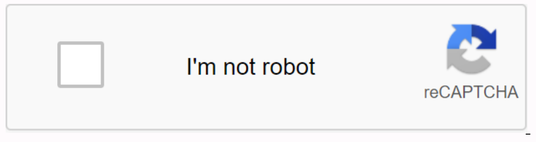

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Pediatric portal hypertension pdf

Primary Care InformationAuor Copyright and License information DisclaimerPediatric portal hypertension management is a team approach between patient, patient family, primary caregiver and specialized providers. Guidelines for evidence-based practice have not been established in pediatrics. This article serves as an overview for primary care np in the management of pediatric portal hypertension, discuss etiology, pathophysiology and clinical presentation of pediatric portal hypertension, diagnostic tests and treatment and management options. Keywords: liver disease, non-cirrh portal hypertension, pediatric portal hypertension, primary care NP can play a key role in pediatric patients with portal hypertension. Subtle signs and symptoms during a routine physical exam may provide clues suggesting significant liver disease and rapid referral to a specialist. It can be either a pediatric gastroenterologist trained in the management of pediatric portal hypertension or a pediatric hepatologist. Depending on the geographical location and the ability of the specialist to manage childhood portal hypertension, either recommendation is appropriate. These specialists can be found in children's hospitals, which are associated with teaching universities or medical schools located in large cities. Some may even have satellite offices where routine follow-up can be done locally, and any procedure or surgery (if warranted) would be performed at an affiliated children's hospital. Once the diagnosis of pediatric portal hypertension is confirmed, a common management plan between the patient, the patient's family, the NP and the specialist will ensure proper care and follow-up for the future. This article discusses etiology, pathophysiology and clinical presentation of pediatric portal hypertension, diagnostic tests and treatment and treatment options. Treatment and treatment of children with portal hypertension has not been well studied and there are no current guidelines. In adults, evidence-based approaches in the treatment and management of portal hypertension are well studied in literature. In May 2010, a workshop Baveno V Consensus was held on the methodology of diagnosis and therapy for portal hypertension; was composed of international experts in this field who reviewed portal hypertension, current evidence and best practices.1 In 2011 at The Children's Hospital of Pittsburgh, international panels of experts convened to review and adapt the results of the workshop, which were then published.1 In 2009, experts gathered at the annual meeting of the American Association for the Study of Liver Disease (AASLD) to discuss primary prophylaxis in children with varicose bleeding in children with portal hypertension.2 The findings from both meetings are findings from both meetings are in this article. Causes of pediatric portal hypertension3Portal hypertension is a clinical manifestation of cirrhosis (scarred) liver disease; however, it may result from other is not associated with cirrhosis. This article focuses on non-cirha pediatric portal hypertension, which is not well documented in the literature. Necirious etiology of portal hypertension are classified as prehepatic, hepatic and posthepatic (see Causes of pediatric portal hypertension).3 Etiology and other examinations are managed by a specialist. NP plays a role in the initial recognition and health maintenance of pediatric patients with portal hypertension. The most commonly identified cause of childhood portal hypertension is extrahepatic obstruction of the portal vein (EHPVO). EEAVO etiology is poorly understood, although studies have identified predisposing factors such as a history of umbilical catheterization, dehydration, trauma or hypercoagulation. More than 50% of pediatric cases are idiopathic.4Portal hypertension is defined as a pathological increase in the pressure of the portal system. Due to the presence of EEAVO, this blockage causes the back flow of blood to connecting organs such as the spleen (causing splenomegaly), esophagus and gastrointestinal (GI) tract (causing enlarged blood vessels).5 (See Portal circulation.) Portal hypertension is a clinical diagnosis identified through anamnesis and physical examination. The two most common clinical manifestations of childhood portal hypertension that may trigger referrals are upper GI haemorrhage (UGIB) and splenomegaly.5 EEAVO can manifest itself from 6 years to adulthood, but is primarily