

Navigating Nutrition: Dietary Management in Children with Mitochondrial Disease

Understanding Mitochondrial Disease

Mitochondrial disease (mito) refers to a group of disorders that result when mitochondria cannot produce enough energy, or an individual doesn't have enough healthy mitochondria in their bodies. This can lead to lower energy in cells, greatly affecting children's growth, health, and how well their bodies work overall. Symptoms can occur across multiple organ systems, such as the brain, muscles, heart, and stomach. Due to the genetic nature and complexity of pediatric mitochondrial disorders, tailored approaches to treatment and care are essential, with nutritional management playing a pivotal role in optimizing energy production and improving the quality of life for children living with mito.



The Role of Nutrition in Mitochondrial Disease

Nutrition plays a crucial role for children with mito for several key reasons:

- 1. Energy Production:** Mitochondrial diseases impair the body's ability to produce energy efficiently. Proper nutrition provides the necessary substrates (like glucose and fatty acids) that the mitochondria need and use to generate ATP. ATP, or adenosine triphosphate, is essentially the energy currency for your body, which allows your cells to perform all their essential functions.
- 2. Growth and Development:** Proper nutrition is critical for regular growth and development, which may be hindered in children with mitochondrial disease. Nutritional deficiencies can worsen growth and developmental delays.
- 3. Immune Function:** Good nutrition supports a healthy immune system. Children with mito may have compromised immune function, making them more susceptible to infections. Proper nutrition can help bolster their immune defences.
- 4. Symptom Management:** Certain nutrients can help manage symptoms associated with mito. For example, antioxidants like Coenzyme Q10, vitamins C and E, and alpha-lipoic acid can help mitigate oxidative stress, a common issue in mitochondrial disorders.
- 5. Muscle Function and Strength:** Mitochondrial disorders often affect muscle function and strength. Adequate protein intake is necessary to support muscle health and prevent muscle wasting.

6. **Brain Health:** The brain requires a significant amount of energy, and dysfunctional mitochondria can lead to neurological symptoms. Proper nutrition, including essential fatty acids and vitamins, is crucial for supporting brain health and cognitive function.
7. **Metabolic Stability:** Metabolic stability refers to the body's ability to maintain a stable internal environment, even when faced with external changes. It involves balancing things like energy levels and the body's chemical reactions. Children with mito can have metabolic instability. Regular and balanced meals can help maintain stable blood sugar levels and prevent metabolic crises.
8. **Quality of Life:** Good nutrition enhances overall well-being and quality of life. It can help improve energy levels, reduce fatigue, and support physical and cognitive activities, leading to a better quality of life.



Nutritional Challenges and Strategies

Managing the nutritional needs of children with mito presents unique challenges due to the variability in symptoms and disease progression. Tailored dietary plans are essential and must be responsive to the metabolic demands and clinical changes in each mito child.

Healthcare professionals, particularly dietitians specializing in metabolic disorders, play a crucial role in developing personalized nutrition plans that cater specifically to a child's type of mito, the symptoms it produces, and how it affects the body. Regular monitoring of the patient's nutritional status through the evaluation of growth and blood tests, looking at nutrient levels and metabolic markers, is vital to ensure that the diet and supplement regimen continues to meet the child's evolving needs.

The following nutritional strategies are typically personalized based on the type of mito a child has, disease progression, and nutritional status. Regular consultations with healthcare providers are crucial to tailor these approaches effectively.

Common nutritional strategies include:

1. **Maintaining a Balanced Diet:** A balanced diet rich in a variety of macronutrients—proteins, carbohydrates, and fats—is important. Proteins not only support muscle maintenance and growth but are also essential for repairing body tissues. Complex carbohydrates (such as whole grains) are preferred over simple sugars (such as table sugar) as they provide a more consistent energy supply, helping the body efficiently convert these nutrients into usable energy without causing rapid spikes in blood sugar levels. Fats are a highly-dense source of mitochondrial fuel, although fat intake may need to be tailored in specific disorders where the metabolism of longer chain fats is impaired.

