



Beyond Leucine: Crafting a Balanced Life and Diet with MSUD

Erin MacLeod, PhD, RD

Director of Metabolic Nutrition - Rare Disease Institute

Assistant Professor of Pediatrics – George Washington University

Pronouns: she/her/hers

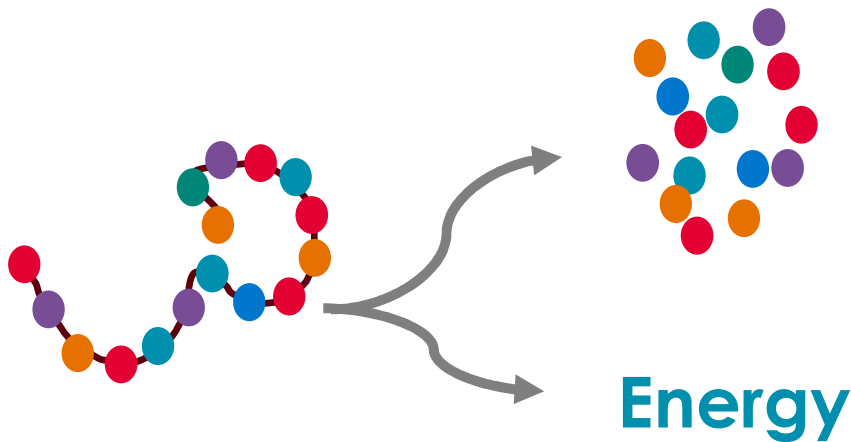
Objectives

- Review protein metabolism and concepts of MSUD diet management
- Discuss overall well-being
- Actions beyond good nutrition to thrive
- Tools for sustainability

Protein Metabolism



Catabolism- the breakdown of protein for energy



Catabolic processes

- Weight loss
- Energy deficit
- Illness

Anabolism: Changing our mindset



Protein requirements and intake for most Americans

Minimum protein needs differ from healthy dietary recommendations

Age	WHO g/kg	DRI g/kg	AMDR (% kcals)
Infants	1.1 – 1.7	1.0 – 1.5	N/A
1-3 years	0.8	0.9	5 - 20 %
4-8 years	0.7	0.8	10 - 30 %
9-13 years	0.7	0.8	10 - 30 %
14-18 years	0.7	0.7	10 - 30 %
Adults	0.7	0.7	10 - 35 %

Typical protein intake for Americans

2-3 year old : 3.5 g/kg

Elementary age: 1.75 – 2.5 g/kg

Teenager: 1 – 1.5 g/kg

Adult: 1.2 – 1.5 g/kg

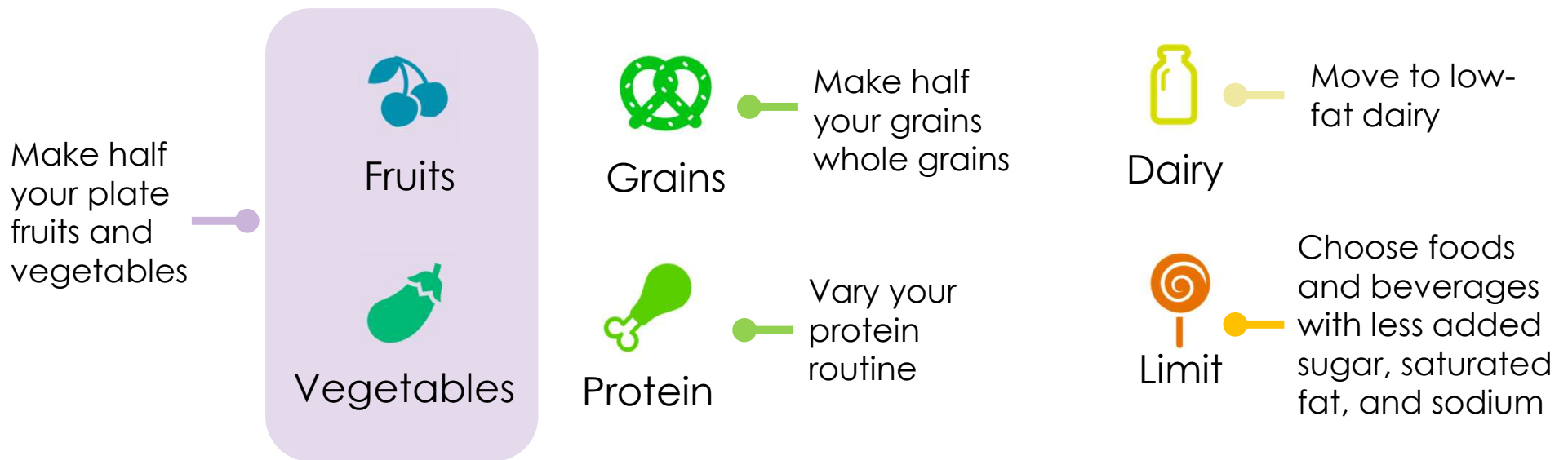
Protein requirements for individuals with MSUD