a childhood disorder.6In children, UGIB is most often the initial clinical manifestation of EEAVO. Esophageal varices occur in 90% to 95% of patients, and gastric varicose varices occur in 35% to 40%.5 Seventy-nine percent of children with EEAVO are believed to have at least one episode of UGIB in their lifetime.5 Varixal bleeding in children is often observed after upper respiratory infection, fever or aspirin ingestion. Throughout the duration of the disease, coughing and sneezing create abdominal pressure; fever increases cardiac output; and the use of non-steroidal anti-inflammatory drugs or aspirin (if medically indicated) to treat symptoms can create ulcers and contribute to the rupture of varicose. In addition, long-term gastroesophageal reflux can contribute to erosion over varicose, which could lead to bleeding.5Splenomegaly is often first discovered during routinephysical tests.5 Children with enlarged spleen are often first referred to as hematologists to exclude possible haematological processes, especially if leukopenia is present. Once haematological causes are excluded, a recommendation to a paediatric gastroenterologist or hepatologist is appropriate.7 Clinical findings may include growth failure; one prospective study showed that 50% of children with EEAVO had growth retardation compared to healthy children who did not have EEAVO laboratory tests.5Diagnostic laboratory tests that np can obtain as an initial examination include complete blood count (CBC) with differential and platelet count (CBC with platelets), gamma-glutamyl transpeptidase, magnesium, phosphorus and international normalized ratio. Laboratory findings of paediate patients with portal hypertension due to EEAVO have normal or near normal liver tests without underlying functional liver disease. Common laboratory findings include leukopenia, anemia, and thrombocytopenia from splenomegaly. In addition, anemia can be caused by chronic blood loss from varicose bleeding.5.6Doppler ultrasonography is the most useful diagnostic tool for differential diagnosis. Aspect of hepatic parenchyma and liver capsule, patency of portal vein or its replacement by cavernoma, The flow pattern of hepatic veins and hepatic arteries, and the presence of splenomegaly or hepatic atrophy are important elements for diagnosing and staging the patient's condition.1,6 For diagnosis of EEAVO, ultrasonography with Doppler has sensitivity and specificity greater than 95%.5Dependent to the sources of practice, TEP can obtain ultrasonography with Doppler before referral to a specialist. Doppler's ultrasonography helps to assess anatomy and blood flow and excludes the presence of any masses. Common findings on abdominal ultrasound in portal hypertension settings may include splenomegaly, the presence of collateral (new veins) and possible reversal of the portal vein of blood flow (more severe cases).3 Patients with EEAVO have an abdominal ultrasound showing obstruction of the main portal vein with cavernous transformation. This appears to be an irregular tangle of blood vessels near the hilum of the liver, which is a sign of chronicity.5 If the child had previous blood tests and/or imaging, parents should be instructed to make copies of the messages and display them on a compact disc for initial consultation with a specialist. Positive histories and physical, blood work, and Doppler ultrasonography are sufficient workup that np can initiate before referral to a pediatric gastroenterologist or hepatologist. Another examination that a specialist can prescribe is contrast computed tomography or angiography magnetic resonance imaging.4 Both are useful for assessing the extent of thrombosis and can serve as an anatomical road map if surgery is required.4.5 Liver biopsy. is standard practice for diagnosis of EEAVO, since hepatic parenchyma is usually normal.1 A liver biopsy would be justified if other underlying liver disease were suspected.1,5Diagnostic of portal hypertension can be further confirmed by measuring portal pressures. This practice is invasive, requires anesthesia and is not a common diagnostic tool used in pediatrics. Portal hypertension is defined as portal pressure greater than 12 mm Hg or gradient greater than 6 mm Hg to 7 mm Hg. Normal portal pressure is between 5 mm Hg and 10 mm Hg.8 Measurement of pressure gradients of the portal is invasive and requires catheterization of the cervical or femoral vein with measurement of right pressure atrial carcinoma, free liver (FHVP) and wedged hepatic venous pressure (WHVP). Hepatic venous pressure gradient is the