Profile of Budd Chiari Syndrome in childrenfrom a north Indian referral unit

Srikanth KP, Lal S, Menon J, Chadha V, Das S, Thapa BR, Das A

Budd Chiari Syndrome (BCS) is a group of disorders due to obstruction of hepatic venous outflow. Pediatric data on BCS is scarce and limited to case reports and small series.

Methodology:

Retrospective chart review of cases with diagnosis of BCS was made from the year 2005 to 2015. Various clinical, labs and radiological variables were documented. Data analyzed by descriptive statistics.

Results

Over the period of 10 years, 35 (male=24) patients were diagnosed, based on either imaging and/or liver histology. There were three patients in infancy. Abdominal distention due to ascites was the predominant complaint seen in nearly 91% of the patients. The mean duration of illness was significantly higher in children above the age of five years (17.54 months) and ten years (21.27 months**)** as compared to preschool children. There was mild elevation in serum bilirubin, with OT and PT elevated up to 3 to 4 times the upper limit of the normal (table 1). The synthetic functions were distinctly preserved. One patient had central diabetes insipidus and another patient had history of pulmonary tuberculosis. **Ultrasonography and MRV were diagnostic**. **Liver biopsy was done in 24 subjects**, based on which four, three and 17 subjects were diagnosed as acute, sub-acute and chronic BCS respectively. In etiological work up **only two patient's** revealed **deficiencies of protein C and protein S**. Warfarin was started after downsizing the varices. Three patients underwent side-to-side portocaval shunting (SSPCS); one

each underwent liver transplantation, middle hepatic vein stenting, transjugular/direct intrahepatic porto-systemic stenting (TIPSS/DIPSS). The median duration of follow up was 17 months. 3% recovered due to recanalization of the veins. Nearly 50 % the patients could not be followed up for more than 6 months. 31% remained asymptomatic and one patient died due to liver failure.

Discussion

BCS in children is an under recognized liver disorder. USG doppler is diagnostic; however MR venography would help in better surgical planning. Along with medical therapy, based on hepatic synthetic reserve patients would be either subjected to minimal invasive interventions like TIPSS, DIPSS or angioplasty. SSPCS would be better options in centers where expertise for the same is available. When all these fail, liver transplantation would be the lost resort.

Table 1: The biochemical & hematological parameters

Age	**Total**(Mean±SD)
Total Bilirubin	1.40±1.26
Conjugated Bil	1.03±0.99
Unconjugated	1.09±0.71
OT	101.07±119.72
PT	172.75±506.49
ALP	264.77±235.16
Total Protein	6.45±0.94
Albumin	3.29±0.86
PT	19.09±7.06
PTI	69.27±16.73
INR	1.64±0.61
aPTT	40.26±8.49
Hb	9.74±2.65
Platelets	192.83±122.58
Follow (Mo)	17.12