

## **ACUTE MANAGEMENT OF MEDIUM CHAIN ACYL CO-A DEHYDROGENASE DEFICIENCY (MCAD)**

- 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
- Check GlucoChecks.
- Insert an IV line and take blood for blood gases, chem 1, Ammonia (NH<sub>3</sub>), lactic acid & CBC, blood C/S (if patient febrile), liver transaminase, CK level and acylcarnitine profile.
- If hypoglycemic <3.5 give 3 ml/kg of D10
- If normoglycemic but patient cannot tolerate orally or if patient has diarrhoea, vomiting, dehydration, acidosis, high fever start 1 1/2 I.V.F as D10 1/2NS +KCl 20meq/l. Re-adjust according to lab results.
- Until the intravenous line is started, it is imperative to provide if possible / safe source of glucose like polycose solution
- Give carnitine 50mg/kg/dose IV then continue same dose divided q6.
- **DO NOT** administer lipids or acetylsalicylic acid.

**For more information please read the attached guidelines for this disorder.**