Portal Diversion for Portal Hypertension in Early Childhood

HENRI BISMUTH, M.D., DOMINIQUE FRANCO, M.D.

From Unité de Chirurgie Hépato-Biliaire, Université Paris-Sud, Hôpital Paul Brousse, 94800 Villejuif, France and Unité d'Hépatologie Infantile Hôpital de Bicêtre, 94270 Le Kremlin Bicetre

Twenty-three children under 6 years of age with portal hypertension were treated by portal diversion. Fourteen had cavernomatous transformation of the portal vein and 9 had an intrahepatic block due to cirrhosis (8) or congenital hepatic fibrosis (1). Portal-systemic shunts were central splenorenal in 20 patients, side-to-side portacaval in 2 and mesocaval in one. In 20 of the 21 peripheral shunts, the veins used for the anastomosis were less than 10 mm in diameter. There was no operative mortality. Thrombosis of the shunt occurred in 3 children (13%) and was responsible for recurrent bleeding in one who was treated later with success by a mesocaval shunt. The two other children with a thrombosed shunt are waiting, at the present time, for a mesocaval anastomosis. The volume of blood flowing through the shunt was small initially and the fall in pressure gradient was slight: therefore intraoperative angiography appeared to be a better way to assess the patency of shunts done at an early age than pressure or flow measurements. The figures recently reported by Clatworthy, with a mortality rate of 12% directly or indirectly related to repeated hemorrhage, are for us a forceful argument for early adequate management of portal hypertension in children. Until now, portal-systemic shunts have been complicated by a high frequency of thrombosis and have given discouraging results. Our results suggest that it is possible to perform portal diversion successfully on diminutive veins (down to 4 mm). From this experience early portal diversion appears to represent the treatment of choice for portal hypertension in childhood.

THE CURRENT ATTITUDE in the management of extra-hepatic portal hypertension in children, following the report of Fonkalsrud et al.,[7] favours conservative measures and weighs against portal-systemic shunts in children less than 8 years old, or when veins are less than 8 mm in diameter. This concept derives from the following assumptions: 1) bleeding episodes are not life-threatening, at least in extra-hepatic portal hypertension; 2) direct attack on varices is fraught with a high rate of recurrent bleeding and 3) thrombosis frequently occurs after portal-systemic shunts when the veins used are small.

Our experience with portal diversion in children less than 6 years old is not in accordance with current doctrine. The purpose of this work is to study the risks and efficacy of portal-systemic shunts in young children with portal hypertension.

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Reprint requests: Dr. Henri Bismuth, Hôpital Paul Brousse, 94800 Villejuif, France.

Patients and Methods

From December 1970 to May 1975, 27 children under 6 years of age were operated on for portal hypertension. Among them, 4 were denied a shunt operation and had variceal ligation (all less than 2 years of age, one with diffuse portal thrombosis); 23 underwent a portal-systemic shunt and constitute the clinical material of the present work.

Age ranged from 18 months to 5 years and 11 months (Fig. 1). The cause of portal hypertension was extra-hepatic in 14 cases and intra-hepatic in 9. In 4 patients with extra-hepatic portal hypertension it was the consequence of thrombosis of the portal vein secondary to catheterization of the umbilical vein at birth. In the 10 other patients the cause of cavernomatous transformation remained unknown. Intra-hepatic portal hypertension was due to a variety of liver diseases. There were 3 with secondary biliary cirrhosis, 2 with cirrhosis and alpha-1-antitrypsin deficiency, 3 cirrhoses of unknown origin and 1 congenital hepatic fibrosis. All patients with cirrhosis were type A in the classification of Child.[3]

There were 18 therapeutic and 5 prophylactic shunts. The number of bleeding episodes in the 18 patients with therapeutic shunts is indicated in Fig. 2. Ten patients were operated on after the first bleeding episode and 8 had had several bleeding episodes before being referred to us. The prophylactic shunts were performed in patients with cir-

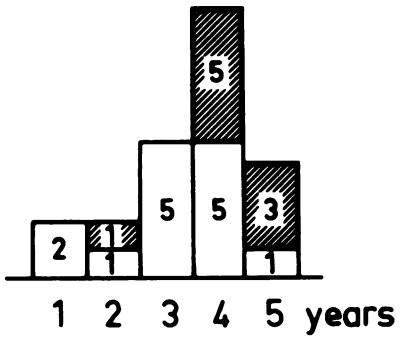


FIG. 1. Age of 23 children with extrahepatic (open block) and intrahepatic (hatched block) portal hypertension treated by shunt operations.

rhosis. All therapeutic shunts but one were performed as elective procedures. In one case, an emergency splenorenal shunt was done together with ligation of gastric varices in an 18-month-old child with massive variceal bleeding.

