

Original Article

FREQUENCY OF INTRA-HEPATIC FACTORS RESPONSIBLE FOR PORTAL HYPERTENSION IN CHILDREN

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Objective: To determine the frequency of intra-hepatic factors responsible for portal hypertension in children.

Methods: This was a descriptive, cross-sectional Study which was conducted from 1st July 2016 to 30th June 2018 in the department of pediatric medicine, Sheikh Zayed Hospital, Rahim Yar Khan. Forty Nine children fulfilling inclusion criteria were included and excluded if they have portal vein thrombosis on Doppler USG/CT scan and not willing for liver biopsy. Ultrasonography abdomen was done in each patient. Eye examination was done to look for KF rings if indicated. Relevant laboratory test were also done. After this liver biopsy was done in patients where other investigations were inconclusive and was sent to for histopathology for presence or absence of intra-hepatic factors responsible for portal hypertension in children.

Results: Mean age was 8.73 ± 3.90 years. Out of the 49 patients, 27(55.10%) were male and 1.2:2 male to female ratio. In my study, intra-hepatic etiology of portal hypertension in children was cryptogenic cirrhosis 34.6%, Wilson's disease in 32.6%, autoimmune hepatitis in 10.2%, congenital hepatic fibrosis 8.1%, biliary artesia in 8.1% and viral hepatitis in 6.1% patients.

Conclusions: This study concluded that most common intra-hepatic cause of portal hypertension in pediatric age group was cryptogenic (idiopathic) cirrhosis followed by Wilson's disease, autoimmune hepatitis, biliary artesia, congenital hepatic fibrosis, and viral hepatitis

Keywords: portal hypertension, cryptogenic cirrhosis and wilson's disease.

Introduction

The normal portal venous pressure is 7-10 mmHg and the hepatic venous pressure gradient (HVPG) is normally 1- 4 mmHg. If this portal pressure increase from 10 mmHg and if gradient is more than 4 mmHg, then it is called as portal hypertension (PH).^{1,19} There are variable causes of portal hypertension. One of the clinical manifestation of PH, is upper GIT bleeding as a result of esophageal varices.² Portal hypertension has intrahepatic, prehepatic & post hepatic causes.¹⁶ Our main emphasis in study was on intrahepatic causes due to their high prevalence. In a study done by Imanieh et al³, 93.3% patients have intra-hepatic etiology while extra-hepatic causes in only 6.7% patients. Doppler ultrasonography help in diagnosis of portal hypertension as it measures increase in portal vein pressure.⁴ Most patients with venous PH have intrinsic liver disease. Hematemesis resulting from esophageal varices is the most common²⁰ presentation in patients with PH, although some patients presents with decompensated liver disease.¹⁷ Other supportive investigations can be ultrasonography (US) and color Doppler imaging (CDI), CT, MRI and endoscopy.⁵ Accurate diagnosis is a pre-requisite for successful management. Portal hypertension and its complications remain a cause

of significant morbidity and mortality. Studies have been conducted for causes and management approaches in adults have been conducted but there is a lacking data in children regarding causes and morbidity and mortality.⁶ The rationale of this study was to document the frequency of intra-hepatic causes implicated in portal hypertension in pediatric age group in our local population. As intra-hepatic etiology constitutes the main bulk of portal hypertension, so this study would give us the magnitude of each intra-hepatic factor responsible for portal hypertension in pediatric age group (set of patients that fulfils operational definition of portal hypertension). The data of this study would not only add statistics of this devastating condition in our local literature but also help the clinicians to design a management protocol for these patients in order to minimize its complications as well as morbidity and mortality.

Methods

This was a descriptive, cross-sectional study conducted in the department of Pediatric Medicine, Sheikh Zayed Hospital, Rahim Yar Khan from 1st July 2016 to 30th June 2018. A total 49 patients were enrolled by non-probability, consecutive sampling after fulfilling inclusion criteria (children presenting

with portal hypertension as per-operational definition of duration >3 months, age 1-15 years, both genders) and excluding the patients <1 and >15 years, patients with portal vein thrombosis (by history, examination and doppler USG/CT scan (if required), patients not willing for liver biopsy and patients not willing for inclusion in study. After permission from the ethical review board, 49 children who were presented to department of pediatric medicine, Sheikh Zayed Hospital, Rahim Yar Khan, fulfilling the inclusion criteria were selected. After obtaining informed consent from parents of patients, serum and urine tests were sent in each patient. Ultrasonography abdomen was done in each patient for hepatobiliary system status. Eye examination was done for KF rings. After this liver biopsy was done (in patients where other test were inconclusive) and was sent for histopathology. All these reports were evaluated by consultant pediatrician for presence or absence of intra-hepatic factors responsible for portal hypertension in children as described in operational definition. A specifically designed proforma was used to document all required data. Statistical analysis was done with help of SPSS version 20.0.

