1. Introduction

In portal hypertension, there are hemodynamic abnormalities. In adults, portal hypertension is generally caused by hepatic cirrhosis, whereas in children it is more commonly caused by extrahepatic abnormalities with normal liver function. Portal hypertension causes hemodynamic abnormalities. Gastrointestinal bleeding is the most severe clinical manifestation of portal hypertension in both children and adults. Pathogenetically, increased pressure in the portal vein can be caused by increased vascular resistance and increased portal blood flow. The site of obstruction can be prehepatic (portal vein obstruction), intrahepatic (presinusoidal: eg congenital hepatic fibrosis; parasinusoidal: cirrhosis, hepatotoxic drug therapy, vitamin A hepatotoxicity; post sinusoidal: venocclusive disease) and/or post hepatic (Budd-Chiari syndrome, constrictive pericarditis).

2. Case Presentation

A boy, FAA, aged 12 years and 2 months came to the emergency room at Dr. M. Djamil General Hospital Padang with the main complaint of vomiting blood, and on physical examination he found splenomegaly. The study reports the results of observations of a case of a boy, FAA, aged 12 years and 2 months who came to the emergency room at Dr. M. Djamil General Hospital Padang with the main complaint of hematemesis and splenomegaly from physical examination. Non cirrhotic portal fibrosis is a cause that is not uncommon in the population in the early second decade of life. Some children with non-cirrhotic portal fibrosis as adults can end up with end stage liver disease. Patients with noncirrhotic or cirrhotic portal hypertension can be assessed using the Child Pugh instrument as an instrument that is still used to determine the survival rate if patients with portal hypertension.
abdominal pain in the pit of the stomach 2 days before entering the hospital, the abdominal pain was followed by brownish vomiting followed by vomiting fresh blood for 2 days SMRS, the frequency of vomiting was around 5x per day, with each time the vomiting could reach 1 glass, the last vomiting was approximately 4 hours before entering the hospital. The child also appeared to have become paler 1 day before entering the hospital. Black stools with a watery consistency 3 hours before entering the hospital. The presence of fever was denied. The patient had previously been treated at the regional hospital 3 times in the period from November 2021 to January 2022 with complaints of vomiting blood. The previous hospital, the patient also underwent an abdominal ultrasound with the results finding splenomegaly with splenic vein dilatation, minimal ascites in the perihepatic area, and gallbladder polyps with mild small intestine dilatation.

On initial physical examination, the patient appeared moderately ill, conscious, with blood pressure 100/70, HR 130 x/minute, temperature 37°C, respiratory rate 26x/minute, SpO2 99% with nasal cannula oxygen 2l/minute, BW 29.3kg, TB 138cm with BB/U 72%; TB/U 90%; BB/TB 87%. Upper arm circumference 18 cm with the impression of malnutrition and normal stature. The abdominal examination did not show palpable liver distension 1/4-1/4 sharp edges, smooth surface, and palpable spleen on Schuffner 2 visible venecation in the abdominal region. There is tenderness in the epigastric region, and bowel sounds (+) are normal. Warm acral, and capillary refilling time < 2 seconds, no edema was found. Puberty status A1 P3 G3.

The results of laboratory examinations are:
Hemoglobin 6.8; leukocytes 17,360; platelets 283,000; diff count 0/0/2/75/16/7; PT 10.4; APTT 21.8 (normal); SGOT 35 (normal); SGPT23 (normal); Na 130; K 5; Ca 7.7; albumin 2.1; total bilirubin 0.4; indirect bilirubin 0.2; direct bilirubin 0.2; LDH 252 (normal); Na 130 (hyponatremia); K 5.1; Cl 98; Gamma GT 38 (normal); medium alkaline phosphatase examination cannot be carried out because the reagent is not available; GDS 119 mg/dl; urea 62; creatinine 0.7; and albumin 2.1. IgM anti-HAV was negative, HBsAg and IgM anti-HBc were negative, anti-HCV was negative, IgM anti-HEV was negative, and a Coombs test was carried out and the results were negative. The results of the urinalysis examination, when the patient was first treated, were found to have slightly increased urine leukocytes of 6-7, urine bacteria was positive and nitrites were also positive, no urine bilirubin was found, and urine urobilinogen was found positive. The patient was given the antibiotics ceftriaxone 2 x 1 gram IV, Ranitidine 2 x 30 mg IV, Omeprazole 1 x 30 mg IV, albumin transfusion, and intravenous calcium correction.

