Original Article

Exploring the Profile and Outcome of Children with Extrahepatic Portal Hypertension: A Cross-Sectional Study

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Abstract

Background: The most typical presentation of extrahepatic portal hypertension is variceal bleeding. Besides this, splenomegaly, ascites, jaundice due to portal biliopathy, and hypersplenic features may be present. Portal vein thrombosis is a common cause of extrahepatic portal hypertension. **Objectives**: To observe the presentation, aetiology, treatment modalities, and hospital outcomes of extrahepatic portal hypertension in children. **Methods**: It was a cross-sectional study conducted at the Department of Pediatric Gastroenterology & Nutrition of Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka. A total of 50 cases of extrahepatic portal hypertension were included in this study with normal liver function tests and no signs of chronic liver disease (CLD). Clinical characteristics, laboratory and imaging profile of the cases, etiologies, endoscopic findings, variceal ligation, complications and hospital management were studied. **Results**: The majority (64%) of patients were male. The mean age at presentation was 9.42 ± 3.55 years. Hematemesis in 46 (92%), Melaena in 45 (90%) and Splenomegaly in 49 (98%) patients were observed. Features of pancytopenia in 18% of patients with portal vein thrombosis in 68% of patients were explored. Endoscopy of upper GIT showed 64% of cases had grade- 4 and 34% grade- 3 esophageal varices. There were no significant complaints in 47 (94%) patients with EVL, except four (8%) patients complained of chest pain. **Conclusion**: Gastrointestinal bleeding and Splenomegaly were two standard clinical features of extrahepatic portal hypertension and portal vein thrombosis was the leading cause of it. This study underscores the importance of further research in this area, and a prospective study with a larger sample size is recommended to strengthen the present findings.

Keywords: Extrahepatic Portal Hypertension, Endoscopic variceal ligation, Splenomegaly in children

Introduction

Portal hypertension (PH) is defined as a portal pressure gradient (PPG) or hepatic venous pressure gradient (HVPG) exceeding five mmHg.[1] Clinically significant portal hypertension occurs when HVPG is 10 mmHg or higher.[2] This syndrome arises from various pathological conditions that result in elevated portal venous pressure. Portal hypertension can be classified as prehepatic, hepatic (which can be presinusoidal, sinusoidal, or post-sinusoidal), or posthepatic.[3]

Extrahepatic portal hypertension (EHPH) includes a range of disorders that feature increased portal pressure due to prehepatic or post-hepatic issues without liver cirrhosis. Extrahepatic portal venous obstruction (EHPVO) and hepatic venous outflow obstruction, such as Budd–Chiari syndrome, are factors that lead to the occurrence of EHPH.[4] Congenital abnormalities, portal vein valves, stenosis, atresia, or agenesis also contribute to EHPH.[5,6] In cases of extrahepatic portal hypertension, the obstruction is located in the portal vein and its branches or in the perisinusoidal area of the liver. In patients with EHPH, the wedged hepatic venous pressure (WHVP) is typically normal or only slightly elevated and is considerably lower than the portal vein pressure.[7]

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In India and other tropical regions, EHPV is a common cause of extrahepatic portal hypertension in children, while in adults, it occurs occasionally.[8,9] EHPVO accounts for 70-80% of all portal hypertension cases in children.[10] Each year, a significant number of EHPVO patients are documented in densely populated countries like Bangladesh.[11]

The primary clinical manifestation of PH, which leads to hospital visits, is gastrointestinal bleeding. Most bleeding originates from esophageal varices, with 2-10% of cases stemming from gastric varices.[12] All patients with EHPH exhibit splenomegaly, while ascites are rarely present.[13] In children, sudden massive hematemesis is accompanied by varying degrees of anemia in cases of EHPH.[12] Ultrasound (US) is the preferred diagnostic method with high accuracy, enabling the detection of splenoportal axis issues and portal thrombosis.[14]

The leading cause of significant gastrointestinal bleeding in children is PH. In EHPH cases without liver disease or systemic illness, most patients tend to have a relatively benign course. The condition primarily results from variceal bleeding, recurrent thrombosis (if a prothrombotic disorder is present), symptomatic portal biliopathy, and features of hypersplenism.[12]

It's crucial to highlight that extrahepatic portal hypertension generally has a favorable prognosis, with mortality mainly linked to variceal bleeding, and there has been virtually no mortality following endoscopic variceal ligation (EVL).[15] This study aimed to assess the clinical presentation, treatment options, and immediate outcomes during hospital admissions, revealing an overall optimistic perspective.

