Surgical management of portal hypertension in children

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The management of children with portal hypertension has dramatically changed during the past decade, with an improvement in outcome. This has been achieved by improved efficiency of endoscopic variceal control and the success of liver transplantation. Emergency surgical shunt procedures are rarely required, with acute bleeding episodes generally controlled endoscopically or, occasionally in adults, by interventional radiological procedures. Portosystemic shunts may be considered as a bridge to transplant in adults but are rarely used in this context in children. Nontransplant surgery or radiological interventions may still be indicated for noncirrhotic portal hypertension when the primary cause can be cured and to allow normalization of portal pressure before liver parenchyma is damaged by chronic secondary changes in some specific diseases. The meso-Rex bypass shunt is used widely but is limited to those with a favorable anatomy and can even be performed preemptively. Elective portosystemic shunt surgery is reserved for failure to respond to conservative management in the absence of alternative therapies.

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Normal portal venous pressure is slightly above that of the inferior vena cava and ranges from 0 to 11 mm Hg in physiological conditions.1-3 The pressure gradient between both venous systems is <10 mm Hg and is low because of the high compliance and low resistance of the portal vein and sinusoids and the outflow of the large hepatic veins. There is also a negative pump effect resulting from direct drainage into the right atrium and its location at the thoracoabdominal interface with its alternating positive abdominal and negative thoracic pressures.

Portal hypertension (PHT) can be defined as pressure that is higher than normal values, but is rarely measured directly. As such, it is usually defined by observation of a series of pathological changes and complications, such as splenomegaly and hypersplenism, variceal development, portal gastropathy, and accompanying observations of cirrhotic changes at ultrasonography, and, even more rarely, the clinical signs of portopulmonary syndromes.

Classification of PHT

PHT can be categorized on the basis of anatomical level of vascular resistance and histology of liver parenchyma into 3 main categories (Table 1) (Figures 1-3).3-5 Arterioportal hypertension has a somewhat different mechanism and will be considered now as a separate entity before returning to the more common causes listed.

Arterioportal hypertension

This is caused by excessive unconstrained arterial inflow into the portal venous system—a closed vascular system limited by volume with a single outflow.6,7 Typically, there is an arterioportal fistula (APF) from a vascular malformation located within the splanchnic area or the liver. The APF can be “simple,” with a single feeding vessel, or “complex,”...
involving multiple arteries ending in a common venous channel or aneurysm (Figures 2 and 3). Although the liver parenchyma is normal initially, it changes with venous barotrauma and capillaries becoming fibrotic with thickening of the intima. At that point, PHT can be regarded as mixed, with a combination of both increased flow and resistance. Managing these patients can be challenging, as they may ultimately need liver transplantation, which in turn can be compromised because of the damaged wall and reduced flow from the portal vein.

This straightforward classification of PHT plays a key role in determination of the correct treatment protocol. Those who have a relatively normal parenchyma and normal hepatic function will have a different surgical strategy compared with those who have severe parenchymal changes and hepatic dysfunction. Nonetheless, secondary damage and parenchymal fibrosis can develop rapidly in patients with postsinusoidal PHT (groups 2c and 3) and arterioportal hypertension (group 4), and more slowly in those with presinusoidal PHT (group 2a). Portal biliopathy changes and secondary biliary cirrhosis can even be seen in patients with portal vein thrombosis and cavernomatous transformation (group 1). Thus, the distinction between “cirrhotic” and “noncirrhotic” PHT may be evident from the original cause, but it may not always be relevant in terms of management for children who are seen at a late stage or who are managed conservatively for too long a period.

Therefore, the surgeon should consider not only the etiology but also the secondary changes of PHT that have occurred in the parenchyma and how this can affect portal flow, the anatomy of the portal venous system, and the underlying health of the liver—the hepatic functional reserve.

