

# Argininemia

## Ages 0-6 months

### Clinic Visits:

- Every 2 months

### Laboratory studies:

- At diagnosis
  - Plasma amino acids
  - Ammonia
  - Possible urine organic acids
  - Possible arginase enzyme assay
  - Possible DNA analysis
- Monitoring of diet
  - Plasma amino acid levels at least every 2 weeks
  - Targeted arginine <300 umol/L
- Clinic visits
  - Possible ammonia, orotic acid, and prealbumin

### During illness:

- Call Metabolic Clinic to alert them immediately if your baby is sick
  - Poor feeding, fever, vomiting, and/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, ammonia and plasma amino acids
- Medication or dialysis may be used to lower your baby's ammonia levels
- Please alert the metabolic clinic if your baby needs surgery

### Evaluations:

- Head CT scan or brain MRI at time of diagnosis if there are concerns of neurological problems
- Possible referral to Liver, Renal (kidney), or neurology if needed

### Topics:

- 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