difference between WHVP and FHVP.8 If a pediatric patient undergoes anesthesia for another purpose, portal pressures can be measured for baseline, but this is not common practice in diagnosis and treatment. A retrospective one-centred study has shown that this practice is safe and feasible in children with acute and chronic liver disease, including those who are critically ill; however, further research is needed regarding its use as a diagnostic tool after the initial presentation.9Measures for pediatric portal hypertension include endoscopic treatment, drug therapy to reduce portal pressures and surgery.10 Treatment and treatment are determined by the root cause of portal hypertension and the expertise of a specialist.1,5Endoscopic treatment. Supervision of esophagogastroduodenoscopy (EGD) can be performed at the outset to stratify the risk of varicose bleeding. EGD is the best available test for varicose diagnosis.3 There are two types of endoscopic modals: endoscopic sclerotherapy (EST) and endoscopic varicose ligation (EVL), also known as endoscopic belt ligation. Both are highly effective in controlling acute varicose bleeding in more than 90% of cases, as well as in eradicating varicose varicose varicose bleeding.5 The use of beta-blockers, EST and/or EVL is considered the primary prophylaxis for varicose bleeding in adults; however, due to the lack of controlled paediatric data, primary prophylaxis remains controversial and practices vary significantly between centres.1,3EST is the use of endoscope and injectable sclerosing agents, such as ethanolamine oleate, inside or around varicose.5,11 Children with EEAVO who received EST for varicose bleeding showed low relapse rates.5 Potential complications associated with EST include ulcers on the esophagus and strictures, esophageal perforation, Motility disorders, reduced low esophageal sphincter pressure and esophageal reflux disease.5 EST has been shown to be useful even in very young children as young as 5 months of age weighing 12 lb (5.5 kg).1,8EVL is performed through upper endoscopy and uses strips to sew the vial to prevent further bleeding. In the last 10 years, EVL has been more widely used and found to be more improved than EST in terms of efficacy, safety, and degree of standardization in adults and children.1,3 However, this is not always possible because currently there are no resources small enough that can be used in pediatric patients. Therefore, est remains the only option for young children.5 EVL is the recommended treatment of acute bleeding from varicial esophagus.1 Recurrence of bleeding is lower in patients treated with EVL, since varicose are eradicated in less endoscopic sessions.5There is very little data on the diagnosis and triage of esophageal testicles in children. The current scoring system has been adopted from adult practice; However, it is not validated in literature.1,2,11 One retrospective study conducted in one academic hospital environment found that the majority of patients with non-punk portal hypertension performed long-term without surgical placements, and treatment focused primarily on the supervision and treatment of varicose.12 However, this practice was not well studied and reviewed in the literature.12Drug therapy to reduce portal pressure. In children, the use of non-seedy beta-blockers (propranolol or nadolol) should be avoided while evidence of appropriate efficacy and safety is expected.1 However, some experts may use them to reduce the hepatic venous pressure gradient by reducing cardiac output (beta1-receptor antagonism) and inducing splanchnic vasoconstriction (beta-receptor antagonism).5 Data on the use of beta-blockers for treatment in children with portal hypertension are rare. , resulting mainly from a number of cases with a limited number of patients. Side effects from the use of beta-blockers in children include significant hypotension due to suppression of normal tachycardial response and hypovolemia due to poor ability to increase stroke volume to support cardiac output, bronchospasm and hypoglycemia.2,5 It is important that np is informed about indications of beta-blocker in the management of hypertension and potential side effects. Surgery. Surgical shunts are usually reserved for patients with thrombocytopenia and recurrent bleeding, when liver transplantation is not considered and endoscopic therapy is ineffective.7 Surgery is performed in a children's hospital with which the specialist is associated. The ideal operation for patients with EEAVO is meso-Rex bypass, which connects the upper mesenteric