ACUTE MANAGEMENT OF PROPIONIC ACIDEMIA (PA)

- Such patients easily and frequently decompensate with minor infections, poor oral intake, vomiting or constipation. The patient should be admitted immediately to ER and managed as triage level II.
- Immediate actions which should be accomplished within 1 hour of arrival:
- Basic life support
- Stop all source of protein central and parenteral nutrition.
- Check GlucoChecks.
- Insert an IV line and take blood for blood gases, chem 1, Ammonia (NH_3), & CBC, liver enzymes, lactic acid and blood C/S (peripheral and central if patient has central line) as **STAT** order.. Other labs as needed.
- Ammonia blood sample should be taken with precaution because of high false positive rate (without tourniquet, in green-top tube, put on ice to the laboratory, separated within 20 minutes of collection and analyzed immediately).
- After taking the blood give Carbaglu® 100-250mg/kg by NGT or oral then continue on 100-250mg/kg/day q6.
- High caloric intake is the main stay of therapy. Therefore, Start 1 1/2 to double maintenance I.V.F as D10 1/2NS + Kcl 30meq/l. Re-adjust according to lab results (Keep GlucoChecks 5-8mmol/L). Consider start **<u>insulin</u>** if hyperglycemia develop at dose **of 0.01-0.05 unit/kg/hour** and titrate up until blood glucose controlled.
- Call the pharmacy hotline# to expedite the delivery of medications .
- If ammonia > 100 umol/l. Start intralipid 20% at 2-3 g/kg/day to provide additional calories.
- **<u>DO NOT DECREASE DEXTROSE RATE</u>** or amount and **<u>DO NOT STOP</u>** calorie delivery in the acute stage for any reason (e.g. medications, addition required fluid bolus, or hyperglycemia) as this can precipitate hypoglycemia and catabolism which will further worsen the patient's condition.
- If Ammonia elevated >100 umol/l and Carbaglu not available:
  - If the patient has central line give ammonul® 250mg/kg IV over 90 minute as loading dose then maintenance dose 250mg/kg/day over 24 hours.
  - If there is no central line gives the patient sodium benzoate 250mg/kg PO/NGT as loading dose then maintenance dose 250mg/kg/day over 24 hours.
- Call pharmacy to expedite the intralipid20% and medications.
- Call metabolic genetics dietitian on-call.
- If ammonia < 100umol/l start 50% natural protein and propiomex formula.
- Give polycose or prophree PO/NGT as tolerated.
- According to clinical evaluation, empirical antibiotics may be started.
- In case of refractory acidosis, give bolus of $NaHCO_3$ and start $NaHCO_3$ infusion as 0.5-2 meq/kg/h.(5)
- Continue on metronidazole at 10–20 mg/kg per day divided Q8.
- Increase Carnitine dose to 300-400 mg/kg/day divided Q8 hours IV, orally or NGT.
- For Nausea and vomiting give serotonin receptor-blocking agent Granisetron 10 to 40 microgram/kg, infused over 3 to 5 minutes.
- Continue on same Biotin dose (if patient still on it)
- For constipation give glycerin suppositories, Dulcolax or Docusate by NGT.
- **Consider dialysis:**
  - Hyperammonemic coma.
  - Dilated pupils.
  - Failure to improve or deterioration within 4 hours of initial treatment
  - Ammonia more than 500micromol/l.
  - Extreme acidosis or electrolytes imbalances.
- Ammonia, electrolyte and blood gases need to be followed at regular intervals during this acceleration of management stage. The frequency is dictated by the patient's condition and the speed at which results can be obtained.

- **<u>DO NOT STOP</u>** other oral chronic medications.
- Proteins should be reintroduce within 24 hours-36 hours of initiation of therapy even if the patients on dialysis.
- Call Biochemical Geneticist (metabolic) on call Tel: 018011111

<u>For more information please read the attached guidelines for this disorder.</u>