

MitoNutrition: Dietary Management for Adults Living with Mitochondrial Disease

Understanding Mitochondrial Disease

Mitochondrial disease (mito) is an umbrella term used to describe over 350 different mito disorders that are most often caused by a genetic mutation(s) in the mitochondrial or nuclear DNA. These genetic changes affect the mitochondria's ability to produce enough energy, either because an individual doesn't have enough healthy mitochondria in their cells or the mitochondria in certain cells have defects in the proteins or pathways needed for the mitochondria to produce energy, this can greatly affect how well organs like the brain, muscles, lungs, eyes or body systems (e.g., digestive system) work.

Because of the genetic nature and complexity of mitochondrial disorders, tailored approaches to treatment and care are essential. Alongside medical management, nutritional strategies can play an important role in supporting energy production, reducing the severity of symptoms, and improving quality of life for adults living with mito.

This MitoGuide is designed to provide you with practical, evidence-informed nutrition knowledge to help you better understand how food, hydration, and supplements can support mitochondrial health.

The Role of Nutrition in Mitochondrial Disease

Nutrition refers to the process of taking in food and fluids to provide the body with energy, support growth, and repair and maintain overall health. Nutrition provides the raw materials mitochondria need to produce energy.

When we eat **carbohydrates**, they are broken down by digestive enzymes into **glucose**, **fats** are broken down by bile from the liver and digestive enzymes into **fatty acids**, and the **protein** we eat are broken down by digestive enzymes into **amino acids**.

Glucose, fatty acids and amino acids (smaller building blocks) are absorbed into the bloodstream and then taken up by muscle, brain and nerve, heart, and liver cells among others.

Inside the cells, mitochondria act as energy factories, using these fuel sources to produce ATP (adenosine triphosphate), the body's main source of energy.



Energy problems can arise for a number of reasons in mitochondrial disease. Some disorders affect different energy-producing pathways, depending on the underlying genetic cause. Some individuals have difficulty using carbohydrates efficiently, others may struggle to use fats for energy, and some may be affected by multiple pathways. Understanding which energy pathways are impacted can help explain why certain nutrition strategies are recommended for specific mitochondrial disorders.

Key Energy-Producing Pathways

Carbohydrate-Based Energy Pathways

What happens: Glucose is processed and ultimately used by mitochondria to create ATP

When this pathway is impaired:

The body has difficulty using glucose efficiently for energy, which can lead to energy drops and make the body more reliant on other energy pathways to produce energy.

HOW ENERGY FROM CARBOHYDRATE IS CREATED

Step	Energy Pathway	What Happens
1	Digestion	Digestive enzymes break carbohydrates down into glucose 
2	Glycolysis	Breaks glucose down into smaller molecules (mainly pyruvate) 
3	Citric Acid Cycle (or Krebs)	Pyruvate is broken down into fuel, releasing electrons 
4	Electron Transport Chain (ETC)	Electrons are carried into the ETC and move through four complexes (or transfer stations), to eventually create a gradient (gradient is like a warehouse) 
5	Oxidative Phosphorylation	Once the gradient (warehouse) starts to fill, it releases to generate ATP. This is the final stage in the energy producing (ATP) process. 

Fat-Based Energy Pathways

What happens: Fatty acids are transported into mitochondria and broken down to create ATP

When this pathway is impaired:

- There's difficulty using fatty acids as a fuel source, which increases reliance on carbohydrates as a backup. During times illness, fasting or missed meals, the body is more likely to run out of usable energy.

HOW ENERGY FROM FAT IS CREATED

Step	Energy Pathway	What Happens
1	Digestion	Bile and digestive enzymes break fat down into fatty acid, which are absorbed into the bloodstream, taken up into cells, then into the mitochondria 
2	Fatty Acid Oxidation	Inside the mitochondria, fatty acids are broken down into acetyl-CoA 
3	Citric Acid Cycle (or Krebs)	Acetyl-CoA that moves through the citric acid cycle, releasing energy-rich electrons 
4	Electron Transport Chain (ETC)	Electrons are carried into the ETC and move through four complexes (or transfer stations), to eventually create a gradient (gradient is like a warehouse) 
5	Oxidative Phosphorylation	Once the gradient (warehouse) starts to fill, it releases to generate ATP. This is the final stage in the energy producing (ATP) process. 