### Strategies for surgery

With prehepatic PHT (group 1), children may have a normal parenchyma and normal liver function for decades, as there is little, or absent, ongoing damage occurring downstream.
to the portal blockage. Such patients may have a patent intrahepatic portal system that is surgically accessible at the level of the Rex recessus and thus may benefit from a restoration of the continuity of the portal venous system by meso-Rex bypass (MRB) (Figure 4). We recommend that reconstruction is done preemptively when the anatomy is favorable, knowing that this can cure the patients’ condition and restore normal physiology. However, children with a thrombosed left portal vein within the Rex recessus who cannot benefit from MRB should be continued on a conventional endoscopic program and conservative management, as this also provides satisfactory long-term results by prevention of bleeding. Such children might become candidates for portosystemic shunt surgery when complications cannot be managed medically or endoscopically, and our preference is for a “selective” shunt, such as a Warren shunt, to preserve some hepatopetal flow through the cavernoma and preserve the hepatotrophic effect of mesenteric blood. Other teams use nonselective H-type shunts (eg, mesocaval and lateral splenorenal) in this scenario and obtain excellent clinical results, possibly because H-type shunts may not cause full diversion of the portal flow. Rather, there may be partial diversion by using a long venous interposition graft with a degree of resistance. We believe that nonshunt surgery should be strictly avoided, at least as primary surgical therapy, as it has little effect on portal pressure and may even seriously worsen the condition of the abdominal cavity for future surgery.

In PHT groups 2c, 3, and 4, the patient may be amenable to a cure if the diagnosis is made early in the progression of the disease, with the liver parenchyma not irreversibly damaged. If the liver parenchyma has been irreversibly damaged, then patients should be managed as those with cirrhosis.

Children with venoocclusive disease (group 2c) may benefit from medical management, with resolution of the problem and normalization of portal pressure and hepatic function. Interventional radiology (transjugular intrahepatic portosystemic shunt [TIPSS]) or transplantation is rarely indicated.

Patients with Budd–Chiari syndrome (group 3) may be amenable to angioplasty and stenting of the hepatic vein and vena cava, which, when combined with anticoagulation, may allow stabilization or recovery of parenchymal damage and normalization of liver function for many years. Ultimately, there is still a possible need for liver replacement.

Patients with arterioportal hypertension and APF (group 4) patients may benefit from embolization if the latter is a simple single tract (Figures 2 and 3). In more complex arterioportal malformations, radiological interventions should be avoided, as they often fail at achieving full devascularization and control of the fistula and, more importantly, repeated attempts result in an abnormal arterial network and may seriously interfere with further surgery. Surgery should be considered if there is either resection of the liver segment involved or direct arteriovenous disconnection-devascularization (depending on the anatomy).

Traditionally PHT shunt surgery was reserved for patients in whom the control of variceal bleeding failed in either the short or long term, with recurrent bleeding episodes. This approach has been relatively successful in adults with well-compensated cirrhosis. However, in children and especially in infants, surgery was associated with a relatively high rate of anastomotic stricture or thrombosis because of the small diameter of the veins involved. Prolongation of a conservative approach, coping medically with recurrent non–life-threatening bleeding, and postponing surgery until the child reached adolescence was not an uncommon practice, as it was thought that deferring the shunt procedure led to a higher success rate in the older adult-sized adolescent. Nowadays, with experience gained in vascular and transplant surgery and the use of microsurgical techniques, reasonably good success rates can be achieved even in the smaller age-group by trained teams, and age should not be a criterion per se for, or indeed...
against, shunt surgery. Although surgeons can now offer excellent technical outcomes even in smaller children, the indication for shunt surgery has dropped to very low figures, with liver transplantation being offered instead in many cases.

The choice of shunt procedure itself may vary from team to team on the basis of the experience of the surgical team. Selective shunts have a relative advantage and should be preferred in children to preserve them from encephalopathy, although it has been shown that these turn out to be less selective and more "centralized" with time. Moreover, non-selective H-type shunts do provide excellent clinical results. It is perhaps more important that the surgical team is expert and confident with one technique that in their hands provides good success rates. It is also crucial that a multidisciplinary team is involved who can propose a wide range of therapeutic options (medical, surgical, radiological, and transplant) to individualize treatment.

