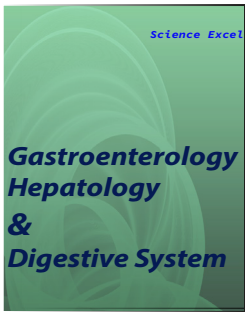
Gastroenterology Hepatology and Digestive System

Case Series



Patency of spleno-pancreatorenal shunt in infant with portal hypertension, case series and literature review

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Patency; shunt; spleno; pancreatorenal; rebleeding; ascites; Portal hypertension

Abbreviations

PHT: portal hypertension; BA: biliary atresia; C.P: child-Pugh class; SPR: spleno-prancreatorenal; CMNSXXI/IMSS: National Medical Center of XXI Century of Mexican Institute of Social Security; IMSS: Mexican Institute of Social Security

Abstract

Introduction: Portal hypertension is the main cause of morbidity and mortality in infant diagnosed of biliary atresia (BA) and cavernous deformation of the portal vein. Portal hypertension evolves as a result of increased intrahepatic vascular resistance, most commonly caused by a chronic liver disease. The clinical manifestations are ascites and bleeding owing the esophageal and gastric varices. The mortality rate per acute episode of bleeding is 5% to 19% in children. The aim of this work was to evaluate the clinical influence of rebleeding, refractory ascites, child-Pugh class (C.P) and shunt patency in infant underwent spleno-pancreatorenal (SPR) shunt in a case series in a pediatric reference medical center.

Patients and Methods: Retrospective and observational work was performed owing clinical surveillance of C.P, shunt patency, rebleeding and ascites formation after SPR shunt procedure from December 2017 to November 2020. Of 16 shunts performed in this period, 3(18.7%) patients had SPR shunt. The three patients were females and the mean weight was 6.5kg.

Outcomes and Results: The search identified three patients underwent surgical shunts of SPR less than 2-year-old. All of three patients had shunt patency in the first three months from the procedure. One patient had thrombosis of the shunt at fourth month from the operation. The three patients had improvement in the C.P class, 1 patient had infectious, rebleeding and ascites between 4 months from the procedure.

Conclusion: The three patients had no experienced shunt complications in the first three months from the procedure.

Introduction

Portal hypertension is the main cause of morbidity and mortality in infant diagnosed of biliary atresia (BA) and cavernous deformation of the portal vein [1-3]. The BA is an Obliterative process that affected the intrahepatic and extrahepatic bile ducts, and it is characterized by a progressive intrahepatic fibrosis. Meanwhile, Portal hypertension evolves as a result of increased intrahepatic vascular resistance. Consequently, PHT is leading to multiple pathological events in the sinusoidal circulation, such as structural disease by fibrosis, microvascular thrombosis, dysfunction of liver sinusoidal endothelial cells, and hepatic stellate [2-5]. Hence, the BA is the main cause of cirrhosis in children and it is occurring in 1:20,000 births. Generally, many reports have observed that two-thirds of long-term survivors with BA will have clinically definable PHT. Therefore, sustained PHT leads to the formation of esophageal, gastric, ectopic and anorectal varices. All of them are associated with hypersplenism and contributing of the development of ascites [3-7]. Portal hypertension is frequent and occurs early in these infants. Esophageal varices are found before age of two years in more than 50% of children, and gastrointestinal bleeding occurred in 22% at a median age of 17 months [3-5]. The mortality rate per acute episode of bleeding is 5% to 19% in children [1-3].

Gastrointestinal bleeding can result in acute ischemic liver necrosis, impairment of liver function or in death. It is calculated that the risk of death or the need for liver transplantation after bleeding in children with biliary atresia is 3 times higher than in children who do not bleed [3-9]. Children with intrahepatic PHT can be treated with portosystemic shunts and finally with liver transplantation as the

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definitive treatment. Hence, the main indications for liver transplantation for biliary atresia are persistent jaundice, liver cell failure as well as cardiopulmonary complications and recurrence ascites. The choice for primary prophylaxis of bleeding lies between endoscopic therapy and rapid liver transplantation, depending on the child´s liver condition [6-10]. Although, Primary prophylaxis of gastrointestinal bleeding is commonly performed in adult, however, it has not endorsed the concept of primary prophylaxis of bleeding for children with portal hypertension. Hence, the presence of deaths reinforced the importance of identifying patients who would benefit with Kasai procedure, endoscopic treatment; primary prophylaxis shunts operation or liver transplantation. Certainly, the liver transplantation is the only definitive treatment [1-10]. The aim of this work was to evaluate the clinical influence of rebleeding, refractory ascites, child-Pugh class (C.P) and shunt patency in infant underwent spleno-pancreatorenal (SPR) shunt procedure for controlling the HTP with hypersplenism, in a case series in a pediatric reference medical center.

