

Isovaleric Acidemia Ages 0-6 months

Clinic Visits:

- Every 2 months

Laboratory studies:

- At diagnosis
 - Urine organic acids
 - Plasma acylcarnitine profile
 - Plasma amino acids
 - Comprehensive metabolic panel
 - Ammonia
 - CBC
 - Urine analysis (for ketones)
 - Possible urine acylglycine profile
 - Possible DNA analysis of IVA gene
 - Possible skin biopsy (enzymatic studies)
- Monitoring of diet
 - Plasma amino acid levels at least every 2 weeks
 - Monitoring not needed in milder form of IVA

During illness:

- Call Metabolic Clinic to alert them immediately if your baby is sick
 - Poor feeding, fever, vomiting, or lethargy
- Give your baby glucose gel or polymer solution if needed (or instructed to do so)
- If your baby 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, phosphorous, ketones, CBC, amylase/lipase (risk of pancreatitis), possible ammonia, possible plasma amino acids, and possible urine organic acids
- IV carnitine may be given to your baby
- If possible, glycine should continue to be given orally
- Please alert the Metabolic Clinic if your baby needs surgery

Topics:

- Biochemistry and genetics of isovaleric acidemia
- Diet/management
 - Leucine-restricted diet
 - Importance of medical formula
 - Carnitine and glycine 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

