

Summary – Nutrition for Mitochondrial Disease Patients

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Introduction The role of nutrition is vital in all patients with mitochondrial disease, regardless of the specifics of the diagnosis. There is not a lot of research about how nutrition affects those with Mito, and as a result much of today's discussion and suggestions are based on experience. The mitochondria are the powerhouse of our cells and allow the body to change food into energy. Any disruption in this energy metabolism process causes the symptoms we see in the group of disorders we call mitochondrial disease. Good nutrition goes hand in hand with the pharmaceutical-based treatments like the Mito cocktail.

Goals The goals of good nutrition for children with mitochondrial disease are to promote proper growth and development, energy production, and provide sufficient protein, vitamin and minerals. Adults require the same nutritional support, but obviously do not have to be concerned with growth & development as do children.

Protein The best way to find out if someone is getting enough nutrients is to ask them (or their parents) to write a 24-hour food recall. Patients tend to have a higher carbohydrate intake and lower protein intake than they need. Protein is stored in the muscles and is needed for strength. Protein will also moderate your blood sugar in concert with carbohydrates. Good sources of protein include:

- Eggs
- Milk
- Soy
- Yogurt
- Cheese
- Beans/legumes
- Nuts/peanut butter
- Meat
- Fish

For every mitochondrial disease patient, every meal (and snack) should contain protein AND a complex carbohydrate. This small step is very important for allowing the carbohydrate to be properly and adequately metabolized into energy.

Another concern is the amount of calories taken in; too many or too few can be a problem. Mito patients often take in too few although patients who use a central line for hydration may be getting too many "empty" calories from IV solutions that contain sugar (i.e., D5W, which is dextrose, sugar and water). Sometimes changing to or alternating with a saline solution (no sugar added) may help reduce the "empty" calories that can contribute to weight gain. Of course, any of these concerns should be addressed with your or your child's physician.

Calcium & Vitamin D Calcium and Vitamin D are often found in dairy-based protein choices, such as milk, yogurt and cheese, but can also be found in broccoli and almonds. Three to four servings of dairy-based protein a day is a great way to ensure

that you or your child is getting Vitamin D and calcium in the daily diet. While we used to believe that milk provided enough of the recommended daily allowance of calcium and Vitamin D, today's recommendations are higher. The amount needed may vary according to a person's age, but adding foods like cheeses and tuna fish will help boost your intake. Keep in mind also that calcium is NOT absorbed unless you have enough Vitamin D, and Vitamin D is not activated unless you have a minimum of 10 minutes exposure to sunlight a day. Supplements of both Vitamin D and calcium are often prescribed by physicians for their mitochondrial disease patients, and can also be found in a good multi-vitamin if tolerated.

Despite knowing and understanding the recommendations, sometimes there are challenging symptoms facing Mito patients that interfere with obtaining good nutrition, such as fatigue, muscle weakness, dysmotility, dysphagia (inability to swallow), nausea and vomiting, ataxia, and reflux.

General Guidelines

- 4-6 small meals a day
- include complex carbohydrate and protein at each meal
- maintain a healthy weight
- eliminate empty calories
- avoid fasting (including avoiding liquids for many hours)
- have a bedtime snack that includes both a complex carbohydrate and a protein

These guidelines may be difficult to follow but should be attempted. Bedtime snacks that are high in sugar or starches should be avoided, some folks find that corn starch added to pudding works well. Since for some people with Mito, especially children, it is difficult to take in enough calories each day. For infants and children special formulas may be used at each meal/feeding to add protein. Again, have a dietician review your 24-hour recall to determine if and what kind of protein supplement you (or your child) might require. Even nutritionists or dieticians not familiar with mitochondrial disease can help you with this.

Supplements of multivitamins and minerals may also be needed depending on your intake. Those with enteral feedings (J tubes or G tubes) can receive extra nutrition via the tube feedings. Those receiving TPN (Intravenous lines) may also receive added nutrition thru their lines. However, the best way to receive nutrition is through the gastrointestinal tract whenever possible.

Low Energy Low energy is a frequent issue for Mito patients. The first step to avoid nutrition pitfalls on "bad days" is to be prepared! Keep healthy, high protein meals frozen and ready to pull out when you need them. That way when you are having a "bad day," you will have high-energy food available. Always combine protein with your complex carbohydrate for a good meal/snack. Keep supplements on hand as well like Carnation Instant and/or Ensure. Before any taxing activity, take a snack and bring one with you. Some people like to use a blender for fruits and veggies, which reduces the fatigue of eating. Don't be afraid to use one! This makes the foods easier to take and

uses less energy to digest - conserving energy and getting the best "bang for your buck" when eating is fundamentally important!

