

# Very Long Chain Acyl CoA Dehydrogenase Deficiency

## Ages 18+ years

### Clinic Visits:

- At least every 6 months

### Laboratory studies:

- Clinic visits
  - Carnitine levels
  - Serum CK (muscle health)
  - Possible acylcarnitine profile
  - Possible liver function tests
- Yearly
  - Comprehensive metabolic panel
  - Acylcarnitine profile
  - B-type natriuretic peptide (heart health)
  - Essential fatty acid profile

### During illness:

- Call Metabolic Clinic to alert them that you are sick
  - Poor feeding, vomiting, 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, ammonia levels, serum CK, acylcarnitine profile, and B-type natriuretic peptide
- Wear medic alert bracelet!**

### Evaluations:

- Yearly developmental questionnaires to be completed by you
- Yearly cardiac evaluation
- Psychiatric screening at 18 years

### Topics:

- Biochemistry and genetics of Very Long Chain Acyl-CoA Dehydrogenase Deficiency
- Diet/management
  - Avoid fasting
  - Management during illness
  - Carnitine supplementation
  - Cornstarch if needed
  - MCT formula
  - Heart healthy diet (lower fat)

*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