

Very Long Chain Acyl CoA Dehydrogenase Deficiency

Ages 0-6 months

Clinic Visits:

- At least every 2 months

Laboratory studies:

- At diagnosis
 - Acylcarnitine profile
 - Comprehensive metabolic panel
 - Carnitine panel
 - Serum creatine kinase (CK) for muscle health
 - Urine analysis
 - DNA analysis of the VLCAD gene
 - B-type Natriuretic Peptide (BNP) for heart health
 - Possible urine organic acids
 - Possible fatty acid oxidation flux studies
- Clinic visits
 - Carnitine levels
 - Serum CK
 - 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

During illness:

- Call Metabolic Clinic to alert them that your baby is sick
 - Poor feeding, vomiting, or lethargy
- Give your baby glucose gel or polymer solution if needed (or instructed to do so)
- If your baby 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 baby needs surgery
- Consider medic alert bracelet

Evaluations:

- Echocardiogram at diagnosis and 6 months

Topics:

- Biochemistry and genetics of very long chain acyl-CoA dehydrogenase deficiency (VLCADD)

- Diet/management
 - Avoid fasting
 - Management during illness
 - Carnitine supplementation
 - 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