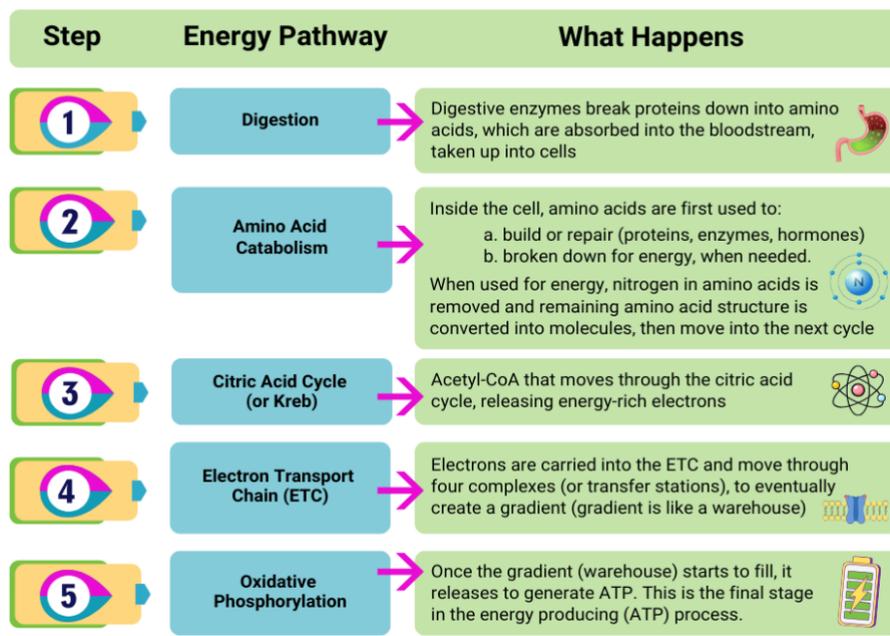
Protein-Based Energy Pathways

What happens: Some amino acids can be used to support energy production, when needed

When this pathway is impaired:

The body has more difficulty using amino acids for backup energy, which can increase fatigue and place extra strain or workload on other energy systems to compensate.

HOW ENERGY FROM PROTEIN IS CREATED



* Protein (amino acid) is **not** the body's preferred fuel source because it's more complex to breakdown, creating more metabolic stress or making cells in the body work harder to create needed energy.

In mitochondrial disease, any of the above energy-producing pathways may be inefficient or impaired. As a result, individuals may experience fatigue, muscle weakness, exercise intolerance, gastrointestinal symptoms, and difficulties maintaining weight or energy balance.

Thoughtful nutrition strategies can help:

- Support more stable energy production
- Reduce metabolic stress
- Prevent nutrient deficiencies
- Manage symptoms such as fatigue, nausea, and muscle pain

While nutrition is **not a cure** for mitochondrial disease, it can be a key component of comprehensive care.

Building a Well-Balanced, Nutrient-Dense Diet

A well-balanced diet provides a steady supply of fuel and essential nutrients that cells rely on to produce energy and maintain cellular function. In mitochondrial disease, where energy-producing pathways may be less efficient, consistent access to high-quality nutrition becomes especially important.

Rather than relying on one primary fuel source, a balanced approach supports multiple energy pathways and helps reduce metabolic stress on the body. Nutrient-dense foods, those that provide vitamins, minerals and energy without excessive calories, can help support energy levels, muscle function, and overall resilience.

Key principles to build a balanced diet:

1. Regular meals and snacks

Eating at consistent intervals helps maintain stable energy availability and may reduce fatigue, dizziness or symptom flare-ups associated with low energy stores.

2. Complex carbohydrates

These are foods that digest more slowly, providing a steady release of glucose, and longer-lasting energy, which many cells rely on for energy. They also help avoid rapid spikes and drops in sugar.

EXAMPLES

Whole Grains	Vegetables	Legumes	Fruits
<ul style="list-style-type: none">• Brown rice• Barley and Farro• Whole-grain bread and pasta• Steel-cut and rolled oats• Quinoa 	<ul style="list-style-type: none">• Sweet potatoes• Potatoes (with skin)• Squash• Corn• Peas 	<ul style="list-style-type: none">• Lentils• Chickpeas• Black beans• Kidney beans• Split peas 	<ul style="list-style-type: none">• Apples• Berries• Pears• Oranges• Bananas 

3. High-quality proteins

Provide essential amino acids that supports muscle maintenance, tissue repair and immune function. Adequate protein intake is important for preserving strength, especially for individuals with muscle involvement.

