

TABLE 1: Common causes of portal hypertension in children and suggested management.

Cause	Treatment	Comment
Portal vein obstruction	(1) Endoscopic	Endoscopic treatment consists on elastic banding or sclerotherapy
	(2) Meso-Rex shunt	
	(3) DSR or mesocaval shunt	
Biliary atresia	(1) Endoscopic	Screening at age of 1, prophylaxis in high-risk varices
	(2) Liver transplantation	
Cystic fibrosis	(1) Endoscopic	Need repetitive anesthetics
	(2) DSR or meso-caval shunt	Risk of pulmonary complications and worsening encephalopathy
	(3) Liver transplantation	When good respiratory function
Congenital hepatic fibrosis	(1) Endoscopic	When recurrent cholangitis (need to consider liver and kidney transplantation)
	(2) DSR or meso-caval shunt	
	(3) Liver transplantation	
Other cirrhosis	(1) Endoscopic	If good liver function
	(2) DSR or meso-caval shunt	
	(3) Liver transplantation	In end-stage liver disease

DSR: distal spleno-renal shunt.

when feasible. Once again, both procedures are equally effective, but when used in an emergency scenario their results are less satisfactory [47]. TIPS has the advantage of avoiding a laparotomy, but its availability is limited to specialized services and is not suitable for small children, especially in cases of portal vein obstruction or biliary atresia, which are the main causes of variceal hemorrhage among pediatric patients [38]. The choice of the surgical technique, on the other hand, depends on the medium-term prognosis of the disease. Shunting procedures are preferred in patients with relatively well-preserved liver function, like those with portal vein obstruction, congenital hepatic fibrosis, or compensated cirrhosis. Liver transplantation needs to be considered for children with more advanced disease.

4.3. Prevention of Rebleeding (Secondary Prophylaxis). Once the first bleeding has occurred, there is a substantial risk for rebleeding in the next years; consequently, eradication of esophageal varices becomes a logical goal. Endoscopic variceal ligation and sclerotherapy have been reported to be equally successful in achieving this. Variceal ligation is usually preferred because of its reported simplicity, lesser number of sessions needed, and a safer profile when compared to sclerotherapy [45, 46]. Both techniques are complementary and have been used even in primary prophylaxis with good results [5, 20].

An observational study in children with portal hypertension, of several different etiologies, showed a benefit of secondary prophylaxis in avoiding esophageal varices bleeding. In this study, the use of propranolol did not affect results of endoscopic prophylaxis [48]. In contrast, a large study including mainly adolescents did not find

differences between propranolol and endoscopic ligation in the recurrence of bleeding [49].

Longer followup of endoscopic treatments is available, showing recurrence of esophageal varices in 40% of the patients, with a tendency to worsening of gastric varices, portal hypertensive gastropathy, and rising incidence of ectopic varices, all of them representing a more difficult problem to solve [50]. Progression of the spleen size and late incidence of complications like portal cholangiopathy in patients with portal obstruction, formerly considered a rare entity, affect children quality of life. Moreover, for these complications endoscopic treatments are clearly unsuitable [51]. In those cases, or when hemorrhagic episodes are refractory to other treatments, surgery becomes the only option [52].

Shunt procedures could be classified as total, partial, and selective. Total portosystemic shunts are those more than 10 mm in diameter, constructed between the main veins of the portal system and the inferior vena cava. They provide excellent control of hemorrhages and ascitis, but at the high cost of encephalopathy, and are rarely used in children. Partial shunts comprises portocaval or mesocaval anastomoses of 8 mm in diameter or less, allowing part of the portal flow to reach the liver sinusoids, and thus reducing the risk of systemic complications without losing efficacy for the prevention of further bleeding. This type of shunts has been widely used in children employing the internal jugular vein as a graft, with excellent results [53, 54]. Selective shunts are constructed by the anastomoses of the splenic vein to the left renal vein, thereby decompressing gastroesophageal varices through the short gastric veins (distal splenorenal shunt), and maintaining portal perfusion to the liver. Spleno-renal shunts achieve good hemorrhagic control and reduce systemic complications.