Portal vein thrombosis in children and adolescents

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Abstract

Objective: To review the literature on portal vein thrombosis in children and adolescents, focusing on its diagnosis, complications and treatment.

Sources of data: The medical literature of the past 10 years was reviewed using the PubMed and MEDLINE search engines, with major focus on portal vein thrombosis and its clinical outcomes. The following keywords or expressions were used for the web search: *portal vein thrombosis*, *extra-hepatic portal vein obstruction*, *prognosis*, *children*, *portal hypertension*, *esophagogastric varices*. Additionally, we also reviewed the articles cited in the references of the initially selected papers, as well as relevant textbooks.

Summary of the findings: Portal vein thrombosis is one of the most common causes of portal hypertension among children. 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 secondary to splenomegaly, growth retardation, and portal biliopathy. The diagnosis is made by abdominal Doppler ultrasonography. Treatment is targeted at the complications and includes primary and secondary prophylaxis against upper gastrointestinal bleeding (which results from the rupture of esophageal varices), and portosystemic shunting in selected cases.

Conclusions: Portal vein thrombosis is one of the major triggers of upper gastrointestinal bleeding in children. Bleeding episodes have a remarkable effect on the quality of life of affected patients. Thus, appropriate diagnosis and treatment are needed in order to reduce morbidity and mortality.

J Pediatr (Rio J). 2006;82(3):171-8: Portal vein thrombosis, portal hypertension, extrahepatic portal vein obstruction, esophageal varices, children, prognosis.

Introduction

Extrahepatic portal vein obstruction (EHPVO) is an important cause of portal hypertension (PH) among children. It is characterized mainly by portal vein thrombosis (PVT), and is detected in 40% of children with upper gastrointestinal bleeding (UGIB) caused by esophageal varices. It may be asymptomatic. However, the mortality and morbidity rates are high, due to UGIB. Approximately 79% of children diagnosed with PVT will have at least one episode of UGIB in their lifetime.[1] The present article aims to build up the profile of children with PVT, to investigate the predisposing factors, complications, and therapeutic approach, by reviewing the medical literature.

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Anatomy and embryology

The liver originates from the anterior primitive intestine, and its primordial form – *pars hepatis* – develops as a duodenal diverticulum in the fourth gestational week. Its full development occurs in the third month. Fetal hepatic blood flow comes from the hepatic artery and from the portal and umbilical veins, which form the portal sinus. At birth, this flow corresponds to nearly 5% of an infant's total body weight, whereas, in an adult, it does not exceed 2.5%. The liver is responsible for hematopoiesis during the fetal period. In the seventh gestational week, hematopoietic cells outnumber functional hepatocytes. After the second month of life, the liver no longer produces red blood cells.[2]

The liver can be divided into two large lobes (right and left), in additional to the caudate and quadrate lobes, which are small. On top of that, there is a functional subdivision, based on blood supply, which divides the liver into eight segments. Hepatic architecture is extremely complex. The basic functions of the liver are carried out by hepatocytes.[3] The liver has a dual blood supply via the hepatic artery and portal vein. The portal vein carries venous blood from the digestive tract into hepatic sinusoids, which is drained by the hepatic veins into the inferior vena cava. The portal vein is formed by the confluence of the

172 Jornal de Pediatria - Vol. 82, No.3, 2006

Portal vein thrombosis in children and adolescents – Schettino GCM et al.

superior mesenteric and splenic veins, ascending behind the common bile duct and the hepatic artery. At the *porta hepatis*, it is subdivided into right and left branches (besides providing the quadrate lobe with an additional branch). These branches ramify to form small vessels that drain into the sinusoids. The portal vein usually receives the left gastric, pancreaticoduodenal, and cystic veins. Therefore, it drains blood from several organs, such as the intestine, spleen, pancreas, and gallbladder.[4]

The valves in the portal system are insignificant or nonexistent; the direction of blood flow is determined by the pressure gradient. Therefore, portal vein obstruction may increase the blood flow in the connections between portal vein tributaries and systemic veins, as well as inversion of blood flow at the following venous sites:

– inferior mesenteric vein and inferior vena cava and their tributaries (superior, middle and inferior rectal veins; common and internal iliac veins);

– gastric veins and superior vena cava and their tributaries (azygos system, vertebral plexus, esophagogastric veins);

– retroperitoneal veins and vena cava and azygos venous systems;

– paraumbilical and subcutaneous veins.