Any therapeutic algorithm must take into consideration the possibility of liver replacement in the medium to long term. Its use may be jeopardized by previous abdominal operations and portosystemic shunt surgery. The indications for shunt surgery in children with chronic liver disease are few because of the successful results of liver replacement. These may include those with well-compensated disease (Pediatric End-stage Liver Disease [PELD] <10), such as those with PHT group 2a causes (ie, congenital hepatic fibrosis) and, less commonly, groups 2c and 3, with well-preserved liver function but symptomatic variceal bleeding. Nonsurgical interventional options such as TIPSS may be more appropriate and should be discussed with the transplant team. This alternative preserves the intact abdomen, and stents can be removed at the time of transplantation. It may even improve the patient’s condition when used as a bridge to transplant by its effect on ascites and PHT.

Hypersplenism is rarely a major issue in children and does not cause clinical concern in most cases, even in those with long-lasting PHT and major splenomegaly (Figure 5). Rarely, a selective portosystemic shunt (distal splenorenal shunt or “Warren shunt”) may be used because it provides regional decompression and effective venous drainage of the spleen with reduction in size and improvements in thrombocytopenia and leukopenia. Reported results have been mixed, and a lot depends on the quality of the flow through the shunt. The option for isolated splenectomy

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**Figure 3**  Schematic representations of vascular malformations with arterioportal fistulae and PHT: complex (A-C) and simple (D) types (CA, celiac trunk; CHA, common hepatic artery; LHA, left hepatic artery; RAHA, right accessory hepatic artery; DHA, diaphragmatic hepatic artery; RPV, right portal vein; L, left portal vein; II-III,-V, liver segments II-III and IV).
must be discussed carefully, and it is rare in clinical practice. It may be indicated for those with recurrent gastrointestinal bleeding and extensive thrombosis of the portal system, including the splenic vein where options for performing a portosystemic shunt are few or nil. Portal pressure does drop for some time after splenectomy. A further indication, at least in our opinion, is the difficult teenager with longstanding prehepatic PHT (group 1) not amenable to an MRB, presenting with huge splenomegaly, who starts refusing medical therapy, medical assessments, and having a lifestyle that puts him/her at risk of difficult-to-manage bleeding either from splenic rupture caused by hazardous sporting practice, or gastrointestinal bleeding far from medical support. These patients may benefit from preemptive

Figure 4  Retrograde portogram (top) and conventional angiograms (bottom) in 2 patients (left and right) showing that intrahepatic portal veins can be ignored on standard imaging and best delineated on portograms (R, Rex recessus; RL, Right liver; SMV, superior mesenteric vein).

Figure 5  Huge splenomegaly of idiopathic origin in a teenager with longstanding extrahepatic PHT.
splenectomy (after an adequate vaccination program), with a central splenorenal shunt being performed at the same operation (to avoid loss of the splenic vein by thrombosis) or a spleno-Rex bypass if the left portal vein in the Rex recessus is patent in a child who was not referred previously for MRB surgery (Figure 5). In rare cases, the occurrence of a portopulmonary syndrome (hypoxia and hepatic-pulmonary syndrome, or pulmonary arterial hypertension, as a complication related to PHT and portosystemic shunting) is an indication for liver transplantation. Again, in a patient with PHT caused by portal cavernomatous change, having a patent Rex, and who was not referred for MRB previously, MRB may also be considered to treat Portopulmonary syndrome, as it can resolve both portal hypertension and the hepato-pulmonary syndrome.

Assessment of the surgical patient

Preliminary assessment must establish the cause of PHT, hepatic function, and “reserve,” with appropriate histology and any existing complications of PHT.

Appropriate preoperative assessment includes coagulation profile, delineation of the portal venous system, Doppler ultrasonography of the neck.

Coagulation profile

Thrombocytopenia and a slightly prolonged international normalized ratio are common, but other anomalies may be identified. These are rarely important for those going for liver transplantation, as the new liver will correct existing anomalies, but can be important when performing vascular nontransplant surgery. Coagulopathy is a risk for intra- and postoperative hemorrhage, and conversely a hypercoagulable state can be associated with an increased risk of thrombosis. Minor changes may be relevant; a deficit in antithrombin III makes the patient insensitive to heparin.

