MADD: Finding the Balance

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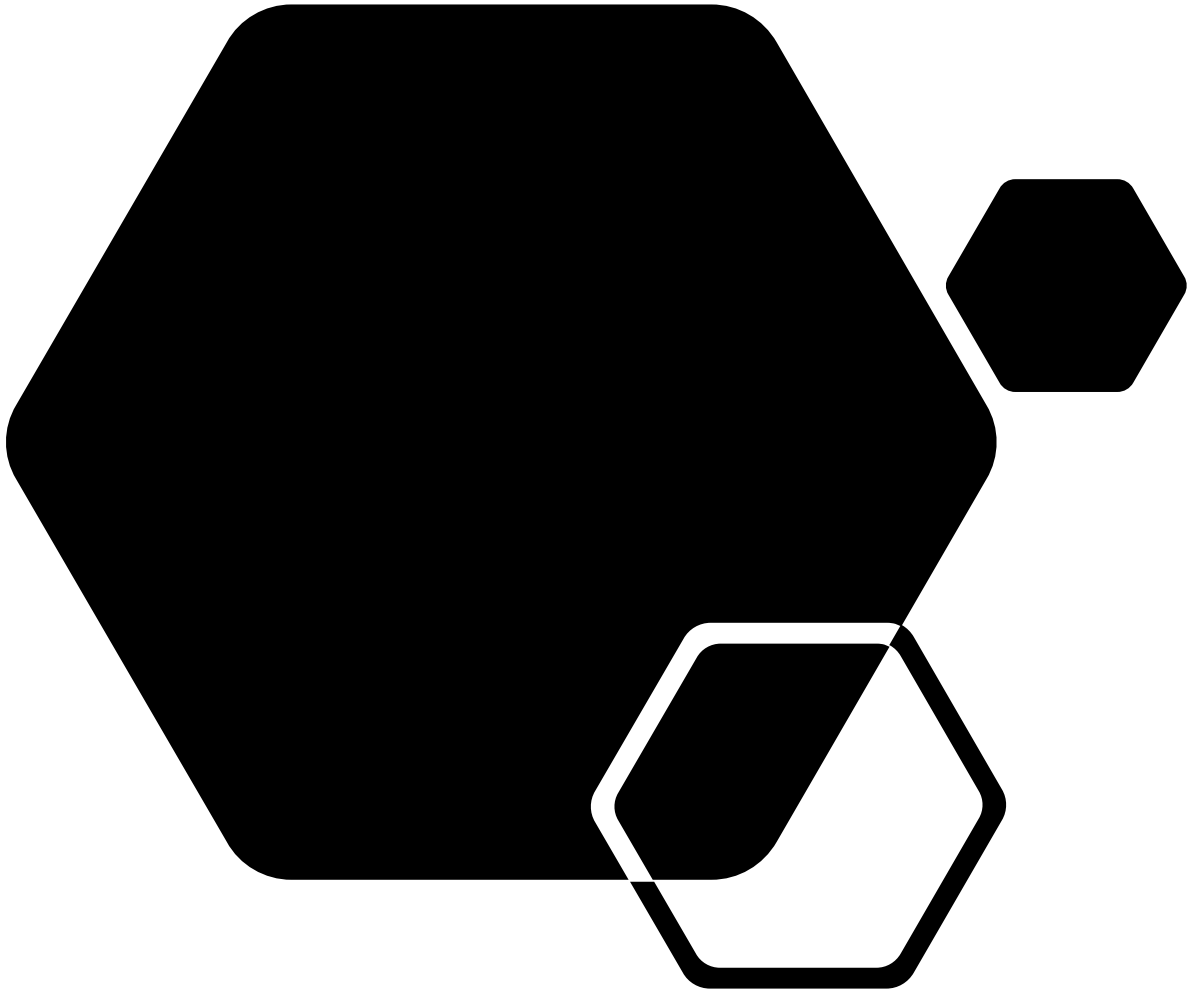


Objectives

- Describe the difference between MADD and other FAOD
- Provide overview of treatment options
- Discuss diet management of MADD through the ages
- Describe the major roles of the different macro- and micronutrients in the diet
- Brief overview of monitoring laboratories

Introduction

- The following presentation is intended for parents/caregivers of individuals with MADD or GAII.
- Specific recommendations for each person's diet are individual and based on their disease severity.
  - Always consult with your health care provider before making any changes to your diet or care.