Portal hypertension

Portal vein is responsible for around two thirds of the blood flow into the liver. It supplies blood that is rich in oxygen, nutrients, growth factors, hormones, among other elements. Although the liver receives a large amount of blood (approximately 25% of the cardiac output per minute), the portal vein only withstands low pressures. PH occurs when portal vein pressure exceeds 10 mmHg.[5,6] Doppler ultrasonography shows a continuous blood flow, with a normal range from 15 to 30 cm/s.[7] An increase in portal pressure results from the oscillation of intravascular pressure (hyperdynamic state) and from the elevated resistance to this flow. Thus, oscillation of pressure within blood vessels is directly proportional to the flow within the portal system and to the resistance to this flow (Ohm's law). In turn, resistance to blood flow is inversely proportional to the fourth power of the radius of the vessel (Poiseuille's law).[2,3,6] Based on these physical laws, one may conclude that small variations in portal vessels cause large increases in pressure.

PH can be extrahepatic (prehepatic obstruction), intrahepatic or post-sinusoidal (Table 1). Another classification takes into account the etiology of PH: cirrhotic and noncirrhotic.[8] The consequences of PH are related to the interactions produced by the development of portal systemic collaterals, which cause UGIB.

Table 1 - Causes of portal hypertension in children

1. Extrahepatic portal hypertension
    Venous obstruction
        Portal vein thrombosis
        Splenic vein thrombosis
    Arteriovenous fistula
    Splenomegaly

2. Intrahepatic portal hypertension
    Hepatocellular disease
        Acute or chronic viral hepatitis
        Autoimmune hepatitis
        Wilson's disease
        Alpha-1 antitrypsin deficiency
        Glycogenosis type IV
        Toxins
    Biliary tract disease
        Biliary atresia
        Cystic fibrosis
        Choledochal cyst
        Sclerosing cholangitis
        Intrahepatic cholestasis
    Congenital hepatic fibrosis
    Schistosomiasis
    Idiopathic portal hypertension

3. Post-sinusoidal portal hypertension
    Budd-Chiari syndrome
    Congestive heart failure
    Veno-occlusive disease

Adapted from Shneider.[8]

Extrahepatic obstruction – portal vein thrombosis

Portal vein obstruction can occur at any site of the portal system. Obstruction is often associated with portal vein thrombosis, extrinsic tumor compression (hepatocellular carcinoma) and compression by an extrahepatic malignant tumor (especially pancreatic adenocarcinoma). In adult patients, PVT is usually secondary to some systemic diseases (e.g.: cancers), which account for nearly 40% of PH cases.[9] In children, portal vein obstruction results from thrombosis and often develops into PH of varying grades.

Thrombosis can affect only the portal vein and its tributaries or the superior mesenteric or splenic vein. The size of the thrombus determines different clinical manifestations. Orllof et al. determined the site of blood flow obstruction in 200 children and young adults. The authors observed that 67% of patients showed only portal vein obstruction, whereas 28% showed portal and splenic vein involvement, and 5% had portal vein and superior mesenteric thrombosis.[10]

Pathophysiology and etiology of portal vein thrombosis

Cessation of blood flow in the portal system causes small clinical problems due to compensatory mechanisms: immediate reflex vasodilatation of the hepatic artery and formation of collateral vessels that envelop and bypass the

Portal vein thrombosis in children and adolescents – Schettino GCM et al.

Jornal de Pediatria - Vol. 82, No.3, 2006 **173**

thrombotic site. As a general rule, initial thrombus formation is asymptomatic, and the only sign may be the formation of new vessels, which, in Doppler ultrasonography, is known as "portal cavernoma" or "cavernomatous transformation," due to the volume of blood at the site.[9,11] These plexuses with hepatopetal flow are not sufficient to release the system's pressure, resulting in hepatofugal flows. These natural (spontaneous) shunts may become pronounced, have exuberant clinical expression and may require surgical correction.

The pathophysiology of thrombus formation in the portal system is still unclear. Admittedly, there may be a combination between prethrombotic factors and local factors. Local factors are found in 25% of individuals aged over 30 years.[12] The causes of PVT can be split into four different groups: direct injury to the portal vein and consequent thrombus formation and obstruction, as occurs in omphalitis or in umbilical catheterization; congenital malformation of the vascular system with involvement of the portal system associated with other cardiovascular disorders; indirect factors that predispose to thrombus formation in the portal system; idiopathic PVT, i.e., the etiology cannot be identified.[9,11-13] Table 2 lists the main causes of PVT.

