Ann & Robert H. Lurie Children's Hospital of Chicago

Diet in Short Gut Syndrome

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Definition of Intestinal Failure

Intestinal failure

- obstruction
- dysmotility
- surgical resection
- congenital defect
- disease-associated loss of absorption

Characterized by the inability to maintain

- protein-energy
- fluid
- electrolytes
- micronutrient balances



Disclosures

- I serve on Nutricia's speaker bureau
- I am the Lurie Children's Hospital of Chicago site PI for the NPS-sponsored safety trial of Gattex®



Common Pediatric Diagnoses

Short bowel syndrome

- Necrotizing Enterocolitis
- Gastroschisis, primary indication for intestinal transplant
- Intestinal Atresias
- Long-segment Hirschsprung's Disease
- Midgut or Intrauterine Volvulus

Motility Disturbances

- Chronic intestinal pseudo obstruction
- Gastroschisis

Mucosal lesions

- Microvillous Inclusion Disease
- Tufting enteropathy



Outline

- Understand the advantages and limitations of carbohydrates, protein, and fat
- Develop regimen to introduce solid foods

 Recognize the optimal timing to refer to a multidisciplinary feeding clinic



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Breast Milk

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- Breast milk always been encouraged
 - 19% of Pediatric Intestinal Failure Consortium (PIFCON) cohort, n=272
 - 20 different infant formulas
- Growth Factors
 - Glucagon like peptide-2
 - Epidermal growth factor
 - Secretory immunoglobulins
 - Lysozyme
 - -Interferon

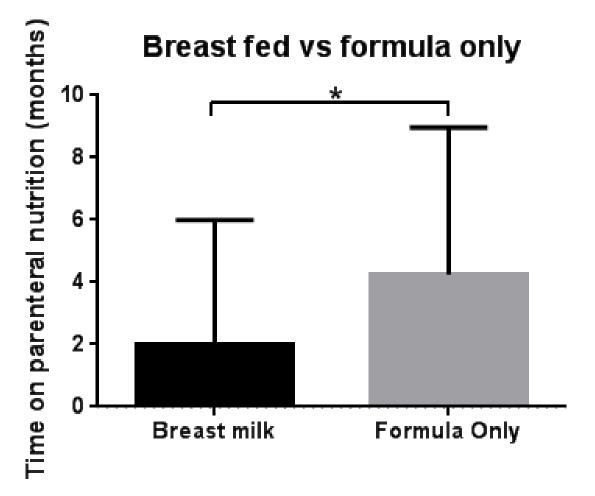


- Improved outcomes with enteral autonomy
 - Mean duration of TPN 290 vs 720 days in non-breast milk infants

Squires et al J Pediatr 2012;161:723-8, Andorsky et al. J Pediatr 2001: 139:27-33,

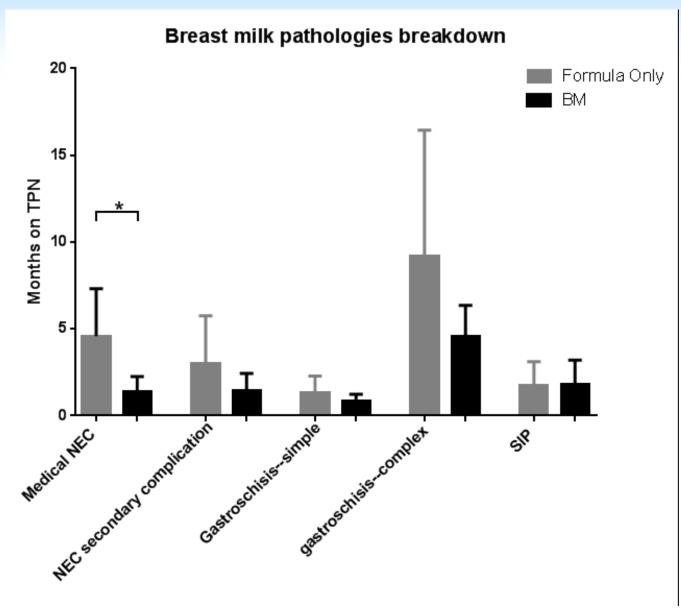


PIFCON



Choquette et al abstract PIFRS 2014





Choquette et al abstract PIFRS 2014



Formula/Protein

- No significant difference in absorption between hydrolyzed and non-hydrolyzed formulas.
- Isosmotic
- Anecdotal data
 - Hydrolyzed and Amino Acid based formulas
 - Evidence of improved absorption in sitting of inflammation
 - Non-IgE mediated milk protein allergy in patients with short bowel syndrome
 - Shorter duration of TPN dependence
- Drawbacks for Amino Acid based formulas
 - Will require calcium/phosphorus supplementation especially in premature infants
 - Expensive