Preoperative angiography (selective arteriography of the celiac axis and superior mesenteric artery and/or spleno portography) was obtained in every child to assess the patency and the size of portal, splenic and mesenteric veins. In 7 patients the mesenteric vein was not visualized preoperatively. In patients with intra-hepatic portal hypertension, all veins were patent. In patients with extrahepatic portal hypertension, the splenic vein was always patent and the mesenteric vein was not patent in two cases. The size of the veins used for peripheral shunts as measured on angiograms is shown in Fig. 3.

The anastomoses performed were central splenorenal in 20, side-to-side portacaval anastomoses in 2 and mesocaval anastomosis in one. Central splenorenal shunts were performed according to the technique of Clatworthy and Boles.[4] The following technical details should be emphasized: the left part of the mesenteric root was opened to approximate the two veins as closely as possible; the renal vein was completely freed and clamped at its two ends in order to allow the excision of a round patch from its anterior wall; the shunt was performed using interrupted sutures (Fig. 4). Mesocaval shunt was performed according to the technique of Marion[12] using the right iliac vein anastomosed end-to-side to the superior mesenteric vein.

Evaluation of the shunt was done at operation by measurement of pressure gradient and of flow through the anastomosis (Square wave electromagnetic flowmeter). In addition, in the last 9 patients intraoperative portography was done. Heparin, 1 mg/kg per day, was infused via a peripheral vein as soon as the veins were clamped, and continued for 10 days.

Disappearance of esophageal varices was documented during the first postoperative month by barium swallow and/or endoscopy. Patency of the shunt was tested by selective arteriography of the superior mesenteric artery in 18 patients one to 6 months postoperatively. All patients were examined within the last six months. The average followup was 2.5 years with a range of 2 months to 4 years and 7 months.

Results (Table 1)

Survival. There was no operative mortality and so far there have been no late deaths.

Postoperative complications. Intra-peritoneal hemorrhage occurred in a 3 year, 5 month-old-child with mesocaval shunt for portal vein thrombosis. This was due to an overdose of heparin and necessitated an emergency laparotomy on the fifth postoperative day. No precise site of bleeding was detected and the rest of the postoperative course was uneventful. There were no other postoperative complications.

Patency of the shunt. Twenty shunts were proven to be patent on late followup controls: 17 splenorenal shunts, 2 portacaval shunts and 1 mesocaval shunt. This was demonstrated in 15 patients by postoperative angiography (Figs. 5 and 6); in the 5 other children, varices had dis-

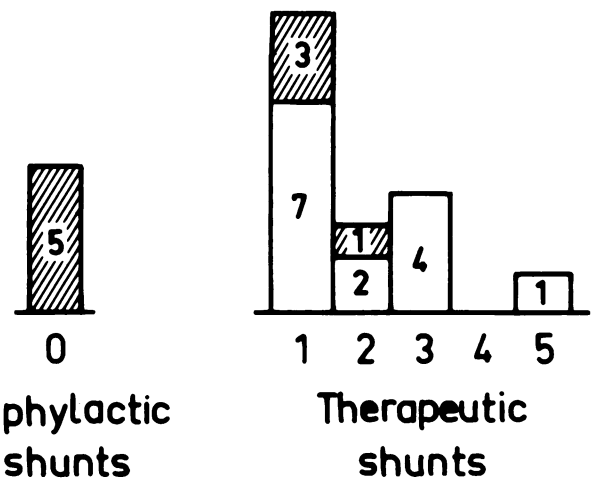


FIG. 2. Number of preoperative bleeding episodes in 23 children with extrahepatic (open block) and intrahepatic (hatched block) portal hypertension.

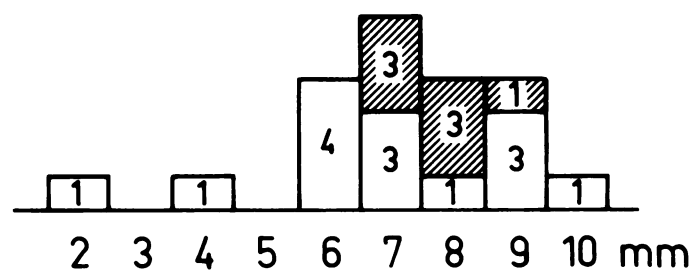


FIG. 3. Size of veins used for peripheral portal diversion (20 splenorenal anastomosis and one mesocaval anastomosis) in 21 children with extrahepatic (open blocks) and intrahepatic (hatched blocks) portal hypertension. In the two patients receiving direct portacaval shunt, the portal vein measured respectively 10 and 15 mm.

