# Glutaric Acidemia, Type II
# (Neonatal/Infantile type)
# (AKA Multiple Acyl CoA Dehydrogenase Deficiency)
# Ages 0-6 months

**Clinic Visits:**

- ☑ Every 2 months

**Laboratory studies:**

- ☑ At diagnosis
  - o Plasma acylcarnitine profile
  - o Urine organic acids
  - o Carnitine levels
  - o Comprehensive metabolic panel
  - o Ammonia
  - o Serum CK (muscle health)
  - o B-type natriuretic peptide (heart health)
  - o Possible DNA analysis of the genes responsible for MADD/GA II
  - o Possible skin biopsy (enzymatic studies)
- ☑ Monitoring of diet
  - o Plasma amino acid levels *every 2 weeks*
- ☑ At clinic visits:
  - o Carnitine levels

**During illness:**

- ☑ Call Metabolic Clinic to alert them that your baby is sick
  - o 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, and B-type natriuretic peptide
- ☑ Please alert the Metabolic Clinic if your baby needs surgery
- ☑ **Consider medic alert bracelet**

**Evaluations:**

- ☑ Abdominal ultrasound if needed
- ☑ CT scan of head if needed

**Topics:**

- ☑ Biochemistry and genetics of GA II /MADD deficiency
- ☑ Diet/management
  - o Low protein, low fat diet
  - o 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