and spleen veins with the left portal vein using the internal cervical graft jump.1,5 This procedure bypasses obstruction and restores nutritious blood flow to the liver. If meso-Rex cannot be performed due to unsatisfactory anatomy, a distal splenorenal shunt should be considered if EST or EVL has not adequately controlled convulsive bleeding. Haemophilus influenzae type B and pneumococcal (pneumococcal 13valent conjugation vaccine and polyvalent pneumococcal vaccine 8 weeks apart). This is in line with the recommendations of the American Academy of Pediatrics for vaccination of patients with chronic liver disease or cirrhosis.13,14 There is a potential for damage to the spleen during short-circuit surgery due to anatomical location and emergency spleen embolization can be performed; Therefore, proper preoperative immunization is important.1Transjugular intrahepathic portosystemic shunt (TIPS), which can be a good technical choice in THE EEAVO, is rarely, if ever indicated as a means of secondary prophylaxis in varicose bleeding for portal hypertension in children. indications are refractor or recurrent varicose bleeding and diuretic resistant ascites in adults.5 The location of TIPS stents can prevent future successful bypass of meso-Rex by permanently blocking access to the intrahepatic vein if it is open.1 However, this practice is rarely used in pediatrics and there is no supporting evidence of its effectiveness in the literature. Reported complications from portal hypertension include varicose bleeding, ascites, hepatopulmonary syndrome, portopulmonary hypertension and hepatic encephalopathy.3 Varmic bleeding is the most serious complication of portal hypertension, which can occur from venous collaterals in the stomach or esophagus. Varicose vargia and ascites are seen when the portal pressure is 12 mm Hg or higher.8 This is not a subtle finding as there is a 30% mortality rate with varicose bleeding.8Varial bleeding can occur at any time with or without the above treatment. Patients with active varicose bleeding should be redirected to the nearest ED, as this constitutes a medical emergency. Upon arrival, the child's parent or guardian should tell the ED provider that the child has UGIB due to portal hypertension. It will drive ED in control and stabilization. Octreotide is a vasopressin analogue (used off-label for varicose bleeding) that is effective in stopping acute varicose bleeding in 95% of cases when administered as an infusion. After stabilisation with octreotide, acid-blocking substances (for PROPHYLAXIS GI) and blood products (red blood cells and/or platelets), the patient should perform EGD within 24 hours.1Portal hypertension: Instructions for parentsIs important that both NP and experts remain up-to-date in the guidelines and recommendations in the management and care of child portal hypertension. Most centers follow the AASLD guidelines. It is important to educate the family about portal hypertension (see Portal hypertension: A Guide for Parents). In addition, limiting excessive salt and water intake to prevent ascites and recognizing UGIB to accelerate emerging intervention is clearly a common charge between primary care IBO and specialist. The team approach of the IBO and the specialist is crucial in the management of patients with portal hypertension. Np is usually a doorman who first identifies the initial signs of portal hypertension, such as splenomegaly for physical examination, and plays a key role in the initial examination, referral, diagnosis and procedure (see Clinical pathway for referral). Children with portal hypertension should receive all normal health care, including vaccination of children and routine examinations. Children with enlarged spleen should refrain from contact sports and may need to wear spleen protection to prevent spleen rupture.1 The IBO should work closely with the child's school nurse to monitor changes in clinical status, such as vital signs control, and help avoid contact sports in the physical education class the child has splenomegaly. The school nurse may also be the first responder if the child can show signs of UGIB and help in obtaining medical care. The specialist will manage the supervision of portal hypertension, such as physical examination, endoscopy, laboratories, possible beta-blockers and imaging. The frequency of laboratory acquisition could be coordinated with a specialist and an IBO. Children with portal hypertension should eat a healthy and balanced diet. The main management of portal hypertension in children focuses on the prevention