Abstract Category:

- HEPATOLOGY

Abstract Title:

Long term clinical outcome of Budd-chiari syndrome in children after radiological intervention

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Body of Abstract:
AIM: Budd-Chiari syndrome (BCS) is an uncommon cause of chronic liver disease in children. The literature on management of pediatric BCS is scarce. Our primary objective was to determine the long-term outcome of patients undergoing radiological intervention for the treatment of BCS. Methods: 32 children diagnosed with BCS between 2004 and 2014 were included. Retrospective data on the course of disease, response to intervention, complications and outcome of the disease was collected. Results: Twenty-five patients who were on regular follow-up were analysed. The median age at presentation was 9 months (4.5-214). The median follow-up was 44 months (5-132). Sixteen patients initially received anticoagulation alone. It was associated with high failure rate of 66%. Twenty patients underwent radiological intervention. Failure rate with

angioplasty was 57%. Hepatic vein stenting was successful in all patients. TIPS was needed in 14 patients. Immediate complications were seen in four patients (28.5%). TIPS patency rate at 2 years was 75%. Four patients developed hepatopulmonary syndrome after a median period of 3 years (1.5 - 5.25) and 1 developed hepatocellular carcinoma. TIPS decreased portal hypertension and hepatic dysfunction and improved growth in all patients. Conclusions: BCS commonly presents during infancy. Anticoagulation alone and angioplasty of the hepatic veins are associated with high failure rate. Hepatic vein stenting or TIPS is feasible and efficacious in improving liver function, portal hypertension and growth. It is associated with good long term outcome and delays the need for liver transplantation, but may not prevent complications like hepatopulmonary syndrome and hepatocellular carcinoma.

Key Words:
TIPS, pediatric, hepatopulmonary syndrome, stenting, angioplasty