NUTRITION AND FATTY ACID OXIDATION DISORDERS: From the Tube to the Table

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Preview

- General nutrition guidelines
- Individual symptom management
- Transitioning from tube to oral intake

NUTRITION SUPPORT GOALS

1. Provide enough macro and micro nutrients to ensure proper growth and development

   - Vitamin and minerals should be provided at the greatest intakes suggested for each age group
   - Make sure to supplement essential fatty acids

NUTRITION SUPPORT GOALS

- 2. Prevent fasting/catabolism
  - Allow infants to fast no longer than 4-6 hours; older children and adults no longer than 8 hours
  - If ill, recommend Pedialyte, GatorAde, fruit juices
  - If unable to take anything orally, hospital admission for IV dextrose

NUTRITION SUPPORT GOALS

3. Correct the primary imbalance in the metabolic pathway
   - Restrict, through diet, the accumulation of the substrate that is toxic
   - Supply products of the blocked pathway
   - Supplement cofactors and ‘conditionally essential’ nutrients

GOALS OF NUTRITION THERAPY

4. Manage individual symptoms

- failure to thrive
- swallowing difficulties
- oral aversions
- GI disturbances (vomiting, diarrhea, constipation)

Monitor

- Measure weight, height, head circumference (as often as weekly weights in infancy)
- Provide disease specific diet, formula, and supplement recommendations
- Blood levels of electrolytes, nutrition indices, essential fats

Case of Scooby Doo

- 10 y/o male with VLCAD
- Identified by NBS and started on treatment at 6 days of life
- Breast fed first week of life, poor suck-reflex and reflux
- Spent weeks in NICU with IVs and NG tube

- Sent home at 1 month of age; bottle fed Pregestimil (contains 55% MCT oil, 45% long chain fats) and Polycose
- Frequent admissions requiring IV dextrose and NG feeds during first few year of life
- By 14 months of age, Scooby only tolerating small amount of high calorie formula, by bottle; very small volume of baby foods. He dropped 2 percentiles on growth chart for weight. Not taking any solid foods.

Failure to Thrive

- Weight in young infants or height in children under 2 that consistently falls below the 3rd %ile for age
- Failure to maintain an established growth curve
- Weight for height that falls below the 10th %ile on growth curve
- Weight deceleration across 2 or more major %iles
- Weight loss of >10% in a previously healthy child

Aspiration indicators

- Coughing and choking during feeding
- Increased congestion
- Increased fussiness during feeding
- Frequent or persistent respiratory illnesses
- Persistent low grade fever
- Wet vocal quality during or after feeding
- Failure to progress to the next developmental level of feeding

Caloric Supplementation

Infants

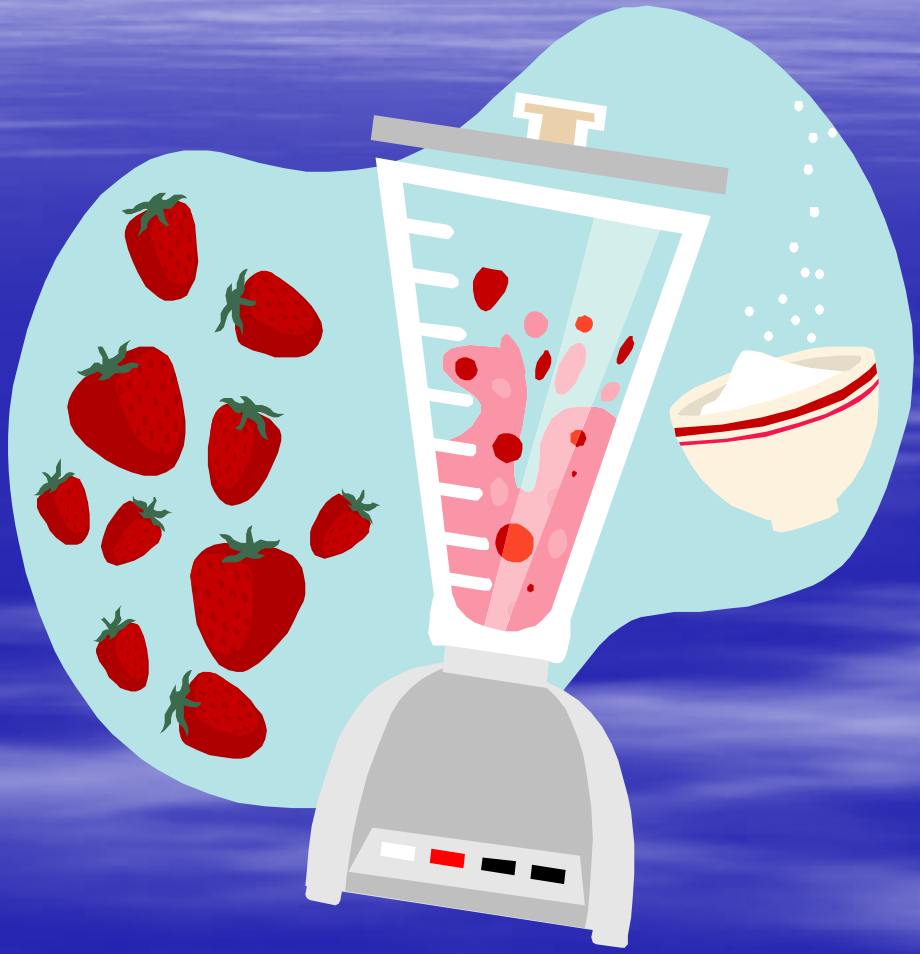
- Formula - increase caloric density of formula
- Strained foods - choose high calorie foods, fortify with infant cereal, Polycose, Benecal, Duocal & oil

