Original Article

Etiological spectrum and clinical profile of Portal Hypertension in Children at A Tertiary Care Hospital

*Baidya M,[1] Mahmud S,[2] Rashid R,[3] Hasan AR,[4] Farhana T,[5] Ahmed SS[6]

ABSTRACT

Background: Definition of Portal hypertension is a clinical syndrome characterized by a pathologic increase of portal venous pressure. This study aims to investigate the clinical, laboratory, and etiological profiles of portal hypertension in children at a tertiary care hospital in Bangladesh. **Methods:** A descriptive type of cross-sectional study was conducted at the Department of Pediatric Gastroenterology, Hepatology, and Nutrition, Bangladesh Shishu Hospital and Institute, from January 2022 to December 2024. A total of 64 children aged 0–18 years diagnosed with portal hypertension, either clinically or radiologically, were enrolled using purposive sampling. Data were analyzed using descriptive statistics by SPSS version 25.0. **Result:** Among the 64 children studied, 65.6% were male, with the majority aged 6–10 years. The most common cause was Portal vein thrombosis (54.7%). Patients with Cirrhotic liver were more frequently exhibited hepatomegaly, jaundice, ascites, coagulopathy, and elevated liver enzymes, while non-cirrhotic patients had higher rates of anemia and thrombocytopenia due to hypersplenism. Esophageal varices and variceal bleeding were common in both groups, but advanced complications like hepatic encephalopathy and hepatorenal syndrome were seen only in the cirrhotic group. **Conclusion:** Extrahepatic portal vein obstruction, cryptogenic cause, Wilson disease, and biliary atresia were the most prevalent causes of portal hypertension in this study. The most common cause of extra-hepatic portal venous obstruction was Portal vein thrombosis. Gastrointestinal bleeding and splenomegaly were found in most of the cases of non-cirrhotic portal hypertension. Hepatomegaly, increased transaminase levels, and synthesis dysfunction were suggestive of cirrhotic Portal hypertension.

Keywords: Portal Hypertension, Children, Esophageal varices, Cirrhosis

Received on: 25th August'25 **Accepted on:** 21st September'25

Author's Affiliation

1. *Madhabi Baidya, Assistant Professor, Department of pediatric Gastroenterology, Hepatology and Nutrition, Bangladesh Shishu Hospital and Institute.
2. Salauddin Mahmud, Professor, Department of Pediatric Gastroenterology, Hepatology & Nutrition Bangladesh Shishu Hospital & Institute
3. Rafia Rashid, Assistant Professor, Department of Pediatric Gastroenterology, Hepatology & Nutrition Bangladesh Shishu Hospital & Institute
4. Ahmed Rashidul Hasan, Assistant Professor, Department of Pediatric Gastroenterology, Hepatology & Nutrition Bangladesh Shishu Hospital & Institute
5. Tanzila Farhana, Assistant professor, Department of Pediatric Gastroenterology, Hepatology & Nutrition Bangladesh Shishu Hospital & Institute
6. Syed Shafi Ahmed, Professor & Head, Department of Pediatric Gastroenterology, Hepatology & Nutrition Bangladesh Shishu Hospital & Institute

Address of Correspondence: *Dr. Madhabi Baidya, Assistant Professor, Department of pediatric Gastroenterology, Hepatology and Nutrition, Bangladesh Shishu Hospital and Institute,
Email: madhabibaidya018@gmail.com

Introduction:

