VERY LONG-CHAIN ACYL-COA DEHYDROGENASE DEFICIENCY (VLCAD)
FREQUENTLY ASKED QUESTIONS

SERN/GMDI Nutrition Management Guidelines **First Edition**

F.A.Q. About Nutrition Management for Individuals with VLCAD[1,2,3] January 2020

NUTRIENT INTAKE	
Can a baby with VLCAD be breastfed?	Babies' diets depend on whether they have mild, moderate or severe VLCAD. Talk to your metabolic team to determine whether or not your baby with VLCAD can be breastfed and/or requires a medical food (formula) (Rec 1.6).
How often do I need to eat?	The amount of time that you can fast (go without eating) depends on your age and your symptoms of VLCAD (Rec 1.7). Your metabolic team will recommend a maximum fasting time. For those with severe VLCAD who do not tolerate extended fasting times, an overnight feeding may be considered if a bedtime snack is not sufficient (Rec 1.9).
What should I have as a bedtime snack?	To meet fasting guidelines and prevent catabolism, a bedtime snack emphasizing complex carbohydrates may be needed (Rec 1.9). If so, your dietitian can provide suggestions for appropriate snacks based on your preferences.
How much long-chain fat (LCF) can I consume?	The amount of LCF that you can consume depends on your age and the severity of your VLCAD (Rec 1.2). Your metabolic team will consider your clinical status and lab results to determine if they need to adjust your LCF goals.
SUPPLEMENTS	
Do I need a medium-chain triglyceride (MCT) supplement?	If you need to restrict your LCF intake, then you may need to take MCT to meet your energy needs (Rec 1.2).
Do I need other supplements?	Your dietitian may also recommend fatty acid supplements such as oils or DHA (Rec 1.8). L-carnitine supplementation may or may not be recommended by your metabolic physician, depending on your symptoms and the free carnitine concentration in your blood (Rec 3.1).
MONITORING	
What blood tests are helpful to assess the impact of VLCAD?	Creatine kinase (CK) and plasma carnitine profile are useful for the routine management of VLCAD (Rec 4.3). Other blood tests that may be done when indicated by special circumstances include a comprehensive metabolic panel (CMP), acylcarnitine profile, essential fatty acids, fat soluble vitamins, and other biochemical markers (Rec 4.4).
How often should blood testing be done?	Routine blood testing (such as CK and plasma carnitine profile) should be measured about one to four times per year, depending on your age, the severity of your VLCAD, the symptoms you have, and your clinical history. Additional monitoring may be required when indicated by special circumstances (Rec 4.3, 4.4).

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

[1] *For the child, teenager, or adult living with VLCAD and their caregivers.*

[2] *Based on the 2019 Nutrition Management Guidelines for Very Long-Chain Acyl-CoA Dehydrogenase Deficiency (VLCAD) by GMDI/SERN: https://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/*

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EXERCISE	
How should I prepare for exercise?	Individuals with VLCAD can participate in normal physical activity for age, as tolerated, with appropriate energy support (Rec 5.1). MCT may be recommended 20 minutes before exercise to improve exercise tolerance (Rec 5.2). Talk to your dietitian about the appropriate dose of MCT.
ILLNESS	
What should I do if I become ill?	Call your metabolic physician for medical guidance. Take your emergency letter with you when seeking urgent medical care (Rec 2.2).
How does nutrition management change during illness?	When ill, fasting times may be shorter, and frequent high-carbohydrate feedings may be recommended to maintain energy intake (Rec 2.3). Talk to your metabolic physician or dietitian for a nutrition management plan during illness.

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