Toddlers

- Increase caloric density:
  - Milk: dry milk or instant breakfast
  - Juices: Calorie supplement
  - Entrees/vegetables: add gravies, sauces, margarine
  - Snacks: add cream cheese, peanut butter

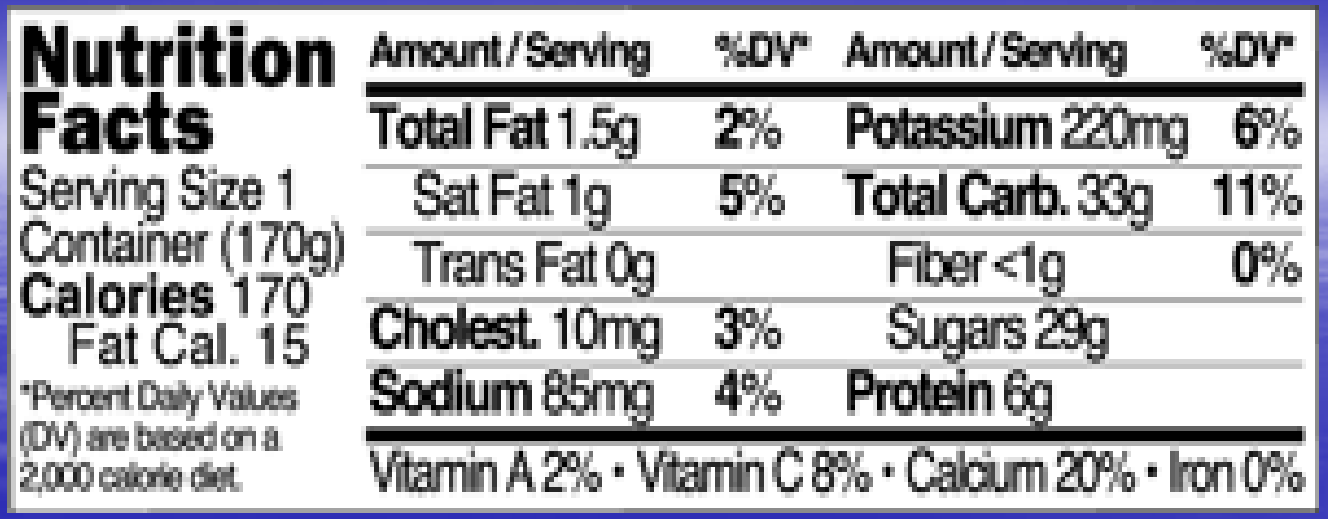
Pureeing Food

- Recommend pureed table foods over baby foods if age appropriate
- Recommend using a blender or food processor for anything the family is eating
- Add extra high calorie fortifiers
- Pour puree into ice cube trays and freeze, most ice cubes are 30 mL



Goal for weight gain

- Focus on highly preferred foods
- Every calorie counts
- Fortify foods to add extra calories
- Choose high calorie food options, look at brands