Patients and methods

Retrospective and observational study of: clinical surveillance, C.P and patency of SPR shunt in infant (minor of 2-year-old) with variceal bleeding and ascites in patients diagnosed with portal hypertension from December 2017 to November 2020. Data collected included portal hypertension, gastrointestinal bleeding, ascites, hepatic disease, hypersplenism, C.P, endoscopic treatment, platelet count (PLT), amylase, lipase, Doppler Ultrasound, Angio tomography, SPR shunt. Patients with other type of shunts and older of 2 years were excluded in this study. Patency of the shunt, it was defined as flow presence of shunt from 24 hours after the surgical procedure and to the time of the last evaluated color Doppler ultrasound. Serum amylase and lipase reference ranges was 0-100U/I and 0-300U/I respectively. All patients had Pneumovax vaccine. Of 16 shunts performed in this period, 3(18.7%) patients had spleno-pancreatorenal shunt. The three patients were females and the mean weight was 6.5kg (Table 1).

Table 1. *Demographic and clinical characteristics of patients with Portal Hypertension (n=3)*

Case	Gender	Age	weight	Diagnostic	Bleeding episodes	Ascites	C.P	hypersplenism	Type of Shunt
1	Female	6 m	5.8 kg	BA	3	high	C	massive	SPR with injert
2	Female	8 m	6.5 kg	BA	4	high	C	Massive	SPR
3	Female	7 m	7.3 kg	Cavernous transformation	3	middle	A	Massive	SPR

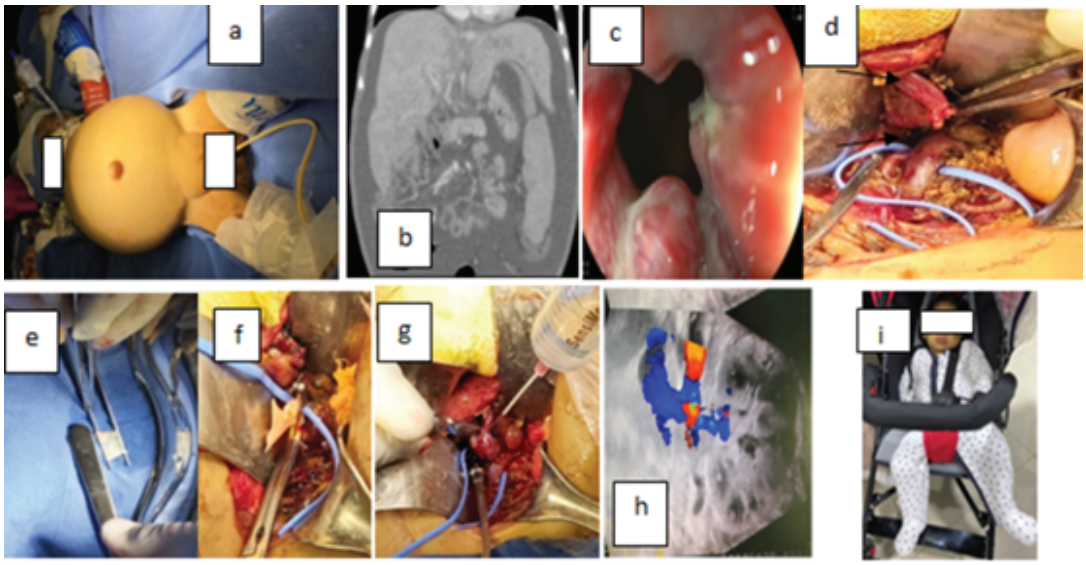

Figure 1. *a: infant of 1/6 year-old, b: varices in endoscopic study, c: renal vein (VR) and splenopancreatic vein very short, d: cadaveric vein injert, e: procedure of injert anastomosis to renal vein, f: shunt performed with the cadaveric vein injert to the splenopancreatic vein., Doppler of shunt patency, i: patient at 18 months of shunt operation.*