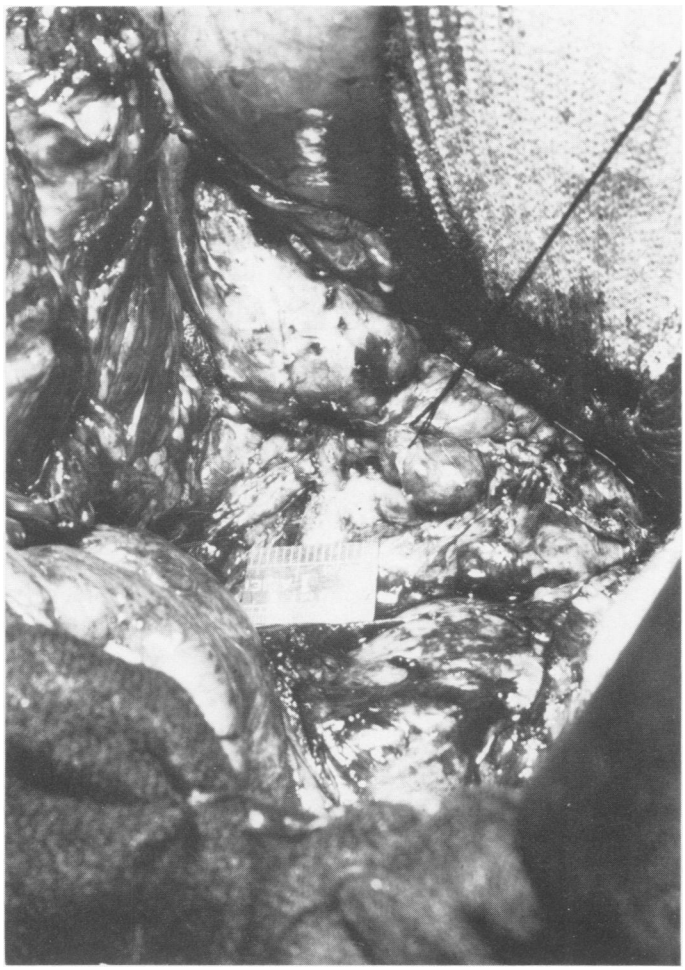


FIG. 4A. Operative view of a splenorenal shunt in a 4 year, 5-month-old boy. The splenic vein was 4 mm in diameter.

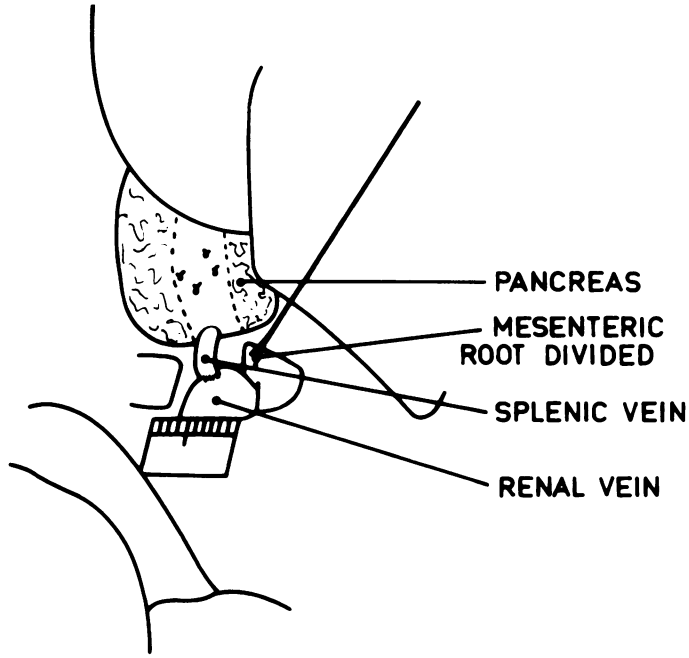


FIG. 4B. Drawing of Fig. 4A.

appeared at radiological and/or endoscopic examinations. None of these 20 children experienced recurrent hemorrhage.