EXAMPLES

Animal-based proteins

- Fish (salmon, trout, sardines)
- Poultry (chicken, turkey)
- Eggs
- Lean meats (beef, pork)

Plant-based proteins

- Legumes (lentils, chickpeas, black beans)
- Soy food (tofu, tempeh, edamame)
- Nuts and seeds (almonds, walnuts, pumpkin, chia)
- Nut and seed butters

4. Healthy fats

Fats provide a concentrated source of energy and support the absorption of fat-soluble vitamins. The ability to use fat for energy varies by mitochondrial diagnosis, so individual guidance from a healthcare provider is important.

EXAMPLES

Fatty fish

- Salmon
- Sardines
- Trout
- Mackerel



Plant-based proteins

- Nuts (almonds, walnuts, pecans)
- Nut and seed butters
- Avocado & avocado oil
- Olive oil



Other sources

- Olives
- Full-fat dairy (yogurt, cheese)
- Eggs



Not all individuals with mitochondrial disease can use fats efficiently and recommendations may differ depending on diagnosis and symptoms.

Because the body relies on multiple fuels and interconnected energy-producing pathways to generate ATP, maintaining a balanced intake of carbohydrates, proteins and fats is especially important in mitochondrial disease. When one or more of these pathways is less efficient, the body benefits from having flexible access to different sources.

For this reason, **highly restrictive diets are generally discouraged unless they are medically indicated and supervised by a healthcare professional**. Limiting entire food groups may increase the risk of nutrient deficiencies and can place additional strain on already challenged energy systems, potentially worsening fatigue and other symptoms.

Hydration for Energy Support

Adequate hydration is an essential component of nutrition and plays a critical role in how the body produces and uses energy. Water is necessary for circulation, digestion, temperature regulation, and cellular metabolism, all processes that are already under increased strain in mitochondrial disease.

At the cellular level, hydration supports the movement of nutrients, electrolytes, and metabolic by-products in and out of the cells. When fluid levels are low, these processes become less efficient, which can further challenge energy production and worsen symptoms.

For individuals living with mito, even mild dehydration may contribute to increased fatigue, dizziness, headaches, constipation, muscle cramps and difficulty regulating body temperature, symptoms commonly reported across many mitochondrial conditions. Dehydration can also place additional stress on the cardiovascular and nervous systems, which may already be affected.

Practical hydration tips include:

- **Sip fluids** regularly throughout the day, rather than drinking large amounts at once, to support steady absorption and avoid sudden fluid shifts
- **Include water-rich foods** such as soups, fruits, and vegetables, which can contribute to daily fluid intake
- **Consider electrolyte-containing fluids**, if recommended by a healthcare provider, especially during illness, hot weather, or periods of increased physical stress.

Individual fluid needs vary. Individuals with cardiac involvement, kidney disease, gastrointestinal symptoms or autonomic dysfunction (e.g., dizziness, temperature regulation, feeling faint, headaches) may require more personalized hydration strategies. Always speak with a healthcare provider before making significant changes to fluid or electrolyte intake.

Strategies to Enhance Energy Intake

Meeting energy needs can be challenging for some individuals living with mito. Reduced appetite, feeling full quickly (early satiety), nausea, gastrointestinal discomfort, or fatigue related to meal or food preparation can all make it difficult to eat enough to meet the body's energy demands. Because energy-producing pathways may be less efficient in mito, even small shortfalls in intake can contribute to increased fatigue or symptom flare-ups over time.

The goal of nutrition strategies is to support steady, realistic energy intake while minimizing additional strain on the body.



Helpful strategies may include:

- Eating **small, frequent meals** rather than large portions, which may feel more manageable and help maintain consistent availability throughout the day.
- Choosing **energy-dense foods** (such as nut butters, avocado, olive oil or full-fat dairy, if tolerated) to provide more calories and nutrients in smaller volumes.
- Adding **healthy fats** to meals to increase energy intake without significantly increasing portion size
- Using **smoothies, soups or oral nutrition supplements** when solid foods are difficult to tolerate or appetite is low

Because nutritional needs and tolerances can vary widely depending on the specific mitochondrial diagnosis, symptoms, and overall health, working with a healthcare provider or dietitian familiar with mitochondrial disease can help tailor these strategies. Personalized guidance can be especially important during periods of illness, stress or unintentional weight loss, when energy needs may change.