Methods

Study Design and Participants

This cross-sectional study took place at the Department of Paediatric Gastroenterology & Nutrition at BSMMU, Dhaka, Bangladesh, from July 2015 to June 2016, involving 50 patients clinically diagnosed with portal hypertension below 16 years of age. These patients exhibited normal liver function tests on investigations and no signs of chronic liver disease (CLD), making this a distinct and focused group for the research. Participants were chosen using a non-probability purposive sampling method, while children who were critically ill were excluded from the study.

Study Procedure

After obtaining informed written consent from each patient or their guardian, comprehensive history-taking and clinical assessments were conducted. Hematological tests, which included serum bilirubin, serum alanine aminotransferase, serum albumin, International Normalization Ratio, Hb%, and platelet count, were performed at the Biochemistry department of BSMMU using an auto-analyzer. A sonographic assessment of the portal vein was carried out to check for patency, thrombus presence, cavernous transformation, organomegaly, liver echotexture, and ascites. Subsequently, upper gastrointestinal endoscopy was conducted to detect the presence of esophageal varices and any related findings using an Olympus video endoscope GIF Q 150 machine. Patients were treated effectively through sclerotherapy, endoscopic variceal ligation (EVL), and medication with propranolol, dosed between 1mg to 5mg/kg/day. The propranolol dosage was adjusted to achieve a 25% reduction in resting heart rate.

Data Collection Technique

Following the treatment, patients were monitored for any episodes of variceal rebleeding, chest pain, or other complications. If there were no complications, patients were patient was discharged from the hospital with advice after one or two days. Data were collected through face-to-face interviews and medical record reviews using a pre-tested, semi-structured questionnaire and checklist. Each patient's socio-demographic details, clinical history, physical examination results, laboratory findings, treatment administered, and outcomes were documented in individual questionnaires.

Statistical Analysis

All collected data were thoroughly reviewed for inconsistencies, followed by coding, categorizing, and tabulating with SPSS software version 25. Descriptive statistics were computed, including mean, median, and standard deviation for numerical data, and frequencies and proportions for categorical data.

Ethical Consideration

Ethical standards were upheld at every stage of the study. Written permission was also secured from the relevant department where the study was conducted, and ethical clearance was granted by the Institutional Review Board of BSMMU, Dhaka. Memo-BSMMU/2015/12901, dated 29/10/2015.

Results

Table 1 presented that the mean (± SD) age was 9.42±3.55 years. The majority, 23 (46%) of patients, were between 6-10 years of age, followed by 17 (34%) were between 11-15 years of age. Regarding the sex distribution of the patients, it was observed that 32 (64%) were male and 18 (36%) were female.

Table 1: Socio-demographic information of the study patients (n=50)

Attributes	Frequency (f)	Percent (%)
Age (Years)		
< 6	8	16
6-10	23	46
11-15	17	34
> 15	2	4
Mean±SD=9.42±3.55		
Sex		
Male	32	64
Female	18	36

Regarding clinical history, it was revealed that hematemesis was present in 46 (92%), melena and abdominal mass were present in 45 (90%) and 10 (20%) of patients, respectively. Physical examination findings detected moderate anaemia in the majority of 20 (40%) and palpable spleen in a maximum of 49 (98%) of patients. Jaundice and ascites were absent, and the liver was not palpable for all patients, as stated in Table 2.

