

Citrullinemia

>18 years

Clinic Visits:

- At least yearly

Laboratory studies:

- Monitoring of diet
 - o plasma amino acid levels at least yearly
- Clinic visits
 - o Comprehensive metabolic panel and liver function test
 - o Possible ammonia, PT, and prealbumin/albumin
- Yearly
 - o Comprehensive metabolic panel with liver function tests
 - o PT/PTT (clotting)
 - o Prealbumin/albumin
 - o Plasma ferritin, transferrin, or iron studies
 - o Possible ammonia
 - o Possible CBC, hemoglobin, and hemacrit
 - o Possible folate and vitamin B12
 - o Possible nutritional labs: zinc, selenium, vitamin D, essential fatty acids, and lipid profile

During illness:

- Call Metabolic Clinic to alert them immediately when you are sick
 - o Poor feeding, fever, vomiting, and/or lethargy
- Give yourself glucose gel or polymer solution if needed (or instructed to do so)
- If you are not able to keep down glucose containing fluids, you will need to go 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 and ammonia
- Medication or dialysis may be used to lower your ammonia levels
- Please alert the Metabolic Clinic if you need surgery
- Wear Medic Alert Bracelet!!!**

Evaluations:

- Psychiatric screening at 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 CIT
- Complications
 - o Elevated ammonia levels

- 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 you have 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