

Long Chain 3-hydroxyacyl-CoA Dehydrogenase Deficiency

Ages 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 you are sick
 - Poor feeding, vomiting, and/or lethargy
- Take glucose gel or polymer solution if needed (or instructed to do so)
- If you are not able to keep down glucose containing fluids, you will need to go 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 you
- Possible cardiac monitoring during hospitalization
- Wear medic alert bracelet!**

Evaluations:

- Yearly cardiac evaluation
- Yearly ophthalmology evaluation
- Psychiatric screening at 18 years

Topics:

- Review 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

Please make sure you have 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