

## Surgical Shunt Procedures in Childhood Portal Hypertension: A Review Article

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Received: 20 Apr 2021  
Accepted: 08 May 2021  
Published: 16 May 2021

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### Citation:

Erikci VS. et al., Surgical Shunt Procedures in Childhood Portal Hypertension: A Review Article. Clin Surg. 2021; 5(10): 1-3

### 1. Abstract

Normal portal pressure is between 0 and 10 mm Hg and the pressure in the portal vein is slightly higher than that of the pressure in the inferior vena cava [1]. Portal hypertension (PH) is usually defined as either a hepatic venous pressure gradient greater than 5 mm Hg, or hepatic venous wedge pressure greater than 10 mm Hg [2]. It is usually encountered as a complication arising from chronic liver disease and cirrhosis. Common presentation of PH in children include catastrophic variceal hemorrhage usually from esophagus. Other common clinical features of PH include splenomegaly, hypersplenism, ascites, encephalopathy, hepatopulmonary syndrome and Porto pulmonary hypertension.

It has been reported that up to 15% of children with PH ultimately require shunt surgery [2]. Traditionally shunt surgery was a treatment option for children in whom control of variceal bleeding failed however it was associated with relatively high rate of anastomotic stricture or thrombosis [1]. Nowadays it has also been reported that as the experience in vascular and transplant surgery together with microsurgical techniques have improved good success rates can be achieved even in small children [1]. In this review article it is aimed to review the surgical treatment options in children with PH with special regard to shunt procedures under the light of relevant literature.

### 2. Shunt Surgery in Children with PH

Indications for shunt surgery in children are depicted in Table 1 [1, 3, 4]. There are variety of surgical techniques available. Common feature of all these surgical treatment options is to decrease the portal pressure in order to avoid complications of PH especially

life threatening esophageal varices [5-8]. Portosystemic shunts may be selective or non-selective [9, 10].

**Table 1:** Indications for shunt surgery in children with PH

Uncontrolled bleeding from esophageal varices*
Bleeding from gastric or ectopic varices
Hypersplenism or massive splenomegaly
Isolated extrahepatic portal vein obstruction
Lack of access to endoscopy
Symptomatic biliary obstruction due to choledocal varices
Neurological impairment
Patient choice

\*No response to at least 2 sessions of endoscopic treatment

### 3. Selective Shunts

For the sake of decompressing esophageal varices, these shunts divert blood from abdominal cavity to the low-pressure systemic venous circulation.

#### 3. Distal Splenoreal Shunt (Dean Warren Shunt)

In this kind of shunt, some Porto petal blood flow is maintained in order to avoid encephalopathy. The technique includes division of splenic vein and anastomosis of distal stump into the left renal vein. Clinical importance is that after performance of distal splenorenal shunt, spleen size decreases, hypersplenism improves and platelet and leukocyte counts normalize [2, 11, 12]. It has been reported that as the time passes selectivity in these shunts is lost and selective shunts progressively centralize [1, 10, 13, 14]. Higher than 90% long-term patency rates have been reported [1].

#### 4. Mesenterico-Left Portal Bypass (Rex Shunt)

This is an example of Porto portal shunt and was first described in 1992 for a patient with portal vein thrombosis following liver transplantation [14]. In this selective shunt surgery, in order to restore blood flow to the liver and bypass portal vein obstruction, a communication using a vein graft between the superior mesenteric vein and left portal vein at the level of Rex recess is constructed. Internal jugular vein is an example of these vein grafts used in these cases resulting in best results [15-18]. It has been proposed recently that this surgical option should be performed earlier in the course of the disease (1). On the other hand, in a more recent meta-analysis comprising 22 studies including 461 children with PH who underwent shunt surgery either with portosystemic (PSS) shunt (n=121) or Rex shunt (n=340), it was reported that Rex shunt was associated with higher post-shunt thrombosis compared to patients who underwent Porto systemic shunt surgery and it was concluded that PSS when compared to Rex shunt might offer advantages to pediatric patients with extrahepatic portal vein obstruction [19].

#### 5. Nonselective Shunts

Common feature of these shunts is the direct communication between portal system and the systemic circulation. Benefit of these shunts is the reduction of the risk of bleeding from varices but compared to selective shunts risk of hepatic encephalopathy is a significant matter.

#### 6. Portocaval Shunt

Direct communication between the portal and systemic circulation in this shunt is performed by the direct anastomosis of lower stump of portal vein after closure near to the liver to the inferior vena cava. Full diversion of blood flow within the portal system to the systemic circulation can be obtained by this surgical intervention but the risk of hepatic encephalopathy is again a significant risk.

#### 7. Proximal Splenorenal Shunt

In this shunt procedure spleen is removed after the splenic vein is divided. Anastomosis between the splenic vein stump and left renal vein is performed end-to-side in fashion [20]. Post splenectomy sepsis is a major concern in this kind of Porto-systemic shunt together with shunt stenosis and thrombosis. Intraperitoneal adhesions and propagation of the thrombosis in the splenic vein into the portal venous system may exacerbate PH. Liver transplantation if necessary in future may also be jeopardized.

#### 8. Mesocaval Shunt

The usual mesocaval shunt is an "H" type inter-positional graft using a prosthetic conduit such as Dacron/GoreTex (Drapanas shunt) or autologous vein graft although this kind of shunt may also be performed direct side-to-side mesocaval anastomosis after a sufficient mobilization of both superior mesenteric vein and inferior vena cava [1, 21]. Auvert shunt is another variant of mesocaval

shunt using anastomosis between iliac veins and superior mesenteric vein [22]. Other nonselective shunts include Sarfeh and Mitra shunts. In Sarfeh shunt a GoreTex graft is used after massive collateral ligation while in Mitra shunt a side-to-side splenorenal anastomosis is performed preserving spleen and the anastomosis is performed between splenic vein and left renal vein [1, 7].

In conclusion, the spectrum of surgical shunts in children with PH has a wide range. All of these procedures focus on the prevention of catastrophic variceal hemorrhage usually from esophagus. Non-selective shunts have the risk of postoperative encephalopathy. Although Rex shunt has a benefit of maintaining the physiologic portal venous flow through the liver, it has been suggested that portosystemic shunts, even if they have higher stenosis rate but lower thrombosis rate with regard to Rex shunts, may be a useful treatment option in children with extrahepatic portal vein obstruction and the long-term outcome of children with PH will be better than those previous cases.

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