Managing Food Triggers

Food-related symptom flare-ups have been a reported experience for some individuals living with mito. Because mito can affect multiple organ systems, including the gastrointestinal tract, muscles, nervous system and autonomic nervous system, responses to food, meal timing and portion size can vary widely from person to person.

Research and clinical care guidelines increasingly recognize that metabolic stress, including prolonged fasting, large energy demands from digestion or rapid shifts in blood sugar, may worsen symptoms in individuals with impaired mitochondrial energy production. Patient reports also highlight that certain foods or eating patterns can contribute to fatigue, gastrointestinal discomfort, headaches, muscle pain or symptom flares.

Commonly reported triggers may include:

- **Skipping meals or prolonged fasting**, which can increase metabolic stress and reduce available energy, particularly important for individuals who rely heavily on regular carbohydrate intake
- **Large, heavy meals**, which may be more difficult to digest and can temporarily increase fatigue or gastrointestinal symptoms
- **Highly processed foods**, which may be lower in essential nutrients and harder for some individuals to tolerate
- **Excessive caffeine or alcohol**, which can affect hydration, sleep, heart rate and nervous system regulation, potentially worsening symptoms in some people.

It's important to note that these are not universal triggers. Foods or patterns that cause symptoms for one individual may be well tolerated by another.



Identifying food and symptom triggers

Keeping a food-and-symptom journal can be a helpful, low-pressure way to notice patterns over time. Recording what is eaten, when meals occur, portion size, and how symptoms change afterward may help individuals and their healthcare providers identify potential triggers and make thoughtful adjustments.

When symptom flare-ups are frequent, severe, or associated with weight loss, dehydration, or declining function, it's important to discuss these changes with a healthcare provider. Dietitians and mitochondrial specialists can help distinguish food-related triggers from other causes and support safe, individualized nutrition strategies.

Mealtime Approaches to Manage Energy Levels

Energy conservation is an important part of daily living with mito. Because energy production may be limited, everyday activities such as planning, preparing and eating meals can contribute to fatigue.

Mealtime strategies that reduce physical and cognitive effort can help preserve energy for other essential activities while still supporting consistent nutrition. Small adjustments in how and when meals are prepared and eaten make a meaningful difference.

Helpful mealtime approaches include:

1. Planning meals in advance

Deciding what to eat ahead of time can reduce decision fatigue and help avoid missed meals on lower-energy days. Simple meal plans or rotating familiar meals can support consistency without added stress

2. Sitting while preparing food

Sitting during food prep helps conserve energy, reduces muscle strain, and support safety, especially for individuals with muscle weakness, balance challenges or fatigue.

3. Using convenience options, when needed

Pre-cut produce, frozen meals, canned food, slow cooker and delivery options can be helpful tools, not shortcuts. These options can help maintain nutrition on days when energy is limited.

4. Timing larger meals during periods of higher energy

Eating more substantial meals at times of day when energy is typically better may help reduce fatigue and support digestion. Lighter meals or snacks may feel more manageable during lower-energy periods.

Care partners play a valuable role in supporting consistent nutrition, particularly during periods of illness, increased fatigue, or symptom flare-ups. Assistance with meal preparation, reminders to eat or drink, and flexibility around mealtime expectations can help reduce pressure and support overall well-being.



The Mito Cocktail: Nutritional Supplements for Mitochondrial Disease

Some mitochondrial specialists may recommend a combination of vitamins and mitochondrial cofactors (nutrients that help enzymes do their jobs), often referred to as the *mito cocktail*. The goal of a mito cocktail is supportive, to help optimize mitochondrial function, reduce metabolic stress, and improve symptom management.

It's important to know that there is no single standard mito cocktail. Supplement choices (if recommended at all) depend on an individual's diagnosis, symptoms, age, medications and lab findings. Clinical experts emphasize that while strong randomized trial evidence is limited for many supplements, supplements are often used in practice because some have biologic rationale, relatively favourable safety profiles when monitored, and may be particularly relevant in specific mito conditions.

Why some specialists prescribe a mito cocktail

Specialists may consider supplements because they can help:

- Support energy production pathways (including oxidative phosphorylation)
- Provide cofactors needed for mitochondrial enzymes
- Help manage secondary effects of mitochondrial dysfunction (like oxidative stress)
- Address suspected or documented nutrient deficiencies

Why Diagnosis Matters for Supplements

In some mitochondrial disorders, specific supplements are more commonly considered because of the underlying biology. For example, the Mitochondrial Medicine Society care standards note that riboflavin may be considered in ACAD9-related myopathy and that a combination of CoQ10 and riboflavin may be considered for ETFDH gene-related myopathy.