The child underwent an abdominal ultrasound on February 15th, 2022, and April 20th, 2022 with results of hepatosplenomegaly. The child also underwent a CT scan of the abdomen with contrast on February 18th, 2022 with results of hepatosplenomegaly and liver cirrhosis, a CT scan with contrast was performed again on April 28th, 2022 with results of hepatosplenomegaly with images of cholangitis and cholelithiasis. The child underwent a laparoscopic cholecystectomy from pediatric surgery, and PA tissue biopsy results showed chronic cholecystitis. EGD was carried out followed by ligation of esophageal varices, and ligation of esophageal varices, namely on 15/3/2022, 5/4/2022, 10/6/2022 (Figure 1), and 2/7/2022. The results of the laboratory examination when the patient was admitted for re-treatment were: Hb 11.8 leukocytes 7780 platelets 115000 type count 0/4/0/69/15/7 albumin 3.4 SGOT 119 (increased 3.1 x) SGPT 130 (increased 3, 1 x) Na 142 K 4.3 Cl 104 Calcium 9 Gamma GT 128 (increased) total bilirubin 0.9 direct bilirubin 0.4 (increased 2x) indirect bilirubin 0.5 (normal) PT 11.6 (increased 1.05x ) APTT 29.2 (increased 1.16x) urinalysis within normal limits, only urobilinogenuria was found.
Figure 1. EGD results (June 10th, 2022). Conclusion: Grade IV esophageal varices, ligation of esophageal varices was carried out in 4 places.

Figure 2. EGD Results (July 13th, 2023). Conclusion: esophageal varices were found in 2 places, and esophageal variceal ligation was performed.

However, during the treatment period, the child increasingly looked jaundiced, when the bilirubin evaluation and urinalysis were repeated, the total bilirubin was 9.3 (increased 9.3x), direct bilirubin 8.2 (increased 41x), indirect bilirubin 1.1 (increased 1, 2x), SGOT 167 (increased 4.3x), SGPT 181 (increased 4.4x), with the results of repeat urinalysis found bilirubinuria +2, urea 9, creatinine 0.5. Fibroscan
examination results showed FIV (cirrhosis) on February 11th, 2023 and July 4th, 2022. The results of the repeat Fibroscan examination (on July 4th, 2022) this semester also still show a picture of FIV (cirrhosis). The child then received therapy according to the cholangitis therapy protocol and a laparoscopic cholecystectomy was performed from pediatric surgery. PA tissue biopsy results showed chronic cholecystitis. Entering observation in semester II, the child returned to treatment in June 2022 with black stools again. The results of supporting examinations found Hb 7.2; leukocytes 4,110; platelets 159000; count type 0/11/0/50/28/11; PT 11.1; APTT 25.9 (increased 1.06x); Na 135; K 4.1; Cl 112; calcium 8.4; albumin 3.2; total bilirubin 0.7; Direct bilirubin 0.4 (increased 2x); indirect bilirubin 0.3; alkaline phosphatase 178 (normal); Gamma GT 29 (normal), Gamma GT increased slightly 1 month later to 79; SGOT 35; SGPT 16; Urea 11; Creatinine 0.5. The results of the EGD examination this semester, namely on November 14th, 2022, still found esophageal varices and ligation of esophageal varices was carried out in several places. Entering the observation in the third semester, the child returned to treatment because there was vomiting blood. Laboratory examination results were obtained this semester with Hb 12.7; leukocytes 13,840; platelets 169,000; count type 0/2/2/76/15/5; PT 11.9 (increased 1.1x); APTT 30.8 (1.17x increase); albumin 3.5; total bilirubin 1.3 (increased 1.3x) with indirect bilirubin 0.7 (increased 1.1x); direct bilirubin 0.6 (increased 3x); Na 139; K 4.2; calcium 8.8; SGOT 103 (up 2.7x); SGPT 90 (increased 2.1 x); urea 34; creatinine 0.5. The EGD results for this semester are explained in Figure 2.