Doppler ultrasonography of the neck

This is to confirm patency and caliber of both internal jugular veins if this is being considered for bypass. Most consider it as the best available material, as it is autologous, grows with the child, and mimics the characteristics of both diameter and length of the portal vein, ensuring adequate flow and low thrombosis rates (certainly by comparison with smaller diameter veins such as the saphenous vein graft). Using 1 of the 2 patent internal jugular veins has a low morbidity, which is not the case when iliac veins are procured or when the vena cava is divided for creation of a shunt (Auvert shunt). The internal jugular vein can be procured as long as 6-8 cm in length and easily matches relatively long gaps needed in mesocaval and meso-Rex bypasses. Neither does it need much in the way of complex graft preparation as, for example, when preparing a spiral saphenous vein graft. In those cases of significant difference in diameter between left and right internal jugular veins, further assessment of the venous anatomy of the brain is recommended to check whether intracranial left/right anastomoses are present and, if so, are large. Lack of this is a relative contraindication for procuring 1 vein.

Feasibility of MRB

Wedged hepatic venous portography via the transjugular approach is safe and efficient where this shunt is being considered (Figure 4). Patency of the left portal branch and Rex recessus is the key observation to success. Nowadays, direct percutaneous portography is not recommended, as the portal radicals are small in caliber and the chance of opacification is lowered.

Shunt surgery

Portosystemic shunts are nonphysiological routes created to divert the portal and mesenteric blood flow into the systemic circulation. A variety of techniques are available, and with relatively simple technical modifications (side-to-side anastomosis vs H-type, caliber of the bypass, length of the graft, use of prosthetic material, site of drainage), the surgeon has plenty of choices for achieving a partial, rather than a full, portal flow diversion. The drop in portal pressure is variable, depending on how much flow is diverted, and varies according to the diameter of the anastomosis, the diameter and length of the interposition graft, and the location of the site chosen for portal blood diversion.

Portosystemic shunts may be either “selective” or “non-selective.” Selective shunts divert selectively a region of the abdomen, for example, the stomach and spleen, to
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decompress esophagogastric varices. Blood is predominately shunted into the low-pressure systemic venous circulation, and the varices then flow at a lower pressure with no residual risk of bleeding.

Selective shunts

Distal splenorenal shunt (“Warren” shunt)
This involves division of the splenic vein and anastomosing the distal stump into the left renal vein, thereby preserving portal and mesenteric blood flow to the liver. However, with the complex collateralization and networking of the splanchic venous system, some communications always persist, and it has been shown that this “selective” shunt progressively “centralizes” with time. This option decreases spleen size, improves hypersplenism, and normalizes platelet and leukocyte counts. Selective shunts have been used with success to treat bleeding varices and hypersplenism and have been shown to reduce postoperative encephalopathy with equivalent long-term mortality and rebleeding rates. Long-term patency rates >90% have been reported.

Nonselective shunts

These create a direct communication between the portal system and the systemic circulation, and depending on the flow, a full diversion can be achieved, with a consequent fall in the portal pressure. All these shunts have been associated with a significant risk of hepatic encephalopathy (compared with selective shunts). Nonselective shunting should be avoided when possible, as preservation of neurocognitive function is particularly important in children. In children, learning disorders or behavioral abnormalities can also be manifestations of encephalopathy.

End-to-side portocaval shunt
In this operation, the portal vein is divided close to the liver and end-to-side anastomosis of the lower stump to the inferior vena cava (IVC) is performed.

Proximal splenorenal shunt
The splenic vein is divided and the spleen is removed. End-to-side anastomosis of the splenic vein stump to the left renal vein is performed.  

Mesocaval shunt
Although it is possible to create a side-to-side mesocaval shunt after mobilization of both the superior mesenteric vein (SMV) and IVC (without the interposition of a graft), the usual mesocaval shunt is an “H type” with the interposition of a prosthetic conduit (Dacron/Gore-Tex) or autologous vein graft. In children, mesocaval shunting with an interposed jugular vein has been preferred by many teams and has been used with acceptable results. The Auvert shunt is a mesocaval shunt fashioned after division of the iliac veins and mobilization of the IVC, which is then anastomosed onto the SMV. The disadvantage is a high rate of late venous complications related to interruption of both iliac veins.

