Very Long Chain Acyl CoA Dehydrogenase Deficiency
Ages 6-24 months

Clinic Visits:

- ☑ At least every 3 months

Laboratory studies:

- ☑ Clinic visits
  - o Carnitine levels
  - o Serum CK (muscle health)
  - o Possible acylcarnitine profile
  - o Possible liver function tests
- ☑ Yearly
  - o Comprehensive metabolic panel
  - o Acylcarnitine profile
  - o B-type Natriuretic Peptide (heart health)
  - o Essential fatty acid profile at 6 months and yearly thereafter

During illness:

- ☑ Call Metabolic Clinic to alert them that your child is sick
  - o Poor feeding, vomiting, 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, ammonia levels, serum CK, acylcarnitine profile, and BNP
- ☑ Please alert the Metabolic team if your child needs surgery
- ☑ 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 VLCADD
- ☑ Diet/management
  - o Avoid fasting
  - o Management during illness
  - o Carnitine supplementation
  - o Cornstarch after age 1 year if needed
  - o Low-fat diet with limited long-chain fats
  - o Medium chain triglyceride (MCT) formula
  - o 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