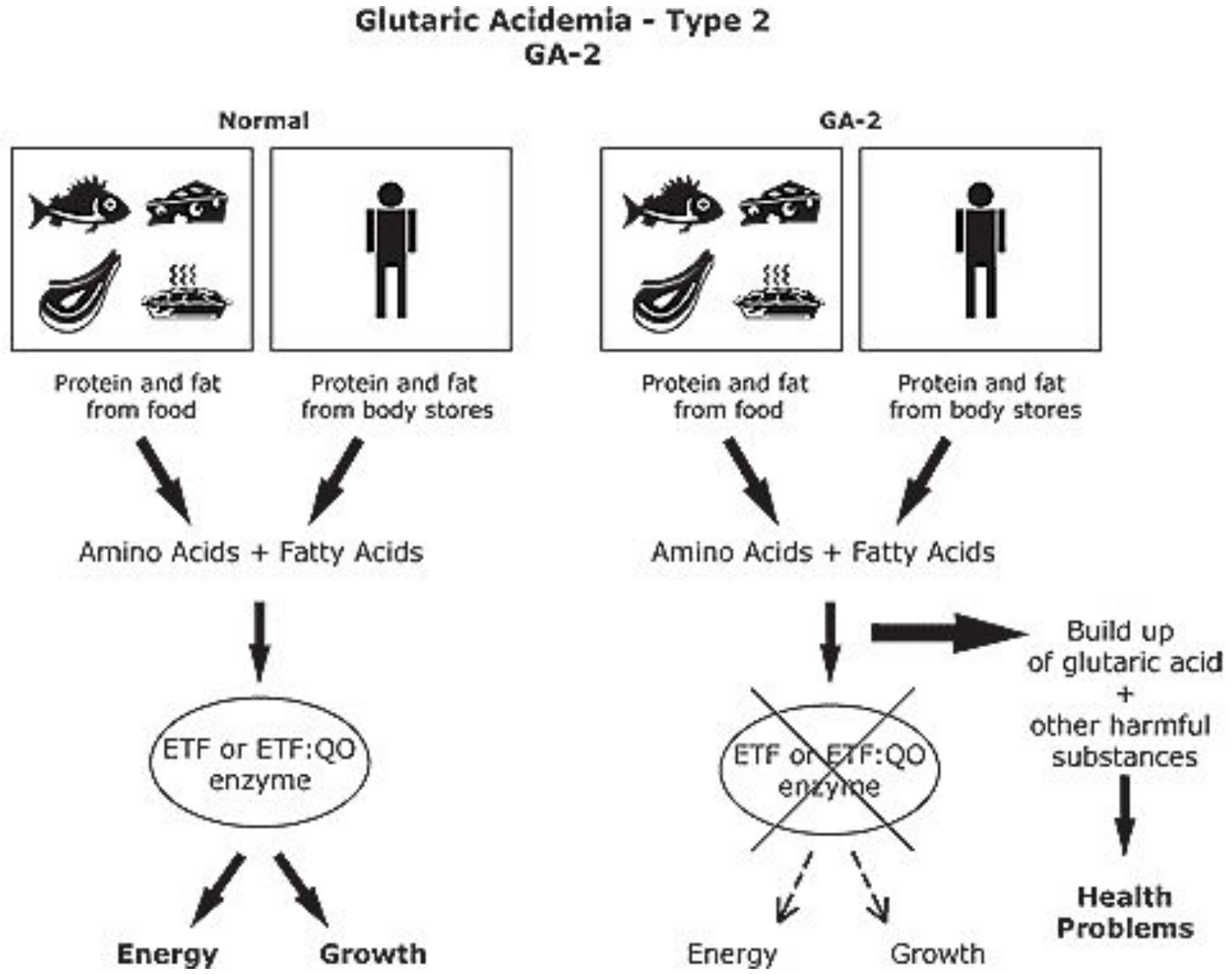
MADD Versus FAOD

MADD

- Energy metabolism disorder
- Must limit protein and fat intake
- More reliant on carbohydrates for energy

FAOD

- Other FAOD include:
  - VLCAD
  - LCHAD
  - CPT1
  - CPT2
    - Limit intake of long chain fats, all other macronutrients can be eaten in unlimited amounts
  - MCAD
    - Avoid concentrated sources of medium chain fats, other fats and macronutrients are not limited



https://www.newbornscreening.info/ga-2-glutaric-acidemia-type-2-2/

MADD Treatment Plan

Fasting Precautions

- Fasting times change based on age
  - Babies – typically starting at 4 hours, increase by 1 hour per month
  - Children, teens, adults - ~12 hours fasting is acceptable
- *will change in times of illness, check with your healthcare provider

Diet

- High in carbohydrates and low in protein and fat
  - Individual diet will vary based on age, severity and should be discussed with your healthcare provider

