

O-009 - PEDIATRIC LIVER TRANSPLANTATION FOR INBORN ERRORS OF METABOLISM

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The aims of this article were to report a single-center experience of pediatric liver transplantation for Inborn Errors of Metabolism (IEM) **MATERIALS AND METHODS:** The medical records of 40 patients younger than 18 years undergoing liver transplantation (LT) from March 1993 to December 2018 at Juan P. Garrahan Pediatric Hospital of Buenos Aires where 811 (LT) were performed, there were reviewed. The variables measures were: Patient demographics, indications, cirrhotics or non cirrhotics, graft type, survival, postoperative complications, growth and schooling. **RESULTS:** 40 patients LT were performed (22 female, 18 male) at a median age of 57 months (range, 1- 217 months). IEM was 5% of the LT in 25 years. The median time follow-up was 3.1 years (range, 0.01- 22.7 yr). LT indication were: cirrhosis complications in 21, quality of life in 10 and 9 acute liver failure. 15 were prioritized to access LT. IEM were 25 cirrhotics and 15 non cirrhotics in the explant. The cumulative 1, 5, and 10-year patient and graft survival rates were 74.4%, 74.4% and 68.6% and 74.4%, 74.4% and 68.6% respectively. For non cirrhotic vs cirrhotic disorders cumulative 1, 5, and 10-year patient survival rates were 80% vs 70.6%, 80% vs 70.6% and 80% vs 64.1%. There were 16 whole grafts and 24 reduced. 1 combined liver-kidney transplantation was performed in methylmalonic acidemia. Biliary and vascular complications were 22% and 20% respectively and one retransplants were done. Five had a liver tumor: 2 adenomas and 3 hepatocellular carcinoma. 55% had acute cellular rejections and 9% developed chronic rejection. Four Post-transplant lymphoproliferative diseases were developed. 52% had growth failure and improved by 37% after LT and 43% had education according to their age. **CONCLUSION:** Liver transplantation is an alternative therapy that improves the survival, growth and quality of life of a group of IEM with different clinical manifestations. The risk assessment and the LT benefit should be comprehensively addressed by an interdisciplinary team before the LT indication and careful monitoring and specific management of the disease is necessary even after LT.