Andorsky et al. *J Pediatr* 2001: 139:27-33, Bines et al. *JPGN* 1998: 27(5):614-616, Degreef et al Journal of Nutrition and Metabolism 2010, Kaufman et al *J Pediatr* 1997;131:356-61



Carbohydrates

- Standard infant formulas 19-20 kcal/oz
- May dilute to 10 or 15 kcal/oz
 - Decreasing the osmotic load to reduce diarrhea
- Avoid fruit juices/fruits
 - Worsen D-lactic acidosis
 - Diarrhea
- If volume sensitive
 - More concentrated formulas 24 kcal/oz or greater
 - Increase caloric intake without increasing volume/fluid load especially in sensitive children
- More concentrated the increased chance of osmotic diarrhea



Fat

- Long chain Triglycerides (LCT)
 - Require bile acids to absorb LCT
 - Ileal resection, loss of enterohepatic circulation
- Medium Chain Triglycerides (MCT) can be directly absorbed
 - Slightly less calories
 - Less helpful adaptation
 - Improved absorption in preserved colon
- Elemental and casein hydrolysate formulas
 - High in MCT
 - More calories from fat even in setting of malabsorption, intestinal resection

DiBaise et al. *Am J of Gastro* 2004;99;1823-1832, Jeppesen and Mortensen. *Gut* 1998;478-483



Randomized Controlled Trial of Early Enteral Fat Supplement and Fish Oil to Promote Intestinal Adaptation in Premature Infants with an Enterostomy

Table III. Nutritional outcomes after bowel reanastomosis*

| | All | | High ostomy | |
|--------------------------------|---------------------|------------------------------------|--------------------|----------------------|
| Infants | Control (n = 17) | Treatment [†] (n = 18) | Control (n = 6) | Treatment (n = 8) |
| Hyperalimentation, d | 13 ± 17 | 10 ± 13 | 27 + 23 | 16 ± 18 |
| Intravenous lipid, d | 11 ± 13 | $6\pm5^{\ddagger}$ | 21 ± 18 | $7\pm7^{\ddagger}$ |
| Total calorie, cal/kg/d | 115 ± 10 | 114 ± 12 | 112 ± 14 | 117 ± 13 |
| Weight gain, g/d | 20 ± 9 | $27\pm11^{\ddagger}$ | 14 ± 4 | $23\pm5^{\ddagger}$ |
| Length gain, cm/wk | 0.9 ± 1.3 | $2.1\pm1.5^{\ddagger}$ | 0.6 ± 0.8 | 2.2 ± 1.6 |
| Head circumference gain, cm/wk | $.1\pm0.7$ | 1.4 ± 1.0 | 0.8 ± 0.9 | 1.2 ± 0.8 |

*Mean \pm SD in the interval between resumption of enteral feedings and attainment of 150 mL/kg/day of enteral feedings.

+Treatment group received early enteral fat supplement and fish oil.

 $\ddagger P < .05$ treatment vs control.

Yang et al J Peds 2014;165:274-9



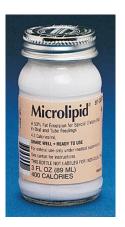
Supplements

- Duocal Powdered carbohydrate ®
 - Hydrolyzed cornstarch 73% Fat supplement 22% (35% MCT)
 - Added to formulas to increase the caloric density

Liquigen

- -42 kcal/Tablespoon
- Microlipid® (100% LCT)
 - Safflower oil
 - -67.5 kcal/tbsp
- Liquigen®
 - Emulsified MCT
 - -67.5 kcal/tbsp







Fiber

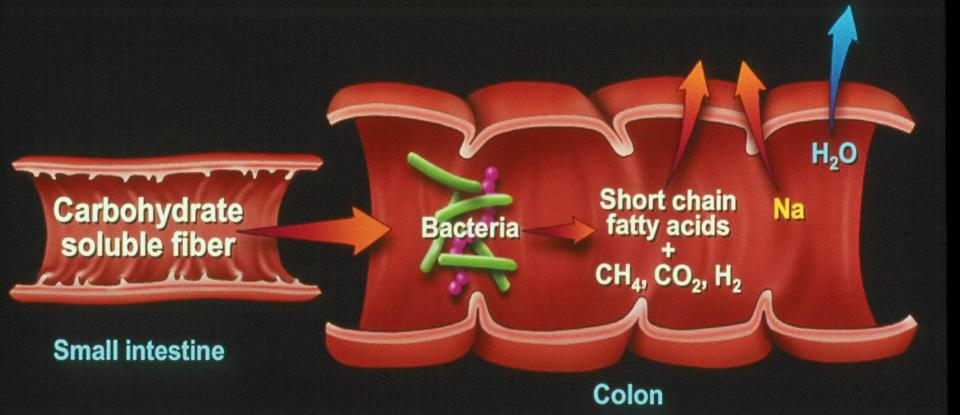
- Soluble fiber is fermented by colonic bacteria
 - Short chain fatty acids; acetate, butyrate, proprionate
 - Colonocyte fuel/health
 - Enterocyte proliferation
 - Water and sodium resabsorption
- Delays gastric emptying
- Decreases gut transit
- Increase fluid absorption decreasing fluid losses





