Diagnosis: Ornithine Transcarbamylase (OTC) Deficiency

(Urea Cycle Defect)

EMERGENCY ROOM MANAGEMENT

for Intercurrent Illness

1. Baby should be triaged as soon as possible upon arrival in the Emergency Room even if he/she does not appear to be ill, because metabolic decompensation can occur very rapidly.
2. **PLEASE NOTIFY METABOLISM SERVICE** at The Children’s Hospital of Sometown **(215) 555-5555** [Ask Page Operator to page Metabolism Fellow On-Call].
3. **COMMON ACUTE COMPLICATIONS:**
* Hyperammonemia, Seizures, Cerebral Edema, Coma

**4. CONTRAINDICATED MEDICATIONS:**

* Systemic Steroids-- ***Unless otherwise specified by Metabolism staff***
* THAM (Tris hydroxymethyl aminomethane)

**5. LABORATORY EVALUATION [STAT]**

* Ammonia, Venous Blood Gas, Comprehensive Metabolic Panel, Bicarbonate
* CBC/differential, PT, PTT, LFTs
* Plasma Amino acids [3 ml, green top tube, sodium heparin]. Send to Metabolism Lab

**6. MANAGEMENT:**

* **STAT:**
* Place Peripheral IV. If unable to get venous access, place nasogastric tube.
* **Bolus:**
* 10-20 cc/kg of Normal Saline bolus, if indicated for dehydration.
* **Continuous IV Fluids**: D 10% with 0.45 NS

**Rate:** x1.5 maintenance **[IF NO INCREASED INTRACRANIAL PRESSURE].**

Please discuss further management with Metabolism Team

**Metabolism Fellow:**

#55555

**Office (weekdays):**

(215) 555-5555

**Nights/Weekends:**

(215) 555-5000 [Operator to page the Metabolism Fellow On-call]