Commonly used mito cocktail supplements

These are often discussed during mito clinic appointments and in patient resources, but not everyone needs them, and dosing/selection should be individualized.

⇒ Coenzyme Q10 (CoQ10)

A naturally occurring compound that plays a key role in mitochondrial energy production as an electron carrier in the respiratory chain.

⇒ L-carnitine

Plays a key role in transporting fatty acids into mitochondria for energy production.

⇒ Riboflavin (Vitamin B2)

Supports energy metabolism by acting as a cofactor for many energy-related enzymes and helps activate other B vitamins.

⇒ Thiamine (Vitamin B1)

Supports carbohydrate metabolism and energy-related enzyme function.

⇒ Niacin (Vitamin B3)

Involved in cellular energy production and supports skin, digestive, and nervous system health.



Note on safety and online supplement advice

There's a fair amount of supplement information online that can sound very convincing. But with mitochondrial disease, the right supplement plan depends on the type of diagnosis and the right clinical context. Supplements can interact with medications, affect lab test results and, depending on the individual, may cause side effects.

Before starting, stopping or changing any supplement, it's strongly recommended that you discuss it with the specialist overseeing your mitochondrial disease care.

Fasting and Mito

Fasting can significantly change how the body fuels itself. When you go without food for longer periods, the body shifts away from using incoming carbohydrates and begins relying more on stored fuels (including fats, protein/muscle). For individuals living with mito, where energy production may already be less efficient, this shift can add metabolic stress and may worsen symptoms for some individuals.

Clinical care standards for mitochondrial disease specifically emphasize preventing catabolism (a state where the body breaks down stored energy and tissue) and recommend avoiding prolonged fasting, particularly around illness, procedures or other stressors.

Important considerations:

- **Fasting is not appropriate for everyone with mito.** The safety of fasting depends on the individual diagnosis, symptoms, medications, nutrition status and risk for low blood pressure (hypoglycemia).
- **Fatty acid oxidation disorders (FAOD):** Fasting is often unsafe. Individuals diagnosed with an FAOD are often advised **to avoid fasting** because they may not be able to use fat efficiently for energy, which increases the risk of metabolic decompensation/crisis (when the body's energy demands are higher than what the mitochondria can produce)
- **Prolonged fasting may worsen symptoms.** Longer periods without food can increase the risk of fatigue, hypoglycemia, dizziness, and muscle breakdown, particularly when energy reserves are already limited
- **Illness and procedures require extra caution.** Expert guidance often focuses on preventing fasting-related stress during surgery/medical procedures and during sudden illness (flu, infection, cold).

Because mitochondrial disorders are highly variable, always consult the specialist overseeing your mitochondrial disease care before trying fasting, intermittent fasting or any restrictive eating approach. The "right" strategy must match the diagnosis and the individual, not what is trending online.



Nutrition and Specific Mito Disorders

As mitochondrial disease represents over 350 different diagnoses, it makes sense that nutrition can play different roles depending on the underlying condition. In some disorders, nutrition strategies are focused on preventing metabolic crisis; in others, the goal may be to reduce catabolic stress, support muscle health, or complement diagnosis-specific therapies. Broad clinical care standards emphasize individualized planning and avoiding metabolic stressors (like prolonged fasting), with diagnosis-specific guidance helping further shape nutrition approaches.

Fatty Acid Oxidation Disorders (FAOD)

For FAOD, nutrition is often a central part of disease management. Because the body may not be able to use fats effectively for energy, maintaining a reliable fuel supply (often emphasizing carbohydrates) and avoiding fasting is critical.

Clinical and guideline literature repeatedly highlights:

- Preventing fasting and providing adequate, uninterrupted caloric intake to reduce the risk of metabolic crisis.
- Many care plans include a higher carbohydrate approach and, for some long-chain FAODs, individualized fat management and/or specialized fats (e.g., MCT or triheptanoin) under specialist direction.

MELAS

MELAS can involve high metabolic demand and multi-system symptoms (including GI and nutrition challenges), so nutrition strategies often emphasize maintaining energy balance and preventing **catabolic stress** during illness or poor intake.

