Non-Bleeding Manifestations in Children with Extrahepatic Portal Vein Obstruction

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ABSTRACT

Background: Extra hepatic portal venous obstruction is the commonest type of portal hypertension (EHPVO) in children[1-3], apart from variceal bleeding[4,5], other features include anemia, hypersplenism[5], protein losing enteropathy, growth retardation[6] and portal biliopathy.

Aim: To study the bleeding and non-bleeding manifestations in South Indian children with EHPVO.

Study Design: A prospective descriptive study on children from 1-12yrs of age with (EHPVO) from a tertiary care pediatric centre in Chennai-India. Study period - one year.

Materials and Methods: Children with EHPVO were recruited based on clinical features, USG abdomen findings, liver function tests with or without varices on upper GI endoscopy. They were divided into 2 groups; Bleeders (group 1) and non-bleeders (group 2).

Statistical Analysis Used: Chi square test, Student t test.

Results: There were 48 children with male, female ratio of 0.9:1. History of umbilical sepsis was present in 20.8%. Recurrent variceal bleed was the common presentation seen in the majority (83.3%) more so in rural children between 5-15 years (p-0.025). Non - bleeding manifestations observed were splenomegaly (95.8%), hypersplenism (37.5%) as age advances (p-0.038), anemia (91.6%), ascites (10.4%), epistaxis (6.25), growth retardation less than 3rd centile (22.7%). Associated comorbid conditions include insulin dependent diabetes mellitus (4%), atrial septal defect (2%).

Conclusions: Upper GI bleeding was the common presentation in majority of children between 5-15 years in group 1 and in group II hypersplenism (37.8%), anemia (91.6%) 2 were common. Deranged liver function was noted in 10.4% and growth retardation in 22.7%. One should look for associated congenital anomalies though rare in children.

Key words: EHPVO, Non-Bleeding manifestations

INTRODUCTION

Extra hepatic portal venous obstruction is the commonest type of portal hypertension (EHPVO) in children and constitutes 70-80% of all types of Portal hypertension in children[1-3], Usual presentation is UGI-bleed (>80%), asymptomatic splenomegaly (<10%) and pain left hypochondriac region. The usual age of presentation is 4-7 years. The signs of portal vein thrombosis (PVT) appear any time from birth till 15 years of age in children. Bleeding occurs in 60-70% children by the age of 6-7years[4,5]. UGI bleed is usually triggered by respiratory infection. Apart from variceal bleeding[4,5], other features include anemia, jaundice, ascites, protein losing enteropathy, hepatic dysfunction, growth retardation[6] and portal biliopathy. Objective is to study the bleeding and non-bleeding manifestations in South Indian children with EHPVO.

SUBJECTS AND METHODS

A prospective and descriptive study was done at Department of Pediatric Gastroenterology at Institute of Child health and hospital for children, Chennai, for one year from September 2007 to August 2008. Children up to 12 years, diagnosed as a case of EHPVO were included and there were 48 children. Demographic details like age, sex, socio economic status, and age at onset of first symptom were recorded in a pre - structured pro-forma. Children with history of hematemesis or melena were grouped as bleeders (Group 1) whereas others without were grouped as non-bleeders (Group 2) which included anemia (Hb < 11g/dl under 6 years of age and <12 g/dl in children under 12years of age), jaundice, abdominal distension, mucosal bleeds other than upper GI bleed like epistaxis. Clinical signs viz splenomegaly, hepatomegaly, ascites, growth retardation (height for age <3rd percentile according to WHO growth chart) were noted. Congenital anomalies if present were recorded.

EHPVO Children who either presented with well tolerated GI bleeding, or with non-bleeding manifestations, and with USG abdomen with Doppler showing cavernoma, splenolmegaly and normal liver echoes were also recruited. Statistical Analysis was done using SPSS-20, Proportions of various outcome measures were arrived. Demographic factors and clinical features among bleeders and non-bleeders as well as those with and without hepatic dysfunction were analysed using chi square test.

RESULTS

Demographic details of two groups of children with EHPVO- Total number of children with EHPVO were

48. 40 children (83.3%) presented with UGIB and 80% of them were from rural area when compared to 3 (37.5%) among non-bleeders. This was statistically significant (p=0.025, Table 1). EHPVO is significantly seen in children < 5 years in this study presenting with GI bleed. Bleeding was common between 5-15 years. History of umbilical sepsis was recorded in 20.8% of our patients. Majority of children (95.8%) presented with splenomegaly, moderate in 21 (45.6%), mild 18 (39.1%) and massive splenomegaly in 7 (15.2%). Hypersplenism was found in 18 out of 48 children. The prevalence of hypersplenism was 37.5 (95% C.I. 23.8, 51.2) with increasing trend as age advances (p=0.038) without any statistical significance in distribution of hypersplenism between male and female children. Anemia was the common finding in the majority 44 (91.6%) with female sex predilection in the age group of beyond 5 years. Ascites was seen in 10.4% of patients, more in male children. Albumin/Globulin reversal was seen in 13(27%). Jaundice was rarely seen and portal biliopathy was not seen during the study period. 2 of our children had recurrent loose stools.

