

NUTRITION IN SMITH LEMLI OPITZ SYNDROME

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June 27, 2019



National Institutes of Health
Clinical Center

OUTLINE

- Nutrition-related issues in Smith Lemli Opitz Syndrome (SLOS)
- Major components of nutrition
- Meeting nutrition needs in SLOS
- Tube feeding options

NUTRITION-RELATED ISSUES IN SLOS

- Poor growth compared to general population
- Feeding problems (cleft palate, weak suck, swallow difficulties, vomiting)
- Oral tactile defensiveness and eating selectivity
- Intestinal dysmotility- reflux, constipation

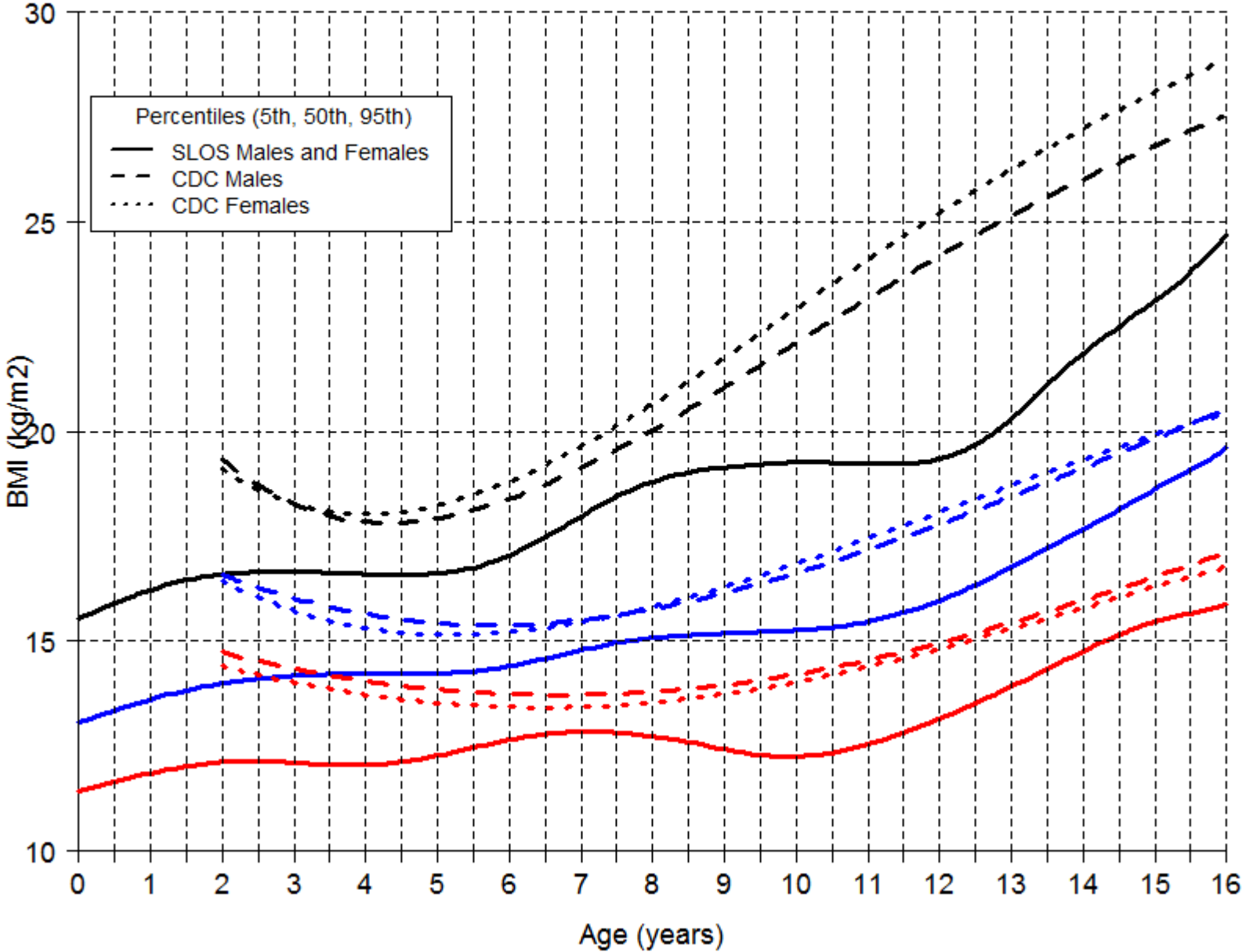
COMPONENTS OF NUTRITION

- Calories/Growth
- Protein
- Fluid
- Fiber
- Vitamins/Minerals
- Cholesterol

CALORIES/GROWTH

- Difficult to predict calorie needs for individuals with SLOS
 - Track growth to determine if calorie intake is appropriate
- Height, weight, and body mass index (BMI) should track parallel to the curve
- SLOS growth charts reflect height, weight, and BMI that are below growth charts of general population
- Avoid overfeeding

Smith-Lemli-Optiz Syndrome, BMI vs. Age (0-16 Years) with CDC Curves



OPTIMIZING CALORIES- FATS/OILS

- Butter, cream, cooking oils, peanut butter, nuts, avocado, hummus, cheese, eggs
- Can be added to many foods: sauces, meats, yogurt, smoothies, pasta/mac n' cheese, soup, mashed potatoes, pancakes, muffins, oatmeal

ADEQUATE FLUID INTAKE

- One factor in managing constipation
- Fluid needs are based on body weight
- How to get the fluid in:
 - Keep fluid accessible throughout the day
 - Set up mini goals for each part of the day
 - Put fluid goals in IEP
 - Use food as a source of fluid
 - Melons, cucumbers, iceberg lettuce, carrots, berries, citrus fruits
 - Soups
 - Gelatin
 - Ice cream, popsicles, sorbet
- Aim for even distribution of fluid delivery across the day



<https://i.ytimg.com/vi/Fll5mmlTrJA/hqdefault.jpg>

FIBER

- Another factor in managing constipation
- Insoluble fiber is most effective for constipation
 - Bran (cereals, muffins)
 - Whole grains (whole wheat bread, brown rice, popcorn)
 - Beans/Lentils
 - Whole fruits (including skins)
 - Sorbitol-containing fruits can help with constipation – apples, pears, cherries, plums, apricots, peaches and their dried versions or juices
 - Vegetables
 - Nuts
 - Seeds

VITAMINS/MINERALS

- Common deficiencies in children: Calcium, iron, vitamin D
- Elimination of entire food groups could lead to other deficiencies
 - Grains- iron, folate, B vitamins, iron, magnesium
 - Fruits- vitamin C, folate, potassium
 - Vegetables – vitamin A, vitamin C, folate, potassium
 - Meat- B vitamins, iron, zinc
 - Dairy- calcium
 - Fish- omega- 3 fatty acids

VITAMINS/MINERALS & MEDICATIONS

- Anti-reflux medications
 - Decreased stomach acid can lower iron and calcium absorption
- Iron-
 - check iron status periodically
 - if supplementation is needed, consider 'every other day' schedule
- Calcium-
 - blood levels do not reflect body calcium status; need bone density test
 - use calcium citrate if supplementation is needed

