

GUIDELINES FOR PROPIONIC ACIDEMIA AND METHYLMALONIC ACIDEMIA PATIENTS UNDERGOING LIVER TRANSPLANT

1. Please order Ammonia and BMP. Ask about PO intake, ongoing illness, vomiting, diarrhea
2. Consult Medical Genetics: The genetics team will evaluate for signs and symptoms of an acute decompensation. Genetics fellow will need to check in with the Research coordinators as soon as the patient is admitted to see if the patient is enrolled in any studies requiring more lab work. Genetics will need to inform metabolic nutritionist
3. **On admission:**
 - a. Labs:
 - i. Ammonia and BMP (see above)
 - ii. Carnitine
 - iii. Acylcarnitine profile
 - iv. **STAT** MMA level, CALL LAB (for patients with MMA only)
 - b. Continue Carnitine at home dose (same dose can be given IV)
 - c. IV fluids with D10 at 1.5 times maintenance plus appropriate electrolytes for age
 - d. Insulin if needed for hyperglycemia per primary team (target glucose within normal limits)
 - e. Intralipids 2gm/kg/day
 - f. **ECHO functional only**
4. **Intra-Operative:** Target glucose within normal limits
 - a. Maintenance fluid should contain dextrose (start with D10) – may need to titrate glucose to D5 after intra-operative steroid bolus
 - b. Insulin infusion if needed for hyperglycemia
 - c. Continue pre-operative Intralipids 2gm/kg/day as needed
 - d. Avoid acidosis – correct with sodium bicarbonate or THAM
5. **Post transplant:**
 - a. Labs:
 - i. Urine organic acids **once** post transplant
 - ii. MMA level once weekly
 - iii. Acylcarnitine profile once weekly
 - iv. Plasma amino acids twice weekly
 - v. Carnitine level prior to discharge
 - b. Continue carnitine IV but transition to PO (1:1 conversion)
 - c. Continue Intralipids 2gm/kg/day
 - d. IV fluids D10 at 1.5 times maintenance plus appropriate electrolytes for age initially plus start TPN ASA
 - e. Protein guidelines:
 - i. POD 1: 0.5 gm/kg/d in TPN

- ii. POD 2: 0.8 gm/kg/d in TPN
 - iii. POD 3: 1 gm/kg/d in TPN
- f. Insulin if needed for hyperglycemia per primary team
- g. Start oral feeds when able with propimex and advance according to nutrition (Nutritionist Nancy Baugh). Goal is to maintain on same diet as pre-transplant
- h. Discharge with oral carnitine
- i. Follow up in Biochemical Genetic Clinic 3 months after discharge.
Liberalization of diet will be done on an outpatient basis.

GUIDELINES FOR UREAC CYCLE DISORDER PATIENTS UNDERGOING LIVER TRANSPLANT

Urea cycle disorders:

1. Ornithine transcarbamylase deficiency (OTC). Most common

2. Carbamoyl phosphate synthase 1 (CPS1) deficiency
3. N-acetyl glutamate deficiency (NAGS): VERY Rare. Not curative
3. Citrullinemia (Argininosuccinate synthase deficiency: (ASS))
4. Argininosuccinic aciduria (ASL)
5. Argininemia (ARG1).
6. HHH syndrome (hyperammonemia, hyperornithinemia, homocitrullinuria).

1. Please order Ammonia and BMP. Ask about PO intake, ongoing illness, vomiting, diarrhea
2. Consult Medical Genetics: Review history of hyperammonemia. The genetics team will evaluate for signs and symptoms of an acute decompensation. Genetics fellow will need to check in with the Research coordinators as soon as patient is admitted to see if patient is enrolled in any studies requiring more lab work.
3. **On admission:**
 - a. Labs: Ammonia. No need to follow in the OR
 - b. IV fluids with D10 at 1 to 1.5 times maintenance plus appropriate electrolytes for age
 - c. Insulin if needed for hyperglycemia per primary team (target glucose within normal limits)
 - d. Intralipids 2gm/kg/day
 - e. *Consider ammonul only if unclear how long the waiting time is for transplant. However it needs to be stopped prior to surgery. This is to be decided on a case-by-case basis.*
4. **Post transplant:**
 - a. Proceed with fluids and diet as you would for any other liver transplant
 - b. Discharge on L- citrulline or L- arginine for OTC, CPS1*
 - c. Discharge ASL and Citrullinemia (ASS) on L-arginine
 - d. Discontinue all nitrogen scavengers: Sodium benzoate and sodium phenylbuterate
 - e. Follow up in Biochemical Genetic Clinic 3 months after discharge.

*Dosages (please refer to dosages prior to transplantation. If not available see below):

Citrulline: Peds: CPS and OCT: 100-200 mg/Kg/day in 3 to 5 doses. Adult: >20 Kg 3.8 g/m2/day in 3 to 5 doses. Max 6g/day

L-arginine Pediatric: 100-200 mg/Kg/day (OTC and CPS); mg/Kg/day on ASL and ASS. Adult: 8.8-15.4 g/m2/day. Max 6g/day