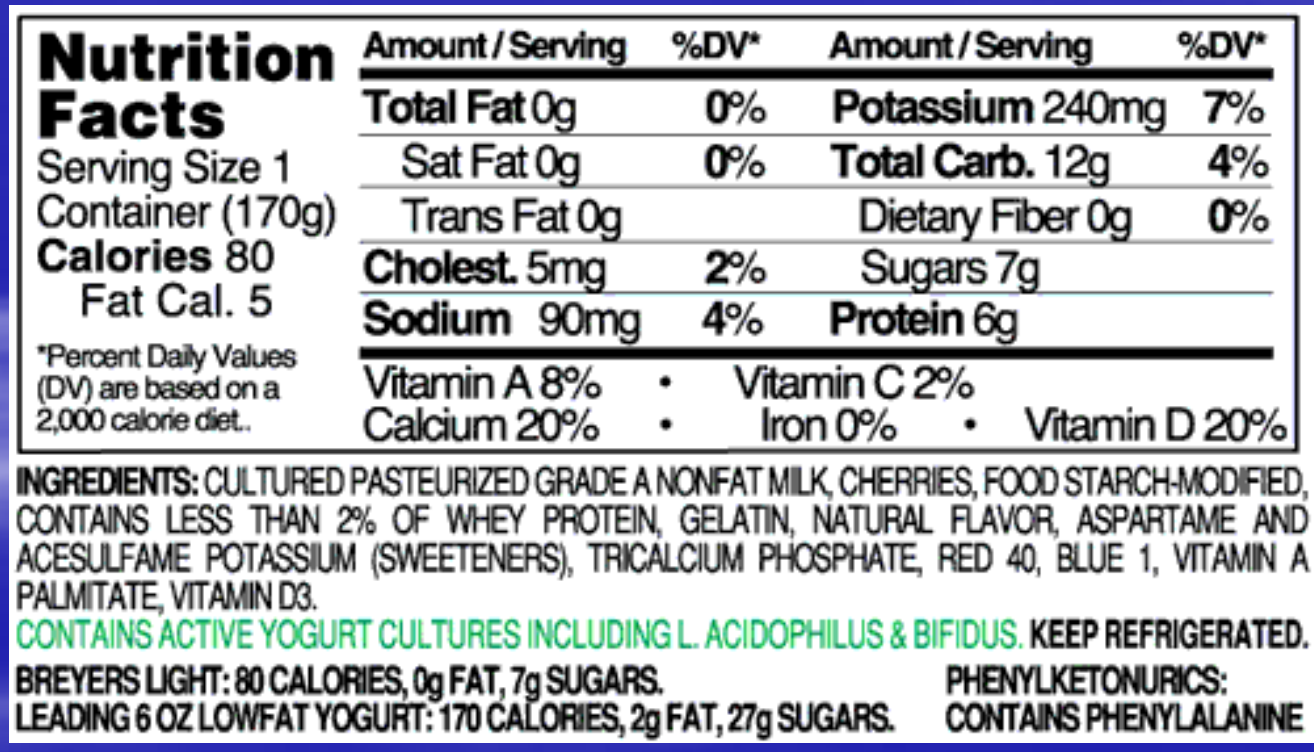
Nutrition Facts

Serving Size 1 Container (170g)
Calories 170
Fat Cal. 15

*Percent Daily Values (DV) are based on a 2,000 calorie diet.

Amount / Serving	%DV*	Amount / Serving	%DV*
Total Fat 1.5g	2%	**Potassium** 220mg	6%
Sat Fat 1g	5%	**Total Carb.** 33g	11%
Trans Fat 0g		Fiber <1g	0%
Cholest. 10mg	3%	Sugars 29g	
Sodium 85mg	4%	**Protein** 6g	

Vitamin A 2% • Vitamin C 8% • Calcium 20% • Iron 0%



Nutrition Facts

Serving Size 1 Container (170g)
Calories 80
Fat Cal. 5

*Percent Daily Values (DV) are based on a 2,000 calorie diet..

Amount / Serving	%DV*	Amount / Serving	%DV*
Total Fat 0g	0%	**Potassium** 240mg	7%
Sat Fat 0g	0%	**Total Carb.** 12g	4%
Trans Fat 0g		Dietary Fiber 0g	0%
Cholest. 5mg	2%	Sugars 7g	
Sodium 90mg	4%	**Protein** 6g	

Vitamin A 8% • Vitamin C 2%
Calcium 20% • Iron 0% • Vitamin D 20%

INGREDIENTS: CULTURED PASTEURIZED GRADE A NONFAT MILK, CHERRIES, FOOD STARCH-MODIFIED, CONTAINS LESS THAN 2% OF WHEY PROTEIN, GELATIN, NATURAL FLAVOR, ASPARTAME AND ACESULFAME POTASSIUM (SWEETENERS), TRICALCIUM PHOSPHATE, RED 40, BLUE 1, VITAMIN A PALMITATE, VITAMIN D3.

CONTAINS ACTIVE YOGURT CULTURES INCLUDING L. ACIDOPHILUS & BIFIDUS. **KEEP REFRIGERATED.**

BREYERS LIGHT: 80 CALORIES, 0g FAT, 7g SUGARS.
LEADING 6 OZ LOWFAT YOGURT: 170 CALORIES, 2g FAT, 27g SUGARS.