Some nonselective shunts are performed to try to limit the hemodynamic effects of the flow diversion either by calibration or by some sort of hemodynamic balance in an attempt to both avoid full rerouting of blood flow away from the liver and reduce the risk of encephalopathy. In return, one expects a reduced effect on the portal pressure, hypersplenism, and possibly a higher risk of thrombosis or occlusion in the long term. Examples of this approach include the Sarfeh and the Mitra shunts. The former is an H-type portocaval shunt using an 8-mm-diameter Gore-Tex graft and is usually done with extensive collateral ligation. The latter, first described by Denton Cooley in the 1960s, is a nonselective side-to-side splenorenal shunt. In this, the spleen is preserved, and the shunt is fashioned between the splenic vein and the left renal vein. It is said to reduce the risk of encephalopathy compared with other nonselective shunts because it does not completely steal portal flow away from the liver. Good results have been reported with this shunt.

TIPSS as (nonsurgical) shunt intervention
An intrahepatic metallic stent is inserted between the intrahepatic portions of the portal vein and the hepatic vein, which creates a (nonselective) portocaval shunt. Calibration can be achieved in some, with a direct pressure gradient measurement during the procedure to allow limited flow diversion, but hepatic encephalopathy can still occur. The advantages of avoiding a major abdominal intervention and preserving the abdomen intact for possible transplantation is balanced by potential complications such as shunt stenosis/thrombosis, bleeding, portal vein thrombosis, and migration of the stent into the right atrium.

Experience in children remains limited to case reports because it is more difficult in small patients. Nonetheless, it should be considered if a surgical shunt is indicated, whether as a bridge to transplant or not. Patients with well-preserved liver function and severe chronic PHT are good indications (ie, children with hepatic fibrosis or cystic fibrosis).

Portal vein thrombosis and the MRB
Prehepatic PHT, and specifically portal vein thrombosis, is the commonest cause of PHT in children. Most have no obvious etiologic cause in their history and are thought of as idiopathic, perhaps even congenital, cases. Direct damage to the portal vein in about 25% of cases can be inferred with a history of umbilical vein catheterization (UVC) during the neonatal period. Less commonly, portal vein thrombosis may be caused by trauma, tumors, or secondary to peritonitis.
The etiologic role of thrombophilic states and/or of single prothrombotic factor remains controversial. Most studies have shown that coagulation abnormalities demonstrable in portal vein thrombosis are secondary, rather than primary, disorders. In posthepatic venous thrombosis in adults, the reverse is probably true. Primary coagulation disorders have been reported but remain uncommon. It seems more likely that coagulation disorders may predispose to thrombosis when associated with other risk factors. Pietrobattista et al observed a correlation between minor thrombophilias (eg, MTHFR-C667T mutation) and portal vein thrombosis. In general, genetic prothrombotic risk factors seem to have only a minor role in prehepatic PHT, with a few exceptions confirming the rule. Diffuse thrombosis of the splanchnic venous system may also be an exception to this general rule.

Children with portal vein thrombosis should have no underlying liver disease, and their liver function is expected to remain normal throughout life. Some, however, may develop a portal biliopathy caused by extrinsic compression of the extrahepatic bile ducts, which worsens with time and may cause progressive cholestasis after a few decades. Such cholestatic change may be inferred by minor dilatation of the extrahepatic bile ducts and an association with biliary sludge or even stones. Portal vein thrombosis may extend variably into splanchnic system, although usually the portal vein trunk has disappeared. Extension upstream into the splenic or mesenteric veins is rare, leaving these vessels available as anastomotic sites. Even in those children with large cavernomas (Figure 1), thrombosis may not extend into the intrahepatic portal venous system. Usually the latter is patent, although hypoplastic with small-diameter (3-4 mm) veins. These may not be visible on conventional imaging (eg, Doppler ultrasonography, angio computed tomography, and angio magnetic resonance imaging, or even conventional angiography) (Figure 4), but it should be possible to image them adequately using retrograde portography. Direct catheter insertion into the reopened umbilical vein can be done intraoperatively but has the disadvantage of needing a laparotomy and the risk of finding a thrombosed left portal vein, therefore making MRB unfeasible.