Riboflavin, Carnitine and Glycine Supplements

- Help with energy metabolism
- Use of ketones

Emergency Treatment

- Call your healthcare provider to discuss home care versus hospital care

Macro- and Micronutrients and MADD

Macronutrients

Main nutrients that make up the foods we eat

Provide energy and the building blocks for growth

Used in large amounts (macro = large)

The three macronutrients are:

Carbohydrates

Protein

Fat (lipids)

Each macronutrient has a unique role and all three are part of a balanced, nutrient-dense diet

Slide courtesy of Mary Sowa and Sandy Van Calcar

Carbohydrates

- Major role: main source of fuel for the body
- 4 calories per gram
- Three main types:
  - Sugars (simple carbohydrates): 1-2 sugar molecules, broken down quickly by the body
  - Starches (complex carbohydrates): long chains of sugar molecules, broken down slowly by the body
  - Fiber: a type of carbohydrate that the body cannot digest – fiber helps to regulate digestion and blood sugar, and promotes fullness and satiety

Slide courtesy of Mary Sowa and Sandy Van Calcar

Carbohydrates

Sources of simple carbohydrates

- Table sugar, soft drinks, candy, white bread/pasta, baked goods

Sources of complex carbohydrates

- Whole grains (bread, pasta, cereals), brown rice, oats, quinoa, sweet potatoes, beans/lentils, starchy vegetables

Sources of fiber

- Fruits, vegetables, whole grains, beans, lentils

Slide courtesy of Mary Sowa and Sandy Van Calcar

Carbohydrates and MADD

- Primary source of calories so should be the base of intake
  - Still contain fats and proteins so read labels!
- Complex carbohydrates contain more fiber and slow digestion
  - Resistant starches – what are they and are they good?
  - Whole grain versions will have more protein
- Simple carbohydrates can still be included, typically contain more fat
- Get creative with fruits and vegetables by cutting into fun shapes, roasting in an air fryer, adding sauces (Walden Farms)

Simplified Diet

Uncounted Fruits

- Apples – fresh and dry
- Apricots – fresh and dry
- Bananas
- Berries (all varieties)
- Cherries
- Cranberries – fresh and dry
- Dates
- Figs
- Grapefruit
- Grapes
- Guava
- Jackfruit
- Kiwi
- Lemons
- Limes
- Mango
- Melon (all varieties)
- Olives
- Oranges*
- Papaya
- Peaches
- Pears – dry and fresh
- Persimmon
- Pineapple
- Plantains
- Plums
- Pomegranates
- Prunes
- Raisins

Uncounted Vegetables

- Acorn Squash
- Beets
- Bok Choy
- Butternut Squash
- Cabbage
- Carrots
- Cauliflower
- Celery
- Chayote Squash
- Cucumber
- Green Beans
- Eggplant
- Jicama
- Leeks
- Lettuce
- Okra
- Onions
- Parsnips
- Peppers (all varieties)
- Pumpkin
- Radishes
- Rutabaga
- Sauerkraut
- Spaghetti Squash
- Summer Squash (zucchini and yellow)
- Tomatoes
- Turnips
- Yucca (Cassava Root)

Please remember your NO foods are still NO foods

For any questions on specific items, please contact your metabolic dietitian.

Developed by IMD Nutrition, Children's Hospital Colorado

Fats

- Major role: provides energy and essential fatty acids, and aids absorption of fat-soluble vitamins
  - MVI timing
- 9 calories per gram
- Classified as saturated, unsaturated (monounsaturated or polyunsaturated), or trans
- Most of the fat in our diet contains long-chain fatty acids (LCFAs)
- Type of fat does not matter in MADD

Slide courtesy of Mary Sowa and Sandy Van Calcar

Maximize Your Fat

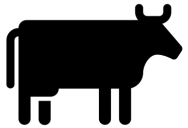
- Essential Fats
  - Must be consumed in the diet – cannot by synthesized
  - Good sources include: flax oil, walnut oil, soybean oil, vegetable oil, fatty fish (if allowed)
  - DHA supplement to maximize food fat
- Meal plan with allotted amounts of fat per meal, leave wiggle room
- Choose low fat carbohydrates and lean proteins to allow for added fats for cooking/flavoring
- Use low fat versions of foods to be able to incorporate more variety

Protein

- Major role: cell and tissue growth
- 4 calories per gram
- Proteins are composed of many different amino acids linked together
- Two types of amino acids:
  - Essential: cannot be made by the body and must be consumed through the diet
  - Non-essential: can be made by the body
- Adequate protein intake is important during periods of growth or increased demand (infancy, childhood, adolescence, pregnancy, breastfeeding)