Drenckpohl et al. *Nutr in Clin Practice* 2005:20:674-7. Alam et al. *Arch Dis Child* 2005;90:195-9

Carbohydrate Salvage







Fiber

- Green beans/Pectin
 - Case series using green beans in 3 children with sbs with increase form in their stool
 - Retrospective cohort SBS/IF n=18 with reduction in stool number and increase in consistency with addition of green beans
 - Stage 2 green beans 1 jar/240 ml of formula
- Benefiber®
 - Guar gum (Benefiber®)
 - RCT in persistent diarrhea decreased duration in children receiving hydrolysed guar gum in chicken diet

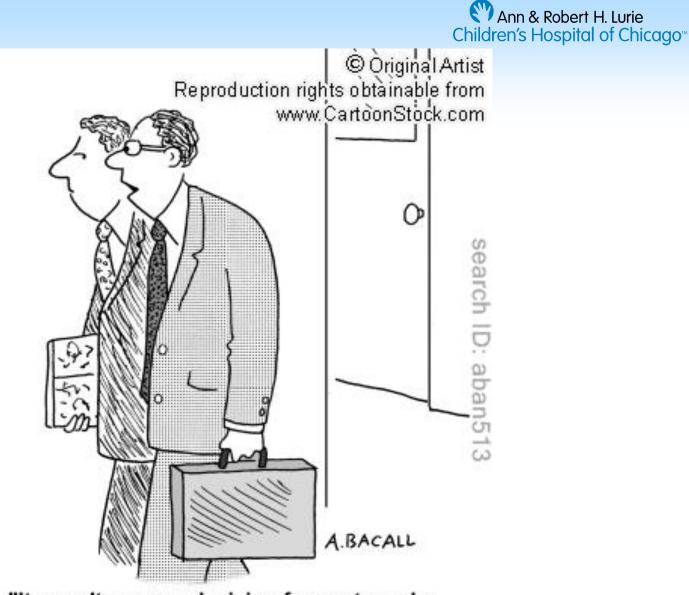
Drenckpohl et al. *Nutr in Clin Practice* 2005:20:674-7. Alam et al. *Arch Dis Child* 2005;90:195-9



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"It wasn't an easy decision for me to make. Lots of coin tossing went into it."



Oral Nutrition

Underlying intestinal physiology

- Remnant length
- Motility disturbances
- Ileum vs jejunum
- B₁₂ deficiency, bile acid diarrhea
 - MCT
- Colonic resection
 - Pectin/Benefiber®/Green beans
- Milk protein allergies

| | Colon Present | Colon Absent |
|------------|------------------------------|--------------------------|
| Carbohydra | ate 50-60% of caloric intake | 40-50% of caloric intake |
| | Complex carbohydrates | Complex carbohydrates |
| Fat | 20-30% of caloric intake | 30-40% of caloric intake |
| | Ensure adequate | Ensure adequate |
| | essential fats | essential fats |
| | MCT/LCT | LCT |
| Protein | 20-30% of caloric intake | 20-30% of caloric intake |
| | High biologic value | High biologic value |
| Fiber | Net secretors | Net secretors |
| | Soluble | Soluble |
| Oxalate | Restrict | _ |
| Fluids | ORS and/or hypotonic | ORS |

DiBaise et al. Am J of Gastro 2004;99;1823-1832



Introduction of foods

Starches

- Complex carbohydrates but maybe bland
- Milk free cereal (Beechnut or Earth's Best)
- Meat
 - Protein absorption in stomach and proximal small intestine
- Vegetables
 - Green beans-pectin
 - Beneficial in the setting of a colon
- Foods to avoid
 - Milk protein in allergic patients
 - Juices
 - Fruits



Taste

- Short bowel syndrome
 - Spicy
 - Sharp
 - Salty
- Tend to avoid bland foods
 cereals











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Normal progression of feeding skill acquisition:

| Breast / Bottle only | 0-4 mos | |
|------------------------|-----------|--|
| Smooth puree by spoon | 4-6 mos | |
| Soft chewables and cup | 6-8 mos | |
| Mashed table food | 8-12 mos | |
| Chopped table food | 12-18 mos | |

Development of Swallowing and Feeding: Prenatal through First Year of Life Delaney & Arvedson, Dev Dis Res Rev, 2008