PHENYLKETONURICS: CONTAINS PHENYLALANINE

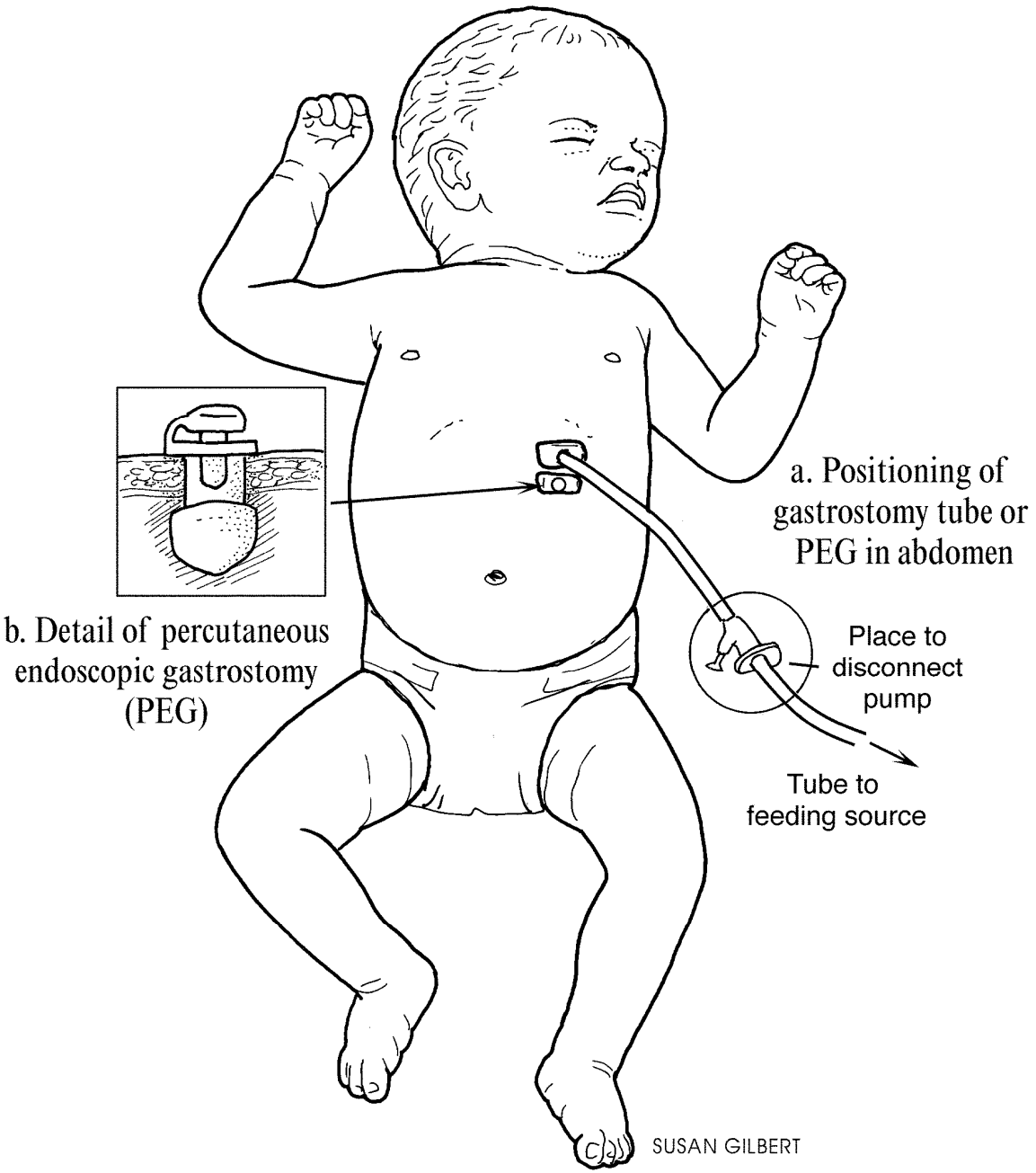
Alternative forms of Nutrition

- IV – provides dextrose (sugar), hydration and electrolytes
- TPN – provides nutrition when GI tract can not be used (most fat mixtures are not appropriate for FAODs)
- Tubes: Oral-gastric, Naso-gastric, Naso-jejunal, Gastric, and Gastro-jejunal (GJ)

Benefits of Gastric Tube Feeding (G-Tube)

- Decrease stress in child and family
- Continue to feed for pleasure, if safe
- Better route of nutrition during crisis
- Provide an excellent source of nutrition
- Can use for medications, supplements, and formulas