The term "portal hypertension" (PH) refers to the presence of portal resistance or a rise in blood flow in the portal venous system, that is, both low baseline portal venous pressure ranging from 7 to 10 mmHg and hepatic venous pressure gradient (HVPG), which is the difference between the wedged hepatic venous pressure and the free hepatic venous pressure, ranging from 1 to 4 mmHg. PH is defined as a portal pressure greater than 10 mmHg or HVPG greater than four mmHg. [1,2] Although it is estimated that portal pressure above 12 mmHg is associated with, it is rarely measured directly. It is instead inferred through events of pathological changes, such as splenomegaly, development of varices, ascites, hepatopulmonary syndrome, and cirrhosis as complications. Portal hypertension is a wide variety of pediatric liver disorders. The common causes of Portal hypertension in children include extrahepatic portal vein obstruction (EHPVO), biliary atresia, alpha-one antitrypsin deficiency, and autoimmune hepatitis.[4] However, the etiology varies from country to country. While intrahepatic causes, such as biliary atresia, are prevalent in developed countries, extrahepatic causes, such as EHPVO, are more common in developing countries.[5] Causes of Portal hypertension are due to a combination of two simultaneously occurring hemodynamic processes: (1) due to cirrhosis, increased intrahepatic resistance to the passage of blood flow through the liver, and (2) increased splanchnic blood flow secondary to vasodilatation within the splanchnic vascular bed. Portal hypertension can be due to many different causes at prehepatic, intrahepatic, and post-hepatic sites.[6] The initial clinical manifestation is characterized either by episodes of upper gastrointestinal bleeding or by splenomegaly on routine clinical examination. The major complications include upper gastrointestinal bleeding, hypersplenism (massive splenomegaly), growth retardation, and portal biliopathy.[7]

In many cases, upper gastrointestinal bleeding due to oesophageal or gastric varices may be the initial and most alarming presentation, prompting further investigation. Diagnosing portal hypertension in children involves a combination of clinical evaluation, laboratory tests, imaging studies, and endoscopic examination. Non-invasive tools such as abdominal ultrasound with Doppler can help assess portal vein patency, liver structure, and splenic size. Endoscopy plays a vital role in identifying varices and planning appropriate intervention.[8] Acute variceal bleeding is a medical emergency and requires prompt stabilization, typically with intravenous fluids, blood transfusions, and medications that reduce portal pressure. To control bleeding and reduce recurrence, endoscopic interventions such as variceal ligation or sclerotherapy are often employed.[9] So, this study aims to evaluate the etiological spectrum and clinical presentation of portal hypertension in children admitted to a tertiary health care centre in Bangladesh.

Material and Methods:

A descriptive type of cross-sectional study was conducted at the Department of Pediatric Gastroenterology, Hepatology, and Nutrition, Bangladesh Shishu Hospital and Institute, from January 2022 to December 2024. A total of 64 children aged 0–18 years diagnosed with portal hypertension, either clinically or radiologically, were enrolled using purposive sampling. Patients with incomplete records, loss to follow-up before diagnosis, isolated splenomegaly without portal hypertension, or non-portal causes of GI bleeding were excluded. Based on clinical evaluation, laboratory investigations, and imaging findings, including abdominal ultrasonography and Doppler studies, patients were categorized into cirrhotic and non-cirrhotic groups. Detailed demographic data, clinical features (such as hepatosplenomegaly, hematemesis, melena, jaundice, and edema), laboratory parameters (including complete blood count, liver function tests, and coagulation profile), and complications related to portal hypertension (e.g., ascites, variceal bleeding, hepatic encephalopathy) were recorded. Etiological classification was based on final diagnosis confirmed through imaging, serology, and relevant investigations. Data were analyzed using descriptive statistics by SPSS version 25.0, and comparisons were made between cirrhotic and non-cirrhotic groups to identify clinical and etiological differences. Ethical clearance was obtained by the institutional review board before the study, and written informed consent was obtained from attendants.

