

Long chain 3-hydroxyacyl-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 CK (muscle health)
 - Possible urine organic acids
 - Possible DNA analysis of the LCHAD gene
 - Possible fatty acid oxidation probe studies
 - Possible B-type natriuretic peptide (heart health)
- Clinic visits
 - Possible carnitine levels
 - Possible serum CK
 - Possible acylcarnitine profile (diet monitoring)
 - Possible liver function tests

During illness:

- Call Metabolic Clinic to alert them that your baby is sick
 - Poor feeding, vomiting, and/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, serum CK, acylcarnitine profile, B-type natriuretic peptide, and possible ammonia
- IV carnitine may be given to your baby
- Possible cardiac monitoring during hospitalization
- Consider medic alert bracelet**

Evaluations:

- Cardiac evaluation at diagnosis and 6 months

Topics:

- Biochemistry and genetics of long chain 3-hydroxyacyl-CoA dehydrogenase deficiency (LCHADD)
- Diet/management
 - Avoid fasting
 - Management during illness
 - Low-fat diet with limited long-chain triglycerides
 - Medium chain triglyceride (MCT) supplementation (oil or formula)

- Supplementation with essential oils
- Possible carnitine supplementation

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