At a Glance
Very Long Chain Acyl Co-A Dehydrogenase Deficiency (VLCAD)

Deficient enzyme: Very long chain acyl-CoA dehydrogenase

Restrict: Dietary long chain fat (LCF)

Clinical presentation, in untreated patients:*

Mild: asymptomatic beyond infancy, tolerates catabolic stressors without decompensation, potential for rhabdomyolysis

Moderate: asymptomatic at diagnosis, hypoketotic hypoglycemia, rhabdomyolysis due to catabolic illness, fasting or exercise

Severe: symptomatic at diagnosis or within first months of life, hypertrophic or dilated cardiomyopathy, pericardial effusion, hypotonia, hepatomegaly, intermittent hypoglycemia rhabdomyolysis

* SERN/GMDI VLCAD Management; https://southeastgenetics.org/ngp/guidelines.php

Nutrient Needs by Age (VLCAD)*

Age	Disease Severity	Total Fat (% of total energy)	Long-Chain Fat (% of total energy)	Medium-Chain Fat (% of total energy)
0-6 months	Severe	40-55	10-15	30-45
	Moderate		15-30	10-30
	Mild		30-55	0-20
7-12 months	Severe	35-42	10-15	25-30
	Moderate		15-30	10-25
	Mild		30-40	0-10
1-3 years	Severe	30-40	10-15	10-30
	Moderate		20-30	10-20
	Mild		20-40	0-10
4-18 years	Severe	25-35	10	15-25
	Moderate		15-25	10-20
	Mild		20-35	0-10
>19 years	Severe	20-35	10	10-25
	Moderate		15-20	10-20
	Mild		20-35	0-10

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Starting a VLCAD Diet

(asymptomatic individuals with mild VLCAD may not need a fat-restricted diet)

1. Determine goals for LCF, MCT, Total Fat, Protein (g), Energy (kcal)
2. Calculate amount of LCF (breast milk, infant formula, food) needed to meet LCF goal.
3. Calculate amount of MCT needed to meet total fat goal.
4. Calculate energy intake from protein and fat sources to ensure total energy needs are met.

Very Long Chain
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Medical Food Therapy

	Mead Johnson hcp.meadjohnson.com	Nutricia NutriciaMetabolics.com	Vitaflo www.VitafloUSA.com
Infant (0-1 yr)	Enfaport™		
Toddler & Young Children		Monogen® Liquigen®	LIPIstart™ MCTprocal® Betaquik®
Older Children & Adults		Monogen® Liquigen®	LIPIstart™ MCTprocal® Betaquik®

Nutrition Supplementation*

Medium chain triglycerides (MCT): dose depends on severity of disease and LCF restriction

-Medical foods (above) contain varying amounts of MCT. MCT oil is also available. These sources of MCT contain even-chain fatty acids with 6 to 10 carbons.

Docosahexaenoic acid (DHA): 60 mg/d- for patients <20 kg; 100 mg/d- for patients >20 kg if normal plasma or RBC DHA concentrations cannot be achieved by diet modification.

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Medical Management

Triheptanoin (Dojolvi®) Ultragenyx Pharmaceutical (Novato, CA): An odd-chain fatty acid containing 7 carbons, used instead of even-chain MCT. Dose: 35% of total energy intake. www.dojolvi.com

Fasting Precautions*

Times between feedings for a well patient; lower end of the range applies to patients with severe VLCAD:

0-4 months: 3-4 hours
4-<6 months: 4-6 hours
6-<9 months: 6-8 hours
9-<12 months: 8-10 hours
>12 months: 10-12 hours

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Laboratory Monitoring

Creatine Kinase[1, 2]
Plasma Carnitine[1, 2]
Plasma Acylcarnitine[1, 2]
Essential Fatty Acids[2]
B-natriuretic protein (BNP)[3]
25-OH Vitamin D[3]
CMP[3]
CBC[3]

[1] Every 3 months
[2] Every 6 months after 1 yr. of age
[3] As indicated

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