Targeted supplementation is sometimes discussed in MELAS, most notably **arginine and/or citrulline** in relation to stroke-like episodes. The literature includes supportive reports and reviews, but consensus guidance also notes limitations and ongoing debate about strength of evidence and clinical use.

In MELAS, nutrition care often focuses on steady intake, illness-day planning, and discussing targeted therapies (like specific supplements) with a mitochondrial specialist because recommendations vary across clinics and individuals.

Mitochondrial Myopathies

When muscle involvement is prominent, nutrition strategies often focus on supporting muscle maintenance and minimizing fatigue by ensuring adequate overall calories and protein, especially if appetite is low or unintentional weight loss is present.

Some research in mitochondrial disease populations (including those with muscle symptoms) suggests that insufficient intake is common, and that higher intakes of macronutrients (including protein) may correspond with better outcomes like muscle strength, lower fatigue, and improved quality of life, though this doesn't mean "more is always better," and personalization matters.



For mitochondrial myopathies, nutrition support often prioritizes “enough energy + enough protein,” using practical strategies (small frequent meals, energy-dense additions, smoothies) when fatigue or GI issues make intake difficult.

POLG-related Disorders

POLG-related disorders can present very differently from one person to another. Nutrition strategies commonly emphasize **preventing catabolic stress** (especially during illness or reduced intake) and maintaining consistent energy availability, an approach aligned with broader mitochondrial care standards.

In some POLG-related conditions, **seizures** can be a major concern, and dietary therapies (such as ketogenic-style approaches) may be considered in specialized contexts (typically for difficult-to-treat ((refractory)) epilepsy) under expert supervision, with careful monitoring and individualized risk–benefit discussions.

For POLG, the key nutrition themes are often “avoid catabolic stress, plan for illness, and create individualize dietary approaches with your mito specialist and care team,” especially if seizures or significant GI/nutrition issues are part of the picture.

Because mitochondrial disease includes many different conditions, nutrition approaches are rarely one-size-fits-all. Some disorders rely heavily on nutrition strategies to prevent metabolic crisis, while others focus on maintaining energy balance, supporting muscle health, or managing symptoms. Across many diagnoses, maintaining consistent energy intake and avoiding metabolic stress are key themes. Individualized guidance from a mitochondrial specialist or dietitian is important to determine the best approach.

Research in MitoNutrition

Mitochondrial disease is complex and highly individualized, so it’s no surprise that nutrition research is moving toward more **personalized, diagnosis-informed approaches**. While we don’t yet have a single “best diet” for mitochondrial disease, growing evidence is helping clinicians better understand *when* nutrition strategies may be helpful, *for whom*, and *why*.

This is an encouraging direction. As research evolves, it strengthens the possibility of more refined nutrition guidance, better symptom management tools, and ultimately, improved quality of life for adults living with mito.



Precision nutrition based on genetic diagnosis

One of the most hopeful shifts in the field is a move away from one-size-fits-all advice and toward precision care, where nutrition strategies are tailored to a person's specific mitochondrial diagnosis, symptoms, and metabolic risks.

This approach recognizes that different genetic conditions can affect energy pathways differently, and nutrition strategies need to match those differences (for example, fasting guidance in FAOD versus other mitochondrial conditions).

Novel mitochondrial-targeted supplements and metabolic therapies

Researchers continue to explore therapies aimed at supporting mitochondrial function more directly, including targeted supplements and “metabolic support” strategies that influence how cells generate and use energy. For example, NAD⁺ biology and NAD⁺-boosting compounds (such as nicotinamide riboside and related approaches) are being actively studied across conditions involving mitochondrial health, with emerging clinical trial results in rare disorders showing mixed but evolving findings. NAD⁺ (Nicotinamide Adenine Dinucleotide) is a coenzyme found in every cell, crucial for energy metabolism, DNA repair and cellular signaling.

It's important to note that many of these approaches are still under study, and what's appropriate can vary widely by diagnosis. This is why specialist guidance remains essential as new options emerge.

The role of gut health and the microbiome

Another rapidly growing area is the connection between gut health, the microbiome, and mitochondrial function. A 2024 review focused specifically on mitochondrial disease highlights the “mitochondria-microbiome” connection and explores how diet and microbe-produced metabolites (small organic molecules produced during metabolism) may influence mitochondrial pathways, opening up new possibilities for supportive therapies and research.

For many adults with mito who experience gastrointestinal (GI) symptoms, this research is especially meaningful because it validates that GI function isn't “separate” from energy, it may be part of the same interconnected system.