Results:

Table 1: Demographic profile of the Study Population (n=64)

Demographic Variable	Category	Number of Patients (n)	Percentage (%)
Sex	Male	42	65.63
	Female	22	34.38
Age Group	≤ 5 years	18	28.13
	6–10 years	24	37.50
	>10 years	22	34.38

Out of the total 64 patients included in the study, 42 (65.63%) were male and 22 (34.38%) were female, showing a male predominance. The majority of the patients were aged between 6 and 10 years (37.5%), followed by >10 years (34.38%) and ≤5 years (28.13%). [Table 1]

Table 2: Clinical Features of the Study Population (n=64)

Clinical Features	Cirrhotic Group (n=15)	%	Non-Cirrhotic Group (n=49)	%
Hepatomegaly	11	73.3	1	2.04
Splenomegaly	13	86.7	35	71.4
Hematemesis and melena	10	66.7	32	65.3
Anemia	9	60.0	47	95.9
Jaundice	10	66.7	0	0.0
Edema	11	73.3	0	0.0

In the cirrhotic group, hepatomegaly (73.3%), splenomegaly (86.7%), jaundice (66.7%), and edema (73.3%) were notably prevalent, whereas jaundice and edema were absent in the non-cirrhotic group. Repeated anemia and gastrointestinal bleeding (hematemesis and melena) were observed in both groups, but in non-cirrhotic patients, they were more common. [Table 2]

Table 3: Laboratory Profile of the Study Population (n=64)

Laboratory Findings	Cirrhotic Group (n=15)	%	Non-Cirrhotic Group (n=49)	%
Leucopenia	4	26.7	30	61.2
Anemia	9	60.0	47	95.9
Thrombocytopenia	5	33.3	45	91.8
Elevated transaminase	15	100.0	5	10.2
Hyperbilirubinemia	13	86.7	0	0.0
Coagulopathy	13	86.7	0	0.0

Cirrhotic patients exhibited classic signs of liver dysfunction, including elevated transaminases (100%), hyperbilirubinemia (86.7%), and coagulopathy 86.7%). In contrast, the non-cirrhotic group had a higher prevalence of cytopenia, especially anaemia (95.9%) and thrombocytopenia (91.8%), which is likely secondary to hypersplenism. [Table 3]

Table 4: Etiology of Portal Hypertension in the Study Population (n=64)

Etiology	Number of Patients (n)	Percentage (%)
Portal vein thrombosis	35	54.69
Splenic vein thrombosis	6	9.38
Wilson disease	5	7.81
Biliary cirrhosis	4	6.25
Autoimmune hepatitis	3	4.69
Budd–Chiari syndrome	1	1.56
Cryptogenic	10	15.6

The most common cause of portal hypertension in this cohort was portal vein thrombosis (54.69%), followed by cryptogenic (15.6%).

Table 5: Portal Hypertension Complications in the Study Population (n=64)

Complications	Cirrhotic Group (n=15)	%	Non-Cirrhotic Group (n=49)	%	p-value
Ascites	11	73.3	0	0.0	<0.001*
Portal gastropathy	5	33.3	10	20.4	0.293
Oesophageal varices	15	100.0	46	93.9	0.553
Variceal bleeding	10	66.7	32	65.3	1.000
Hepatorenal syndrome	1	6.7	0	0.0	0.217
Portal biliopathy	2	13.3	0	0.0	0.075
Hepatic encephalopathy	2	13.3	0	0.0	0.075
Hepatopulmonary syndrome	1	6.7	0	0.0	0.217

Table 5 shows that ascites was significantly more common in cirrhotic patients (73.3%) compared to none in non-cirrhotic patients ($p < 0.001$). Esophageal varices and variceal bleeding were prevalent in both groups, with no significant difference. Complications in advanced stages, such as hepatic encephalopathy, portal biliopathy, and hepatorenal syndrome, occurred only in cirrhotic patients but were not statistically significant, likely due to small numbers. Overall, cirrhotic patients experienced more severe complications associated with liver dysfunction. [Table 5]

Discussion:

This study was carried out in the Pediatric Gastroenterology, Hepatology & Nutrition department of Bangladesh Shishu Hospital & Institute. Patients aged 0-18 years who present with GI bleeding with or without splenomegaly are included. The most affected age group in our study was 6–10 years (37.5%), consistent with the multicenter analysis by Martinez et al.[10], who found that children in this age range are frequently diagnosed incidentally or during evaluations for complications, such as variceal bleeding. In the present study, the male-to-female ratio was 1.9:1. Therefore, portal hypertension was more commonly observed in males than in females. Mahmud et al.[11] found a male-to-female ratio of 1.5:1. However, Imanieh et al.[8] reported a different result. Their ratio was 1.01:1. Regarding etiology, our study identified portal vein thrombosis (PVT) as the leading cause of portal hypertension (54.69%). Mahmud et al.[11]also found that out of 32 children, it was observed in 20 (62.5%) cases. Podder et al.[12] and Arora et al.[13] also reported similar results. Present study results further showed that splenomegaly, anemia, and in both groups variceal bleeding were common. However, features such as jaundice, edema, ascites, hyperbilirubinemia, and coagulopathy were significantly more common in the cirrhotic group. Similar findings are supported by Sarma and Medhat et al.[14], who highlighted that such features are hallmarks of hepatic decompensation and poor synthetic function. Biochemical findings in cirrhotic patients, including 100% elevated transaminase levels and 86.7% hyperbilirubinemia, indicate extensive hepatocellular injury, as similarly documented by Ferrarese et al, 15, who found that these markers correlate strongly with liver disease progression and risk of complications.

In contrast, non-cirrhotic patients demonstrated significantly higher rates of cytopenia, especially anaemia (95.9%) and thrombocytopenia (91.8%), likely secondary to hypersplenism. This finding aligns with the observations of Dutta et al.[16], that is haematological profile in children with extrahepatic portal vein obstruction. Ascites was significantly more prevalent. In contrast, oesophageal varices and variceal bleeding were found in both groups, with no significant difference, implying that both intrahepatic and extrahepatic causes can lead to portal pressure elevation and variceal formation. Mushtaq et al.[17] similarly noted that varices and upper GI bleeding are common presentations regardless of the underlying cause of portal hypertension. In advanced complications such as hepatic encephalopathy, portal biliopathy, and hepatorenal syndrome, only cirrhotic children experienced these complications, albeit with no statistical significance due to the small sample size. Saadah et al.[18] described similar trends, noting that such complications typically appear only in advanced cirrhosis with a prolonged disease course. Elevated liver enzymes, prolonged INR, and jaundice in our cirrhotic patients further reinforce the conclusions drawn by Poddar et al, [12,] who described these parameters as reliable indicators of progressive pediatric liver disease.

Limitations of the Study

The study was conducted in a single centre with a small sample size. So, the results may not accurately represent the entire community.

Conclusion:

This study highlights the distinct clinical, laboratory, and etiological profiles of cirrhotic and non-cirrhotic portal hypertension in children. When portal vein thrombosis emerged as the leading cause of non-cirrhotic PHT, cirrhotic patients exhibited more severe hepatic dysfunction and advanced complications such as ascites, encephalopathy, and coagulopathy. Non-cirrhotic cases, although presenting with significant variceal bleeding and hypersplenism-related cytopenia, had better-preserved liver function.

Recommendation:

Early identification of the underlying cause of portal hypertension in children, especially distinguishing between cirrhotic and non-cirrhotic types, is crucial for timely and appropriate intervention. Routine screening for varices, regular monitoring of liver function, and tailored management strategies such as Porto-systemic shunt surgery for non-cirrhotic patients and consideration of liver transplant in cirrhotic Patients are recommended to reduce complications and improve prognosis in pediatric patients.

Funding: No funding sources

Conflict of interest: None declared

Reference:

1. Gugig R, Rosenthal P: Management of portal hypertension in children. World J Gastroenterol. 2012, 18:1176-84.

2. Chapin CA, Bass LM: Cirrhosis and portal hypertension in the pediatric population. Clin Liver Dis. 2018, 22:735-52.

3. de Ville de Goyet J, D'Ambrosio G, Grimaldi C: Surgical management of portal hypertension in children. Semin Pediatr Surg. 2012, 21:219-32.