Table 2 - Causes of portal vein obstruction

Direct injury
    Omphalitis
    Umbilical catheterization
    Neonatal peritonitis
    Abdominal trauma
    Surgical trauma
    Cysts and tumors at the *porta hepatis*
Portal vein abnormalities
    Stenosis/atresia/agenesis
Indirect factors
    Neonatal sepsis
    Dehydration
    Multiple exchange transfusions
    Hypercoagulable states
Idiopathic

Sarin & Agarwal[11] compared seven studies that assessed the etiology of portal vein obstruction in infants and children (Table 3). In most cases, the cause could not be identified and, when it could be, the majority of cases showed direct injury to the umbilical vascular system (omphalitis and umbilical venous catheterization), or intra-abdominal or umbilical sepsis. However, there seems to be a relationship between the various causes, suggesting the coexistence of a prethrombotic state.[11]

With regard to umbilical catheterization, the predisposing factors are: later insertion, catheter dwell time over three days, catheter misplacement (either

subdiaphragmatic or peripheral), trauma on catheter insertion, and type of solutions infused.[8]

In adults, the main cause of thrombosis is thrombophilia secondary to malignant neoplasms. Janssen et al.[14] assessed 172 patients, aged between 14 and 91 years, in a cohort study. They found that 84% had at least one risk factor for EHPVO, such as malignant hepatobiliary diseases, inflammatory abdominal diseases, intra-abdominal surgeries, hypercoagulable states, and myeloproliferative syndrome.

A different category includes cirrhosis-related PVT. In this case, PVT seems to be associated with the low venous blood flow secondary to the increase in intrahepatic resistance, presence of portal lymphangitis, and fibrosis. About 10% of children with biliary atresia also have PVT.[6] The prevalence of PVT in adults with cirrhosis ranges between 0.6 and 17%.[15,16]

Clinical manifestations

PVT patients initially present with UGIB or splenomegaly on routine clinical examination in asymptomatic individuals.

The initial presence of hematemesis is usually dramatic in a previously healthy child, with past history of morbidity, often without remarkable intercurrent events. Melena may also be observed, but it is less common than hematemesis. The child can be lethargic, with signs of orthostatic hypotension. The clinical examination revealing splenomegaly in a child with UGIB indicates esophageal varices as the most probable site for the bleeding.

Less frequently, the diagnosis can be based on the investigation of a child with abdominal pain or with complications related to hypersplenism. The signs and symptoms of PVT vary considerably (Table 4).

The physical examination may reveal splenomegaly; hepatomegaly is not common in children with PVT without underlying liver disease, as well as stigmata of chronic liver disease. Skin pallor may be observed.

The episodes of UGIB usually recur while varices are not completely obliterated or if secondary prophylaxis is not initiated. These episodes are often precipitated by upper airway infections or use of acetylsalicylic acid (ASA). Mortality rate secondary to UGIB in patients with isolated PVT is not so high as that of cirrhotic patients, who have more complications, such as impaired liver function, refractory ascites, and encephalopathy.

Several congenital abnormalities associated with PVT have been described, such as malformations of the heart, of large vessels, of the biliary tract, and of the renal system.[1]

Transient ascites may develop after episodes of UGIB. However, patients with long-established PH secondary to PVT may have ascites without any predisposing factors. This same group of patients may have low albumin levels,

Table 3 - Etiology of extrahepatic portal vein obstruction in children

Etiology	Shaldon & Sherlock, 1962	Maddrey et al., 1968	Webb et al., 1979	Househam et al., 1983*	Boles et al., 1986	Cardinet et al., 1992	Stringer et al., 1994
n	16	37	55	32	43	61	31
Umbilical sepsis	56%	14%	22%	0	19%	11%	45%
Intra-abdominal sepsis	6%	0	20%	22%	7%	31%	6%
Umbilical catheterization	6%	0	11%	6%	2%	8%	0
Thrombophilic disorders	0	0	0	0	0	0	0
Pancreatitis	0	5%	0	0	0	0	0
Trauma	0	3%	0	0	0	0	3%
Idiopathic	31%	78%	47%	53%	72%	50%	45%

