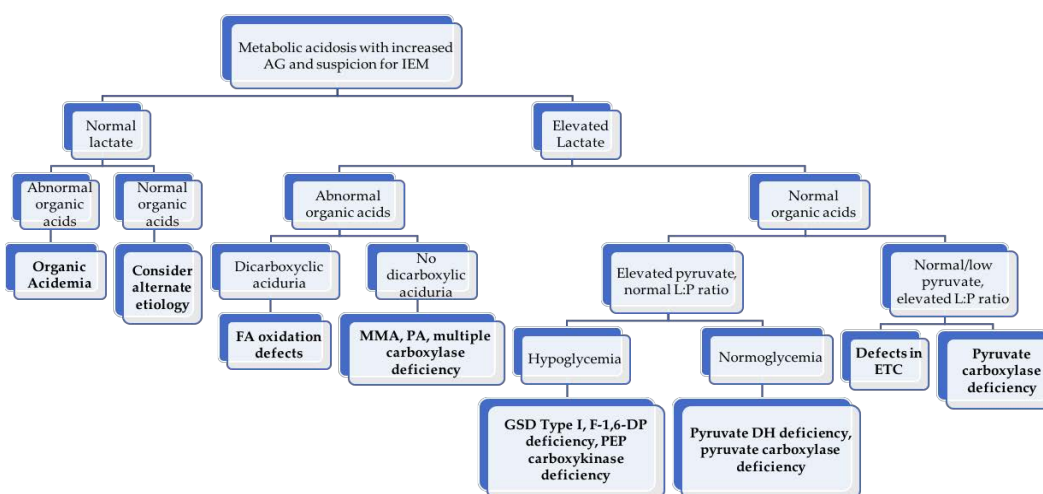


## Management of Metabolic Crises

### Metabolic Acidosis (when due to IEM)

|                         |   |
|-------------------------|---|
| <b>PowerPlan</b>        | Metabolism Lactic or Metabolic Acidosis NOS Admit Plan  |
| <b>Definition</b>       | Arterial blood gas with pH < 7.35, pCO <sub>2</sub> < 35, bicarbonate < 22  |
| <b>Etiopathogenesis</b> | Inherited: organic acidurias, primary lactic acidemias, renal tubular acidosis; <b>ANY</b> metabolic crisis, if left untreated long enough, will progress to metabolic acidosis   |
| <b>Presentation</b>     | Acute vomiting, dehydration, lethargy, and rapid, shallow breathing, often h/o protein load   |
| <b>Physical Exam</b>    | <b>Organic acidurias:</b> limb hypertonia/axial hypotonia, large amplitude tremor, myoclonic jerks, pedaling, sustained paraspinal contraction (opisthotonic posturing)<br><b>RTA:</b> Failure to thrive, polyuria, and rachitic changes<br><b>PDH deficiency:</b> blindness, hypotonia, DD, narrow forehead, frontal bossing, wide nasal bridge, long philtrum, and anteverted nostrils  |
| <b>Treatment</b>        | Hydration, caloric intake of 120-140kcal/kg/day, stop proteins initially (esp stop all BCAAs if MSUD is suspected), maintain glucose 100-150 (using high GIR +/- insulin), avoid hypoNa, cerebral edema<br>If serum bicarb < 14 meq/L and pH < 7.2, give IV bolus NaHCO <sub>3</sub> as 2.5 meq/kg over 30 minutes, then 2.5 meq/kg/day until serum bicarbonate is 24-28 meq/L<br>HD = last resort but may be lifesaving in severe refractory cases (especially neonates) |



### Seizures (when due to IEM)

|                  |  |
|------------------|--|
| <b>Etiology</b>  | Alteration of intracellular <b>osmolality</b> , depletion of substrates needed for <b>cellular metabolism</b> or <b>membrane function</b> , and/or intracellular accumulation of <b>toxic substances</b>   |
| <b>DDx</b>       | DDx of 'seizures in a newborn' is large, including many IEMs with poor prognosis. <b>Rare but potentially treatable etiologies:</b> <b>pyridoxine responsive</b> seizures, <b>folinic acid responsive</b> seizures, <b>serine responsive</b> 3-phosphoglycerate DH deficiency, sz from <b>hypoglycemia</b> , <b>biotin responsive</b> holocarboxylase synthetase deficiency, biotinidase deficiency.                   |
| <b>Treatment</b> | <b>See neurology section for treatment of status epilepticus;</b> avoid AEDs that block mitochondrial fxn (VPA, chloral hydrate) - c/s fosphenytoin, BZDs, and/or levetiracetam. Correct fever, electrolyte issues, acidosis, hypoglycemia. If refractory, c/s empiric pyridoxine (100-200 mg IV x1), folinic acid (2.5-5 mg PO once daily), L-serine (200-600 mg/kg/d div 6x/day), or biotin (5-20 mg PO once daily). |