P-008 - LONG TERM FOLLOW UP OF LIVER TRANSPLANTATION IN PATIENTS WITH GLYCOGEN STORAGE DISEASE TYPE I IN A TERTIARY HOSPITAL IN BUENOS AIRES, ARGENTINA

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INTRODUCTION: Glycogen storage disease type I (GSDI) is an inborn error of carbohydrate metabolism. GSDI affects liver and kidney. Additionally, GSDIb have impaired neutrophil function and inflammatory bowel disease. Treatment is based on avoiding hypoglycemia and its complications. OBJECTIVES: Descriptive analysis of GSDI 4 patients who underwent liver transplantation (LT) indications and outcome. PATIENTS AND METHODS: Retrospective cases were obtained from Garrahan records. From a total of 32 GSD, we analyzed 4 GSDI patients who underwent LT between 2006 and 2016. RESULTS: We described 4 GSD patients: 3 Ib and 1 la. GSD Ib have symptoms at an earlier age than la (7 months vs 16). All GSDIb had prior to LT neutropenia and inflammatory bowel disease. None had renal involvement. Median age at LT was 9.5 years (7-12 years). All received cadaveric liver transplant. Immunosuppressive regime consisted of steroids, tacrolimus, sirolimus, mycophenolate and cyclosporine. The anatomical pathology of liver explant revealed adenomas, steatosis, fibrosis and hepatocellular carcinoma (1). The indication for LT included hepatic adenomas (3) and poor metabolic control (3). Time follow-up was 5.5 years (2-12). Global survival was 100%. Metabolic control normalized in all patients and in those with growth-retardation, catch-up was achieved post transplant (3/3). Short-term complications (<1 year) were: hypertension (1), CMV infection (2) tacrolimus encephalopathy (1), acute liver rejection (2), acute renal insufficiency (1), vena cava thrombosis (1) and lymphoproliferative disorders (1). Long-term complications in GSDIa was obesity and microalbuminuria. In GSD Ib were proteinuria, nefrocalcinosis, microalbuminuria, thyroid carcinoma, neutropenia. Renal dysfunction developed after LT: microalbuminuria (2) and proteinuria (1). Persistent neutropenia was the most important complication (3/3). Two persisted with splenomegaly and thrombocytopenia. CONCLUSIONS: LT allows to correct intolerance to fasting, release the diet improving the quality of life and reduce the risk of malignancy. However LT is not an innocuous procedure, it can bring with it complications related to the procedure, use of immunosuppressive medication adding comorbidity. The benefit of LT depends on the extent of extrahepatic manifestations. Renal involvement is a documented complication in LT. The use of nephrotoxic medications has been proposed as one contributing factor added to the disease progression. It remains unclear why neutropenia improves after LT in some patients and persists in others.