

NUTRITION FOR LBSL AND OTHER LEUKODYSTROPHIES

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TOPICS

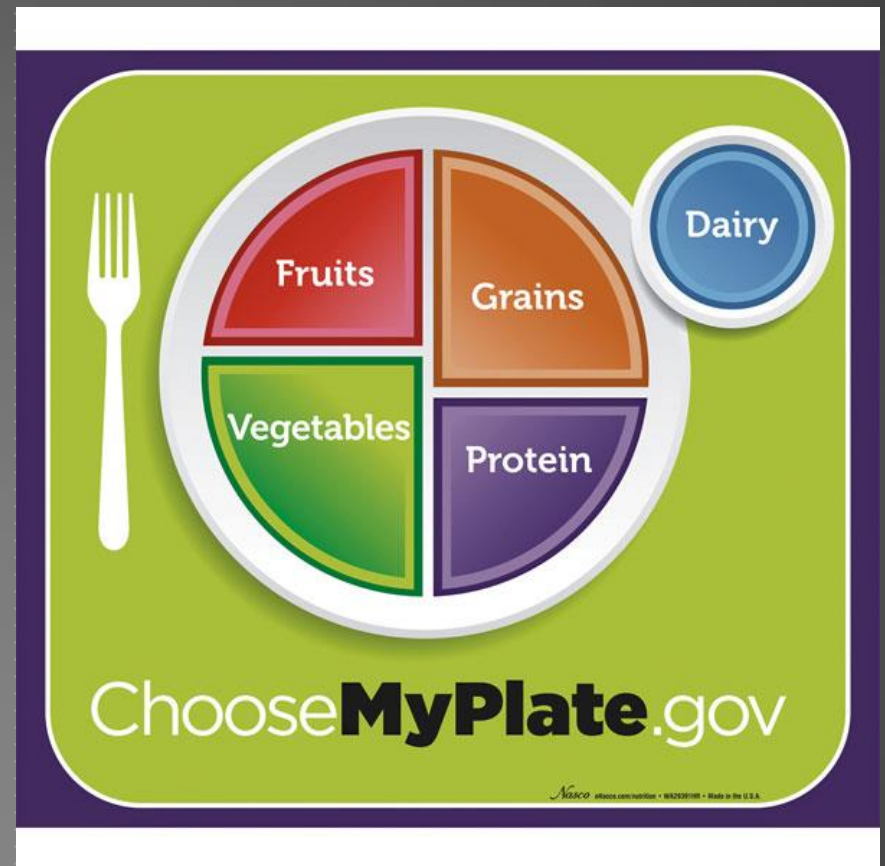
- ▶ General Principals for Nutrition Health
- ▶ Common Nutrition Concerns for Patients
 - ▶ Altered nutrition needs
 - ▶ Oral-motor difficulties
 - ▶ Swallow safety
 - ▶ GI dysfunction
 - ▶ Constipation
 - ▶ Picky eating
 - ▶ Specialized diets

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?
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QUESTIONS? COMMENTS?

- ▶ This is your time



REFERENCES

- ▶ Sumit P, Saneto R, et al. A modern approach to the treatment of mitochondrial disease. *Curr Treat Options Neurol*; 2009 November; 11(6):414-430
- ▶ Kohlschutter A and Eichler F. Childhood leukodystrophies: a clinical perspective. *Expert Rev Neurother*; 11(10); 1485-1496 (2011)
- ▶ Van der Knaap MS and Salomons GS. Leukoencephalopathy with brain stem and spinal cord involvement and lactate elevation. <https://www.ncbi.nlm.nih.gov/books/NBK43417>. accessed 4/3/18