P-013 - LIVER TRANSPLANT IN PEDIATRIC PATIENTS WITH INBORN ERRORS OF METABOLISM: IMPACT ON NUTRITIONAL TREATMENT

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i. Liver transplant (LT) is a therapeutic alternative for different inborn errors of intermediate metabolism (IEM), whose treatment is based on specific dietary restrictions and supplementation with medical foods. This therapy can significantly improve the quality of life of those patients who suffer severe manifestations of the disease or metabolic decompensations that threaten life despite adequate medical and nutritional treatment. ii. To describe the nutritional treatment, feeding route and growth of pediatric patients with IEM pre and post liver transplantation. iii. A retrospective review of 9 pediatric IEM patients with specific nutritional therapy who underwent liver transplant in Garrahan Hospital. They were divided into 2 groups according to the type of pathology. 5 patients had IEM of protein/amino acid metabolism (UCD, TYR type 1, MSUD, MMA) and 4 with Glycogen Storage Disease type 1 (GSD1). Anthropometry (z score W/Y, H/Y and BMI/Y), dietary intake, type of diet and feeding route were analyzed. iv. 9 children (2 male/7 female) were transplanted at median age 7.6 (1.8-13.6). Group 1: 60% (n=3) of pre-transplant patients had normal BMI and 40% were overweight (n=1) and obese (n=1). Post-transplant, last two patients improved their nutritional status. Pre transplant, all of them were treated with medical food and protein restricted diet according to each disease. 40% needed enteral nutrition support complement to the oral intake, 20% required same and 40% fed only orally. Post-transplant, 4/5 gradually released the protein-restricted diet. Only 1 case who received a combined liver kidney transplant should continue with slight protein restriction. Group 2: 75% (n=3) of patients presented normal BMI and 25% obesity (n=1), after BMI was normalized. All of them were treated with uncooked cornstarch therapy tolerating up 3 hours of fasting. All patients required feeding tube and fructose and lactose free diet. Post-trasplant, all the children were released from their diet and tolerated prolonged fasting. Only 1 patient required protein control due to renal failure. v. LT has been a successful therapeutic approach for our population that prevents metabolic decompensation, dietary restrictions and the improvement of quality of life, achieving adequate growth.