NUTRITION FOR LBSL AND OTHER LEUKODYSTROPHIES

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TOPICS

- General Principals for Nutrition Health
- Common Nutrition Concerns for Patients
  - Altered nutrition needs
  - Oral-motor difficulties
  - Swallow safety
  - GI dysfunction
  - Constipation
  - Picky eating
  - Specialized diets
TOPICS

- Supportive Nutrition Approaches
  - Micronutrient therapy (mito cocktail)
  - Ketogenic diet
  - Antioxidant-rich diet
GENERAL NUTRITION PRINCIPLES

- **Adequate** but not excess calories, macronutrients, vitamins, minerals and FLUID
- **Balanced**: fruits, vegetables, proteins, carbohydrates including whole grains, dairy or dairy substitutes
ALTERED NUTRITION NEEDS

- Higher calorie needs/chronic underweight
  - Caloric supplements, increased meal frequency, avoid fasting, enteral or parenteral feeding
- Metabolic dysregulation
  - May need restrictions of carbohydrates, protein or fat
- Excess weight gain
  - Limited/decreased mobility, poor dietary choices, medications
CONCERNS

- **Oral-motor difficulty (may be progressive): chewing, fatigue**
  - Include soft foods, decrease texture, small, frequent meals, high-calorie liquid supplements; may require a feeding tube

- **Swallow safety: choking, aspiration**
  - Swallow study
    - modify drink consistency or food texture, avoid risky foods (nuts, grapes, hot dogs); may be advised to limit volume, avoid drinks, or perhaps eat only for pleasure; may require a feeding tube
  - Oral-motor therapy (OT or SLP)
CONCERNS

- GI dysfunction:
  - GER:
    - pain, throat discomfort, food coming back up, vomiting
    - treat with positioning, medications, avoiding trigger foods
  - Slow esophageal or gastric motility:
    - food “gets stuck,” full after a small volume, vomiting
    - small frequent meals, emphasize liquids/chase solids with drinks, modify texture, pro-motility medicines
CONCERNS

- **Constipation**
  - infrequent stools, small hard stools or large painful stools, smearing, bloating, nausea, vomiting, poor intake
  - manage with adequate fluid, dietary fiber, exercise/movement.
  - may require medication because of poor tone/motility: stool softeners, laxatives, suppositories, enemas; bowel program
CONCERNS

- Picky eating:
  - limited variety, food refusal, poor fluid intake
  - caution for specialized diets that limit variety even more
  - refusal to eat a medically recommended diet
  - dietary non-compliance

- Treatment: behavioral therapy, food chaining, creative presentation of foods
CONCERNS

- **Specialized diets** (ketogenic, low-fat/low saturated fat)
  - May be unappetizing
  - May create other health risks (nutrient deficiencies, abnormal blood lipids or glucose)
  - Further limits already picky eaters
  - Difficult to maintain compliance
  - Extra cost and/or preparation
SPECIALIZED NUTRITION

- **Ketogenic diet**: high-fat diet, primarily used to treat seizures refractory to medication
  - Contraindication: errors of fatty acid metabolism
- **Pharmacologic treatment**: “Mito cocktail” – goal is to replace or increase compounds that metabolize energy
  - Some of the compounds are disease specific, or their efficacy has not been elucidated
OTHER DIETARY STRATEGIES

- **Antioxidant supplements**: goal is to reduce reactive oxygen molecules. There is such a thing as “too much of a good thing.”

- **Healthy eating plans**: DASH diet, Mediterranean diet, avoiding processed foods, avoiding sugar. Goal is to support optimal health, manage symptoms, avoid secondary mitochondrial dysregulation.
FAQ

- Should certain foods be avoided?
- Should certain foods be emphasized?
- Will eating organic foods improve health?
- Should sugar be avoided? If so, how stringently?
- What about the sugar content in nutrition supplements?
- What are the pros and cons of probiotics?
QUESTIONS? COMMENTS?

- This is your time ….
REFERENCES

- Kohlschutter A and Eichler F. Childhood leukodystrophies: a clinical perspective. Expert Rev Neurother; 11(10); 1485-1496 (2011)