Monogen[®] A low fat, high MCT formula for the dietary management of long chain fatty acid oxidation disorders now available from Nutricia North America

Ulrike Reichert, MS Nutricia North America

Fatty acid oxidation disorders recommended for newborn screening

Fatty acid oxidation disorders (FAO) are increasingly better-known disorders within the medical community. They have also attracted more attention recently due to the expansion of newborn screening and efforts to make screening more uniform throughout the United States. Medium and long chain fatty acid oxidation disorders, trifunctional protein and carnitine uptake deficiency are all part of the recommended core panel of disorders for which every newborn in the US should be screened¹.

Nevertheless, even with earlier detection, these disorders pose a challenge to families and medical teams alike. Nutritional therapy based on a restricted diet can be challenging to create and manage. Nutricia North America is committed to partnering with families and healthcare professionals in finding solutions to these dietary challenges via innovative products, programs and tools. Since our very beginning, we have continuously and diligently worked with families and the medical community to improve the quality of life and care for metabolic patients. From these efforts, *Monogen* was developed, and it is now available to families in the United States and Canada.



Monogen - a new formula available for patients with long chain FAO disorders

Monogen is a medical food formula for the dietary management of long chain fatty acid oxidation disorders. These disorders are rare—long chain 3-OH acyl-CoA dehydrogenase deficiency, for example, is estimated to have an incidence of 1:50,000 to 1:200,000.² Individuals with these disorders cannot metabolize long chain fatty acids (LCFA), a certain group of fats commonly found in fat-containing foods. These disorders result from defects in enzymes needed to convert LCFAs to

smaller fatty acids—a process necessary to produce energy in the body.

The main goals of dietary management are to reduce dietary long chain fatty acids and avoid fasting. Tolerance of dietary LCFA is individual and depends on the remaining activity of the blocked enzymes. The actual recommended amounts of restricted dietary LCFA and total fat may differ from clinic to clinic. Typically, 10% or less of the daily required calories is needed and tolerated from LCFA. Recommendation of total fat intake may be reduced to as low as 25% of total energy intake.

In order to reach the goal of reduced total fat intake with limited LCFA and to provide adequate calories, medium chain fats (MCT) are used in formulas or directly added to the diet. Medium chain

^{1, 2} American College of Medical Genetics, *Newborn screening: Toward a uniform screening panel and system*. Rockville, MD, 2005.

fats bypass the enzymes that are blocked in long chain fatty acid oxidation disorders, and they can be used as alternative sources of fat.

However, MCTs do not contain any of the long chain essential fatty acids needed by the body. Consequently, the amount and type of long chain fatty acids in the diet must be carefully considered to provide sufficient but not excessive amounts of these important fatty acids.

Monogen – optimized for use in long chain fatty acid oxidation disorders

Monogen is an ideal formula for use in long chain fatty acid oxidation disorders. Here are some of the unique features and benefits of the product:

- *Monogen* provides 25% of calories from fat as recommended for a low fat diet
- Monogen has a high content of MCT (90% of fat is MCT), which bypasses the blocked enzymes
- In contrast to other formulas, *Monogen* offers a balanced ratio of essential fatty acids, which is important in avoiding essential fatty acid deficiencies
- **Monogen** also provides a complete nutrient profile, including trace elements, to minimize the risk of nutrient deficiencies.

In order to make **Monogen** suitable for most patients, the essential fatty acid content is 1.2% of total energy. This allows for a tailored approach to the individual needs of patients. Dietary essential fatty acids can be easily increased by adding a prescribed amount of walnut or canola oil to **Monogen**. In long chain FAO disorders, monitoring of essential fatty acids and micronutrients is generally recommended.

Contact Nutricia North America for samples of Monogen

Families, please have your metabolic dietitian or physician contact Nutricia North America. We can send a sample of *Monogen* conveniently to your home.

Healthcare professionals, please contact our Nutrition Services Department or your regional Nutricia representative.



Monogen is an excellent choice when considering a formula for a child with a long chain fatty acid oxidation disorder.

For more information, please call our Nutrition Services Department at 1-800-365-7354. Monday – Friday 8:30 am – 5:00 pm EST or visit one of our Web sites, <u>www.myspecialdiet.com</u> and <u>www.nutricia-na.com</u>. We are here for you.

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