

Isovaleric Acidemia Ages 6-12 months

Clinic Visits:

- Every 3 months

Laboratory studies:

- Monitoring of diet
 - Plasma amino acid levels at least monthly
- Yearly
 - Comprehensive metabolic panel and phosphorous
 - Amylase and lipase (risk of pancreatitis)
 - Carnitine levels
 - Prealbumin/albumin
 - Ferritin, transferrin, or iron studies
 - Possible CBC, hemoglobin, and hematocrit
 - Possible folate and vitamin B12
 - Possible nutritional testing: zinc, selenium, vitamin D, essential fatty acids, and lipid profile

During illness:

- Call Metabolic Clinic to alert them immediately if your child is sick
 - Poor feeding, fever, vomiting, 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, 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 child
- If possible, glycine should continue to be given orally
- Please alert the Metabolic Clinic if your child needs surgery
- Consider Medic Alert bracelet

Evaluations:

- Yearly developmental questionnaires to be completed by parents

Topics:

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