P-121 - HYPERAMONEMIA AND PROPIONIC ACIDEMIA: CARGLUMIC ACID AND TIME OF AMMONIA REDUCTION DURING METABOLIC DECOMPENSATION.

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**INTRODUCTION:** Propionic acidemia (PA), caused by propionyl-CoA carboxylase deficiency, is responsible for the catabolism of isoleucine, threonine, methionine and valine, odd-chain fatty acids, and the three-carbon side chain of cholesterol. As a result, PA patients have several disruptions in different metabolic pathways including the TCA cycle, urea cycle, and glycine synthase complex, resulting in lactic acidosis, hyperammonemia, and ketotic hyperglycinemia. Hyperammonemia in PA is explained by several complex mechanisms. Propionyl-CoA competitively inhibits N-acetyl glutamate synthase, the allosteric activator of carbamoyl phosphate synthase, the rate-limiting enzyme of urea cycle, resulting in hyperammonemia. High ammonium values can cause irreversible damage. Carglumic acid is a synthetic analog of N-acetyl glutamate, furthermore stimulates CPS 1.

**MATERIALS:** A descriptive retrospective study of metabolic decompensation with hyperammonemia in patients with a diagnosis of PA together with an analysis of the pharmacological treatment used between 2014-2018 were performed. The medical records of the hospital were reviewed including the biochemical data and time lapse of ammonia exposure.

**OBJECTIVES:** To evaluate the decrease in ammonia values; with respect to time and total value. To analyse if the use of this treatment reduces the time of exposures to high ammonium values.

**DESCRIPTION:** This medication was used in 4 patients. During decompensation, the caloric was increased, the protein intake was decreased. The intravenous dose was given at 200mg/kg/d carnitine. A total of 15 decompensations were found, 12/14 used carglumic acid and in 2/14 only sodium benzoate was used. The average age was 37.4 months (range 19-65). The average ammonium on admission was 174μmol/L (range 364-156μmol/L). During hospitalization, where sodium benzoate was used as an ammonium scavenger, the drop in time was 43-181h (mean 91h). With the use of carglumic (100mg/kg/day), the average drop in time was between 11-40h with an average of 28.5h.

**CONCLUSION:** Consistent with the literature, it is evident that carglumic acid has greater effectiveness in terms of ammonium reduction and the time in which this reduction occurs. It is emphasized that the treatment of decompensation with high caloric intake and low protein intake is essential, as well as treating the decompensation trigger.