VITAMINS/MINERALS & MEDICATIONS

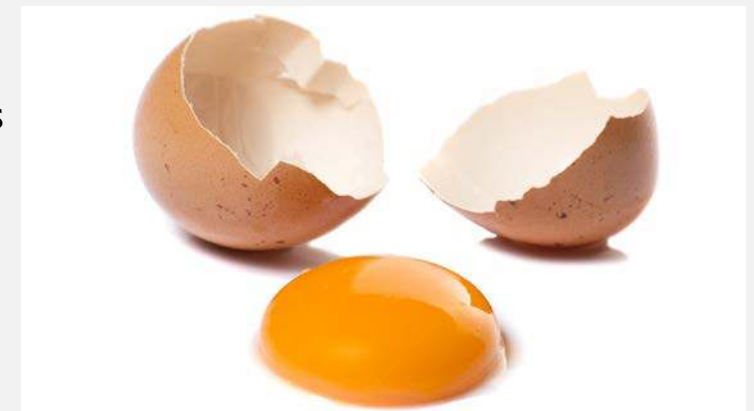
- Anti-seizure medications
 - Can reduce status of calcium, vitamin D, folate, vitamin B6, vitamin B12, and/or carnitine

VITAMIN/MINERAL SUPPLEMENTATION

- Dietary supplements are not regulated pre-market by the FDA
- Liquid and gummy multivitamins tend to be less complete than tablets
- Few multivitamins contain a significant amount of calcium, phosphorus or magnesium
- Avoid mega doses (unless otherwise indicated)

CHOLESTEROL SUPPLEMENTATION

- Food:
 - Liver, Egg Yolks, Shrimp are biggest sources
 - Pasteurized eggs (In-the-shell, Liquid, Frozen, Powdered)
 - OHSU SLOS cookbook: SLO Foundation Website → FAQ's → Books and Learning Websites
- Cholesterol Suspension:
 - Crystalline form dissolved in aqueous or oil-based solutions



https://media2.s-nbcnews.com/i/newscms/2016_06/969866/eggs-today-160211-tease_e3414da28431d69ea44f9eb36aa7667a.jpg

CHOLESTEROL SUPPLEMENTATION

- Medical Food-

CholexMAX™ (Solace Nutrition)
- not readily soluble; can be mixed into foods/ thick liquids

Cholextra t/f™ (Solace Nutrition)
- powder is added to formula for administration via feeding tube



- Dietary supplement-

- Sonic Cholesterol™ (New Beginnings Nutritionals)- crystalline powder in capsule form










MyPlate Plan

Find your Healthy Eating Style

Everything you eat and drink matters. Find your healthy eating style that reflects your preferences, culture, traditions, and budget—and maintain it for a lifetime! The right mix can help you be healthier now and into the future. The key is choosing a variety of foods and beverages from each food group—and *making sure that each choice is limited in saturated fat, sodium, and added sugars*. Start with small changes—“MyWins”—to make healthier choices you can enjoy.

Food Group Amounts for 1,200 Calories a Day

				
1 cup	1 1/2 cups	4 ounces	3 ounces	2 1/2 cups
Focus on whole fruits	Vary your veggies	Make half your grains whole grains	Vary your protein routine	Move to low-fat or fat-free milk or yogurt
Focus on whole fruits that are fresh, frozen, canned, or dried.	Choose a variety of colorful fresh, frozen, and canned vegetables—make sure to include dark green, red, and orange choices.	Find whole-grain foods by reading the Nutrition Facts label and ingredients list.	Mix up your protein foods to include seafood, beans and peas, unsalted nuts and seeds, soy products, eggs, and lean meats and poultry.	Choose fat-free milk, yogurt, and soy beverages (soy milk) to cut back on your saturated fat.

HOW TO IMPROVE DIETARY BALANCE & VARIETY

- Two-Pronged Approach
 - Long-term improvement in diet quality
 - Exposure, exposure, exposure
 - Avoid battles over food
 - Consider feeding therapy
 - Short-term
 - Provide foods in the most palatable form

HOW TO GET THE FRUITS/VEGGIES IN

- Increase palatability by changing texture, mixing with other food items, adding preferred flavors
 - Try textures that are soft (cooked vs. raw vegetables) or crispy (kale chips)
 - Add pureed vegetables/fruits to sauces (pizza sauce), smoothies, pancakes, muffins
 - Finely dice vegetables and bake into muffins, pancakes, meatloaf or hamburgers
 - Try strong flavors- balsamic vinegar, lemon, or spicy flavors, if preferred
 - Consider using dips with fruits/vegetables

