in children



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Objectives

- Definition
- Epidemiology
- Etiology
- Symptomatology
- Medical management
- Surgical management



Definition

- Short gut <25% SB for age
- Intestinal failure - reduction of **functioning** intestinal mass below the amount necessary for adequate absorption of nutrients, micronutrients, fluid and electrolytes to allow **growth**
- It's not just how much bowel you have, but whether what you have works.....



Epidemiology

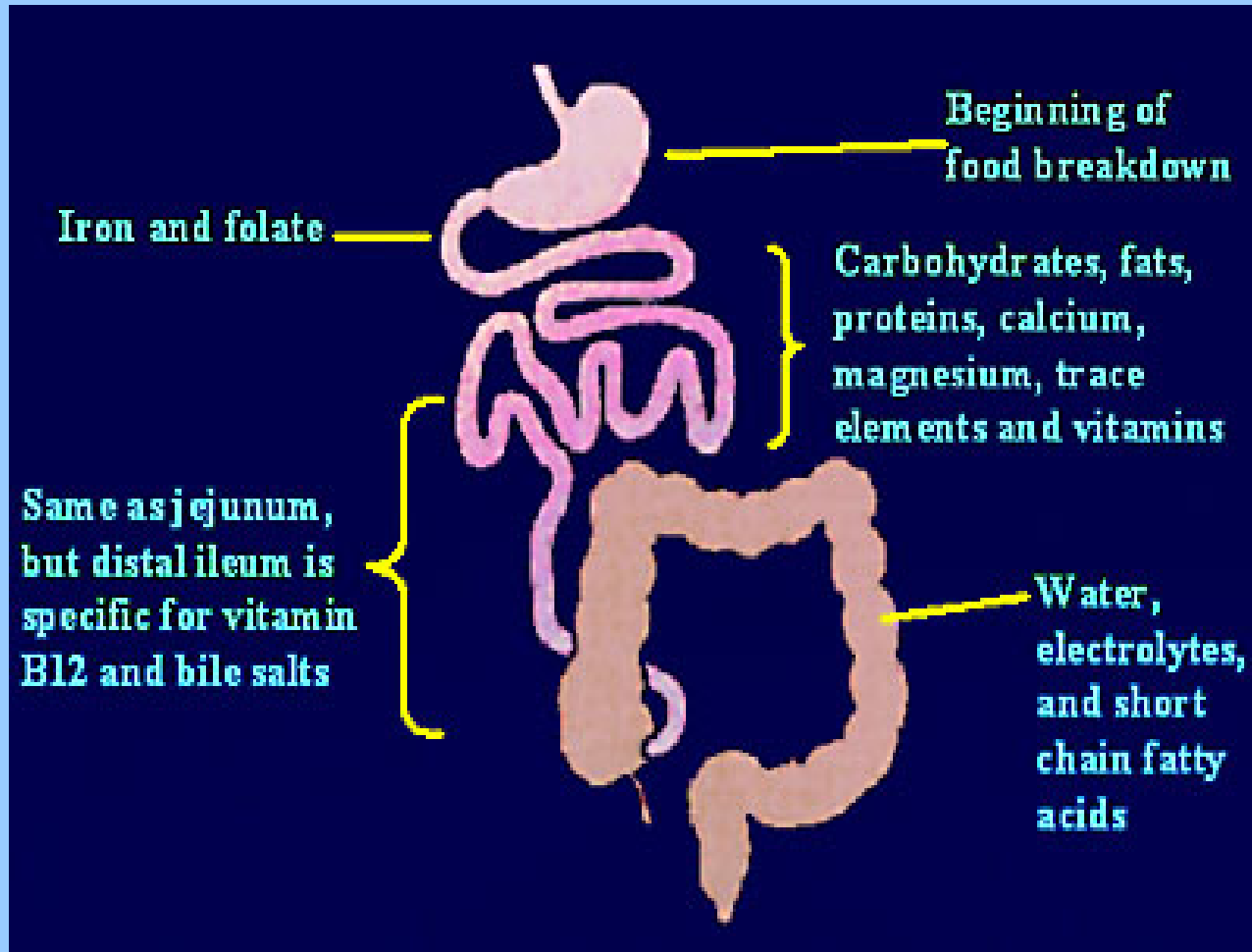
- Estimated incidence; 2-5 per 100,000
- Majority of children with IF are weaned off TPN within 12 weeks, only small proportion require long term TPN
- Increased prevalence due to advances in neonatal intensive care , pediatric surgical techniques, improved PN