of decompensation, which includes the control and prevention of ascites and bleeding from portal hypertension.5 If the child is placed on a beta-blocker or if the child is in a dose by a titration specialist, the child can be scheduled for subsequent visits with np to monitor heart rate and BP. Monitoring with a specialist is individual for each patient. Parents of children with portal hypertension can reach a center where the child is managed for support and support groups that may be available to them locally. More research is needed in the field of esophabic varicose control in children with portal hypertension. On the contrary, in adults, treatment was challenged with a number of studies that were conducted in the Baveno V Consensus Workshop. Most children's centers adapt the practice to what has been found to be successful in adult studies.11 Depending on the preferences of the center and the clinical condition of a child with portal hypertension, treatment can vary from routine endoscopy to the use of beta-blockers as primary prophylaxis and surgical options as secondary treatments. At present, management relies on expert opinion, poor quality paediatric studies and extrapolation of results observed in studies in the adult population. High-quality paediatric studies are needed to help lead the practice.11 Understeering the pathophysiology of portal hypertension due to EEAV in paediatric patients is important to help manage governance, prevention and overall outcomes. Effective cooperation between the IBO and the specialist is the key to ensuring optimal childcare with portal hypertension. Management of portal hypertension may include medications, endoscopic and surgical options that ensure a good long-term prognosis, and routine maintenance of health care, including vaccination of children and routine examinations.1. Shneider BL, Bosch J, de Franchis R, et al. Portal hypertension in children: an expert pediatric opinion on the Baveno V Consensus Workshop report on the methodology of diagnosis and therapy in portal hypertension. Pediatrician's transplant. 2012;16(5):426–437. It's all right. [Google Scholar] 2. Ling SC, Walters T, McKiernan PJ, Schwarz KB, Garcia-Tsao G, Shneider BL. Primary prophylaxis of varicose bleeding in children with portal hypertension: a framework for future research. J Pediatrician Gastroenterol Nutr. 2011;52(3):254-261. [PMC free article] That's fine. [Google Scholar] 3. Ling SC. Portal hypertension in children. Liver Dis. 2012;1(5):139–142. [Google Scholar] 4. Feldman AG, Sokol RJ. Non-cirhátic portal hypertension in the paediatric population. Clin Liver Dis. 2015;5(5):116–119. [Google Scholar] 5. Giouleme O, Theocharide E. Control of portal hypertension in children with portal vein thrombosis. J Pediatrician Gastroenterol Nutr. 2013;57(4):419–425. It's all right. [Google Scholar] 6. Khanna R, Sarin SK. Non-cirrhosis portal hypertension-diagnosis and management. J Hepatol. 2014;60(2):421–441. It's all right. [Google Scholar] 7. Gugig R, Rosenthal P. Management of portal hypertension in children. World J Gastroenterol. 2012;18(11):1176-1184. [PMC free article] That's fine. [Google Scholar] 8. Mileti E, Rosenthal P. Management of portal hypertension in children. Curr Gastroenterol Rep. 2011;13(1):10-16. [PMC free article] That's fine. [Google Scholar] 9. Woolfson J, John P, Kamath B, Ng VL, Ling SC. Measurement of hepatic venous pressure gradient is possible and safe in children. J Pediatrician Gastroenterol Nutr. 2013;57(5):634–637. It's all right. [Google Scholar]10. Grimaldi C, de Ville de Goyet J, Nobili V. Portal hypertension in children. Clin Res Hepatol Gastroenterol. 2012;36(3):260–261. It's all right. [Google Scholar]11. D'Antiga L. Medical management of esophageal varicose and portal hypertension in children. Semin Pediatrician Surg. 2012;21(3):211–218. It's all right. [Google Scholar]12. Wojcik BM, Zahid S, Cai S, Englesbe MJ. Progression of noncirrhal portal hypertension in the child population. J Clin Exp Hepatol. 2011;1(3):169-176. [PMC free article] That's fine. [Google Scholar]13. Centers for Disease Control and Prevention. Intervals between PCV13 and PPSV23: recommendations of the Advisory Committee on Immunisation Practices (ACIP). 2015. www.cdc.gov/mmwr/preview/mmwrhtml/mm6434a4.htm#Tab. [PubMed]14. Centers for Disease Control and Prevention. Recommended immunization plan for adolescents 18 years of age or younger, United States, 2017. 2017. www.cdc.gov/vaccines/schedules/hcp/imz/child-adolescent.html. www.cdc.gov/vaccines/schedules/hcp/imz/child-adolescent.html.