POTENTIAL INDICATIONS FOR FEEDING THERAPY

- Limited or poor oral intake
- Food refusal/selectivity; Sensory problems
- Inadequate or slow weight gain
- Suspected or identified problems with airway protection and swallowing
- Chewing difficulties; problems moving food around in the mouth
- History of gagging
- Lack of oral feeding experiences

TARGETS OF THERAPY

- Oral motor skill development
- Expanding the amount/types of foods consumed
- Development of positive associations with food

FEEDING THERAPY OPTIONS

- Feeding Therapist (speech language therapist (SLP), occupational therapist (OT), sensory integration specialist)
- Feeding Program
 - More intensive therapy
 - Day programs, inpatient programs
 - Multidisciplinary team (behavioral psychologist, OT, SLP, dietitian, social worker, gastroenterologist)
 - Caregiver training

PRE-THERAPY DISCUSSIONS

- Agree on the specific therapy goals
- Understand the methods to be used in therapy

NUTRITION VIA FEEDING TUBE

- Formulas
- Blenderized feeds

BENEFITS OF FORMULA

- Provide complete nutrition
- Many products are ready to feed (minimal preparation needed)
- Can be delivered continuously or as bolus feeds
- Negligible risk of clogging even small tubes
- Usually covered by insurance
- Formulas can be specialized (allergies, malabsorption)

FORMULA SELECTION FACTORS

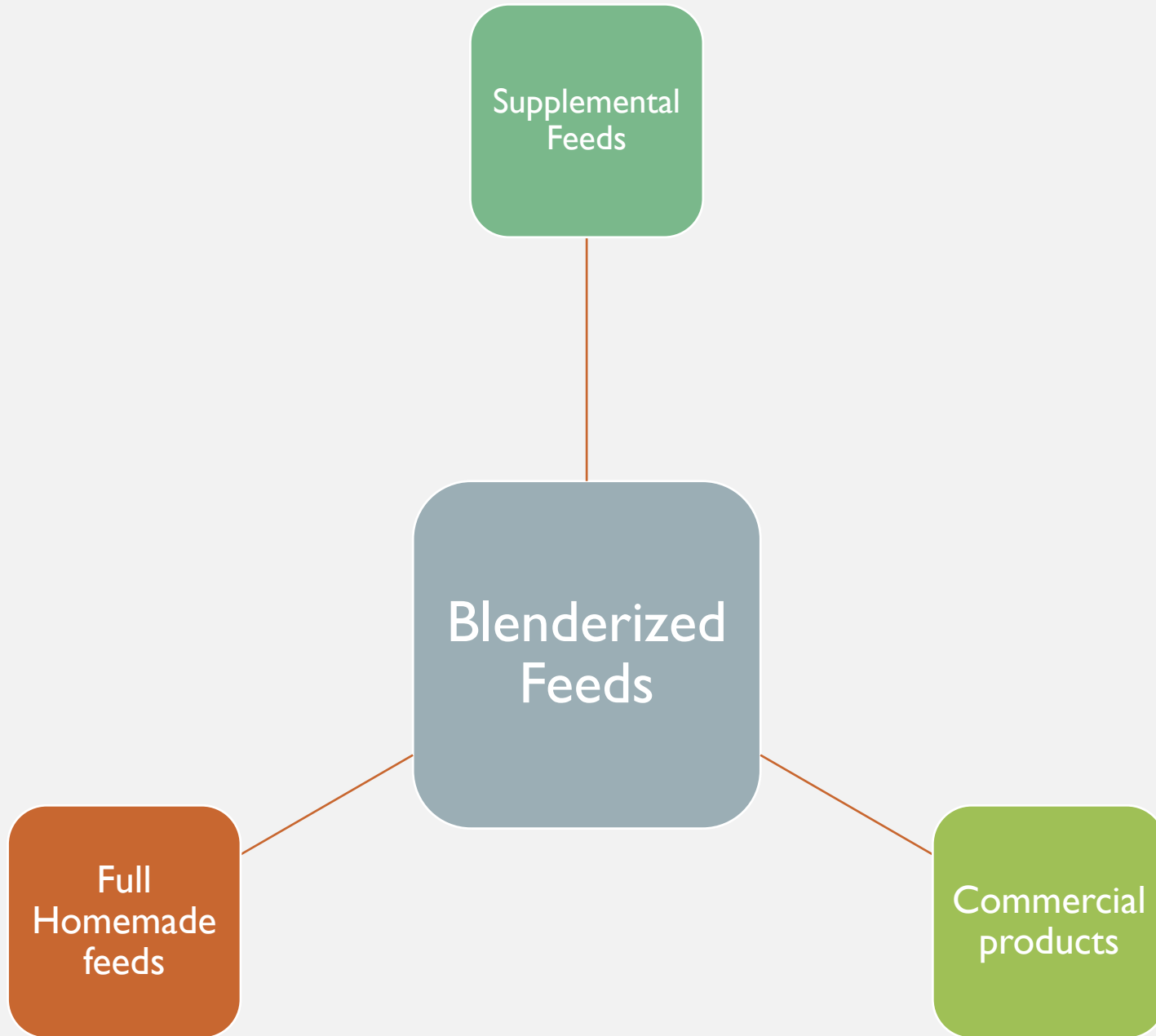
- Age
- Gastrointestinal function and symptoms
 - Osmolality
 - Fiber content
 - Easily digested nutrients
- Food allergies
- Calorie needs
- Volume tolerance