A hopeful but grounded takeaway

This research is promising, but much of it is still evolving. The most consistent message across literature is that **individualized care matters**, and that nutrition strategies are most helpful when they are diagnosis-informed, symptom-aware, and guided by a specialist team.



Conclusion

Nutrition is a powerful tool for supporting daily living with mitochondrial disease.

By understanding the principles of MitoNutrition, balanced meals, adequate hydration, symptom-aware strategies, and appropriate supplements, individuals can make informed choices that help support energy, resilience, and overall quality of life.

As with all aspects of mito care, nutrition works best when it is personalized and team-based, guided by healthcare providers who understand both the diagnosis and the individual. Because every mitochondrial condition, and every person living with mito, is different, there is no single approach that fits all.

You are not alone on this journey. Small, thoughtful nutrition steps, taken one at a time, can add up to meaningful support over time, helping you navigate daily life with greater confidence and care.

This resource was developed with the support of an unrestricted education grant from Ultragenyx.



References:

Clinical Care Standards & Consensus Guidance:

Parikh S, Goldstein A, Koenig MK, et al.

Patient care standards for primary mitochondrial disease: A consensus statement from the Mitochondrial Medicine Society. Genetics in Medicine. 2017.

Camp KM, Krotoski D, Parikh S, et al.

Nutritional interventions in primary mitochondrial disorders. Molecular Genetics and Metabolism. 2016.

Nutrition, Energy Intake, and Quality of Life:

DiVito D, Hirano M, et al.

Optimized nutrition in mitochondrial disease correlates with improved muscle fatigue, strength, and quality of life. Neurotherapeutics. 2023.

Rinninella E, Cintoni M, De Lorenzo A, et al.

Nutritional support in mitochondrial diseases: The state of the art. European Review for Medical and Pharmacological Sciences. 2018.

Fatty Acid Oxidation Disorders (FAOD)

Peña-Quintana L, Martín-Hernández E, et al.

Nutritional management of patients with fatty acid oxidation disorders. Nutrients. 2024.

Merritt JL II, Norris M, Kanungo S.

Fatty acid oxidation disorders. Annals of Translational Medicine. 2018.

British Inherited Metabolic Disease Group (BIMDG).

Adult Emergency Management Guidelines for Fatty Acid Oxidation Disorders

MELAS and Targeted Nutrition Therapies

El-Hattab AW, Emrick LT, Hsu JW, et al.

Arginine and citrulline therapy in MELAS syndrome: A systematic review. Molecular Genetics and Metabolism. 2017.

Ng YS, Bindoff LA, Gorman GS, et al.

Consensus-based statements for the management of mitochondrial stroke-like episodes.

Wellcome Open Research. 2019.



Stefanetti RJ, et al.

Efficacy of L-arginine in MELAS stroke-like episodes: A systematic review. Neurology. 2022.

Mitochondrial Myopathies & Muscle Health

DiVito D, et al.

Nutrition and muscle outcomes in primary mitochondrial disease. Neurotherapeutics. 2023.

POLG-Related Disorders & Dietary Therapies

Pedersen ZO, Jeppesen TD, et al.

Efficacy and safety of dietary therapies in POLG-related disorders: A systematic review.

Epilepsy Research. 2022.

Na JH, et al.

Therapeutic approaches to epilepsy in mitochondrial disease. Yonsei Medical Journal. 2025.

Supplements & Mitochondrial Cofactors

NIH Office of Dietary Supplements.

Dietary Supplements for Primary Mitochondrial Disorders-Health Professional Fact Sheet.

Barcelos I, et al.

Mitochondrial medicine therapies: Rationale, evidence, and dosing considerations.

Frontiers in Neurology. 2020.

Journal of Inherited Metabolic Disease.

Global vitamin and cofactor prescribing practices in mitochondrial disease. 2024

Hydration, Autonomic Dysfunction, and Symptom Management

Parikh S, et al.

Patient care standards for primary mitochondrial disease. Genetics in Medicine. 2017.

Gut Health, Microbiome, and Emerging Research

Zachos KA, et al.

The interplay between mitochondria, the gut microbiome, and mitochondrial disease.

Frontiers in Physiology. 2024.

Yusri K, et al.

The role of NAD⁺ metabolism and its modulation in mitochondrial disease.

International Journal of Molecular Sciences. 2025.