Results

Age range in this study was from 1 to 15 years with mean age of 8.73 ± 3.90 years. Majority of the patients 68 (37.78%) were between 6 to 10 years of age. Out of the 49 patients, 27 (55.10%) were male and 22 (44.89%) were females with male to female ratio of 1.2-1. Mean disease duration was 7.39 ± 2.67 months. In my study, intra-hepatic etiology of portal hypertension in children found as cryptogenic cirrhosis in 17(34.6%), Wilson's disease in 16(32.6%), autoimmune hepatitis in 5(10.2%), congenital hepatic fibrosis 4 (8.1%), biliary atresia in 4 (8.1%) and viral hepatitis in 3 (6.1%) patients.

Table-1: Frequency of intra-hepatic factors responsible for portal hypertension in children.

Intra-hepatitis factors	Frequency	Percentage
Cryptogenic cirrhosis	17	34.6%
Biliary atresia	04	8.1%
Wilson's disease	16	32.6%
Congenital hepatitis fibrosis	04	8.1%
Autoimmune hepatitis	05	10.2%
Viral hepatitis	03	6.1%

Discussion

Portal hypertension is classified based on the anatomical location into extrahepatic, and intrahepatic. Intrahepatic PH is further categorized into pre-sinusoidal, sinusoidal, and post-sinusoidal PH. Numerous vascular and hepatic diseases cause portal hypertension in pediatric age group, which has fatal complications, including symptomatic esophageal varices, pulmonary involvement, encephalopathy and ascites. To improve outcome of such children, proper preventive measures and effective management is important. This study documented the frequency of intra-hepatic factors responsible for portal hypertension in children.

In this study age was from 1-15 years with mean age of 8.73 ± 3.90 years. Majority children (37.78%) had age 6-10 years. Intra-hepatic etiology of portal hypertension in children was found as cryptogenic cirrhosis in 17 (34.6%), Wilson's disease in 16(32.6%), autoimmune hepatitis in 5 (10.2%), congenital hepatic fibrosis 4 (8.1%), biliary atresia in 4 (8.1%) and viral hepatitis in 3 (6.1%) patients.

Western studies about pediatric portal hypertension demonstrated that 68-81 % cases were attributed to intrahepatic etiology while 9-27 % to extrahepatic factors and 5-10% to BuddChiari syndrome.⁷ In one study conducted in India, extrahepatic factors were in 6876% of cases and intrahepatic factors in 2428%.^{8,9}

In a study done by Imanieh et al³, a total of 45 patients included in the study, with mean age of 7.6 ± 4.7 years. Male were 23 (51.1%) and 22 (48.8%) were females. Intra-hepatic causes (cryptogenic cirrhosis in 26.6%, biliary atresia in 24.4%, Wilson's disease in 17.7%, congenital hepatic fibrosis 6.65% and autoimmune hepatitis in 6.6 % patients) of portal hypertension in pediatric age group were found in 93.3% patients while extra-hepatic causes in only 6.7% patients.

Studies had shown the difference in etiology of portal hypertension different age groups as intrahepatic causes was more in adults and extra-hepatic etiology in pediatric age group.^{10,18} One South Indian study showed that amongst the common etiological factors of portal hypertension in children were extra-hepatic portal venous blockage and then cirrhosis.¹¹ Bernard et al. stated that 51% of cases were due to and 34 % because extra-hepatic venous blockage.¹² In any case, debate exists with respect to the etiology and presentation of portal hypertension in pediatric age and adult patients and it is different in various regions. In certain researches performed in the West, frequency of intrahepatic causes was more in pediatric age group.^{13,14} while studies performed in India reported

That extra-hepatic etiological factors were more in frequency in children.¹⁵

Conclusion

This study concluded that most common intra-hepatic cause of portal hypertension in pediatric age group was cryptogenic (idiopathic) followed by Wilson's disease then autoimmune hepatitis, biliary arteria, congenital hepatic fibrosis, and viral hepatitis

in a set of patients that presented with portal hypertension. So, we recommend that special attention should be given in these patients for early diagnosis, effective and proper management steps/guidelines of the these conditions for better prognosis.

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