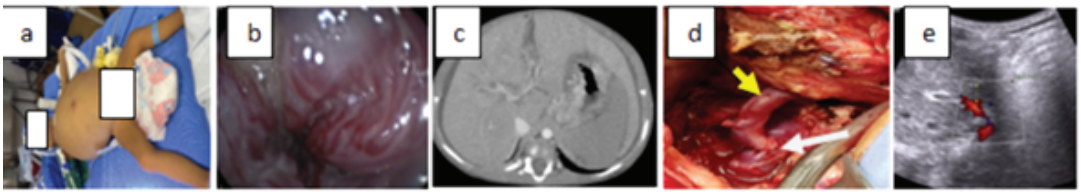

Figure 2. *a: patient 2 of 8-months-old with ascites refractory, b; endoscopic study, c; TC with liver cirrhosis, hypersplenism and refractory ascites, d; pancreas vein (yellow arrow) anastomosed to renal vein (white arrow), e; Doppler ultrasound demonstrated of shunt patency.*

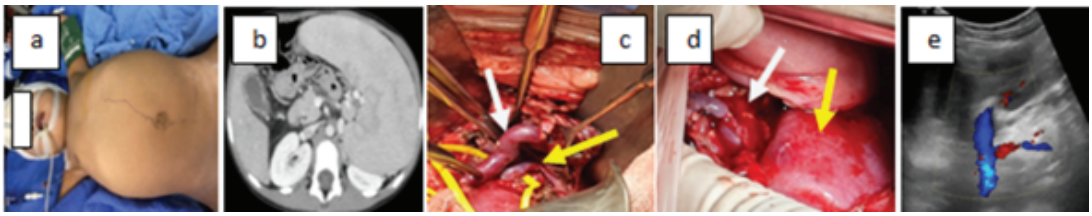

Figure 3. *a: patient 2 of 8-months-old with ascites refractory, b; endoscopic study, c; TC with liver cirrhosis, hypersplenism and refractory ascites, d; pancreas vein (yellow arrow) anastomosed to renal vein (white arrow), e; Doppler ultrasound demonstrated of shunt patency.*

Case 1

This patient had three episodes of bleeding before the shunt procedure. She had umbilical and right inguinal hernias with cirrhosis due to BA. She presented severely clinical manifestations of PHT with class C of C.P. She had the procedure with platelet level lower of 40 G/L. The levels of amylase and lipase serums before the procedure were adequately (Figure 1). In the second Case, the patient had BA as diagnosis with C.P class C and refractory ascites (Figure 2). The third case had cavernous transformation with class A of C.P, she had predominantly a massive hypersplenism, respiratory distress, and platelets counts were lower of 36 G/L. The three patients had esophageal varices (100%), gastrointestinal bleeding (100%) and refractory ascites in 2 patients (66.6%) as primary manifestations of portal hypertension (Figure 3). Combination of vasoactive drugs, antibiotics and endoscopic variceal band ligation were the managements of the three Patients without improvement.

Technical

Technique of spleno-pancreatorenal shunt: Although significant efforts have focused to ameliorate and maintained an adequate portal pressure, shunt thrombosis is a major complication after the shunt procedure. Underwent this perspective, this technique was performed by a left subcostal incision to introduce of the abdominal cavity. The hilum spleen was dissected to remove the spleen and kindly the splenic vein was dissected to the pancreatic vein, through its longitude into the pancreas tail and this part of the pancreas was opened as a book to gain a good size and longitude of the pancreatic vein (Figures 1- 3). Subsequently, the left renal vein was dissected and the shunt anastomoses were performed in the vascular bifurcation zone of the spleno-pancreatic veins to the renal vein (Figures 1-3). For the anastomosis, sutures of PDS (polydioxanone suture) or Prolene 6(0) or 7(0) were used. All patients had Doppler ultrasound control at 4 and 24 hours of the procedure and follow-up at three, six and twelve months from the shunt procedure. The surveillance of patients was observed also under endoscopic reviewing at six months from the post operatory. Exceptionally, a Cadaveric patch vein was placed to the pancreatic vein to perform the anastomosis in the case 1 due to the short longitude of the pancreatic vein (Figure 1). At the end of all shunts procedures liver biopsies were obtained.