4. D'Amico G, Luca A. Natural history. Clinical-haemodynamic correlations. Prediction of the risk of bleeding. Baillieres Clin Gastroenterol 1997; 11:243-56.

5. 5. D'Amico G, Pagliaro L, Bosch J. The treatment of portal hypertnsion: a meta-analytic review. Hepatology 1995; 22:332-54.

6. Merli, M., Nicolini, G., Angeloni, S., Gentili, F., Attili, A.F., Riggio, O. (2004). The natural history of portal hypertensive gastropathy in patients with liver cirrhosis and mild portal Hypertension. Am J Gastroenterol, 99, 1959-6.

7. Hyams JS, Treem WR. Portal hypertensive gastropathy in children. J Pediatr Gastroenterol Nutr 1993; 17:13 18.

8. Imanieh MH, Dehghani SM, Khoshkhui M, Malekpour A. Etiology of portal hypertension in children: a single center's experiences. Middle East J Dig Dis 2012; 4:206-10.

9. Shashidhar H, Langhans N, Grand RJ. Propranolol in prevention of portal hypertensive hemorrhage in children: a pilot study. J Pediatr Gastroenterol Nutr 1999; 29:12-7.

10. Martínez-Martínez LM, Rosales-Sotomayor G, Jasso-Baltazar EA, Torres-Díaz JA, Aguirre-Villarreal D, de León IH, Páez-Zayas VM, Sánchez-Cedillo A, Martínez-Vázquez SE, Tadeo-Espinoza HN, Guerrero-Cabrera JP. Acute liver failure: Management update and prognosis. Revista de Gastroenterología de México (English Edition). 2024 Jul 1;89(3):404-17.

11. Mahmud S, Ahmed SS, Gulshan J, Tasneem F. Etiology of por tal hypertension in children-an experience in a tertiary cen tre of Bangladesh. Bangladesh j child health. 2015;39(1):14–7.

12. Poddar U, Reddy DV. Non-Cirrhotic Portal Hypertension in Children: Current Management Strategies. Current Hepatology Reports. 2023 Sep;22(3):158-69.

13. Arora NK, Lodha R, Gulati S, Gupta AK, Mathur P, Joshi MS et al. Portal hypertension in North Indian children. Indian Pediatr 1988; 65: 585-91.

14. Sarma MS, Seetharaman J. Pediatric non-cirrhotic portal hypertension: Endoscopic outcome and perspectives from developing nations. World Journal of Hepatology. 2021 Oct 27;13(10):1269.

15. Ferrarese A, Senzolo M, Cattelan AM, Sasset L, Battistella S, Zanetto A, Germani G, Russo FP, Gambato M, Pelizzaro F, Vio S. Bacterial Infections in End-Stage Liver Disease: Implications for Liver Transplantation. Transplantology. 2024 Jun 28;5(3):129-39.

16. Dutta S, Kumar A, Sharan S, Prasad U, Prakash J. Role of transient elastography in pediatric patients with portal hypertension: A cross-sectional, single-center study. Journal of Indira Gandhi Institute of Medical Science. 2023 Jan 1;11(1):86-90.

17. Mushtaq I, Cheema HA, Malik HS, Waheed N. Indicators of Hepatopulmonary Syndrome in Patients with Portal Hypertension. Its Various Aetiologies, Clinical Presentations and Outcome. Journal of Ayub Medical College Abbottabad-Pakistan. 2021 Jan 1;33(1).

18. Saadah OI, Khayat A, Alsufyani HA, Bin-Taleb Y. Paediatric autoimmune liver diseases: A descriptive study of patients from Saudi Arabia. Arab Journal of Gastroenterology. 2021 Jun 1;22(2):146-50.