Taste/Easting

- Oral feedings
 - Physiologic
 - Pleasurable, part of culture
 - Secretion of GI trophic factors
 - Minimizes feeding disorders
 - Introduced as soon as medically stable
 - Dipping pacifier into formula
- Non-nutritive sucking appears to facilitate development of sucking behavior and may improve the transition from tube to bottle feedings

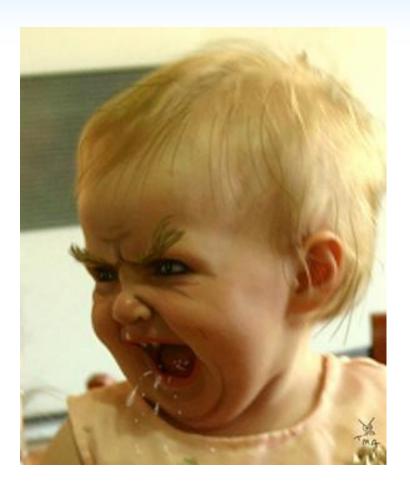
Negri et al. JPGN 2011;53:601-605, Beachamp et al. Dev Psychobiol 1986:19:17-25



27

Oral Feedings





Wessel and Kocoshis et al. *Semin Perinatol* 31:104-111. Goulet et al. *Clinical Nutrition* 2013; 32: 162-171



Oral Aversion

- May be a direct result from prolonged intubation, nasogastric tube feedings, hospitalizations
- Eating is not pleasurable and maybe associated with very negative feelings
- -Solids are typically introduced 4-6 months (corrected for gestational age)
 - Significant delay in SGS patients
- -Small amounts of formula on pacifier to try to minimize



Speech Therapist

- Evaluate child's oral motor skills
- Must be able to master suck/swallow/breathe sequence or unable to advance
 - Breast or bottle feed** foundation of all eating
 - Forward and backward tongue movements
- Spoon feed
 - Close their lips and draw food into the mouth and subsequently to the back of their mouth
- Table foods
 - Move tongue to side of the mouth and place food onto their molars



Oral Aversion

- Occupational Therapist
 - Evaluate your child's ability to process sensory information
 - Create a treatment program to help improve tolerance of sensory input
 - Helps parents to understand and be able to help the child enjoy feedings



Timing of referral

- Depends on your resource availability
- Expertise in eating/feeding disorders ASAP
 - -Speech Therapist
 - -Occupational Therapist
- GI MD
 - -Vomiting
 - Milk protein allergy
 - Delayed gastric emptying/motility disorder
 - –Choking and gagging
 - -GER



Specialized Feeding teams

MDs

- -GI, ENT, general pediatrician
- Ensure no other medical problem is contributing to the feeding disorder
- Speech Therapist
 - Feeding disorders that pertain to oral motor skills and swallowing
- Occupational Therapist
 - Sensory processing disorder or feeding disorder
- Registered Dietician
 - Nutritional needs
- Behavioral Psychologist
 - Guide caretakers with their interaction during meals



Nutritional and Psychosocial Outcomes of Gastrostom Tube–Dependent Children Completing an Intensive Inpatient Behavioral Treatment Program

*Alan H. Silverman, *Midge Kirby, *Lisa M. Clifford, [†]Elizabeth Fischer, [‡]Kristoffer S. Berlin, *Colin D. Rudolph, and *Richard J. Noel

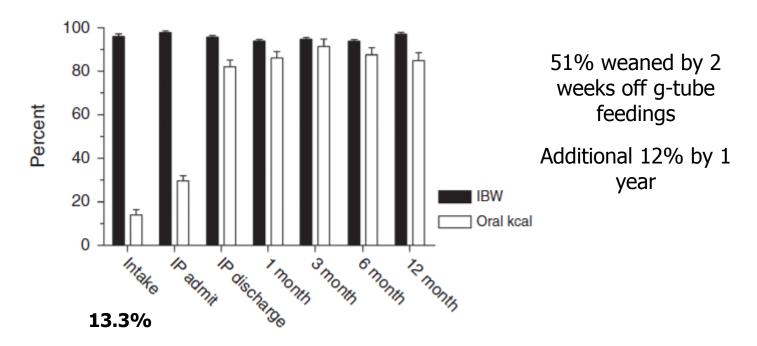


FIGURE 2. Longitudinal nutritional status of study participants.

Silverman et al. JPGN 2013:57:668-72



Conclusion

- Underlying pathophysiology of the patient
 - Remnant length
 - Colonic resection
- Foods
 - Vegetables, meats
 - Avoid high sugar containing foods
 - Spicy, salty
- Oral Aversion
 - Common in these patients
 - Speech Therapy/Occupational Therapy
 - Multidisciplinary Feeding Teams beneficial if available