

Long chain 3-hydroxyacyl-CoA dehydrogenase deficiency

Ages 2-18 years

Clinic Visits:

- At least every 6 months

Laboratory studies:

- Clinic visits
 - Possible carnitine levels
 - Possible serum CK (muscle health)
 - Possible acylcarnitine profile (diet monitoring)
 - Liver function tests
- Yearly
 - Comprehensive metabolic panel
 - Acylcarnitine profile
 - Essential fatty acid profile at 6 months and yearly thereafter
 - 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
- Wear medic alert bracelet!**

Evaluations:

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

Topics:

- Review of biochemistry and genetics of long chain 3-hydroxyacyl-CoA dehydrogenase deficiency
- 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

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