2. **Managing Calorie Intake:** Managing caloric intake is also vital. Both undernutrition and overnutrition can worsen symptoms and make it more difficult to manage mito. Through regular monitoring, healthcare providers can suggest adjustments to calorie intake based on the child's activity levels and metabolic needs that are specific to their disorder. Furthermore, to prevent hypoglycemia—a common risk in mitochondrial disorders—eating small, frequent meals throughout the day may be recommended. Snacks between meals help avoid prolonged fasting periods, which can be particularly harmful. These strategic dietary adjustments are essential to address the energy production challenges inherent in mitochondrial disorders, ensuring that affected children maintain stable health and energy levels.

3. **Customized Mito Cocktail:** Physicians may advise the use of a combination of vitamins and mitochondrial co-factors, sometimes known as the “mito cocktail,” which is designed to support mitochondrial function, reduce symptoms, and improve overall energy levels in children and adults with mitochondrial disease. The specific supplements and dosages, which typically include ingredients like Coenzyme Q10, L-carnitine, and various B vitamins, are tailored to meet the child's needs based on their particular mitochondrial disorder and overall health profile. It is important to always consult healthcare providers for personalized advice and treatment plans.

Common supplements that comprise a mito cocktail are:

- Coenzyme Q10 (CoQ10) is a naturally occurring substance that is crucial in producing energy within cells. It is found in the mitochondria, which helps convert nutrients into energy through ATP (adenosine triphosphate). This process is vital for the functioning of all the body's cells and systems.
- L-carnitine plays a crucial role in energy production by transporting fatty acids into the mitochondria, which can be burned for energy. This process is vital for many bodily functions, particularly for heart and muscle movement and for overall energy metabolism.
- Riboflavin (Vitamin B2) plays a crucial role in the growth and development of body cells, aiding in converting nutrients into energy. It is vital for maintaining healthy skin, eyes, and nerves and ensures the mucous membranes in the digestive system function properly. Additionally, riboflavin enhances the absorption and effectiveness of other essential nutrients such as iron, folate, and other B vitamins, making it a key component in overall health and nutrition.
- Thiamine (Vitamin B1) is an essential vitamin that plays a crucial role in energy metabolism, particularly in converting carbohydrates into glucose, which the body uses as energy. It's also vital for the proper function of the heart, muscles, and nervous system.
- Niacin (Vitamin B3) supports the digestive system, skin, and nerves. It's also important for converting food into energy. One of its major functions is helping the body produce various sex and stress-related hormones in the adrenal glands and other parts.



4. Avoiding Fasting: This is crucial for children with mitochondrial disease, as prolonged periods without food can exacerbate symptoms and lead to metabolic crises. It is important for them to have regular meals and snacks to maintain stable energy levels. Small, frequent meals may be recommended to prevent energy dips and manage gastrointestinal symptoms, ensuring stable blood sugar levels and a continuous energy supply. This approach helps sustain their overall health and well-being by providing a steady source of nutrients and energy.



5. Staying Hydrated: Proper hydration is crucial for children with mitochondrial dysfunction, as dehydration can exacerbate symptoms; therefore, children must be encouraged to drink fluids throughout the day regularly.

Research and Future Directions

Continued research will enhance our understanding of how specific nutrients and diets can improve mitochondrial function. For instance, studies indicate that ketogenic diets, which are high in fats and low in carbohydrates, may be effective in managing epilepsy linked to mitochondrial dysfunction. These findings can point to potential broader uses for diet in treating mitochondrial diseases.

Conclusion

Nutritional management in pediatric mitochondrial disease is a dynamic and critical component of a child's overall care strategy. It requires a personalized approach, guided by ongoing research and clinical insights, to optimize dietary interventions that support energy production and enhance quality of life.

Collaboration between dietitians, physicians, and caregivers is essential to implement effective nutritional strategies that are adapted to the needs of each young mito patient. Staying informed about the latest dietary recommendations and scientific findings is crucial in navigating the complexities of mito conditions.

About Us

Since 2010, MitoCanada has been supporting Canadians living with mitochondrial disease with information and opportunities to inspire, empower and connect while enhancing quality of life through our research, education and awareness programs.

MitoCanada is the only Canadian charity dedicated to mitochondrial disease. We are the voice of Canadians living with or at risk of developing mitochondrial disease.



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