Figure 61: Gastrostomy Tube and PEG



G-tube

Mickey Button

Types of Administration

- Continuous
- Bolus
- Night time

Continuous Feedings

- Run on a pump at a set rate for up to 24 hours per day
- Pros:
  - Often times most easily tolerated
  - Will prevent fasting catabolism
- Cons:
  - Does not allow for normal hunger/satiety cycle
  - Individual must be connected to tubing 24 hours per day

Night Feedings

- Run on a pump but over night only, may be anywhere from 8-12 hours
- Pros:
  - Allows for time disconnected from tubing during the day
  - Often times well tolerated
  - Eliminates night-time catabolism
- Cons:
  - Does not allow for normal hunger/satiety cycle

Bolus Feedings

- Set volume is given over a short period of time, may be given via syringe push, gravity or run on pump at high rate
- Pros:
  - Allows for normal hunger satiety cycle
  - Allows for time disconnected from tubing
  - Can subtract amount consumed orally
- Cons:
  - May not be tolerated by some individuals
  - Must be given frequently enough to prevent catabolism

- Gets G-tube at 20 months of age
- Starts with continuous feeds, advances to continuous feeds at night only with day-time bolus feeds
- Shorter stays in hospital
- Scooby starts growing and gaining weight well

Important Therapy for Tube Fed Child

- **Feeding therapist:**
- Modify utensils (smaller spoons, cut- out cups, larger/smaller nipple holes)
- Chew/swallow techniques (chin tuck, towling, proper feeding positioning)
- Determine appropriate consistency for foods and liquids
- Reduce oral aversions through play and behavior modification

- Age 6, Scooby enters school and makes friends
- Takes interest in eating foods
- Voices desire to have tube removed….
- What does Scooby need to do??

Progression from Tube Feedings to Oral Feeding

- Must consistently consume 60 mL (2 ounces) of solids per day
- Family is educated on fortifying foods to 1 calorie per mL
- Adjust tube-feedings to allow for time off prior to oral feedings
- If possible, adjust to all daytime bolus feedings and cut down on night feedings
- Family or care takers record all intakes

Progression from Tube Feedings to Oral Feeding

- Tube feedings are decreased as oral intake increases
- May require frequent visits with clinic for nutrient intake assessment, body weight, and blood work monitoring
- May recommend appetite stimulant, such as Periactin, if agreeable with MD
- Recommend tube removal after two bouts of illness without use and consistent weight gain and growth

Goal to decrease tube-feeds

- Volume is more of a goal than variety
- Boost oral calories by fortifying foods
- Focus on preferred foods
- Easier textures often go more quickly

Goal to increase volume

- Focus on set meal times rather than grazing
- Work towards a set, daily meal schedule that is consistent
- Utilize preferred foods at all meals
- Remember, a calorie is a calorie

Caloric Supplementation

Infants

- Formula - increase caloric density of formula
- Strained foods - choose high calorie foods, fortify with infant cereal, Polycose, Benecal, Duocal, essential fatty acid supplements & MCT oil

Toddlers

- Increase caloric density:
  - Add flavoring to formula (strawberry syrup, KoolAde, Sunny D)
  - Fruits and Juices: Calorie supplement and/or sugar, maple syrup, non dairy whip
  - Entrees/vegetables: add oils, sauces
  - Snacks: add jelly to crackers, low fat whip to pudding, sugar to cereal

Typical diet recall

- Breakfast: ½ cup oatmeal, 4 ounces whole milk, 4 ounces fruit juice, ½ banana
- Lunch: 2 slices bread, 2 slices turkey, 1 slice cheese, 4 ounces salad/dressing, 1 apple, 8 ounces water
- Dinner: 4 ounces chicken, ½ cup rice, ½ cup green beans, 8 ounces water

Calorie Fortified Meal Plan

- Add coconut oil and sugar to oatmeal, add dry powdered milk to skim milk, add Duocal to fruit juices
- Add in between meal and before bed snacks of yogurts, puddings, sugared fruit purees
- Substitute juices for water
- Find out if commercial calorie boosters are ok for diet Can add an extra: 850 kcals

Real Food Fortifiers

- Coconut and Palm oil, naturally high in MCT (medium chain triglyceride)***
- Sugar

- Wheat germ
- Maple syrup or honey
- Powdered milk

- ***Used only in long chain fatty acid oxidation disorders

Commercial Fortifiers

- Polycose – liquid or powder
- Duocal
- MCT Procal
- MCT oil
- Beneprotein
- Benefiber
- Thick it

MCT Procal