Thrombosis was observed in 3 splenorenal shunts (13%). A 4 year, 5 month-old-child with cirrhosis of unknown origin had recurrent bleeding 2 months following a splenorenal shunt. Thrombosis of the shunt was proven by an angiogram (Fig. 7). He then underwent a side-to-side portacaval shunt. Recurrent bleeding again occurred a month later at which time the second shunt was also proven to be thrombosed. A mesocaval shunt was done; the shunt is still patent. In the two other patients thrombosis of the shunt was discovered by systematic angiography. Detailed observations concerning these three children are given in Table 2. A satisfactory explanation for the obstruction of the shunt could be offered for each case. In the first patient already described thrombosis of both splenorenal and portacaval shunts was due to thrombocythemia (platelet count: 800,000/mm^3), platelet clots being found on the vein walls even before the end of each procedure. Eventual patency of the mesocaval shunt was obtained in this child by intensive heparin therapy. In a second case, there was compression of the anastomosis by the root of the mesocolon at the splenic flexure. In the third case, thrombosis of the shunt occurred in an 18-month-old girl operated upon as an emergency, in whom the diameter of the splenic vein was 2 mm.

Recurrent bleeding. One of the three patients with thrombosed shunts rebled (Case 1, Table 2). None of the other patients has had recurrent bleeding.

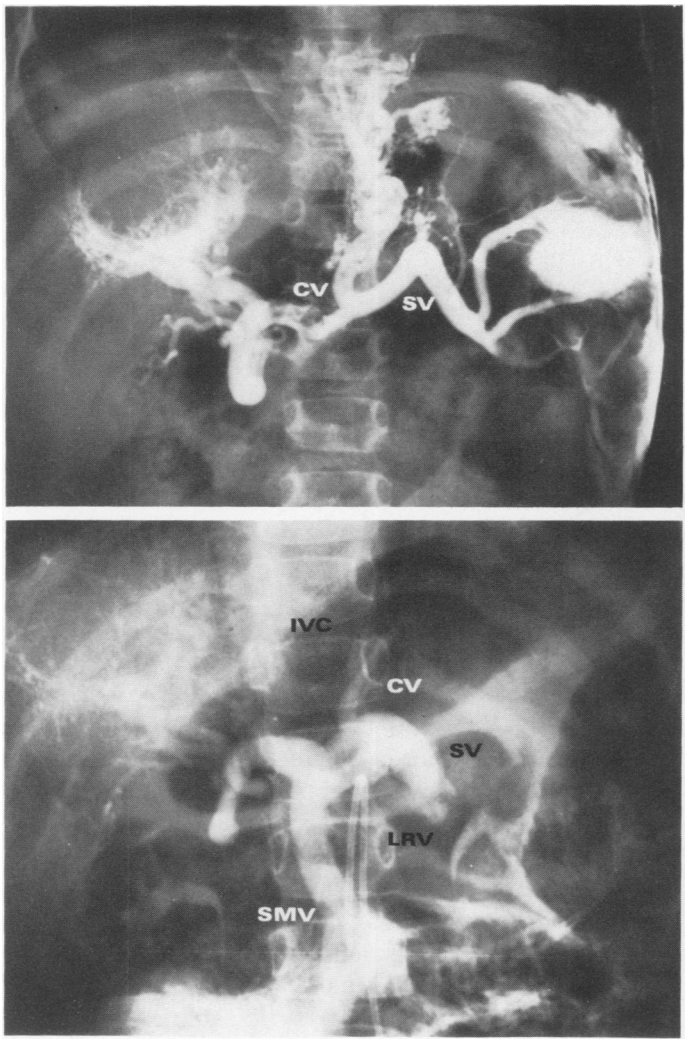
Neuropsychic disorders. In this group of patients, no sign of encephalopathy has been detected in any child.

Comments

In this series of children under age 6, 21 out of 23 (91%) have at the present time patent anastomoses and two others with thrombosis are waiting for a second shunt. Splenorenal anastomosis was preferred for several reasons; 1) In 8 of the 14 children with extra-hepatic block, the splenic vein was either the widest vein or the only

TABLE 1. *Results of Portal-systemic Shunts in 23 Children Under 6 years of Age with Portal Hypertension*

Postoperative complications	
Mortality	0
Morbidity (intra-abdominal hemorrhage)	1
Late results	
Thrombosis of the shunt	3 (13%)
Recurrent bleeding	1 (4%)
Encephalopathy	0



FIGS. 5A and B. Pre- and postoperative angiographic studies in a 30-month-old girl with cavernomatous transformation of the portal vein. (Top) Preoperative splenoportography: the diameter of the splenic vein (SV), measured at the left border of the spine, is 6 mm; there is a large coronary vein (CV) leading to esophageal varices. (Bottom) Venous phase of superior mesenteric arteriography, 3 years after splenorenal shunt. The contrast medium opacifies mainly the superior mesenteric vein (SMV), the central part of the splenic vein (SV), the left renal vein (LRV) and the inferior vena cava (IVC). Collateral channels towards the liver are still slightly opacified. There is a faint visualization of the coronary vein (CV) which now is small. Esophagoscopy did not show varices at that time.