Causes of IF in children

- NEC
- Intestinal atresia
- Malrotation with midgut volvulus
- Gastroschisis
- Total intestinal aganglionosis
- Complicated intussusception
- IBD
- Radiation enteritis
- Vascular events-arterial/venous
- Intestinal obstruction-adhesions
- Trauma
- Extensive angioma, other tumors

Sites of nutrient absorption





Symptoms

- Diarrhea, (fat), dehydration and electrolyte abnormalities
- Malnutrition and poor growth
- Gastric acid hypersecretion and peptic disease
- Bacterial overgrowth and **infection**
- **Liver disease** (TPN cholestasis)
- Bone disease
- Biliary and renal stones
- Vascular complications



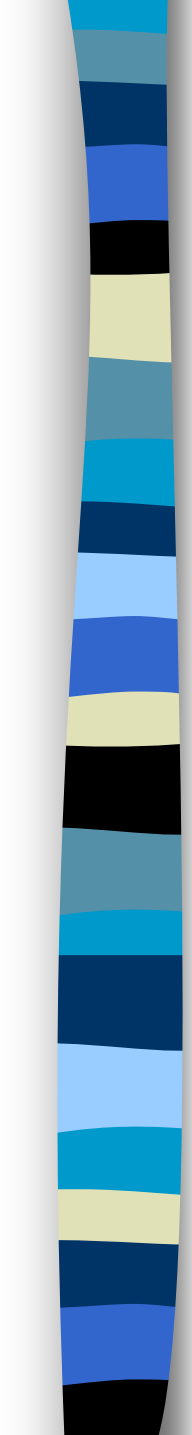
Medical Management

- Enteral feed
 - Oral
 - Tube
- Parenteral feed (TPN)
- Medications



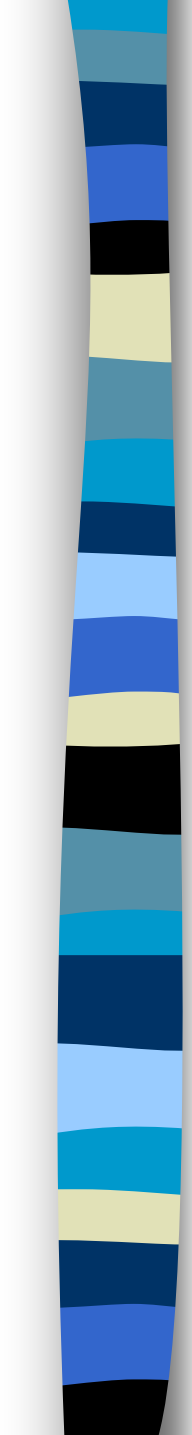
Pediatric IF

- Initial Assessment
 - Define anatomy & length of residual bowel
 - Define functionality
 - Nutritional status and growth
- Establish vascular access/ Initiate TPN
- Establish enteral access
 - Gastrostomy
 - Oral stimulation



Feeding-How?

- Enteral feeding promotes adaptation
- Optimal nutrition -oral feeds: small frequent
- Continuous enteral feeding via NG or gastrostomy
 - slow, "trophic feeds"
 - overnight CEF
 - daytime bolus



Feeding-What?