Insulin-dependent diabetes mellitus was noticed in 2 (4%), congenital heart disease (ASD) in 1(2%) and encephalopathy in 1(2%) children. The cause of encephalopathy may be due to extensive collaterals with resultant portosystemic shunts. Growth retardation (less than 3rd percentile) according to WHO was observed in 11 out of 48 children with a prevalence rate of 22.9 (95% C.I. 11.0, 34.8) without any statistical significance between various age groups and sex.

DISCUSSION

Portal hypertension is one of the common problems in children attending paediatric gastroenterology clinic. EHPVO constitutes 70-80% of all types of portal hypertension in children[1-3]. In a study from North India on portal hypertension in children conducted by N.K Arora, et al[2], 76.5% children had EHPVO, whereas remaining 23.5% were due to intra hepatic and post hepatic causes.

The usual clinical presentation in children with EHPVO is recurrent UGI bleed with splenomegaly. However a percentage of children can present without GI bleeding, splenomegaly, anemia, growth retardation, hypersplenism, jaundice and liver cell dysfunction. Bleeding was the major presenting feature in (83.3%) of our study children which is comparable to Poddar U, et al[7] from Chandigarh, where 85% had bleeding as the presenting feature (Table 2). In our study splenomegaly was seen in 46(95.8%) in comparison to Shah S.R et al[8] from Mumbai where splenomegaly was seen in 82%. Hypersplenism was seen in 37.5% in our study as against 22% by Shah S.R et al[8].

Ascites develop in small proportion of children following haemorrhage or surgery, and are often transient. Ascites was found in 10.4% of our children in comparison to 16% of Shah S.R, et al[8] study from Mumbai and 21% of Rangari M et al[9] study from New Delhi. Webb and Sherlock[10], in their series of 97 patients, observed ascites as a presenting symptom in the absence of bleed in 13(13.4%) patients. Development of ascites is probably secondary to loss of albumin in the bleed with coexisting portal hypertension.

In our study, growth retardation was seen in 22.9% children in comparison to 51% in a study by Sarin SK et al[6] from New Delhi. Reduced portal blood supply to the liver, resistance to action of growth hormone, and reduced insulin-like growth factor have been implicated as the causative factors of growth retardation. Out of 48 children 2(4.1%) had elevated bilirubin, 3(6.25%) had elevated SGPT in comparison to the study by Khuroo MS, et al[11] from Kashmir of 21 children of whom 14(66.6%) had elevated bilirubin and 8(38%) had elevated SGPT. Jaundice is a rare presenting feature in EHPVO and is due to portal biliopathy, which refers to abnormalities in intrahepatic and extra hepatic bile ducts in patients with portal hypertension. Portal biliopathy is present in >80% patients of EHPVO in ERCP studies. ERCP is not recommended in the routine work-up of children with EHPVO. A therapeutic ERCP procedure should be planned only if there are features of cholangitis or obstructive jaundice.

In our study, insulin dependent diabetes mellitus was seen in 2 cases of EHPVO in comparison to Alexander J et, al[12] from Mumbai who reported 3 cases EHPVO with diabetes mellitus. No causal association has been found so far between these two conditions.

Congenital anomalies-Cardiac anomaly (ASD) was seen in 2% in our study in comparison to Odievre M, et al[13] and various others who reported in 19-26% cases. Ectopic Varices in EHPVO are reported in 27–40% of patients with EHPVO, and are commonly seen in the duodenum, anorectal region and gallbladder bed. Bleeding from the former two locations is not uncommon. In our study 2(4.16%) had ectopic-duodenal varices. Ectopic varices can be managed with pharmacotherapy, shunts or TIPSS. Table 3 summarises the various anomalies associated in our study compared with other studies.

Table 1: Demography between two groups

Parameter	N	Bleeder		Non-bleeder		p-value*
		n	%	n	%	
Area						
Urban	13	8	61.5	5	38.5	0.025
Rural	35	32	91.4	3	8.6	
Age						
<5 years	6	4	66.7	2	33.3	0.391
5 – 10 years	22	18	81.8	4	18.2	
>10 – 15 years	20	18	90.0	2	10.0	
Sex						
Male	23	17	73.9	6	26.1	0.130
Female	25	23	92.0	2	8.0	
Age at onset of first symptom						
<5 years	19	16	84.2	3	15.8	0.603
5 – 10 years	25	20	80.0	5	20.0	
>10 – 15 years	4	4	100.0	-	-	
Umbilical sepsis						
Yes	10	6	60.0	4	40.0	0.047
No	38	34	89.5	4	10.5	

*Chi-square test

Table 2: Comparison of features with other studies

S. No.	Features	Present study	Other studies
1	Splenomegaly	95.8%	82% -Shah S.R, et al [8]
2	Hypersplenism	37.5%	22% -Shah S.R, et al[8]
3	Ascites	10.4%	16%- Shah S.R, et al[8] 21%-Rangari M, et al[9] 13.4%-Webb&Sherlock[10]
4	Growth retardation	22.9%	51%-Sarin S.K, et al[6]
5	Raised S.bilirubin	4.1%	66.6%-Khuroo M.S, et al[11]
6	Elevated SGPT	6.25%	38%- Khuroo M.S, et al[11]

Table 3: Comparison of Associations with EHPVO with other studies

S. No.	Associations with EHPVO	Present study	Other studies
1	IDDM	2 cases	3 cases-Alexander J, et al[12]
2	Congenital anomalies	2%	19-26%-Odievre M, et al[13]
3	Ectopic varices	4.16%	27-40% in other studies.

