

Long chain 3-hydroxyacyl-CoA dehydrogenase deficiency

Ages 6-24 months

Clinic Visits:

- At least every 3 months

Laboratory studies:

- Clinic visits
 - Possible carnitine levels
 - Possible serum CK (muscle health)
 - Possible acylcarnitine profile (diet monitoring)
 - Possible liver function studies
- Yearly
 - Comprehensive metabolic panel
 - Acylcarnitine profile
 - Essential fatty acid profile at 6 months and then yearly
 - B-type natriuretic peptide (heart health)
 - Possible nutritional labs

During illness:

- Call Metabolic Clinic to alert them that your child is sick
 - Poor feeding, vomiting, and/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, 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:

- Yearly developmental questionnaires to be completed by parents
- Cardiac evaluation at 6 months and 12 months
- Ophthalmology evaluation yearly

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
- Possible cornstarch after 1 year old

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