



VERY LONG-CHAIN ACYL-COA DEHYDROGENASE DEFICIENCY (VLCAD) CONSUMER SUMMARY

SERN/GMDI Nutrition Management Guidelines

First Edition

Nutrition Guidelines for Individuals with VLCAD^{1,2,3}

August 2019

This information is intended for individuals with VLCAD and/or their caregivers (hereafter referred to as “you/your”). This summary includes current management recommendations and suggests topics that you can discuss with your metabolic team to help identify a plan that is best for you.

- Individualized treatment plans are based on the severity of VLCAD, age, weight and height, activity level, lab results, and whether you are sick or healthy.
- Work with your metabolic clinic to always have a sick-day plan and an emergency letter to use when seeking urgent medical care.
- Follow your metabolic dietitian’s instructions to meet your needs for fat, protein, calories, and other nutrients.
- You may be recommended a diet that is restricted in long-chain fat (LCF). Supplemental medium chain triglycerides (MCT) are often needed when long chain fat is restricted.
- For infants with VLCAD, breast-feeding may be acceptable. However, a formula containing MCT is often used to supplement or replace breast milk.
- Your treatment plan will include a maximum fasting time (that is, the amount of time that you can go without eating). A bedtime snack of complex carbohydrates (i.e. whole grains) is sometimes recommended.
- Discuss how to meet your energy needs during physical activity. MCT, with or without foods/beverages containing carbohydrate, is often recommended as a source of energy before physical exercise.
- Ask your metabolic team if you need any supplements such as carnitine, vitamins or essential fatty acids.
- During clinic visits, your growth and nutrient intake will be assessed, and laboratory tests may be performed. Routine laboratory tests often include creatine kinase and plasma carnitine profile (total, free, esterified carnitine). Plasma acylcarnitines, essential fatty acids, and other laboratory values are sometimes measured.
- If you are a female, obtain information from your metabolic team about the impact of puberty, menstruation, and pregnancy, and potential changes to diet, medications and supplements that may be needed during these times.

This document is not meant to substitute for the medical advice provided by your doctor.

¹ For children, adolescents, and adults with VLCAD, or their caregivers

² Based on the 2019 Nutrition Management Guidelines for Very Long-Chain Acyl-CoA Dehydrogenase Deficiency (VLCAD) by GMDI/SERN available at: http://southeastgenetics.org/ngp/guidelines_vlcad.php

³ The Management Guidelines Advisory Committee used the nationally standardized condition abbreviation of VLCAD; curated by the US National Library of Medicine for this and related guideline products: <https://newbornscreeningcodes.nlm.nih.gov/>