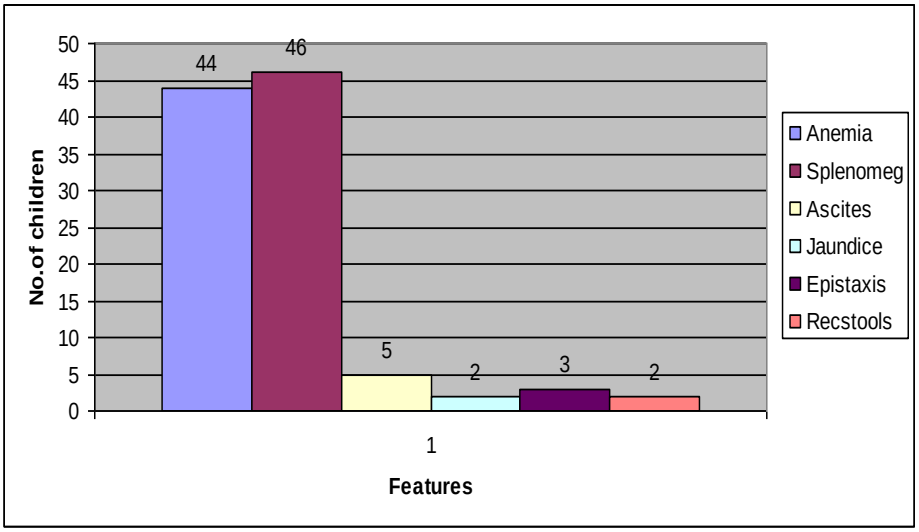

Fig. 1: **Proportion of non-gastrointestinal bleeding manifestations in group II**

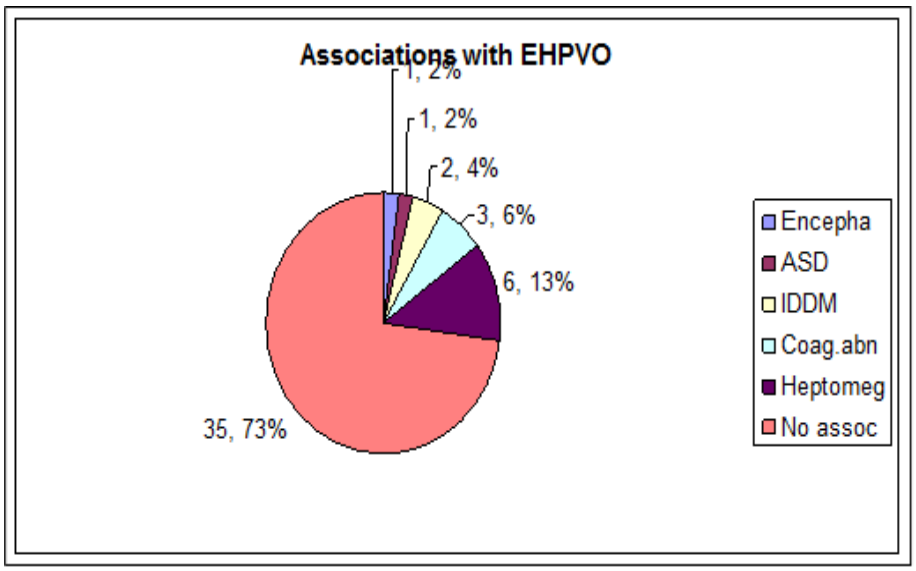

Fig. 2: Associations with EHPVO

CONCLUSION

Upper GI bleeding was the presenting feature in most of our children (83.3%). Of the non-bleeding manifestations, splenomegaly was seen predominantly (95.8%), followed by anemia in 91.6% children. Ascites was found in a small proportion of children (10.4%). Bleeding from other mucosal sites (epistaxis) was noted in 6.25% children.

Of the complications, hypersplenism was commonly seen which accounted to 37.5% and it showed an increasing trend with advancing age. Only the occasional symptomatic patient merits surgery. Splenectomy without shunt surgery should not be performed in these children. Growth retardation occurred as a complication in 22.9% of our children and there was no significant difference among various age range and sex. Hepatic dysfunction was seen in 39.5% children. Of this Albumin/Globulin reversal was seen in 27% cases, raised SGOT in10.4%, raised SGPT in 4.1% and raised S.bilirubin in 4.1% children. Of the associations hepatomegaly was noted in 13% children and IDDM in 4% of our study children.

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