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OUTCOMES FOLLOWING PARTIAL EXTERNAL BILIARY DIVERSION (PEBD) IN PATIENTS WITH PROGRESSIVE FAMILIAL INTRAHEPATIC CHOLESTASIS (PFIC)

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Aim: PFIC is a family of bile acid transport (disorders PFIC1, 2 and 3) that may result in serious liver disease requiring transplantation. We reviewed our experience with PEBD as a method to improve liver function, ameliorate symptoms and avoid transplantation.

Methods: All patients with PFIC were reviewed. Outcomes included changes in serum bile acids (BA), improvement in symptoms permitting conversion to ileal bypass and stoma takedown, and survival without transplantation. All results were quantified according to PFIC subtype. Statistics were obtained using paired t-test and Wilcoxon test. $p<0.05$ was considered statistically significant. IRB approval was obtained for the study.

Main Results: Thirty-five patients with PFIC were identified. Data is available in 24. Twenty-four children (12 males) underwent PEBD: 10 PFIC1, 13 PFIC2, and one PFIC3. BA levels decreased in PFIC1 patients from 1724.5±3215.3 to 11.5±6.2 um/L ($p<0.05$), but in PFIC2 patients, BA levels did not significantly decrease (192.9±99.2 pre diversion to 140.65±118.5 post diversion, $p=0.17$) (figure 1). In the single PFIC3 patient BA decreased from 821 to 11.2. Seven patients were converted from PEBD to ileal bypass: 2 PFIC2 patients were internalized for electrolyte imbalances from fluid loss, and went on to transplantation. However 5 PFIC patients were converted to ileal bypass because of life-style choice and improved symptoms. There were no significant changes in bile acid levels following conversion. Five year transplant free survival was 100% (11/11) in the PFIC1 and PFIC3 children, whereas 5/13 (38%) PFIC2 patients survived without a transplant ($p=0.002$, chi-square).

Conclusion: PEBD is an effective procedure to reduce total bile acid levels in PFIC patients and improve their symptoms. However, it appears to be less effective in PFIC2 patients. Those higher bile acid levels could contribute to ongoing damage to the liver, and contribute for the higher liver transplant rate in the PFIC2 group.