3. Discussion

In portal hypertension, there are hemodynamic abnormalities. In adults, portal hypertension is generally caused by hepatic cirrhosis, whereas in children it is more commonly caused by extrahepatic abnormalities with normal liver function.\(^1\) Gastrointestinal bleeding is the most severe clinical manifestation of portal hypertension in both children and adults. Etiologically, in adults, portal hypertension is generally caused by cirrhosis, while in children, approximately half of it is caused by extrahepatic portal vein obstruction.\(^2\) Pathogenetically, increased pressure from the portal vein can be caused by increased vascular resistance and increased portal blood flow. The location or site of increased resistance depends on the disease process. The site of obstruction can be prehepatic (portal vein obstruction), intrahepatic (precirrhal: eg congenital hepatic fibrosis; parasinusoidal: cirrhosis, hepatotoxic drug therapy, vitamin A hepatotoxicity; postsinusoidal: venocclusive disease) and/or posthepatic (Budd-Chiari syndrome, constrictive pericarditis).\(^3\) Non-cirrhotic portal fibrosis is a cause that is not uncommon in the pediatric population in the early 2nd decade of life. Children with this diagnosis always show signs of splenomegaly and growth retardation, as well as episodic and tolerable esophageal variceal bleeding. In this disease, the HVPG (hepatic vein-pressure gradient) and Liver Stiffness Measurement values in some cases can overlap with patients with cirrhosis, so to exclude cirrhosis, hepatic histopathological examination should still be carried out to confirm the diagnosis. Thus, every patient who presents with evidence of portal hypertension (splenomegaly, esophageal varices with or without bleeding) and whose liver function is well maintained should be evaluated for the possibility of non-cirrhotic portal fibrosis.\(^4\) The literature on adults shows that the onset of non-cirrhotic portal fibrosis in adults is in the 3rd and 4th decades of life. Vikrant et al in a study in India, of 19 children with non-cirrhotic portal fibrosis showed that males were dominant over females, and most sufferers were male, also reported in other previous studies, with an average onset of symptoms at the age of 10 years, which the youngest is 2 years old. The main manifestations were discomfort in the left upper quadrant of the abdomen (16/19), symptomatic hypersplenism (7/19), and bleeding esophageal varices (3/19).
On physical examination, growth retardation was found in 14/19 children. Laboratory examination showed manifestations of hypersplenism, namely platelet levels <100,000/mm$^3$ and/or leukocyte levels <4000/mm$^3$ (in 17/19 children). All patients with preserved liver function. Endoscopy found esophageal varices in 13/16 children, and portal hypertension gastropathy was found in 12/16 children. Abdominal imaging, namely ultrasound and CT scan of the abdomen with contrast, showed signs of portal hypertension (splenomegaly and/or porto-vein collaterals) in all patients. Gallbladder calculus was found in 3 patients, splenic infarction in 2 patients, and evidence of portal biliopathy in 1 patient. Liver function is usually normal or near normal in pediatric patients unless there is a slight decrease in serum albumin during bleeding episodes due to esophageal varices. Patency of the portal vein is not the basis for the diagnosis of non-cirrhotic portal vein fibrosis, this is because of the high incidence of portal vein fibrosis. Secondary vein thrombosis with or without cavernoma formation in patients with non-cirrhotic portal fibrosis. The clinical manifestations of portal hypertension generally appear suddenly and look very dangerous. Approximately two-thirds of children with portal hypertension present with melena hematemesis due to rupture of esophageal varices. Gastrointestinal bleeding can also occur from portal hypertensive gastropathy, gastric antral vascular ectasia (GAVE), or from gastric varices, duodenum, periostomal, or rectum. Bleeding can be very severe and life-threatening. Upper gastrointestinal endoscopy is more readily available than HVPG (Hepatic Vein-Pressure Gradient) measurement and is the preferred method for assessing portal hypertension. With endoscopy, it can be assessed whether there are gaster and esophageal varices, the size of the varices, and portal hypertensive gastropathy.

Abdominal Doppler USG examination can indirectly detect signs of portal hypertension, besides that it can calculate the portal vein flow velocity (is there a slowdown in flow?), the direction of portal flow (is there a slowdown in flow?), the direction of portal flow and the presence or absence of turbulence. In this patient, when an EGD was performed in the fourth semester, a picture of gastropathy ec portal hypertension was found. The currently generally approved therapy in adults for the prevention of rupture of medium-large varices is propanolol. Beta blocker therapy in portal hypertension provide primary effect of inhibiting beta 2 receptors in the splanchnic bed which results in uninhibited stimulations of adrenergic receptors thereby reducing splanchnic and portal perfusion. Apart from that, beta-blockers also reduce heart rate due to the inhibition of beta-1 adrenergic receptors, thereby reducing cardiac output and portal perfusion. Propanolol is also known to reduce collateral circulation such as blood flow in the azygos vein. The main side effects of using propanolol are heart block and asthma exacerbation. Beta-blockers also have the potential to interfere with the physiological response to hypoglycemia so these drugs should not be used in children with diabetes. Patients with noncirrhotic portal hypertension or cirrhosis with Child-Pugh A (Figure 5) can be expected to have a good long-term liver function and a life free of bleeding after splenic renal shunt. This patient with a Child-Pugh A score of 6 is expected to have a good long-term liver function and a life free of bleeding after a splenic renal shunt. This interpretation of Child-Pugh A scoring is also useful for determining patient survival rates, where at a low score of 5 – 6, the 2-year survival rate can reach 85%. The operative action rather than a splenorenal shunt is an action taken to reduce blood flow to the portal vein, where the veins in the spleen are channeled to the veins in the left kidney, thereby creating a new blood flow. In the conditions of liver cirrhosis, spontaneous splenoral shunt has also been reported. In this patient, at the last EGD on 19th October 2023, there was no longer a picture of pulsating esophageal varices, but instead, a picture of hyperemic gaster was found.

