

## Review of Meso-Rex Bypass in Pediatric Management of Portal Hypertension: Case Report

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### Abbreviations:

PH: Portal Hypertension; PHPH: Prehepatic Portal Hypertension; MRB: Meso-Rex Bypass; National Medical Center of XXI Century of Mexican Institute of Social CMNSXXI/IMSS; PSS: Portosystemic shunt

## 1. Abstract

**1.1. Introduction:** Portal hypertension (PH) is defined as pressure that is higher than normal values in the portal vein. Grand parts of the alteration of PH are founded in: the portal resistance, the flow, and the hepatic disease. Prehepatic portal hypertension (PHPH), related to thrombosis and the cavernous transformation are the major causes of PH in children that accounting for up to 75% of cases. Meso-Rex Bypass (mRB) is the best treatment of PHPH and had never been performed in this Pediatric Hospital Center. Herein, in this work we print the case report following the outcome of a female patient under this procedure.

**1.1.1. Case Presentation and Follow-up:** A 7-year-old girl presented with PHPH from cavernous transformation with grade 3 esophageal varices, massive hypersplenism, thrombocytopenia, encephalopathy and progression of liver dysfunction. The protocol of mRB was performed with Doppler Ultrasound of the recessus, jugulars veins and the angiotomography revealed the diagnosis of cavernous transformation. She went on to undergo the mRB and liver biopsy. Postoperative control and monthly, ultrasounds Doppler revealed a patent shunt. Splenic size decreased slightly and the platelet count normalized after 14 months of follow-up, the patient has not had any further of upper gastrointestinal bleeding episodes. The last Doppler ultrasound and angiotomography revealed a patent shunt and clinically

she is doing well without postoperative complication.

**1.2. Conclusion:** Management of PHPH has changed drastically over the past decade after the widespread introduction of the mRB. This surgical procedure might be restoring normal hepatic circulation and control the PH.

## 2. Introduction

Portal hypertension (PH) is defined as pressure that is higher than normal values in the portal vein. In fact, this pressure is rarely measured directly. Over the physiopathology of the PH, grand parts of the alteration are founded in: the portal resistance vein, the flow, and the hepatic parenchyma disease. Eventually, there are also negative pump effects that resulting from direct drainage into the right atrium and its location at the thoracoabdominal interface with its alternative positive abdominal and negative thoracic pressures [1-4]. Prehepatic portal hypertension (PHPH), related to thrombosis and the cavernous transformation or portal cavernoma are the major causes of portal hypertension in children, accounting for up to 75% of cases in developing countries. Since, the extrahepatic portal vein obstruction due to thrombosis or cavernoma formation of portal vein is a chronic entity with a frequency of 1-3 % of patients with portal hypertension from liver cirrhotic. Moreover, it can be secondary to the direct damage related at the neonatal catheterization of the umbilical vein, the latter condition represents <25% of the etiology [1-4].

Congenital or acquired extrahepatic portal vein obstruction is not an uncommon cause of portal hypertension. Even less commonly, it related to regional trauma, tumors, liver transplantation, infection as peritonitis and abscess [2-5]. Few cases of portal thrombosis may present after liver transplantation and, it is estimated that 1 to 8.5% of the pediatric patients will develop portal vein complications. The processes will more observed than likely start in an inverse manner, with the thrombus initiating within the liver as a consequence of the direct damage of the intrahepatic vein and, more precisely the left portal system; overall in childhood who had catheterization of umbilical vein [1-5].

Typically, idiopathic extrahepatic portal vein obstruction is more frequent in childhood that presents symptoms of portal hypertension and moderate gastro esophageal hemorrhage. Usually, this hemorrhage is related to: variceal gastrointestinal bleeding, hypersplenism, protein losing enteropathy, malabsorption, neurocognitive dysfunction, and growthretardation [3,4]. The treatment included in the first step medical management with beta blocker and endoscopic procedure to control varices. In the second step, occasionally splenectomy may be performed for refractory hypersplenism with portosystemic shunt when conservative measures are failed. Never lest, this procedure did nothing restored normal portal circulation [1-6]. The mesenteric-left portal veins bypass meso-Rex Bypass (mRB) or Rex Shunt introduced a curative approach to extrahepatic portal vein obstruction and changed the treatment strategy advocate by an expert consensus. As any other procedure, mRB is not exempt from complications. Failure of this procedure due to occlusion has been reported to occur in 10% to 40% of patients at a 6-month follow-up. Early thrombosis leading to MRB failure is believed to be related to a variety of factors as: inadequate graft type for bypass creation, undiagnosed hypercoagulable disorders that can lead to intra luminal thrombus formation, poor patient selection, bypass contraction or kinking, decrease of the intraluminal area, and portal perfusion steal phenome [2-6]. However, meso-Rex Bypass is the best treatment of EHPH and had never been performed in this Pediatric Hospital Center. Herein, in this work we print the case report following the outcome of a female patient with PHPH under this procedure.