QUESTIONS

How much hydration is needed for Mito patients?

This is difficult because it is individual. Typical adult recommendations of 64 oz per day is probably not enough for most people with mitochondrial disease. The best way to tell is to look at urine output, osmolarity, and then Blood Pressure. In reality, this will all depend on the individual's autonomic nervous system. Some Mito patients have very high hydration needs, while others might be more "normal."

Hydration is a great concern for Mito patients, however, and more often than not, people are under hydrated rather than over hydrated. Use the color of your urine as a guide to how concentrated it is; the darker the urine, the more concentrated it is, and this means you need more fluids. (Unfortunately, some of the supplements used in the Mito cocktail may discolor your urine, so this method may not always work.)

Determine what volume is your goal for fluids each day as a minimum, and then make it a priority to take in what you need. Some parents and adult patients find that filling a container at the beginning of the day helps to simplify the guesswork and keeps them on track to finish the container of fluids by the end of the day. (Drinking slowly throughout the day is better than taking in huge gulps). Remember that anything that melts at room temperature is considered fluid - like popcicles and ice cream. As simple as it seems, hydration is vital to the health of the cell, and so important for anyone with Mito!

Why is protein intake so important?

Carbohydrates are more appealing and easier to eat, especially for kids. However, if you overload the body with carbs it can have a negative effect on the mitochondria which help break down glucose into ATP for cellular function. Protein helps to prevent or minimize the glucose "spikes" which are not helpful in this process. Patients often report "crashes" after a high carbohydrate meal. For children often just one(1) ounce of protein, like 2 tablespoons of peanut butter or one (1) egg is all that is needed to balance their snack or meal.

Children with a G-Tube often have little or no appetite - what can I do?

This is a common issue for children with Mito - especially those with decreased gastric motility. It takes so much longer for food to go through their GI tract, specifically their stomach, so they are never hungry - they always feel "full". Sometimes the timing of g-tube feedings helps. For example, giving the feeding at night might allow the child to wake up full but have an increased appetite as the day goes on. Little feedings throughout the day may also help, savoring every bite, and taking time on each and every bite of food.

Someone with Mitochondrial myopathy who is generally on a high protein, low sugar

diet finds that when she needs an IV, switching to dextrose (sugar) and water rather than saline makes her feel better - why is this?

It may be that the IV Dextrose is a "quick fix" - that is your body can metabolize this very quickly with very little energy expenditure. Carbohydrates by mouth have to be broken down. Remember, the response is so individual!

What do you suggest for adults with motility issues?

Some foods are very difficult to digest, especially proteins like meat. Increased fat in foods can also slow down motility so you should avoid protein foods that are also high in fat. Some believe that after a certain time in the day, they need to take in only softer foods or even just liquids. Keeping a journal to see what works best for you is also a good idea. It is important to work with your physician and nutritionist on all these issues. Obviously, fruit, vegetables and fluids are always the first course of action plus decreasing fat and increasing carbohydrates to those with whole grains and lots of fiber. If this doesn't work, then medication may be needed, but work with your PCP on this.

Dysautonomia in mitochondrial disease comes from the autonomic system not working - signals not being sent or received. Continue to use good judgment about eating and fluids. Consider reading the summary of Dr. Flores' talk called "Tummy Troubles." (see link in comments below).

Is my 2 year-old getting enough fluids?

In one example, a child with mitochondrial disease is taking in 2 cans of Pedialyte a day, also has a fatty acid problem and some other delays. It is difficult to get her to eat anything other than some crackers or potatoes (child uses bottle only - no sippy cups). To begin to tackle this common problem, first take a 24-hour recall and seek the advice of a dietician or nutritionist to calculate the dietary intake for calories, ounces of fluid, protein, etc. Allergies are also a common concern and should be systematically ruled out using an elimination diet approach as recommended by the child's pediatrician. Children like this can benefit from more fluids throughout the day, which could be tried in different form (juices, soups, popsicles, water). Gagging or the inability to swallow or chew can be addressed by **speech therapists**. Parents should seek guidance from speech pathologists who specialize in oral-motor-feeding therapy (this could also be a great resource for adult patients if this is an issue.) SpEech therapists are found in schools, in the community (ask for early intervention support from your doctor) and in rehabilitation hospitals or centers, and their services are often covered by insurance. This can be a helpful resource for Mito patients.

Other Recommendations

Glucerna, a supplement used by diabetics, may be useful to Mito patients because it helps boost energy but has a lower carb content than other liquid supplements. Don't get discouraged! Meeting your needs for good nutrition high in protein and with plenty of fluids can be a challenge for anyone with mitochondrial disease. Take small steps, small bites, drink plenty of fluids and keep a food journal in order to get started.

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