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Effects of partial internal biliary diversion on long-term outcomes in patients with progressive familial intrahepatic cholestasis : Experience in 44 patients

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Abstract

Purpose: Progressive familial intrahepatic cholestasis (PFIC) is a hereditary disease characterized by cholestasis, which may cause jaundice, severe pruritus, and cirrhosis in the later stages. By the invention of biliary diversion methods, these patients were prevented from undergoing liver transplant. Using biliary diversion techniques, the entero-hepatic cycle was interrupted. This lowers the bile acid pool and resolves the pruritus.

Herein, we report 44 cases of PFIC who underwent partial internal biliary diversion (PIBD) and long-term follow-up of these children. This comprises the largest case series of PIBD.

Methods: All patients were diagnosed by liver biopsy as PFIC before the operation. All underwent cholecystocolic bypass by jejunal interposition due to severe pruritus unresponsive to medication. Laboratory blood tests, sonography, and physical exam were done before and after the operation once every three months. Besides, a questionnaire was designed to ask the patients about the symptoms after the operation, and a pruritus score was measured using the 5D-itch scale.

Results: 44 children (25 boys, 19 girls), between 1.75 to 27.5 years (at the time of this study) were followed for a median period of 54 months. Age at operation ranged from 2 months to 18 years, with a median of 29 months. Of these children, 14 were lost to follow up. Results showed a significant decrease in pruritus and sleep disturbance after the surgery ($p < 0.001$). Also, jaundice decreased from 82.1 % before to 7.1% following the surgery. 50 % of the patients became medication-free at follow-up.

Conclusion: PIBD is a safe procedure which helps non-cirrhotic children preserve their liver function. Therefore PIBD prevents them from undergoing liver transplant. Effective results were achieved in terms of severe pruritus and jaundice, and children were able to regain their sleep patterns. It also avoided external stoma, which is more convenient from the patient's point of view.