Identification of the specific cause of cirrhosis requires clinical information from the history of the disease, findings on physical examination, and
supporting examinations. The presence of hypersplenism in the patient and the appearance of dilated splenic veins, vascular changes, and venoocclusive disease are still possible in this patient, this could be due to a previous hypercoagulable condition. Hypercoagulability conditions can be caused by systemic infections such as previous COVID-19 infections. The diagnosis of liver cirrhosis can be confirmed by several examinations, including liver function test, coagulation test, complete blood count, serological test for the causative virus, and liver biopsy. In this patient the viral serological test was also found to be negative, this proves that viral hepatitis may not be the cause of the patient’s cirrhosis. SGOT and SGPT levels in cirrhosis patients are generally slightly increased, alkaline phosphatase and gamma GT levels are usually normal, and bilirubin in cirrhosis is also generally normal, but if it increases, this indicates that the cirrhosis process is ongoing especially biliary cirrhosis. Anemia is common and often has multifactorial causes, can be microcytic due to chronic gastrointestinal bleeding, and can be macrocytic due to folic acid deficiency or hemolysis and hypersplenism. The thrombocytopenia that occurred in the patient could also be caused by hypersplenism, where the patient’s spleen during the fourth-semester observation was found to be increasingly enlarged. A liver biopsy is important to establish the diagnosis, as a clear diagnosis will lead to better management and outcomes. Nonalcoholic steatohepatitis (NASH), which is often associated with obesity, diabetes, or metabolic syndrome, can be seen on an ultrasound scan but still requires a liver biopsy for confirmation. Idiopathic Portal Hypertension or Non-cirrhotic portal fibrosis can show features of portal fibrosis, abnormal portal veins, and lobular regeneration. In general, the prognosis for children with non-cirrhotic portal fibrosis is quite good. Children with idiopathic portal hypertension have a high incidence of failure to thrive due to reduced portal supply, malabsorption due to portal hypertension gastropathy, secondary anemia due to hypersplenism and massive splenomegaly which will compress the stomach, and reduced appetite. The anemia that occurs can be microcytic hypochromic anemia (due to gastrointestinal bleeding) or it can also be normocytic normochrome (due to hypersplenism). Leukopenia in children with hypersplenism also causes children to be more susceptible to infection. Primary management is to prevent bleeding from the varices themselves. Various therapeutic options to prevent bleeding, such as EVBL, administration of beta blockers, sclerotherapy, and transjugular intrahepatic portosystemic shunt. The literature discussing shunt surgery for non-cirrhotic portal fibrosis is still very limited, with post-procedure there being a minimal risk of hepatic encephalopathy, glomerulonephritis, arteriovenous fistula, and ascites. A case study reported by Javier et al stated that the patency of intra-abdominal organ veins can be confirmed with an abdominal CT scan with 3-phase contrast. It can be concluded that to assess whether the patient’s intra-abdominal veins are patent or not, a 3-phase abdominal CT scan can be used, although the presence or absence of signs of thrombosis in the portal vein cannot exclude the possibility of a diagnosis of non-cirrhosis portal fibrosis. Portal vein thrombosis and suprahepatic vein thrombosis can occur secondary to non-cirrhotic portal fibrosis or in patients with cirrhosis, but it can also occur in patients with malignancy and thrombophilia. A liver biopsy is still performed to confirm liver parenchymal tissue damage. Some children with non-cirrhotic portal fibrosis as adults can end up with end-stage liver disease. 4

4. Conclusion
Portal hypertension in children is generally caused by extrahepatic abnormalities with normal liver function. The site of obstruction can be prehepatic (portal vein obstruction), intrahepatic (presinusoidal: congenital hepatic fibrosis; parasinusoidal: cirrhosis, hepatotoxic drug therapy, vitamin A hepatotoxicity; postsinusoidal: venoocclusive disease) and/or posthepatic (Budd-Chiari syndrome, constrictive pericarditis). A liver biopsy is important and still
performed to confirm liver parenchymal tissue damage. Primary management is to prevent bleeding from the varices themselves. Various therapeutic procedure to prevent bleeding, such as EVBL, administration of beta blockers, sclerotherapy, and even surgical procedures such as transjugular intrahepatic portosystemic shunt.

5. References