Table 2. *Outcomes after spleno-pancreatorenal Shunt*

Case	Type of PHT	Time of shunt Complication from surgery (month)	C.P	Shunt complication details
1	Intrahepatic	none	A	None complication (infection, pancreatitis, rebleeding, stenosis, occlusion, refractory ascites and thrombosis) at 24 months of shunting
2	Intrahepatic	4 months	B	Rebleeding, Thrombosis, occlusion and refractory ascites
3	Prehepatic	none	A	None complication (Infection, pancreatitis, rebleeding, stenosis, occlusion, refractory ascites and thrombosis) at 14 months of shunting

PHT; Portal Hypertension, C.P; Child-Pugh Class

Outcomes and Results

The search identified three patients underwent surgical shunts of SPR, Who were under 2 years. The median age was 7 months (range 6 to 8 months). One of the select patients had a shunt with a cadaveric vein patch. All patients had shunt patency in the first three months of the operation. Patient number 1 continues with good flow of shunt patency at 24 months post-surgery, she had child-Pugh class A with increased platelet count (> 150 G/L). Patient 2 had shunt patency at 4 hours and three months of the procedure, she had platelet count >250 G/L; she presented an acute infectious at the fourth month with bleeding and thrombosis of the shunt. Patient 3 had patency of the shunt with good flow at 4 hours and 3, 6 and 14 months after the procedure; this patient continue with Child-Pugh A and normal platelet count (Table 2). In the postoperative setting the three patients had not experienced shunt complication (occlusion or stenosis) in the first three months.

Discussion

The portal venous system is the unique circulatory system which connects two systems of capillary beds. Hence, portal hypertension is depended of the resistance increase of the portal blood flow owing at the morphological changes that occurring in chronic liver disease and the flow. Furthermore, it has demonstrated that factors such as intrahepatic vascular resistance, blood flow in the splanchnic and systemic circulation in liver cirrhosis had contributed to portal hypertension. On the other hand, Hepatic endothelial dysfunction occurs early in the course of chronic liver disease as a consequence of inflammation and oxidative stress. Hence, the normal phenotype of liver endothelial cells becomes: proliferative, prothrombotic, Proinflammatory and vasoconstrictor effects [9-12]. Moreover, The mechanism that involve in the cirrhotic liver are: 1- distortion of the liver vascular architecture that causes structural abnormalities (nodule formation, remodeling of liver sinusoids, fibrosis, angiogenesis and vascular occlusion), 2- increase of the hepatic vascular tone due to sinusoidal endothelial dysfunction, which results in a defective production of endogenous vasodilators, mainly nitric oxide, and increased of the production of vasoconstrictors as thromboxane A2, cytenil leukotrienes and angiotensin II [10-12]. All of the anterior contributes to the pathogenesis of portal hypertension and those are present in biliary atresia of the cases 1 and 2 in this work [12-17]. On the other hand, Portal vein Obstruction is the single most common etiology of portal hypertension in children and representing roughly 50% of all cases in the majority of series. Portal obstruction in children is usually detected early in the first decade, because the splenomegaly and gastrointestinal bleeding, or both are manifested very early as it was observed in the case 3 of this study. There is a prevalence of bleeding in patient with grade II or III of varices, as it was observed in the case 3 with diagnosis of cavernous transformation of portal vein. Probably, bleeding is directly correlated with the size of varices as seen on endoscopic images. However, variceal bleeding is generally well tolerated, owing a normal function of the liver [18-21].

Portal hypertension is frequent and it is associated to early esophageal varices that are found before two years-old in more than 50% of children, followed gastrointestinal bleeding in 22% at a median age of 17 months [4,18-20]. Similarly, in this work the severely clinical manifestations of PTH were presented before the first two years old. Over more, gastrointestinal bleeding can result in death, acute ischemic liver necrosis or impairment of liver function. In fact, the patient of the case 2 had rebleeding at 3 and 4 months of the procedure and she had thrombosis of the shunt after the second episode of rebleeding. Hence, it observed in this study the mentioned in the literature, that the risk of death or the need for liver transplantation after bleeding in children with biliary atresia is 3 times higher than in children who do not bleed [4-8,21]. Hence, the main indications for liver transplantation for biliary atresia are persistent jaundice, liver cell failure as well as cardiopulmonary complications and refractory ascites [7,21]. On the other hand, esophageal variceal bleeding (EVB) is a severe complication of portal hypertension with significant morbidity and mortality. In fact, 10-20% of the patients will have persistent or recurrent bleeding that required an alternative rescue as it was observed in this case series [4,18-20]. The choice for primary prophylaxis of bleeding lies between endoscopic therapy and rapid liver transplantation, depending on the child´s liver condition [1-6,18-20]. Even though, in few cases, it is inappropriate to perform the liver transplantation. Owing this condition, it is important to find alternative treatments as the early use of TIPS or shunt procedure. However, shunt procedure had high risk of thrombosis in pediatric patients at 2 years from the shunt operation, moreover in infant patients [1-5,19]. Performed spleno-prancreatorenal shunt at this age may be used as an alternative treatment in this group of patients with significant short time of rebleeding and high rate of mortality, as it was observed in the case 2 of this work. In fact, the main concern in the management of variceal bleeding is to reduce the recurrence of bleeding episodes.

