

Argininemia

1-18 years

Clinic Visits:

- 12-24 months - every 3 months
- 2-18 years- every 6 months

Laboratory studies:

- Monitoring of diet
 - 1-6 years – plasma amino acid levels at least every 3 months
 - 6-18 years – plasma amino acid levels at least every 6 months
- Clinic visits
 - Possible ammonia, orotic acid, and prealbumin
- Yearly
 - Comprehensive metabolic panel
 - Prealbumin/albumin
 - Ferritin, transferrin, or iron studies
 - Possible ammonia
 - Possible orotic acid
 - Possible CBC, hemoglobin, and hemacrit
 - Possible folate and vitamin B12
 - Possible nutritional labs: zinc, selenium, vitamin D, essential fatty acids, and lipid profile

During illness:

- Call Metabolic Clinic to alert them immediately when 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, and plasma amino acids
- Medication or dialysis may be used to lower your child's ammonia levels
- Please alert the metabolic clinic if your child needs surgery
- Wear 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
- Psychiatric screening at 18 years
- DEXA scan (bone health) at 9 and 18 years
- Possible Head CT scan or brain MRI if there are concerns of neurological problems
- Possible neurology evaluation as needed

- Possible Liver or Renal evaluation as needed

Topics:

- Review biochemistry and genetics of argininemia
- Complications
 - Elevated ammonia levels
 - Spasticity
- Diet/management
 - Low protein diet
 - Importance of medical formula
 - Medications to control ammonia levels
 - Possible liver transplant
 - 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