P-181 - INTERVENTION AND MEDICAL AND NUTRITIONAL TREATMENT OF A NEWBORN WITH HYPERAMMONEMIA. A CASE REPORT.

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INTRODUCTION: In Colombia, patients with Inborn Errors of Metabolism (IEM) are intervened after presenting clinical manifestations of the disease; most of them are detected at a later stage due to the absence of a neonatal screening program. The neonatal manifestation of the disease that leads to suspect an IEM is the non-improvement of its clinical status together with the limited diagnostic methods in health institutions, thus the quantification of ammonia could be a fundamental tool to direct the initial and subsequent intervention diagnosis of these pathologies. 26 years old mother infant, not consanguine parents, received exclusively breastfeeding, on the fourth day shows oral rejection, vomiting, drowsiness and jaundice. Emergencies is contacted, shows distal coldness, drowsiness and ventilatory failure; ventilatory support and antibiotic therapy is initiated. Presents altered state of consciousness, bilirubin encephalopathy is suspected. The child presented convulsive events, suspending phenobarbital and phototherapy. Transferred; without sedation, no response to tactile or pain stimuli. No respiratory effort and is supported on dopamine. On clinical examination: icteric, mild hepatomegaly, isochoric pupils, transaminase and elevated nitrogen compounds, leucocytosis, thrombocytosis, severe hyperammonemia with ammonium of 1261 mmol/L. Due to the metabolic state and lack of dialysis in the institution, it was decided to perform exchange transfusion, dextrose fluids, fasting. Post-exchange ammonium of 158 and 315 after 24 hours. Sodium benzoate, sodium phenylbutyrate, lactulose and metronidazole decreased ammonium progressively in 72 hours (287-60). Nitrogen compounds in improvement and normalization of liver function. Finding cerebral edema in NMR, dynamic management of Nutrition, Parenteral Nutrition (PN) with dextrose and lipids, full contributions and enteral contribution of initial protein 0.25 g/k/d Increments of 0.25-0.5 g/k/d. Improvement of neurological response Scheduled extubation is achieved. URCmed A, weaning of PN is started. Serum ammonium monitoring, achieving stability with protein intake 1.6 g/k/d and weight gain. Metabolic studies report amino acid chromatography: Lysine elevation, normal pyruvate lactate ratio. Organic acids not conclusive. CONCLUSION: Stabilization of the patient with exchange transfusion was achieved, although it is not recommended. In many of our regions where dialysis is not available, it could be an option for the treatment of metabolic emergencies.