Slide courtesy of Mary Sowa and Sandy Van Calcar

Low Fat Protein Sources



Animal-based: lean meats and fish, low-fat or fat-free dairy (milk, yogurt, cheese, cottage cheese)

Plant-based: beans, lentils, peas, powdered peanut butter, protein powder, lite tofu

Always consult with your health care provider before introducing any new foods

Slide courtesy of Mary Sowa and Sandy Van Calcar

Maximize Your Protein

1

Choose plant proteins over animal proteins

- Lower in fat and provide more bulk for amount of protein
- Jack fruit is a great low protein/fat meat alternative

- Essential amino acid supplements versus food protein

2

Use proteins as compliments to meals rather than main component

- Stir-fries, topping on salad, mixed with pasta and vegetables, ground turkey/beans with added vegetables for tacos

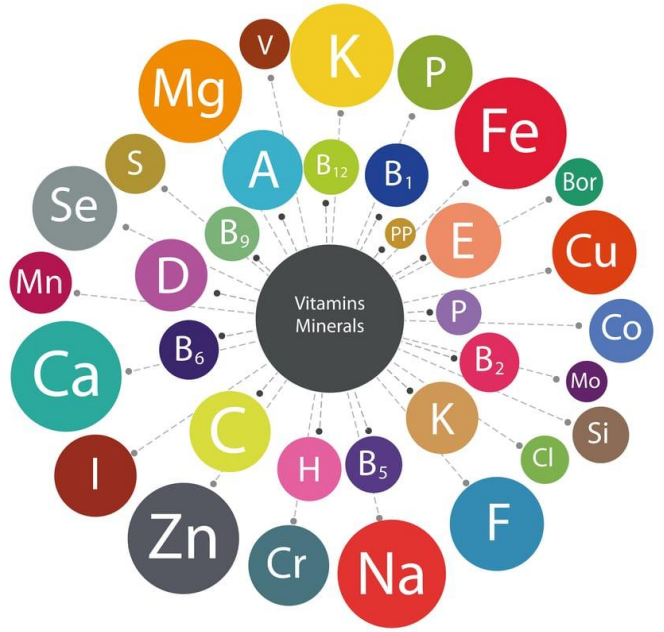
3

Iron needs – can be concerning with limited protein intake

- Use cast iron pans
- Incorporate dark leafy greens with source of Vit C (Ie: spinach salad with mandarin oranges)
- Supplements – slow-release options

Micronutrients

- Vitamins and minerals needed in small amounts (micro = small)
- Major roles: essential for production of enzymes, hormones, and other substances required for normal growth, development, and function
- Micronutrient deficiencies can cause visible and dangerous conditions, but may also cause more subtle/mild impacts on health
- Important to stay in close contact and provide accurate diet records to your healthcare provider to ensure adequate intake



Slide courtesy of Mary Sowa and Sandy Van Calcar

Other Thoughts…

- Focus on meal planning
- Snacks – low fat, gluten free
- Recipe modification – use your dietitian! That's what we are here for
  - Replacing a portion of the protein with vegetables to reduce protein and fat
- Transitioning from g-tube to oral intake
  - Swallow study – will help with tolerated texture and safety
  - Work with a feeding therapy specialist!
  - Offer foods prior to next feed

Foods to Change It Up

Meal Ideas

- Walden Farms sauces/dips
- Rice noodles
- Mung bean noodles
- Portabella mushroom burger
- Curry with lite coconut milk
- Different spices to add flavor
- Great creative with fruits and vegetables by cutting into fun shapes, roasting in an air fryer, adding sauces

Snack Ideas

- Air popped popcorn
- Dairy Free Go-Gurt
- GF bread with jelly
- GF cereal with rice milk
- Rice cakes
- Juicy Gels
- Cook and serve pudding with rice milk
- Fat free whipped topping on fruit
- Fruit snacks
- Pretzels (GF, crisps, etc)
- Apple chips

Laboratory Monitoring

- Quantitative serum amino acids – used to ensure adequate protein intake
  - Low – need additional protein
  - High – need to assess protein intake as well as energy intake
- Urine organic acids – used in diagnosis but not quantitative so not used for ongoing management
- Ketones – measured 90 minutes after consumption to ensure adequate dosing
- Other possible labs – comprehensive metabolic panel, complete blood count, prealbumin

References and Kudos

- https://www.newbornscreening.info/ga-2-glutaric-acidemia-type-2-2/
- Image: https://smpnutra.com/what-are-vitamins-and-minerals

- Special thanks to Mary Sowa, Sandy Van Calcar and Aaina Kochhar for their expertise on this topic

Questions?

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