FORMULA TYPES

- Standard formulas with intact protein
- Semi-elemental or partially hydrolyzed formulas
 - Designed for those with impaired digestion
 - May use whey for more rapid gastric emptying
 - Proteins are partially broken down
 - May include MCT oil, which is more easily absorbed
- Elemental
 - Designed for milk protein allergies
 - Free amino acids (proteins are all the way broken down)
 - Often include MCT oil

BENEFITS OF BLENDERIZED TUBE FEEDS

- Fiber and phytochemical content
- Flexibility in foods used (vegan diet, avoiding allergens)
- Management of GI symptoms?
- Exposure to food aromas and flavors



Benefits

- Flexibility in foods provided
- Lower cost than commercial formulas (if not covered)

Considerations

- Not calorically dense- may need high volume to meet needs
- Bolus feeds through G-tube, pushed via syringe
- Nutrient deficiencies more likely
- Risk for clogging tube: 14 French tube size or larger
- Need a good blender
- Food safety considerations
- Requires preparation time



Benefits

- Flexibility in schedule
- Less concern for meeting nutrient needs from blenderized feeds
- Can be a source of some fiber/phytochemicals when full blenderized feeds are not an option (focus on fruits/vegetables)
- Formula supply is available when unable to provide supplemental feeds (vacation)

Considerations

- Bolus feeds through G-tube, pushed via syringe
- Risk for clogging tube: 14 French tube size or larger
- Food safety considerations