Consideration should be given to perform a liver biopsy to confirm normal liver histology in older children, those with abnormal portogram findings (eg, partial thrombosis, wall irregularities, and poorly defined portogram findings that may indicate associated hepatoporal sclerosis), and those who have undergone transplant because fibrotic livers due to chronic graft dysfunction or cholestasis may compromise flow.

MRB (also known as mesenterico-left portal bypass or, more simply, the Rex shunt) was first used for portal vein thrombosis after transplantation before being applied to those with idiopathic prehepatic PHT (group 1). Ana-
tomically, the umbilical fissure is a natural place for entering the liver outside the porta hepatis, and has relatively good access to the portal system. Ideally, outflow is by a patent umbilical portion of the left portal vein within the Rex recessus, and flow should be from left to right within a patent intrahepatic portal venous system. The easiest inflow is from a patent SMV immediately below the edge of the pancreas; however, alternative sites have been proposed with success, such as the coronary vein, the splenic vein, the IMV, or even a large gastroduodenal vein. The conduit itself is preferably an autologous vein of adequate diameter, and the best results have been described in children in whom the internal jugular vein has been used. Other alternatives are more complex (eg, spiraled saphenous vein) and associated with less favorable outcomes (eg, recanalized umbilical vein, cryopreserved allogeneic vein, or prosthetic GoreTex).

Portosystemic collaterals tend to close spontaneously after the procedure when the bypass is patent, with adequate flow redirected into the liver and hepatopetal flow restored (Figure 6). Restoration of normal physiology (Figure 7) also corrects the secondary coagulation abnormalities, improves neurocognitive ability, and may correct subclinical encephalopathy. It improves growth and body mass index if the child has failed to thrive, and isolated reports suggest it reverses hepatopulmonary syndrome and adenomatous transformation of the liver. Moreover, the procedure has not been associated with significant morbidity, and no mortality has been reported.

More recently, MRB has been proposed as the optimal procedure for this condition, with a suggestion that it should be performed earlier in the course of the disease. Children with portal vein thrombosis have been usually managed conservatively, under the assumption that by having good parenchymal liver function they are not at risk and that intermittent bleeding is relatively well tolerated. Furthermore, it has been often stated that complications are less common both in frequency and in severity as the child grows into adulthood. The report from a workshop in Baveno, Italy, stated that the common outcome in these children includes a “relatively low” risk of bleeding (on average, 1.3 episodes/year), and the risk of rebleeding in the long term after obliteration of varices is low, but there is an increased risk of hepatic dysfunction, hepatitis B and C infection (repeated transfusions), severe hypersplenism related to massive splenomegaly, and progressive symptomatic portal biliopathy with prepubertal growth retardation in about half of all cases. Of course, such patients do much better compared with a comparable cohort of patients with cirrhosis, but they have a diminished quality of life compared with healthy children. Although a 90% survival rate has been reported in long-term follow-up studies with conservative management, similar or even better overall outcomes can be achieved when MRB is done early in life. We believe MRB should be proposed as early as reasonably possible after diagnosis. The operation is rather standard-
ized and in trained hands can achieve excellent results in children from any age and any weight: the best weight/vein diameter balance is from about 10 kg of weight upward. Here the Rex fossa is easy to approach; the mesenteric vein is of a good diameter; and the jugular vein has an optimal length/diameter ratio. The lower limit is difficult to define, but infants of approximately 4 kg have successfully undergone surgery. In older children and in adults, longstanding PHT and the development of numerous collaterals and a huge cavernoma may render the operation technically more demanding.

Overall, we believe that MRB should be performed sooner rather than later, perhaps as soon as the diagnosis has been confirmed in children, even if there is complete absence of complications. Only children in whom the left portal vein can be shown to be thrombosed or damaged should the procedure be postponed and a more conventional strategy pursued. The management of some children with intermediate conditions (eg, abnormal portogram, difficult access to the portal system because of extensive thrombosis, no autologous material available) needs to be individualized. Liver transplantation remains exceptional, and most reported cases are adults with portal vein thrombosis complicated by cirrhosis secondary to portal biliopathy.