* 19% of patients with multiple causes.
Adapted from Sarin & Agarwal.[11]

increased prothrombin time, and elevated aminotransferase levels. This hepatic dysfunction can be attributed to the prolonged decrease in portal circulation and/or to the development of portal biliopathy, which shows that the disease may have a progressive course.[17]

Table 4 - Clinical manifestations of portal vein thrombosis

Growth retardation
Portal biliopathy
Coagulation disorders
Hypersplenism
Immunological disorders
Rectal and esophagogastric varices
Gastrointestinal bleeding
Recurrent abdominal pain
Ascites
Anemia
Hepatopulmonary syndrome

Hepatopulmonary syndrome is rare in PVT without cirrhosis, as well as encephalopathy, but it might occur.[18] Gupta et al. assessed 54 adults diagnosed with cirrhosis and 50 with EHPVO; only two patients with prehepatic obstruction developed hepatopulmonary syndrome without any detectable predisposing factors.[19]

Laboratory results show normal aminotransferase and albumin levels and normal coagulation profile, except for patients with associated cirrhosis. These results may also be abnormal in patients with long-established PVT, even in the absence of cirrhosis.[17] Albumin levels may be low and associated with ascites shortly after the episodes of UGIB. Thrombocytopenia and leukopenia are common findings.

Growth retardation

In general, children diagnosed with long-established PVT are more prone to showing growth deficit. The factor that predisposes to this manifestation is still unknown. It has been assumed that chronic anemia (secondary to losses caused by bleeding and/or hypersplenism), intestinal venous congestion with secondary malabsorption and abdominal distension may interfere with growth rate. Another hypothesis is that the low supply of blood to the liver due to the formation of systemic collaterals in PVT patients may cause hepatotrophic hormone deprivation, thus interfering with the child's global growth.[11,20,21]

Mehrota et al.[22] assessed 33 children with PVT; 55% of them were below the 5th percentile for weight-for-age and height-for-age. Based on the high serum levels of growth hormone and of somatostatins (IGF-I), they concluded that there was some resistance to the action of the growth hormone. On the other hand, Bellomo-Brandão et al. studied 24 children with EHPVO and did not observe growth retardation in any of them. They suggested that pediatric care associated with endoscopic sclerotherapy favored growth, since they reduced the consequences of PH.[23]

Portal biliopathy

The natural history of PVT encompasses the formation of numerous collaterals at the *porta hepatis* and around bile ducts, which may cause compression on bile ducts. With the progression of the disease, there may be stenosis and formation of calculi in the common bile duct and in its branches, as well as in intrahepatic bile ducts, causing secondary biliary cirrhosis. Portal biliopathy appears in adulthood. Jaundice and pain are rare, except in the presence of cholangitis. Its course is silent and progressive and is detected because of its most common complication: biliary cirrhosis.[17,24] Escalation of alkaline phosphatase to levels that are two times to five times greater than the highest benchmark value helps with the diagnosis of this biliary complication of portal vein thrombosis. Endoscopic retrograde cholangiopancreatography (ERCP) can identify

Portal vein thrombosis in children and adolescents – Schettino GCM et al.

Jornal de Pediatria - Vol. 82, No.3, 2006 **175**

portal biliopathy in 80 to 100% of affected patients, by detecting abnormalities in intrahepatic and/or extrahepatic bile ducts (localized dilatations or strictures) and in the gallbladder (wall disorders, kinking and lithiasis).[11] Therefore, according to the extension of the injury found on ERCP, it is possible to classify portal biliopathy into three types, of which type I is the mildest, as shown in Table 5.

Table 5 - Classification of endoscopic retrograde cholangio-pancreatographic findings secondary to portal biliopathy

Types	Sites of biliary involvement
I	EHBD
II	IHBD
IIIa	unilateral EHBD and IHBD
IIIb	bilateral EHBD and IHBD

EHBD = extrahepatic bile duct; IHBD = intrahepatic bile duct.
Adapted from Sarin & Agarwal.[11]