- Maternal milk (immune and growth factors)
- Elemental formula (reduces antigenic load)
- Semi-elemental and complex protein
- Minimize concentrated simple sugars, maximize complex carbohydrate.
- High fat diet –MCT vs. LCT
- $\frac{1}{2}$ - $\frac{2}{3}$ of consumed calories are absorbed
- Start dilute concentration and increased gradually
- Individualized formula



Type of Diet

- Soluble fiber (pectin)- fermented by bacteria to short chain fatty acids in pt. with intact colon.
- Initially (72h) worsens diarrhea
- Higher carbohydrates if intact colon
- Separate fluids from solids
- Cooked vegetables and peeled fruit
- Limit hypotonic fluids, ORS are optimal



ORS

1 Liter water,
8 tsp table sugar
1 tsp salt



Stoma and tube site care

- Skin care
- Stoma color
- Stoma production
- Stoma anchorage
- Leakage
- Prevent accidental dislodgment



TPN

- TPN as bridging to intestinal adaptation
- Compensating for intestinal fluid and nutrient loss via stoma or diarrhea to maintain positive energy balance
- “Custom made” TPN
- Some patients may need TPN during growth spurts



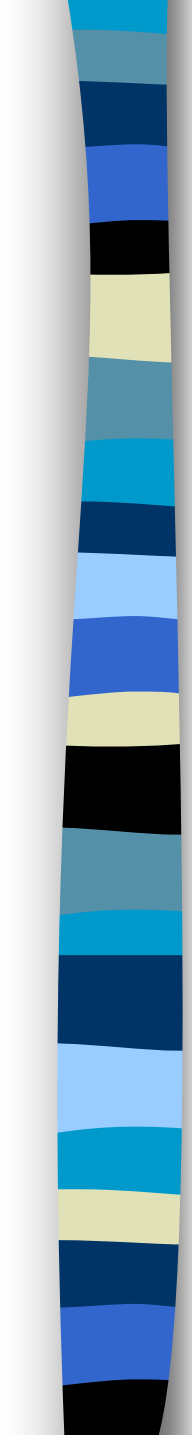
Line care

- PICC or tunneled (broviac, port)
 - Infection
 - Migration
 - Occlusion/thrombosis
- Lock therapy
 - Ethanol
 - Antibiotic
 - Antimicrobial-coated catheters



Prevention of liver disease related IF

- Increase oral intake
- Decrease dextrose infusion
- Decrease lipid infusion, supply carnitine, omega- 3 FA
- Decrease aluminium, manganese and copper
- Cycling of TPN infusion
- Early discharge and home TPN to decrease line infections
- CCK injection
- Abx to decrease BO
- Ursodeoxycholic acid
- Taurine, Choline (investigational)
- Anti-oxidants



Omegaven- Fish oil IL

- TPN cholestasis is thought to be a consequence of exposure to soy fat
- First monotherapy use in 2002, in a child with soy allergy (Gura et al.)
- Uncontrolled studies: reduction of TPN cholestasis 1-18 patients at 1-1.5 mg/Kg/d (Diamond et al.)
- Anti-inflammatory and anti-oxidant
- Used under compassionate protocols



Jury is still out....

- Similar improvements by withholding or reducing standard lipid emulsions $<0.5\text{gr/Kg/d}$
- Concerns about inadequate supply of n-6 fatty acids.
- ClinOleic- 80: 20 mix of olive and soybean oils
- SMOFLipid 30: 30: 25: 15 mix of MCTs, soybean oil, olive oil and fish oil
- Efficacy and safety of Omegaven vs. Intralipid in prevention of PNALD are currently investigated in a randomized, controlled, double-blind clinical trial



QOL

- Emedo et al, King's college 2010 found that children on home TPN developed resilience, positive outlook, and coped well with illness-related demands
- Families can continue to function well.