Role of portal stenting for prehepatic PHT

Several authors have reported attempts at percutaneous transhepatic angioplasty/stenting of a thrombosed portal vein. Gauthier reported a series of 10 patients in Bicêtre, France, with success in only 1 case with medium-term (4-year) follow-up. Similarly, Cwikiel et al reported a successful experience in a single case of the 5 in which it was attempted. It appears that such cases were different from common idiopathic prehepatic PHT cases in that the intrahepatic portal system was large enough in diameter to allow transhepatic access and intravascular maneuvers. This is made clear by Cwikiel et al in their report, where they observed that although transjugular puncture of the intrahepatic portal vein was possible in 4 of 5 cases, full recanalization of the portal trunk was possible in only 1, with difficulties attributable to the small caliber of veins. Semiz-Oysu et al confirmed the technical difficulties and problems in children. As so few stents have been inserted, it is difficult to predict the long-term outcome, and there must be the possibility of revision because of growth.

Interventional radiological attempts should probably be kept in reserve for patients with an identifiable intrahepatic portal system and those with a limited (short in length) portal vein thrombosis, or better, those with limited strictures of the portal vein.

Nonshunt surgical options

A variety of nonshunt surgical procedures have been proposed over the years. Although some of these procedures are rather simple and easy to perform, others are complex and aggressive. The simplicity of the former does not imply that they have a role today or are preferred to other treatments. Many were conceived in a time when alternative more modern strategies were not available, or were proposed to manage complex, sometimes last-chance, clinical conditions. Most probably, they now have no place, particularly in children.
Surgical ligation of varices and esophageal transection

Direct ligation of the varices via a thoracic or transgastric route is historically the “ancestor” of endoscopic variceal eradication, although the latter has proved much more efficient at a less invasive cost. Transection of the esophagus or the upper stomach, with direct reanastomosis (Sugiura procedure and others), interrupts the intramural vein and variceal network, immediately separating esophageal varices from the splanchnic venous system and minimizing bleeding. It was originally proposed as an alternative to shunt procedures in adults. A number of modifications of the Sugiura operation have been proposed for children, and its use has been reported largely from developing countries with limited surgical and medical management options and no access to liver transplantation. Although satisfactory long-term results are reported, these have been mostly in patients with prehepatic PHT. The procedure is palliative and has a high incidence of recurrence in the medium term; it also creates numerous adhesions that make further abdominal surgery more difficult.

Splenic vein embolization

Splenectomy or splenic artery embolization may be advocated in some rare cases of severe hypersplenism. It does reduce the arterial inflow to the portal venous system and PHT, and therefore may reduce the frequency of bleeding esophageal varices. This is usually regarded as temporary, and recurrence is the rule. Additionally, there are other complications, such as postsplenectomy sepsis and intraperitoneal adhesions, together with propagation of thrombosis in the splenic vein into the portal venous system, exacerbating PHT and rendering further shunt surgery limited or hazardous. Splenectomy must be considered a last resort in children and should not be proposed if other management options are available. If splenectomy is to be performed in a child with PHT, the surgeon should also consider performing a proximal splenorenal shunt at the same time to achieve portal decompression and retain splenic vein patency.

Partial splenectomy has been proposed recently with preservation of some splenic flow combined with an omentopexy. This seems a reasonable alternative, as this helps preserve the splenic vein without performing a portosys-
temic shunt. Partial splenic embolization is a similar compromise, but the effects on PHT are limited, although it can have a significant effect on hypersplenism. It should be used with caution, as a significant embolization (necessary to obtain a clinical effect) correlates with increasing complications, such as pain, necrosis, and abscess formation, and again may make further regional surgery more demanding.