Coagulation disorders

Protein C deficiency, an endogenous tissue factor pathway inhibitor, is common among children and adolescents with PVT, but it does not seem to have a genetic etiology or to be the main cause of thrombosis. According to a study undertaken by Universidade Federal do Rio Grande do Sul, in Brazil, protein C deficiency is secondary to consumption resulting from portosystemic shunts of PH and to the possible reduction in the liver synthesis of anticoagulant factors as a result of decreased blood flow into the liver in PVT.[25]

Hypersplenism

Splenomegaly is the second major clinical manifestation of PVT. Moderate hypersplenism can be detected by leukopenia and thrombocytopenia in approximately 40 to 80% of the patients. Although there is a decrease in platelet count, platelets are functionally normal.[11,18]

Immunity

Patients may have normal humoral immunity. However, some will show abnormal cell-mediated immunity due to the sequestration of T lymphocytes by the spleen. Nevertheless, manifestations related to immunological disorders are rare among children.[11]

Rectal and esophagogastric varices

The main clinical manifestation of PVT at diagnosis is variceal bleeding. Nearly 90 to 95% of PVT patients have esophageal varices, and 35 to 40% have gastric varices. The frequency of hypertensive gastropathy, which is lower in PVT patients than in cirrhotic ones, seems to be related to the severity of liver disease.[11,26] Mortality from gastrointestinal bleeding secondary to variceal rupture amounts to approximately 2 to 5% in PVT patients.[14,18]

Anorectal varices are found in 80 to 90% of patients with portal vein obstruction. They usually are of grade III (larger than 6 mm and protruding into the intestinal lumen). They rarely bleed, but when they do, they cause severe complications. All children with hemorrhoids should be investigated as to the probability of PH.[1] Colopathy secondary to PH affects approximately 54% of patients.[27]

Usually, PVT patients have variceal bleeding in their first years of life. The risk for gastrointestinal bleeding does not seem to decrease with age. Adolescents with PVT who did not receive adequate treatment are at a high risk for bleeding in their twenties.[28]

Ascites

Ascites can be observed in extrahepatic portal vein obstruction even without being related to gastrointestinal bleeding or other triggering factors. It develops in patients with long-established disease.[17]

Diagnosis

PVT should be suspected in all children with splenomegaly, without hepatomegaly and hematemesis, with normal liver function test results. Liver biopsy is normal in children without associated cirrhosis.[17]

Abdominal Doppler ultrasonography is the most widely used diagnostic exam in pediatric patients, with a high sensitivity (s = 94-100%) and specificity (sp = 90-96%), even though it is an operator-dependent diagnostic method.[11,18] Chronicity of PVT is defined by Doppler ultrasonography by means of visualization of the formation of new vessels around the thrombus (cavernoma). Some diagnostic exams should not be routinely used in pediatric patients due to their risk: splenoportography and arterial portography, nuclear magnetic resonance (angiography), computed tomography (portogram).

All patients must be submitted to upper gastrointestinal endoscopy to check for the presence of esophagogastric varices, which will allow for a better planned therapeutic approach.

Laboratory exams show normal liver function in most patients, except in those who have a prolonged decrease in portal circulation, or portal biliopathy. Assessment of associated cirrhosis depends on abnormal histological findings in biopsied liver samples. In adults, associated diseases, such as myeloproliferative disease, malignant tumors, coagulation disorders, among others,[17] should be ruled out. There is a high predictive diagnostic value of PVT in cases of UGIB without jaundice.[29]

Differential diagnosis

It is important to establish the differential diagnosis with other causes of splenomegaly and pancytopenia, such as oncohematologic diseases, and infectious parasitic diseases.

The distinction between the various causes of PH is of paramount importance, especially between cirrhosis and PVT, so that the appropriate therapeutic approach can be implemented. Quite often, PVT patients are younger, have normal liver function, show better tolerance of variceal bleeding, do not lapse into hepatic coma, and their hemoglobin levels are lower than those of cirrhotic patients.[11,17,29]

Treatment

Treatment of acute thrombosis

In newborns submitted to umbilical vein catheterization or children with some trauma or intra-abdominal injury that predisposes to portal vein thrombosis, the first step is to determine the level of involvement of the blood vessel, so that the best therapeutic approach can be chosen.

If the child does not show any symptoms of intestinal ischemia, the following procedures should be performed:

– assess the removal of umbilical venous access in newborns;

– start thrombolytic therapy with urokinase (in case of small and localized thrombus) or heparin (in case of systemic injury). Thrombolytic therapy prevents intestinal ischemia in the short run, and extrahepatic PH in the long run.[17,18] This therapy should last for at least 6 months.

