

Glutaric Acidemia, Type II
(Late or Mild onset)
(AKA Multiple Acyl CoA Dehydrogenase Deficiency)
Ages 0-4 years

Clinic Visits:

- Every 6 months

Laboratory studies:

- At diagnosis
 - Plasma acylcarnitine profile
 - Urine organic acids
 - Carnitine levels
 - Comprehensive metabolic panel
 - Ammonia
 - Possible DNA analysis of the genes responsible for MADD/GA II
 - Possible skin biopsy (enzymatic studies)
- At clinic visits:
 - Carnitine levels
- Yearly
 - Comprehensive metabolic panel

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
- Please alert the Metabolic Clinic if your child needs surgery
- Consider medic alert bracelet**

Evaluations:

- Possible brain MRI is needed
- Yearly developmental questionnaires to be completed by parents
- Developmental evaluation at 3 years old

Topics:

- Biochemistry and genetics of GA II /MADD deficiency
- Diet/management
 - Avoid fasting
 - Management during illness
 - Carnitine supplementation
 - Riboflavin supplementation

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