Table- 2: Clinical history and physical examination of study patients (n=50)

Attributes	Frequency (f)	Percent (%)
Clinical history (Multiple response)		
Hematemesis	46	92
Melaena	45	90
Abdominal distension	7	14
Abdominal mass	10	20
Consanguinity	1	2
Umbilical infection	2	4
Physical Examination		
Anaemia		
Mild	18	36
Moderate	20	40
Severe	12	24
Jaundice		
Present	0	0
Absent	50	100
Liver		
Palpable	0	0
Not palpable	50	100
Spleen		
Palpable	49	98
Not palpable	1	2
Ascites		
Present	0	0
Absent	50	100

Haemoglobin level was found to be more than 9 gm/dl in 40 (80%) patients. WBC count was within 4000-10000 cell/cumm (normal range), and platelet count was also within the normal range in the majority 41 (82%) and 41 (82%) of patients, respectively. Considering the USG of whole abdomen findings, thrombus with cavernous transformation occurred in 31 (62%) patients, and only thrombus found in 3 (6%) patients and 16 (32%) patients were normal. Endoscopic findings showed oesophagal varices present in all patients. The majority of the patients, 31 (62%), had grade-4, followed by 17 (34%) patients who had grade-3 esophageal varices (Table-3).

Table 3: Hematological Parameters of Study Patients (n=50)

Laboratory Investigations	Frequency (f)	Percent (%)
Hb (gm/dl)		
<6	1	2
6-9	9	18
> 9	40	80
WBC (cell/cumm)		
<4000	9	18
4000-11000	41	82
Platelet Count		
Normal	41	82
Low	9	18
USG of Portal Vein		
Normal	16	32
Thrombus with cavernous	31	62
Thrombus without cavernous	3	6
Endoscopy of Esophageal Varices		
Grade-1	1	2
Grade-2	1	2
Grade-3	17	34
Grade-4	31	62

Among all patients majority, 42 (84%) did not need supportive treatment, 7 (14%) required PRBC, and 1 (2%) required normal saline along with whole blood transfusion. As a specific treatment, 2 to 4-band EVL was done in 45 (90%) patients, and more than four-band EVL was done in 2 (4%) patients. Sclerotherapy was given in 1 (2%), vasopressin in 3 (6%), and octreotide in 1 (2%) of patients and all 50 (100%) patients were given propranolol (Table-4).

Table 4: Supportive and Specific Treatment Given to the Study Patients (n=50)

Variables	Frequency (f)	Percent (%)
Supportive Treatment		
Normal saline, along with whole blood transfusion	1	2
Blood transfusion (PRBC)	7	14
No need for supportive treatment	42	84
Specific Treatment		
Vasopressin (stat)	3	6
Octreotide (stat)	1	2
Endoscopic Variceal Ligation (EVL)		
2 to 4-band	45	90
> 4 band	2	4
Not needed	2	4
Others Treatments		
Sclerotherapy	1	2
Propranolol given	50	100

Table 5 shows that a maximum of 46 (92%) patients had no complaints, followed by chest pain in 4 (8%) patients.

Table 5: Distribution of patients by in-hospital outcome (n=50)

Outcome	Frequency (f)	Percent (%)
No complaints	46	92
Chest pain	4	8
Rebleeding	0	0
Total	50	100

Discussion

The present study was carried out to observe the clinical profile of extrahepatic portal hypertension (EHPH) in children. A total of 50 patients with EHPH were included in the study. In the present study, among the fifty patients, 46% were in the 6-10 years age group, 34% were in the 11-15 years age group, 16% were aged less than six years, and only 4% were more than 15 years age group. The mean age was 9.42±3.55 years. The lowest age at presentation was 15 months. No case was observed in infants under the age of one year. About 64% were male and 36% female. Several studies correspond to the recent findings. One study found that among all respondents, 59.7% were male patients, and another study among 63 children found that the average age of patients was 5.14 years (33 boys, 52.38%).[16,17,18]

