

Glutaric Acidemia, Type II
(Neonatal/Infantile type)
(AKA Multiple Acyl CoA Dehydrogenase Deficiency)
Ages 2-18 years

Clinic Visits:

- Every 6 months

Laboratory studies:

- Monitoring of diet
 - Plasma amino acid levels
 - 2-6 years: at least every 3 months
 - 6-18 years: at least every 6 months
- At clinic visits:
 - Carnitine levels
- Yearly
 - Comprehensive metabolic panel
 - Essential fatty acid profile
 - B-type natriuretic peptide (heart health)
 - Prealbumin / albumin
 - Plasma ferritin, transferrin, or iron studies
 - Possible complete blood count (CBC), hemoglobin and hematocrit
 - Possible nutritional laboratory studies

During illness:

- Call Metabolic Clinic to alert them that your child is sick
 - Poor feeding, fever, 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, ammonia, serum CK (muscle health), and B-type natriuretic peptide (heart health)
- IV carnitine may be given to your child
- Please alert the Metabolic Clinic if your child needs surgery
- Wear a medic alert bracelet!**

Evaluations:

- Yearly developmental questionnaires to be completed by parents
- Developmental evaluation at age 3 and 6 years
- Neuropsychological evaluation at age 9 years
- Yearly brain MRI
- Yearly cardiology evaluation with echocardiogram

Topics:

- Biochemistry and genetics of GA II/ MADD deficiency
- Diet/management
 - Low protein, low fat diet
 - Importance of medical formula

- Carnitine supplementation
- Riboflavin supplementation
- Avoid fasting
- Management during illness

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