Metabolic Transplant Guidelines
Maple Syrup Urine Disease (MSUD)

PRE-TRANSPLANT:

- ☐ Contact Children's National Genetics: call **202-476-5000** then pager number **50009**and provide call back number to inform of the upcoming transplantation
- ☐ Start peripheral IV STAT upon arrival
- ☐ Start D10 NS with 20 mEq KCl at 1.5x maintenance = _____ mL/hr
- ☐ Start Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ Continue home diet of low protein food and metabolic formula for as long as acceptable
- ☐ Continue home enteral isoleucine and valine doses (often mixed into metabolic formula)
- ☐ NPO for minimum acceptable period prior to surgery; no longer than 24 hours. If longer, must start protein containing TPN.
- ☐ LABS: Plasma amino acids (to be sent STAT to Children's National)

DURING TRANSPLANT:

- ☐ Continue D10 NS with 20 mEq KCL at 1.5x maintenance = _____ mL/hr
- ☐ Continue Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ Check baseline ammonia and one after reperfusion. Additional ammonia levels should be drawn for unexplained metabolic acidosis. BS should be managed with insulin boluses. Do not decrease D10 and IL rate of infusion.

POST-TRANSPLANT:

- ☐ Contact Children's National Genetics: call **202-476-5000** then pager number **50009**and provide call back number to inform them patient is out of surgery.
- ☐ Discontinue low protein diet and formula
- ☐ Discontinue isoleucine and valine
- ☐ Initiate routine TPN, including FULL routine protein needs, until gut is cleared for solids
- ☐ Once full enteral diet is possible initiate normal diet with no protein restriction
- ☐ LABS: Plasma amino acids (to be sent STAT to Children's National) within 12-24 hours post-op. Continue to send plasma amino acids every other day up to 7 days post-op and weekly till one mo
- ☐ Contact Children's National Genetics: call **202-476-5000** then pager number **50009**and provide call back number to inform them when patient is discharged.

LONG TERM FOLLOW UP:

- ☐ Pending revision

For questions call: call 202-476-5000 then pager number 50009and provide call back number

Metabolic Liver Transplant Guidelines
Propionate Pathway disorders

Indicate diagnosis:

- ☐ Propionic acidemia
- ☐ Methylmaonic aciduria B12 responsive (MMA-B12)
- ☐ Methylmalonic aciduria (MMA)

PRE-TRANSPLANT:

- ☐ Contact Children's National Genetics: call **202-476-5000** then pager number **50009**and provide call back number to inform of the upcoming transplantation
- ☐ Start peripheral IV STAT upon arrival
- ☐ D10 ½ NS with 20 mEq KCL at 1.5x maintenance = _____ mL/hr (KCL to be added if not in renal failure)
- ☐ Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ Continue home diet of low protein food and metabolic formula for as long as acceptable
- ☐ NPO for minimum acceptable period prior to surgery; No longer than 24 hours, If longer must started protein containing TPN.
- ☐ **LABS:** Baseline ammonia, lactate, and plasma amino acids (to be sent STAT to Children's National)
- ☐ If MMA and responds to vitamin B12, then 1 mg IM/SQ hydroxycobalamin each day (including after transplant).

DURING TRANSPLANT:

- ☐ Continue D10 ½ NS with 20 mEq KCL at 1.5x maintenance = _____ mL/hr
- ☐ Continue Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ For propionic pathway disorders, check ammonia levels q 1 hr.. BS should be managed with insulin boluses. Do not decrease D10 and IL rate of infusion.

POST-TRANSPLANT:

- ☐ Contact Children's National Genetics: call **202-476-5000,** then pager number **50009**and provide call back number to inform them when patient is out of surgery.
- ☐ Initiate routine TPN, including protein to start at 1 g/kg/day and will be adjusted by Children's National Genetics, until gut is cleared for solids
- ☐ Diet will be determined by Children's National
- ☐ **LABS:**
  - Ammonia every 12 hours for first 24 hours post-op then discontinue if normal
    - If ammonia is > 100 umol/L, contact Children's National Genetics who will give recommendations for ammonia scavengers or calories (call 202-476-5000, and ask for the Geneticist on-call)
  - Plasma amino acids (to be sent STAT to Children's National) within12-24 hours post-op. Continue to send plasma amino acids every other day up to 7 days post-op
  - Lactate at 12 hours post op and daily. Contact if >3mmol/L
- ☐ Contact Children's National Genetics: call **202-476-5000,** then pager number **50009**and provide call back number to inform them when patient is discharged.

LONG TERM FOLLOW UP:

- ☐ *PENDING REVISION*

For questions call: call 202-476-5000 then pager number 50009and provide call back number

Metabolic Transplant Guidelines
Urea Cycle Disorders

Indicate diagnosis:

- ☐ CPS1 deficiency
- ☐ OTC deficiency
- ☐ HHH deficiency
- ☐ Citrullinemia
- ☐ ASL deficiency
- ☐ Arginase deficiency

PRE-TRANSPLANT:

- ☐ Contact Children's National Genetics: call **202-476-5000** then pager number **50009**and provide call back number to inform of the upcoming transplantation
- ☐ Start peripheral IV STAT upon arrival
- ☐ D10 ½ NS with 20 mEq KCL at 1.5x maintenance = _____ mL/hr
- ☐ Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ Continue home diet of low protein food and metabolic formula for as long as acceptable
- ☐ NPO for minimum acceptable period prior to surgery
- ☐ Ammonul (prefer central access)
  - Pharmacy to dilute with D10W to final concentration 10 mg/mL
  - ↑ Patient weight 0-25kg: 250mg/kg/day = 25 mL/kg/day (after dilution) = ____ mL/day
  - ↑ Patient weight > 25kg: 5500 mg/m2 = 55 mL/m2/day (after dilution) = _____ mL/day
  - Infuse calculated dose continuously over 24 hours = _____ mL/hour
  - STOP ammonul infusion at START of transplant procedure
- ☐ Arginine infusion (central line only) *Do NOT give arginine to patients with Arginase deficiency*
  - Infuse continuously over 24 hours.
  - For OTC and CPS1 deficiency:
    - ↑ Patient weight 0-25kg: 200 mg/kg/day = _____ mg/day
    - ↑ Patient weight > 25 kg: 4000 mg/m2/day = _____ mg/day
  - For citrullinemia and argininosuccinic aciduria:
    - ↑ Patient weight 0-25kg: 600 mg/kg/day = _____ mg/day
    - ↑ Patient weight > 25 kg: 12000 mg/m2/day = _____ mg/day
- ☐ **LABS:** Baseline ammonia and plasma amino acids (to be sent STAT to Children's National)

DURING TRANSPLANT:

- ☐ STOP ammonul infusion at liver perfusion
- ☐ Continue D10 ½ NS with 20 mEq KCL at 1.5x maintenance = _____ mL/hr
- ☐ Continue Intralipids (20%) 2g/kg/day = 15 mL/kg/day = _____ mL/hr
- ☐ Continue arginine infusion
- ☐ For urea cycle defects, check ammonia levels q 1 hr.. BS should be managed with insulin boluses. Do not decrease D10 and IL rate of infusion.