Hematemesis was the most common presenting symptom (58%) of extrahepatic portal hypertension, found in a study.[16] There are so many studies that also found that variceal bleeding was the most common symptom of extrahepatic portal hypertension.[4,8,17,18] This current study also found that hematemesis (92%) and melaena (90%) were the most common presenting symptom. Approximately 20% of patients had a history of abdominal mass, which corresponds to another study's findings.[8]

In the cases of a recent study, anaemia was detected in all cases: 24% severe, 40% moderate, and 36% mild. This can be explained by the extent of gastrointestinal bleeding or hypersplenism. The latter can account for leukopenia and thrombocytopenia. These are similar to a study that found anaemia was detected in 85% of cases.[16] On laboratory investigation, this study showed 80% of patients had Hb% >9 gm/dl, 18% had 6- 9 gm/dl, and 2% were severely anaemic with Hb% level <6mg/dl. Leukopenia was present in 18% of cases, and thrombocytopenia in 18% of cases. These features are consistent with other studies. The current study found that on abdominal examination, the spleen was palpable in 98% of cases. In many studies, they described that Splenomegaly is the most common feature of extrahepatic portal hypertension. A study showed that 31 children (49.21%) experienced upper gastrointestinal bleeding, 22 children (34.92%) had Splenomegaly, and 44 children (69.84%) had thrombocytopenia.[17] Another study explored that anaemia was present in 84.6%, thrombocytopenia in 59%, and leukopenia in 48%.[16]

USG findings showed in our study that 32% of cases were normal, thrombus with cavernous transformation was 62%, and only thrombus was 6%. Another survey by El-Karaksy et al. (2015) found that portal vein collaterals were in 72.2% of cases, and portal vein thrombosis was in 11.8%.These findings were nearly similar to those of our study.[16]

In this study, children with gastrointestinal bleeding received treatment with octreotide infusion. Dissimilar treatment was given in a study; the portosystemic shunt was carried out in 11 children (17.46%), while the Meso-Rex shunt was performed in 4 children (6.35%).[17]

Endoscopy findings of upper GIT showed that esophageal varices grade 3 in 34% of cases and grade 4 in 62% of cases. A study in Brazil found that endoscopy findings of esophageal varices were grade 3 40% and grade 4 25.5%. In the current study, there were more grade 4 oesophagal varices. It may be due to the late seeking of medical attention.[19]

EVL is one of the effective treatments for variceal bleeding. In the present study, EVL was done in almost all patients except younger children. EVL was conducted in 94% of cases, and sclerotherapy was given to one patient aged 15 months. A study conducted in Egypt found that sclerotherapy was given in 79.7% of cases, and EVL was done in 15.8%.[16] In another study, 43.8% of cases underwent treatment with sclerotherapy,

31.3% with variceal band ligation and in 8 patients, 25% required an association of both treatments.[19] Variceal ligation was performed in 26 children (41.27%) and sclerotherapy in 5 children (7.94%).[17] Endoscopic variceal injection sclerotherapy and ligation concluded that EVL was superior to EVS regarding rebleeding and complications. A study was done by Dai et al. in 2015 on endoscopic variceal ligation compared with endoscopic injection sclerotherapy for the treatment of oesophagal variceal bleeding and found that EVL is better than EIS in terms of lower rates of rebleeding, complications, and higher rate of variceal eradication.[20] There are several limitations in this study. Follow-up of the patients could not be done due to the study design. Time and resources were limited, so the study included a small sample size.

Conclusion

Gastrointestinal bleeding and Splenomegaly were two standard clinical features of extrahepatic portal hypertension and portal vein thrombosis was the leading cause of it. This study underscores the importance of further research in this area, and a prospective study with a larger sample size is recommended to strengthen the present findings.

Declarations

Consent for Publication

All authors have approved this manuscript for publication.

The datasets supporting this article's conclusions are included within the article. Datasets generated during and/or analysed during the current study are available from the corresponding author upon reasonable request.

Competing Interests

The authors declare that they have no competing interests.

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