The ultimate shunt procedure: Liver transplantation

Management has dramatically changed during the past 2 decades for children with cirrhotic PHT. This has been because of the efficiency of endoscopic variceal control in the chronically ill, but stable, patient, and the wide use and success of liver transplantation in those who deteriorate. Children with poor synthetic liver reserve and PHT are primarily candidates for liver transplant. In these, the high rate of complications after shunt surgery outweighs its potential short-term benefits. One-year patient survival after liver transplant approaches 95% in experienced centers, and the procedure is curative for the primary disease; this has influenced the strategy of a no-shunt choice for the stable patient by many teams.

The role of portosystemic shunting in children with a diseased liver is reduced to highly selected cases with very good residual hepatic function that is likely to persist unchanged for a long period and may typically include cases of hepatic fibrosis and cystic fibrosis. Even here, the role of surgical shunting has probably been displaced by comparison with a TIPSS procedure, as this is easy to follow, revise, tailor, and even remove when necessary (transplantation).

Complications and outcome after shunt and bypass

Successful shunt surgery achieves relief of PHT and correction of most of its complications. When performed by skilled surgeons, it is an efficient and safe procedure. High long-term patency rates, ranging from 89% to 97%, have been reported. Failure to achieve satisfactory outcomes is mostly related to unfavorable anatomy of the portal venous system and diffuse thrombosis. Along with prevention of gastrointestinal hemorrhage, other benefits include correction of splenomegaly and hypersplenism, correction of biliary anomalies (ie, portal biliopathy), and transient acceleration of growth leading to a significant improvement in quality of life.

Shunt thrombosis is a serious problem, more frequent when using a conduit, and can be related to technical problems, such as compression and kinking, or a hypercoagulable state. Experienced centers report low rates of shunt thrombosis (<5%). Doppler ultrasonographic assessment should be done during the first week to detect early problems. Postoperative prophylaxis is recommended, with low-dose heparin during the first week, followed by antiplatelet agents (aspirin and dipyridamole) for 3-6 months. In cases of shunt thrombosis, the shunt may be salvageable either by surgical revision (preferably if early thrombosis), or by radiological intervention to attempt percutaneous recanalization and/or balloon angioplasty. Such procedures were successful in 9 of 10 cases in the Bicêtre series.

Chylous ascites owing to disruption of retroperitoneal lymphatic channels by surgery are uncommon after a successful shunt operation that allows good portal decompression. Persistence may imply an insufficient drop in portal venous pressure, maintaining high pressure and flow in the lymphatic system.

Encephalopathy is a problem after conventional shunt surgery in patients with cirrhosis, yet occurs infrequently in
those without parenchymal disease.\textsuperscript{95-97} Nonetheless, this affirmation may have to be reviewed in light of recent reports suggesting that subclinical encephalopathy is much more common than thought even in those with preserved liver function, and further studies are necessary.\textsuperscript{72,98-100}

Portopulmonary syndrome (hepatopulmonary syndrome and pulmonary hypertension) is rare in children and can be observed in those with either cirrhotic or noncirrhotic PHT.\textsuperscript{95,101} French research suggests a rate of 1 case of pulmonary arterial hypertension among more than 100 children with portosystemic shunts for prehepatic PHT.\textsuperscript{47,80,95} These complications may occur late after shunt surgery, and long-term cardiopulmonary follow-up is recommended (eg, echocardiography, resting and stressed \( \text{O}_2 \) saturation measurements).

Liver nodules (liver cell adenomas, focal nodular hyperplasia) may develop in the long term in some postshunt patients, but the risk appears to be modest. It is seen mostly in those patients with prolonged survival who have not required transplantation. Ultrasound-based screening over the long term is suggested\textsuperscript{102} (Figure 8).

Conclusions

The management of children with PHT has changed dramatically in the past decade with an improved outcome partly based on the efficiency of endoscopic variceal control in the acute scenario and the success of liver transplantation. Emergency shunt procedures should now be extremely rare, with acute bleeding generally controlled endoscopically or, less commonly in the older child, by a TIPSS procedure. The concept of a short-term surgical shunt as a bridge to transplant is rarely considered in children. Surgery is indicated for complicated cases with noncirrhotic PHT, with MRB proposed, even preemptively, for those with favorable anatomy.

References