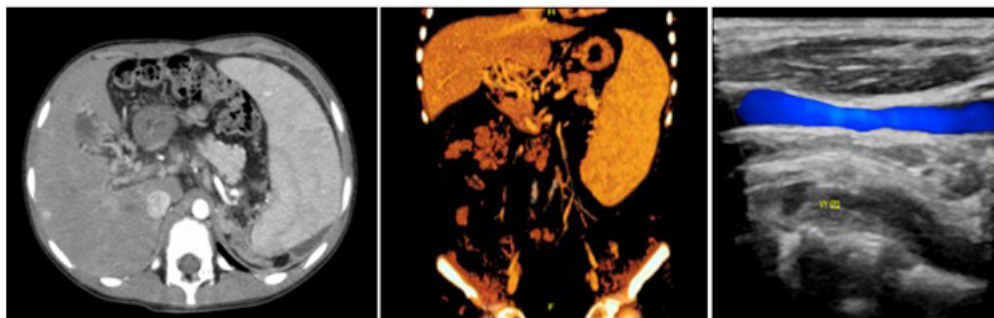
### 3. Presentation of Case

A 7-year-old woman presented for evaluation of recurrent bleeding, encephalopathy. At age 6, she was diagnosed as having portal vein thrombosis due to cavernous transformation. She had medi-

cal treatment underwent numerous endoscopic procedures, including banding and sclerotherapy of varices. The patient had massive hypersplenism, with moderate thrombocytopenia, portal-systemic encephalopathy. At the physical exploration she had data of portal hypertension and massive hypersplenism. Protocol studies were performed with Doppler ultrasound that revealed permeability of internal jugular veins and the angiogram confirmed the diagnosis of cavernous transformation. Portal venous perfusion of the liver was graded angiographically on the venous phase. Liver biopsy was taken at the time of the operation and it was examined for hepatitis, cirrhosis, fatty change and atrophy. The patient required a meso-Rex bypass (MRB) procedure that should be redirect the portal blood flow into the intrahepatic portal system with a minimum risk of portal encephalopathy as treatment, this procedure should also diminish and normalized the portal hypertension at the same time.

### 4. Technique

Step one. Commonly in this procedure a media-line incision is adequate. However, in this case the incision was bilateral sub costal upper umbilicus. After division of the round ligament and the falciform ligament, the liver edge is then suspended by 2 large stitches placed on the right and left of the umbilical scissure, exposing the surgical site for easier preparation of the Rex-recessus. As that point, a needle inspection and portography were performed operatively, this ensures that the vein is patent. Even when the scissure is open with no parenchyma bridge, a portion of the liver (from both segments III and IV) has to be resected (Figure 2). This is done to avoid compression of the bypass, by the edges of the scissure, after the completion of the bypass and closure of the abdomen. Careful dissections provide excellent exposure of the vein, 3 to 4 cm in length without any damage to the biliary or arterial branches that cross in the hilar fibrous plate and that are gently pushed away during the maneuver. Step two. The mesenteric vein was identified and dissected. The left intern jugular vein was identified and resected as auto graft for the bypass. Subsequently, the anastomosis end to side to this graft with mesenteric vein was performed with 7-0 monofilament suture. The mesocolon window was performed to pass the graft under the transverse colon and the anastomosis end-to-side of the SMV to the recessus with previous clamping of the last, was performed using 7-0 continuous monofilament sutures. At unclamping hepatopetal flow was observed immediately. On time, biopsy of the liver and ligation of short stomach veins were performed and, Doppler ultrasound revealed the flow of the bypass. Finally, the abdominal pared was closed.



**Figures 1:** images a and b revealed cavernoma transformation and massive hypersplenism, image c revealed the permeability of the jugular vein.