Otherwise, if the patient is symptomatic, with intestinal ischemia, immediate surgical treatment should be implemented by transjugular catheterization or laparotomy with thrombectomy.[11]

Gastrointestinal bleeding and esophagogastric varices

The approach to UGIB includes the following: 1) primary prophylaxis (patient at a high risk for UGIB, but who has never had bleeding episodes); 2) management of acute bleeding and; 3) secondary prophylaxis (to eliminate varices in patients with history of UGIB).

The use of drug therapy or endoscopy in children with esophageal varices before the first bleeding episode is controversial and there is no consensus about it in the medical literature.[6] In adults with cirrhosis and large varices, beta blockers reduce the risk of bleeding and seem to reduce the mortality associated with UGIB.[30-32] This type of treatment is already established for this age group. Esophageal variceal band ligation (EVBL) has

proved efficient as primary prophylaxis in adults, whereas the use of sclerotherapy has been discontinued due to the high incidence of complications.[33,34] The current trend consists of the use of primary prophylaxis in pediatric patients, even though there is a paucity of studies involving children.[26,35]

Due to the risks of any extensive bleeding, patients with upper gastrointestinal bleeding should be immediately sent to a referral hospital for a more adequate approach. First, the patient receives cardiorespiratory resuscitation and then bleeding varices are treated.

Children with this condition are admitted to the emergency room and necessitate blood derivative transfusion (if hemoglobin < 9 mg/dl), continuous octreotide infusion (dose of 1 to 2 µg/kg/h) and endoscopic treatment by way of elastic ligature or sclerotherapy. If bleeding persists after these measures, an option is to use the Sengstaken-Blackmore tube or a second emergency endoscopic treatment before the decision to operate on the patient.[36]

Preventing the recurrence of UGIB secondary to variceal bleeding is crucial in the follow-up of patients with PH. The treatment involves drug therapy with, for instance, propranolol (1-6 mg/kg/day, given orally, twice or three times a day), endoscopy (sclerotherapy or elastic ligature) and portosystemic shunts.[18]

Beta blockers are the only pharmacological agents with proven efficacy in reducing the risk of recurrent bleeding in adults. Studies involving children are scarce.[37,38] The dose should be gradually increased until 25% of the baseline heart rate at rest is reached. In children, the initial dose usually corresponds to 1 mg/kg/day.[38] Adverse effects and the necessity of a long-term and continuous therapy may be a limiting factor.

The management of children with bleeding complications caused by PH has changed considerably with the improvement of endoscopic techniques in the last 2 decades. Endoscopic sclerotherapy has therefore been the initial treatment of choice for children with variceal bleeding. Recently, EVBL has been more frequently used, replacing sclerotherapy, since it has a lower incidence of complications.

There are only non-controlled descriptive studies with children, comparing the efficacy of endoscopic treatments for varices (ligature and sclerotherapy). Celinska-Cedro et al. assessed 37 children with PH (15 with PVT) and concluded that elastic ligature is a safe and efficient as primary prophylaxis, regardless of the cause of PH.[39]

Zargar et al.[36] assessed recurrent bleeding rates in a randomized trial with 49 children with extrahepatic portal vein obstruction. They concluded that recurrent bleeding rates were higher in the group submitted to endoscopic sclerotherapy than in the elastic ligature group (25 and 4%, respectively, with p = 0.049). After variceal eradication,

Portal vein thrombosis in children and adolescents – Schettino GCM et al.

Jornal de Pediatria - Vol. 82, No.3, 2006 **177**

17.4% of the children submitted to elastic ligature showed recurrence of esophageal variceal bleeding, and 10% of those submitted to sclerotherapy had recurrent bleeding (p = 0.67). EVBL seems to eradicate esophageal varices in a shorter time frame, with fewer sessions, with less rebleeding and with fewer complications.[36,40]

It is recommended that an annual endoscopic follow-up be performed during the first 4 years after variceal eradication, due to the risk of rebleeding.[41]

Patients should be advised not to take ASA.