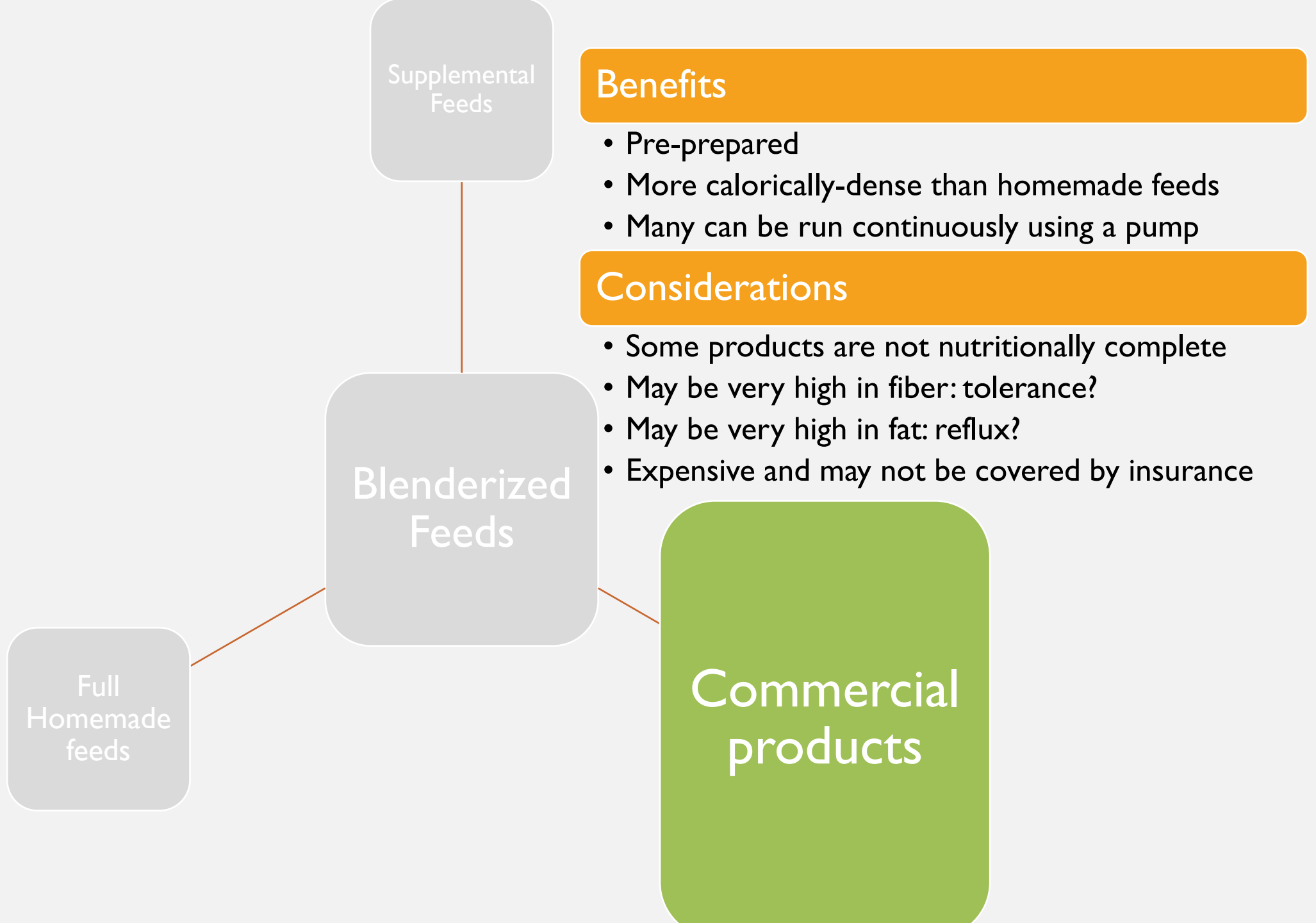
Supplemental Feeds

(using some blenderized feeds to supplement formula)

Blenderized Feeds

Full
Homemade
feeds

Commercial
products



RESOURCES

- Primary physician and Gastroenterologist
- Registered Dietitian Nutritionist
- https://oley.org/page/TFTips_LandingPage

CONCLUSIONS

- Getting optimal nutrition can be challenging in SLOS
- The best method for optimizing nutrition may differ from family to family

REFERENCES

Gallagher K, Flint A, Mouzaki M, et al. Blenderized enteral nutrition diet study: feasibility, clinical, and microbiome outcomes of providing blenderized feeds through a gstrictube in a medically complex pediatric population. *J Parenter Enteral Nutr.* 2018;42(6):1046-1060.

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QUESTIONS