#### 4.1. Follow-Up

Intraoperative Doppler ultrasound for evaluation of the vessel flowing anastomoses revealed excellent flow. Postoperative recovery was rapid without variceal recurrent, intramuscular anticoagulant therapy was discontinued and replaced by acetylsalicylate orally. The average hospital stay was seven days. Clinical follow-up of meso-Rex bypass were observed in resolution of portal hypertensive bleeding and hypersplenism, as well as changes in liver synthetic function, ammonia levels, somatic growth, Platelet count and, ammonia level. Respectively, the patient had a good outcome later of two days after the procedure, concerning the variables of interest in this case, as platelet count, serum ammonia level, INR, AST and ALT. Splenic size decreased slightly after the procedure. The girl had 16 months from the procedure without episode of variceal bleeding recurrence, her endoscopic review report grade 1 of variceal that not required banding of themselves. She continued with low doses of B blocker (propranolol) and, she may continue with her activities without dyspnea. Grow parameters are going better about her weigh-for-age and height-for-age standard derivation. Neurologically, she is going well at the school with improvement, without clinical and vascular complications. Control Doppler ultrasound examinations were performed on day 7, at 1, 3, 6 and 12 months after the surgical procedure and each sixth month thereafter. The last was to assess the shunt patency at the mesenteric anastomosis, the left portal vein anastomosis. GI endoscopy was performed at 6 and 12 months from the procedure and continuous each year thereafter with an angiotomography.

#### 5. Discussion

The portal hypertension is usually defined by observation of series pathological changes and complications. More than half of patients with Portal vein obstruction present before the age of 15 years with related symptoms such as hematemesis or signs of splenomegaly. Portal vein thrombosis as an underlying cause of PHPH and cavernoma formation might be related to umbilical vein catheterization, septicemia, advanced omphalitis and, exchange transfusion and splenectomy trauma. Furthermore, Portal hypertension might be presented as an isolated finding or in combination with biliary atresia and cirrhosis. Therefore, consequences of extrahepatic portal vein obstruction include variceal bleeding and hypersplenism related to the portal hypertension as well as metabolic abnormalities secondary to impaired venous circulation [2-5,7]. In fact, this patient presents

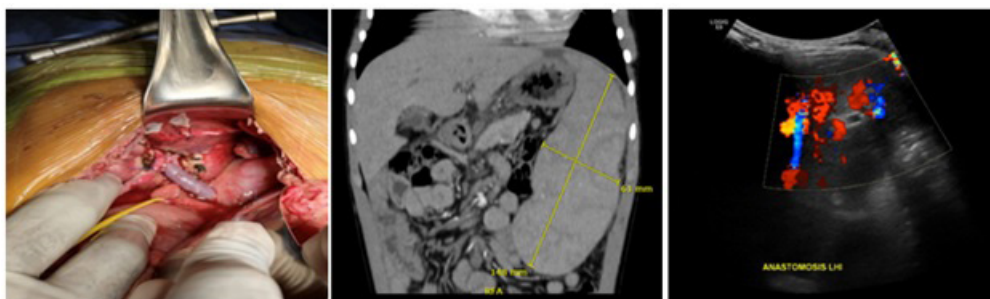
cavernoustransformation with portal hypertension and more than four episodes of variceal hemorrhages. On the order hand, some authors advocated mental changes which followed portal-systemic shunting in children with thrombosis of portal vein with normal liver [3-5,8]. Furthermore, Impaired synthesis of liver dependent coagulation factors is a well –reported alteration in portal blood likewise affects the ability of the body to detoxify false neurotransmitters as ammonia. Studies have shown that between 25% and 45% of children and young adults with PHPH have low-grade or minimal hepatic encephalopathy testing. Hepatic encephalopathy correlates with serum ammonia levels, causes cerebral edema and objective cognitive decline that persists for a long-term in the vast majority of untreated patients. Furthermore, patients with obstruction of portal vein have an elevation of Proinflammatory cytokines, which correlates with the extent of hyper ammonia. In fact, the patient of this work had grade 1 of encephalopathy, somatic grow impairment and massive hypersplenism as it might observed in figures 1 and 3. Even though, the extent of liver dysfunction in children with EHPV is variable, and may be influenced by the amount of collateral circulation that exists between the cavernous transformation and the intrahepatic portal system. Symptoms of liver dysfunction can include abnormal synthesis of coagulations factors, elevated serum ammonia with or without symptoms of hepatic encephalopathy, and /or somatic growth impairment [4,5,8-10]. Usually, the treatment underwent Beta-blockers, with their unproven value and repeated endoscopic procedures such as, sclerotherapies or bandings in some reports had been observed that there are neither curative as it was observed in the preoperative outcomes of this patient. However, some authors may choose these palliative procedures over curative surgery in the vast majority of children with symptoms [3-7,11,12]. The meso-Rex bypass (mRB) is the effective surgical treatment. This accomplishment is performed by using a conduit to connect the mesenteric venous system to the left portal vein in the space of Rex. Furthermore, evidence suggests that mRB have the additional benefit of improvement in platelet count international normalized ratio (INR) and somatic growth; as it was observed in the follow-up of this patient. Since, this autologous venous conduit is preferred in mRB, with the most common conduit being the internal jugular vein; however other options such as an autogenous saphenous vein, splenic vein, right gastroepiploic vein, inferior mesenteric vein or umbilical vein may have used. Therefore, in this patient the left intern jugular vein was used with good improve-