Age	Leucine Requirement (mg/kg)	Natural Protein (g/kg)	Leucine Free Protein (g/kg)	Total Protein (g/kg)
0-1 yr	40 – 100	0.5 - 1.2	2.0 – 3.0	2.5 – 3.5
1-3 yr	40 – 70	0.5 – 1.0	1.0 – 2.0	1.5 – 2.5
4-8 yr	35 – 65	0.5 – 0.8	0.8 – 1.5	1.3 – 2.0
9-13 yr	30 – 60	0.4 – 0.8	0.8 – 1.4	1.2 - 1.8
14-18 yr	15 – 50	0.2 – 0.6	1.0 – 1.5	1.2 – 1.8
> 19 yr	15 – 50	0.2 – 0.6	0.9 – 1.5	1.1 – 1.7

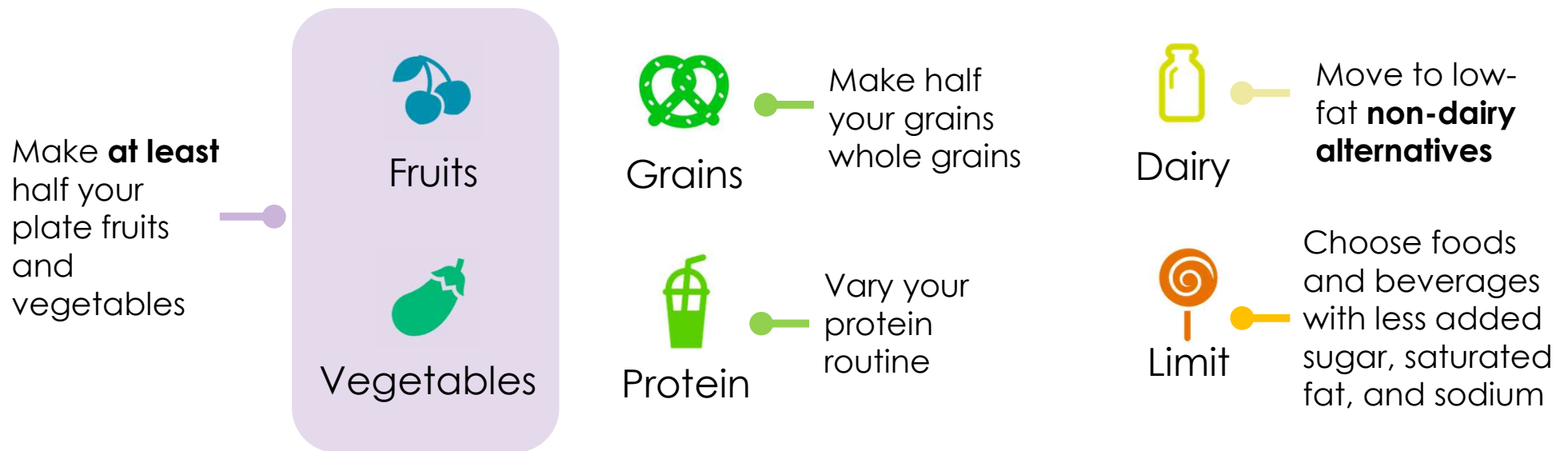
Supplementation of isoleucine and valine is also required.

Adapted From GMDI MSUD Nutrition Guidelines
<https://managementguidelines.net/guidelines.php>

MyPlate Guidelines for Healthy Americans



MyPlate Guidelines for Healthy Americans with MSUD



Make at least half your plate fruits and vegetables



Uncounted Fruits and Vegetables

- Most fruits and vegetables have a low amount of leucine per gram of food.
- Each clinic may have a different uncounted list of foods.

Vary your protein routine

- To build protein all amino acids must be available, at the same time.
- A complete protein has an appropriate proportion of all essential amino acids in once food source.
 - Animal proteins
 - Soy and quinoa
 - Complementary proteins
- Typical population recommendations: lean meats, fish twice a week, incorporate meatless meals into your routine with beans/lentils

Vary your protein routine – MSUD Style

- For MSUD Protein requirements are met from two primary sources
- Natural protein – Leucine containing foods
 - Dependent on individual tolerance
 - Must be enough to meet baseline leucine needs
 - Should be varied for improved nutritional quality – discuss with your dietitian
- Leucine free protein
 - Formula is the primary protein source
 - When consumed with a meal/snack consider it a complete protein

Food and formula should be spaced throughout the day



Vegetarian - beware



This Photo by Unknown Author is licensed under [CC BY-ND](https://creativecommons.org/licenses/by-nd/4.0/)