Follow up

- Monitor I/O, stool < 50cc/kg/day,
- Urine output
- weight gain
- linear growth
- diaper rash
- bone density
- Electrolytes, BUN, Cr
- minerals and trace elements
- vitamins
- liver enzymes
- sexual maturity



Vitamins

- B12
- Fat soluble vitamins
- Iron
- Trace elements
 - Zinc
 - Copper
 - manganese



Medication

- Anti-diarrhea
- Pro-motility
- Anti-acids
- Enzymes (pancreatic)
- Hormones
- Bile acid binding resin- cholestyramine
- Antimicrobials



Motility agents

- Anti diarrheal to prolong intestinal transit (30 min before meal)
 - Loperamide (Immodium)
 - Diphexylate (Lomotil)
 - Clonidine
 - codeine
 - opium tincture
- Promotility agents
 - Reglan
 - Cisapride
 - Erythromycin
 - Augmentin



Gastric Hypersecretion

- Common in 50% of short gut
- May lead to fluid, electrolyte loss, malabsorption and peptic disease
- H2 blockers
 - ranitidine
 - famotidine
- Proton pump inhibitors
 - omeprazole
 - lansoprazole



Hormones

- Somatostatin analogue -octeotride
 - Decreases output and motility
- GH and IGF-1 and Glutamine
 - increase SB adaptation
 - More rapid TPN wean and weight gain
 - Use of GH in adults remains controversial with high side effects
- Glucagon-like peptide 2 (GLP-2)
 - adaptation and decreases motility
 - Given SQ and improved lean body weight



Bacterial Overgrowth

- Residual intestine becomes dilated, dysmotile with stasis
- Mucosal injury, malabsorption, nutrient competition, sepsis, anorexia, vomiting, abdominal distention, FTT, hepatotoxicity and d-lactate
- Diagnosis is empiric and confirmed via duodenal or fecal Cx, hydrogen breath test, D-lactic acidosis
- Therapy;
 - ABX-cycled mostly anaerobic (Flagyl, nitazoxamide)
 - non-absorbed (Rifaximin)
 - Probiotics (In patients without CL)
 - discontinue anti-motility agents



D-lactic acidosis

- Produces by anaerobic bacteria and cleared slowly
 - Confusion
 - Somnolence
 - Dementia
 - Ataxia
 - Seizure
- Therapy
 - Antibiotic
 - Low carbohydrate diet



Are children small adults?

- Oral aversion
- Reflux
- Constipation
- Diaper rash
- Development
- Linear growth
- Weight gain
- Prematurity sequelae
- Immunization
- Sexual maturity
- Growth spurts
- Socialization
- Intentional tube/line pulling
- Cholestasis
- Compliance
- Psycho-social factors
- Family dynamics
- Independence
- Education
- Transition to adult



Multidisciplinary Management of SBS- It takes a village

- Gastroenterology
- Hepatology
- Surgery-general, colorectal, transplant
- Neonatologist
- General Pediatricians
- Specialized nurse
- Interventional radiology
- Pediatric intensivist
- Home nursing
- ID control
- Feeding therapy
- Stoma therapy
- Physical therapy
- Occupational therapy
- Nutritionist with home nutrition support
- PN pharmacist
- Social worker
- Psychologists



Prognosis

- More than 80% of children with IF survive on medical therapy
- Reassess as TPN may be weaned or need to be increased
- Goal is oral independence



Surgical Treatments for Intestinal Failure

- Establishing continuity
- Eliminating stasis
- Prolonging transit time
- Enhance mucosal surface area
- Non transplant surgery
- SB Transplant



Indications for Intestinal Transplantation

- Impending or overt liver failure due to PN associated liver injury (Bil>3, portal HTN, coagulopathy, stomal bleed, cirrhosis)
- Loss of access (2 major vessel thrombosis)
- Recurrent line sepsis, fungal , septic shock , ARDS
- Non-reconstructible GI tract and irreversible IF (ultra short, mucosal disease)
- Recurrent severe dehydration
- Patients must fail PN