On the order hand, the role of lymphatic vessels in the liver and the formation of ascites are largely unknown. However, few reports mentioned that the lymphatic system plays a central role in ascites and edema formation. Nevertheless, ascites occurs when osmotic and hydrostatic pressure within hepatic and mesenteric capillaries exceeds the drainage capacity of lymphatics and excess fluid accumulates in the peritoneal space. The prior is manifested as weight gain, abdominal distension, a fluid wave and ballotable spleen [17,18]. In fact, the three patients of this series had different levels of refractory ascites and infection that would be associated to the development of refractory ascites prior at the shunt procedure [18-21]. Moreover, it has been known that the bacterial translation is closely related to the development of ascites and infection. In this work; infectious, refractory ascites and hepatic encephalopathy had not observed during the follow-ups after the Spleno-pancreatorenal shunt. In fact, it is considered as the consequence of dilated peribiliary venous plexus (cavernous) in the wall of biliary ducts. Furthermore, affected patients as they were observed in cases 1 and 2 of this work; exhibit disturbance of the liver with dilated bile ducts, mainly intrahepatic [1-3].

In all of three Patients, blood flow signals were notes in the first fourth months and thrombosis was not observed in the shunt operation throughout five months after spleno-pancreatorenal shunt. Hence, the surgical procedure to controlling the portal hypertension could be classified as total, partial, and

selective. Total shunts have a diameter more than 10 mm that constructed between the main veins of the portal system and the inferior vena cava. This shunt provides excellent control of hemorrhages and ascites, however all of them at the high cost of encephalopathy. Partial shunts comprise portocaval o mesocaval anastomoses of 8 mm in diameter or less, allowing part of the portal flow to reach the liver sinusoids; finally, the selective shunts that are constructed by the anastomoses of the splenic vein to the left renal vein reduce systemic complication. Hence, in this work it was observed that the diameter of the confluence of the pancreas and splenic veins was around 7-8 mm that permits to perform the shunt at this age. Consequently, three mains' factors influence the therapeutic Choice of this work: 1- ages of the patients, 2-etiology of the liver disease and 3-weight of the patient. Indeed, three patients between the ages of 5 and 7 months had managements underwent this type of shunt. In fact, cases 1 and 2 were biliary atresia of diagnosis and case 3 was cavernous transformation; the middle weight of the three patients was 6.5 kg. Hence, the portal hypertension was controlled after the shunt procedure. Although, cases 1 and 2 display signs of cirrhosis with persisting clinical and biochemical signs of cholestasis within 4 and 18 months respectively from shunt procedure [1-6,19-21]. On the other hand, Splenorenal shunts archive good hemorrhagic control and reduce systemic complication. However, in infant this type of shunt presented high frequency of thromboses due to the diameter of vein size [19-21]. Hence, the spleno-pancreatorenal shunt performed of this work demonstrated good patency higher of 18 months of the follow-up in 66.6% of patients and it is observed improvement of clinical signs of rebleeding and ascites formation. In this study, one patient had thrombosis at the quarter month from the surgical procedure. However, the number of patients is too small to statically assess the shunt vessel obstruction.

Conclusion

Complications of portal hypertension cause significant morbidity and mortality in children moreover in infant. Unavailable the liver transplantation, it is important to find surgical options with low rates of morbidity and mortality as the spleno-pancreatorenal shunt to ameliorate portal hypertension as it was observed in this work, moreover in the patients 1 and 2. The three patients had improvement in the C.P class and clinical manifestations of the portal hypertension at three months of the shunt procedure.

Site

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Conflicts of interest

There are no conflicts of interest to disclose

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