Thinking past food

Well-being is more than just staying out of the hospital



Physical Wellness



Hydration and
nutrition



Physical activity



Adequate sleep

Physical Activity

Activity Goals

- General public: Recommendations 30 minutes of moderate exercise 5 or more days per week or 20 minutes of vigorous exercise 3 days or more per week.
- Persons with disabilities: aerobic exercise 30-60 minutes 3-5 days per week at moderate intensity and resistance training 1-2 sets of 8 to 12 reps 2 to 3 times weekly
 - Moderate intensity: Getting to 64 – 76% MHR
 - Resistance: at least twice weekly

Outcome benefit

- Improved/stabilized metabolic control
- Increased energy/ functional fitness
- Improved protein/leucine tolerance

Ideas to increase physical activity

- Explore a gym memberships
- Take the stairs
- YouTube videos
- Ask for a prescription
- Smart Watch
- Pedometer (phone)
- Indirect calorimetry (formally, gym)
- Keep a record
- Set realistic goals
- Try not to get discouraged
- Understand limits and listen to your body
- Have Fun!

Some is better than none!

Timing and macronutrient required in relation to exercise

- Prior to exercise have energy provided through carbohydrates.
- After exercise have a high protein source.
 - Leucine free formula
 - If very active consider additional formula
- Try not to fast before exercise.



Example prescriptions: Plan must be appropriate for underlying physiology

MSUD: 78 kg deconditioned adult male

Goal: get conditioned

- NO fasting before, must have eaten meal at least 2 hrs before and immediately after
- 60-70% MHR for 3 sessions weekly for 10 minutes each x2 weeks
- 60-70% MHR for 3 sessions weekly for 15 minutes each x2 weeks
- 60-70% MHR for 3 sessions weekly for 20 minutes each for 2 weeks.
- 60-70%MHR for 3 sessions weekly for 25 minutes each for 2 weeks
- 60-70% MHR for 3 sessions weekly for 30 minutes (may increase to 4 and 5 sessions weekly after 2 weeks)

Being Mindful about Sleep

GOOD HABITS LEAD TO BETTER HEALTH OUTCOMES

- Better circulation
- Stronger immune system
- Improved focus

WHEN AND HOW MUCH IS IMPORTANT

- Sleeping at night vs daytime
- Children need 9-12 hours
- Teenagers need 8-10 hours
- Adults need 7-9 hours

DAYTIME HABITS WILL AFFECT SLEEP

- Get outside
- Eating and taking formula regularly throughout the day

DEVELOP A GOOD BEDTIME ROUTINE

- Turn off screens
- Chilled room
- Nightly gratitude

Making MSUD just a part of your life

Family Mealtime

- ❑ Set realistic expectations for everyone
- ❑ Enlist help and encourage involvement
- ❑ Avoid distractions
- ❑ Talk about something other than food
 - ❑ Keep conversation positive
- ❑ Model fine motor skills, social skills
- ❑ Associated with increased academic performance, higher self-esteem and sense of resilience
- ❑ Adolescents who eat with families are less likely to engage in early risky behavior.



How to avoid becoming a short order cook

- ❑ Aim to serve/consume mostly the same meal for everyone
 - ❑ Family brainstorm
 - ❑ Dinner King / Queen rotation
 - ❑ Plan theme days/weeks

- ❑ Consider your own preferences and opinions on food

- ❑ Make fruits and vegetables the backbone of a meal
 - ❑ Think of meat/high protein items as a side dish
 - ❑ It is often easier to add protein rather than remove it

Something for everyone

Pasta Night

- have meat as an add-in
- add hummus to sauce for a boost
- consider higher protein pasta for others

Grain Bowl Night

- Diversify main grain
- Use what's in the fridge
- Grain bowl “deconstructed”

Tex-Mex Night

- vegetable for side/filling
- diversify tortillas

Breakfast for dinner

- Waffles or pancakes
- DIY toasts
- yogurt with fruit

Pizza Night

- English muffin pizzas

DIY Ramen Bowls

- Same base vary fillings
 - pasta
 - shredded chicken
 - hard boiled egg

Utilize the freezer!

Sustainability is not perfection – MSUD is hard

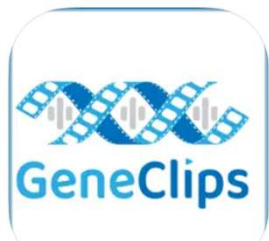
- ❑ Start the conversations early
 - ❑ Build confidence and self empowerment
 - ❑ Don't hide it
- ❑ Build Connections – Find your people
- ❑ Ask questions
- ❑ Validate the daily burden
- ❑ Don't fall to the guilt



Resources

Series of short videos on:

- Genetics / Inheritance patterns
- Macronutrient metabolism
- Genetic testing



MSUD and Non-MSUD specific



Thank you



Questions?

Please feel free to reach out
emacleod@childrensnational.org