Surgical treatment

There has been a consensus agreement on the conservative treatment of patients with extrahepatic PH, thus avoiding surgical interventions. However, the following aspects should be taken into consideration: risk of death (albeit low), high risk for post-transfusion hepatitis, children's and parents' anxiety towards possible new episodes of UGIB, and cost of hospitalizations. Sclerotherapy and EVBL, although efficient in the prophylaxis against UGIB, do not eliminate PH and also have complications. Moreover, multiple procedures are often necessary for variceal obliteration, with risk of recurrent bleeding until the varices can be totally eradicated. Conversely, portosystemic shunts directly decompress the portal venous system, reducing the risk of rebleeding and of other complications, such as hypersplenism and growth retardation. In some international referral services, portosystemic shunts are widely recommended, offering good results, fewer complications and shorter hospital stay. Distal splenorenal shunt procedure and Rex shunting have been preferred.[20,42-44]

The aim of shunting procedures is to transfer blood from portal to systemic circulation. As a result of a decrease in portal pressure, there may be a reduction in blood supply to the liver and development of hepatic encephalopathy, at a lesser frequency than in cirrhotic patients.[20,42,44] The following surgical procedures should be considered:

– portosystemic shunts: distal splenorenal shunt, mesocaval shunt with jugular vein interposition, proximal splenorenal shunt with splenectomy, inferior mesorenal shunt, portocaval shunt and Rex shunt;

– TIPS: transjugular intrahepatic portosystemic shunt;

– splenectomy;

– gastrosplenic decompression.

The major indications for surgery are: 1) persistent bleeding following endoscopic treatment; 2) prominent splenomegaly, with symptomatic hypersplenism; 3) growth retardation; 4) symptomatic portal biliopathy. The distal splenorenal shunt is still the treatment of choice in cases in which endoscopic treatment does not yield good results.[45]

An alternative is the use of Rex shunting, a type of portosystemic shunt with a left mesenteric portal bypass that restores blood flow to the liver, supplying it with hepatotrophic substances and other nutrients.[43]

Orloff et al. carried out a retrospective 35-year study of 162 patients with PVT (74% aged between 1 and 16 years) submitted to portosystemic shunting (distal splenorenal shunt, proximal splenorenal shunt with splenectomy or mesocaval shunt). The cause of thrombosis was not identified in 110 of these patients (68%). Approximately 96% showed hematemesis, 94% revealed melena and 100% had anemia. All of them had at least two episodes of UGIB (and at least one of the bleedings required blood transfusion) and were referred to the authors due to prolonged treatment, without satisfactory therapeutic response. The survival rate after a 10-year postoperative follow-up amounted to 96%; 98% of the patients reported that their quality of life was remarkably improved. Only 2% had complications, such as shunt thrombosis and recurrent bleeding. No case of encephalopathy secondary to portosystemic shunting was observed.[42]

Children with scheduled splenectomy must be immunized with pneumococcal vaccine before surgery.[44] Some surgical complications, which result from the invasive procedure, include:

1) Central venous catheter infection, sepsis, atelectasis, among others, in the immediate postoperative period.

2) Recurrent variceal bleeding due to stenosis at the anastomosis or venous thrombosis, hepatic encephalopathy, intestinal obstruction, in the late postoperative period.

Despite the lower risk of encephalopathy inherent to the shunting procedure, PVT patients are at a greater risk for stenosis or thrombosis at the surgical anastomosis.

Portal biliopathy

Depending on where gallstones are located or on the site of stenosis, the clinical manifestation will determine the surgical approach that should be used. If cholangitis or cholelithiasis is present, cholecystectomy, biliary stenting or endoscopic sphincterotomy is recommended. If the patient is asymptomatic, portosystemic shunting should be scheduled in order to prevent future complications that may arise from the progression of biliopathy.[17,24]

Growth deficit

There is a paucity of studies on the growth and development of children with EHPVO secondary to PVT. Surgically treated children (portosystemic shunting) often show improved growth. Some authors recommend surgical decompression in selected children in order to recover their growth.[20,44]

178 Jornal de Pediatria - Vol. 82, No.3, 2006

Portal vein thrombosis in children and adolescents – Schettino GCM et al.

Conclusions

PVT is one of the most important causes of gastrointestinal bleeding among children. The bleeding episodes are characterized by high morbidity: frequent hospital admissions, increase in school absenteeism, emotional stress for the children and their families, in addition to high hospital expenses. Therefore, it is necessary to approach these patients appropriately so that the quality of life of children with portal vein thrombosis and of their families can be improved.

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