

TREATMENT OF ORGANIC ACIDEMIA

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Immediate Management

Infants and children with acute metabolic crisis require immediate treatment to prevent further acute deterioration and long-term sequelae. Early and appropriate treatment before confirmation of the diagnosis is life-saving.

Eliminate intake of the precursors of possibly toxic metabolites.

This applies most often to suspected inborn errors of amino acid or organic acid metabolism. In both cases, dietary or parenteral intake of protein and amino acids should be eliminated immediately an organic acidemia is suspected.

Administer glucose a simple source of calories at least 8 mg / kg / min to suppress mobilization of endogenous sources of the metabolites.

This is achieved by the intravenous administration of 10% dextrose supplemented by Intralipid.

Specific Therapy/disease

NPO: Minimize intake and endogenous production of toxic metabolites.

Hydration: Administer high-calorie, high-carbohydrate intravenous fluids: 10% dextrose in 0.2% NaCl at 1.5 times calculated maintenance, and add KCl .

Alkali Therapy: Bicarbonate is generally not indicated unless the plasma bicarbonate is <10 mmol/L; deficits should be only half corrected.

Hemo or peritoneal dialysis: If just described measures fail to induce clinical & biochemical improvement ,hemo or peritoneal dialysis is indicated to Accelerate elimination of toxic metabolites.

If hyperammonemia exists:

1.5 × Maintenance D/W 10 % & intravenous lipids 1 g/kg 24 h

Sodium benzoate 250 mg/ kg to be added to 20 cc/kg of 10% glucose & infused within 1-2 h (priming dose)

Continue infusion of sodium benzoate 250-500 mg / kg / 24 h following the above priming dose & should be added to daily intravenous fluids

Initiate peritoneal dialysis or hemodialysis :if above treatment fails to produce an appropriate decrease in plasma ammonia.

Peritoneal dialysis is too slow. Exchange transfusion brings the plasma ammonium down quickly, but rebound hyperammonemia occurs just as quickly.

It may be useful in some circumstances as an adjunct to hemodialysis.

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Cofactors Therapy

Administration of cofactors may be indicated in organic acidemia :

Biotin 10 mg / day

Carnitine 100 / mg / kg / day in three divided doses IV or orally

Cobalamin (vitamin B12) 1 mg SC or IM

B6 100 mg IV or Activated B6 10 mg/kg IV, should be given to neonates with seizures unresponsive to conventional anticonvulsants; if there is no response to B6 and Activated B6 ,folinic acid , should be administered for possible folinic acid responsive seizures.

Keywords: Treatments; organic acidemia; Children.