ment in the surgical procedure. Fortunately, this girl continued with improvement of the abdominal distension for the hypersplenism, the portal hypertension and, she had progressive increase of platelets account [1-5,8-13]. The Rex shunt restores hepatopetal portal flow and avoids the neurologic side effects of portosystemic shunts. In fact, it is essential that newer techniques, such as the MRB, achieve similar success at preventing rebleeding. This relatively new surgical procedure is the first time that it was performed in this Pediatric hospital Center. Even though, in 1992, de Ville De Goyet was the first person to perform a mesentericoportal (Rex) shunt for managing portal hypertension in a liver transplant patient [1,4,9,10]. Comparative studies demonstrated that mRB and portosystemic shunt (PSS) effectively relieve symptoms of portal hypertensive bleeding in children with PHPH, although the mRB relieves better hypersplenism. By restoring normal portal venous circulation flowed with metabolic benefits [1,4, 9-13]. In fact, the autologous left internal jugular vein was used as conduit in this case and, the capacity of a primitive intrahepatic portal venous system was sufficed to accommodate the increase blood flow immediately after mRB redirecting blood flow back into the low-pressure system of large collateral vessels (Figures 2). Other modalities of portal shunt as splenorenal shunt are not recommended as the first selection treatment. Since, this shunt might perform as a conventional portosystemic shunt and it reduces liver perfusion and it is associated with a risk of portal encephalopathy. On the other Hand, the classic shunts (portocaval, mesocaval and central splenorenal) control variceal bleeding by greatly reducing the portal pressure and shunting the portal flow away from the variceal bleeding. Unfortunately, this physiologic change also shunts portal flow away from the liver and increases the absorption of ammonia and other substances from the gastrointestinal tract. The physiologic superiority of the selective shunt is, that it maintains hypertension in the main

portal venous bed. This ensures continuing perfusion of the liver via the portal venous collaterals, and further inhibits the development of encephalopathy by its effects on intestinal absorption [2-4,12-14]. The meso-Rex bypass was recently recommended as the first-choice surgery for children with extrahepatic portal hypertension, a healthy liver, and a patent Rex Recessus, overall TIPS (trans jugular intrahepatic portosystemic shunt) placement is considered difficult in children. The mRB procedure restores the hepatopetal portal flow and intrahepatic portal system flow that reverses the commonly observed coagulopathy [4-9,12,15]. Moreover, this bypass normalizes hyperammonemia, improves neurocognitive ability, reverses encephalopathy caused by portosystemic connections, and improves somatic growth in patients who had growth retardation, reversed hepatopulmonary syndrome and related renal disturbance. This procedure might also prevent formation of liver nodules and adenoma related to portosystemic shunting. In fact, those improvements had been observed in the outcome of this patient within her first year from the surgical procedure. On the other hand, symptomatic patients whose anatomy is not amenable to the mRB, such as those with an occluded left portal vein, and patients who develop uncorrectable shunt stricture or thrombosis after mRB, are offered a distal splenorenal shunt and in rare cases a mesocaval shunt [4,8,15]. Therefore, by restoring mesenteric blood flow to the liver, mRB has been shown to normalize a number of these liver metabolic functions indices [4,11]. However, portosystemic shunts in contrast, are expected to further divert mesenteric blood flow way from the liver, exacerbating the signs and symptoms that are improved after mRB [4,8-10]. hence, the symptoms of hypersplenism and recurrence of variceal bleeding are not relieved. Therefore, only the restorative mRB has the potential to prevent or even reverse complications owing to abnormal portal circulation, as it was observed in the outcome of this patient [11-15].



**Figures 2:** images a and b revealed the position and measure of the left intern jugular vein; image c, demonstrate the jugular vein anastomosed.



**Figures 3:** postoperative image A shows the shunt with jugular vein and the image b shows decreased dimensions of the spleen(arrow) and the patency of the shunt in the Doppler ultrasonography(arrow).

## 6. Conclusion

The management of prehepatic portal hypertension has changed drastically over the past decade after the widespread introduction of the mRB. This surgical procedure might be restoring normal hepatic circulation and control the portal hypertension as it mentioned in the literature. We now advocate a very proactive approach to management of children with PHPH may be evaluated for mRB as soon as after diagnosis. Portosystemic shunts remain a reasonable alternative in children with advanced symptoms of portal hypertension with anatomic constraints preventing mRB. This mRB is the first performed in our institution and in the country and it is a feasible treatment option as we may observe in the literature of portal hypertension.

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