

Very Long Chain Acyl CoA Dehydrogenase Deficiency

Ages 2-18 years

Clinic Visits:

- At least every 6 months

Laboratory studies:

- Clinic visits
 - Carnitine levels
 - Serum CK (muscle health)
 - Possible acylcarnitine profile
 - Possible liver function tests
- Yearly
 - Comprehensive metabolic panel
 - Acylcarnitine profile
 - B-type Natriuretic Peptide (heart health)
 - Essential fatty acid profile at 6 months and yearly thereafter
 - Possible nutritional labs

During illness:

- Call Metabolic Clinic to alert them that your child is sick
 - Poor feeding, vomiting, or lethargy
- Give your child glucose gel or polymer solution if needed (or instructed to do so)
- If your child is not able to keep down glucose containing fluids, you will need to take him or her to the emergency room for IV 10% dextrose (glucose)
- Be sure to bring Emergency Room letter with you to the hospital
- Labs during illness include: comprehensive metabolic panel, ammonia levels, serum CK, acylcarnitine profile, and BNP
- Please alert the Metabolic team if your child needs surgery
- Wear medic alert bracelet!**

Evaluations:

- Yearly developmental questionnaires to be completed by parents
- Yearly cardiac evaluation
- Developmental evaluation at 3 and 6 years
- Neuropsychology evaluation at 9 years

Topics:

- Biochemistry and genetics of VLCADD
- Diet/management
 - Avoid fasting
 - Management during illness
 - Carnitine supplementation
 - Cornstarch if needed
 - Low-fat diet with limited long-chain fats
 - Medium chain triglyceride (MCT) formula

- Essential oils

Please make sure your child has clinic visits and levels done on time. Call the Metabolic clinic at _____ to make an appointment, schedule a blood draw, or with any other questions. If you urgently need to reach a Metabolic physician, call _____ and ask to page the Metabolic physician on call.

Thank You,
Your Metabolic Team