available. 2) In many children huge splenomegaly with hypersplenism was present. Splenectomy appeared to be a useful adjunct since, as recently demonstrated by Mutchnik et al.[14] the size of the spleen does not decrease much following other types of shunt and hypersplenism persists. 3) Our group has a large experience with splenorenal shunts in adults.[2] However, if the mesenteric vein is much larger than the splenic vein, mesocaval shunt may be preferred as in one of our patients. In children, division of both iliac veins in order to construct a cavo-iliomesenteric shunt as described by Farge and Auvert[6a] does not carry the same risk of lower extremity edema that it does in adults.

The mean diameter of the veins used for shunting was 6.5 ± 2.5 mm. Progress in microvascular surgery has enabled surgeons to perform vascular anastomoses successfully on such tiny vessels, and our own laboratory experience in small animals was of great benefit.[10,11]

Thrombosis of the anastomosis was uncommon, 3 out of 23 (13%), and occurred in our first 10 cases. In only one case the size of the splenic vein (2 mm) appeared directly responsible for the thrombosis of the anastomosis. Although the absence of heparin therapy may have

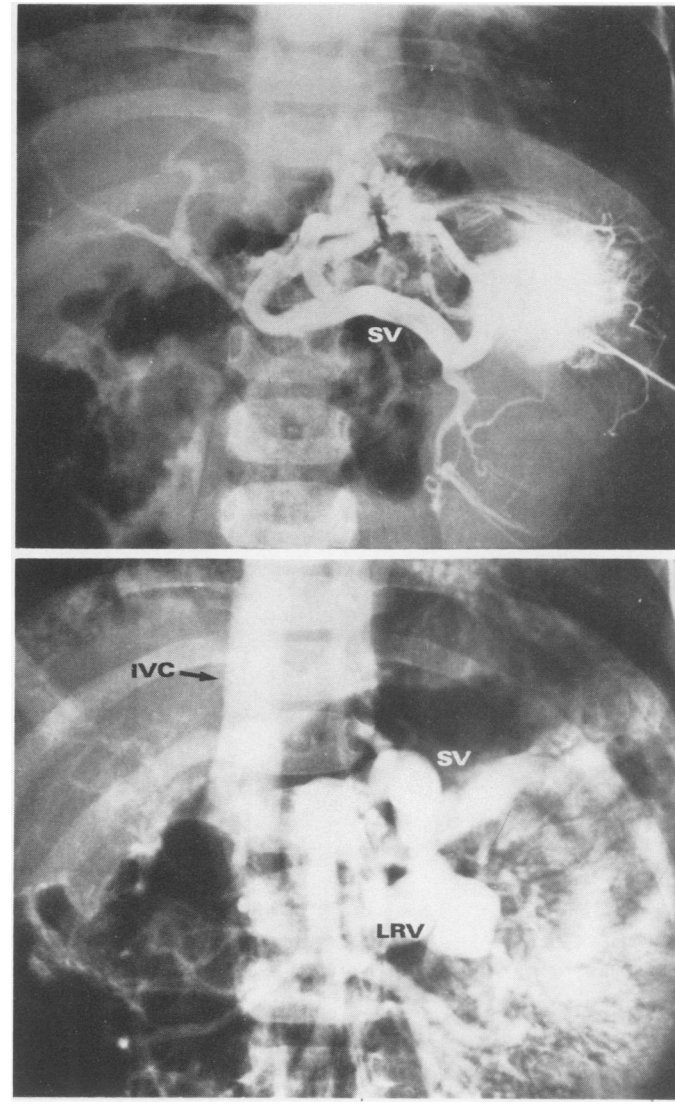


FIG. 6. Pre- and postoperative angiographic studies in a 42-month-old boy with thrombosis of the portal vein secondary to neonatal catheterization of the umbilical vein. (Top) Preoperative splenoportography: the splenic vein (SV) is 6 mm in diameter; most of the contrast medium flows through collateral channels into esophageal varices. (Bottom) Venous phase of superior mesenteric arteriography obtained one year after splenorenal shunt in the same child. Collateral channels have disappeared and the contrast medium fills the inferior vena cava (IVC) through enlarged splenic (SV) and left renal (LRV) veins.

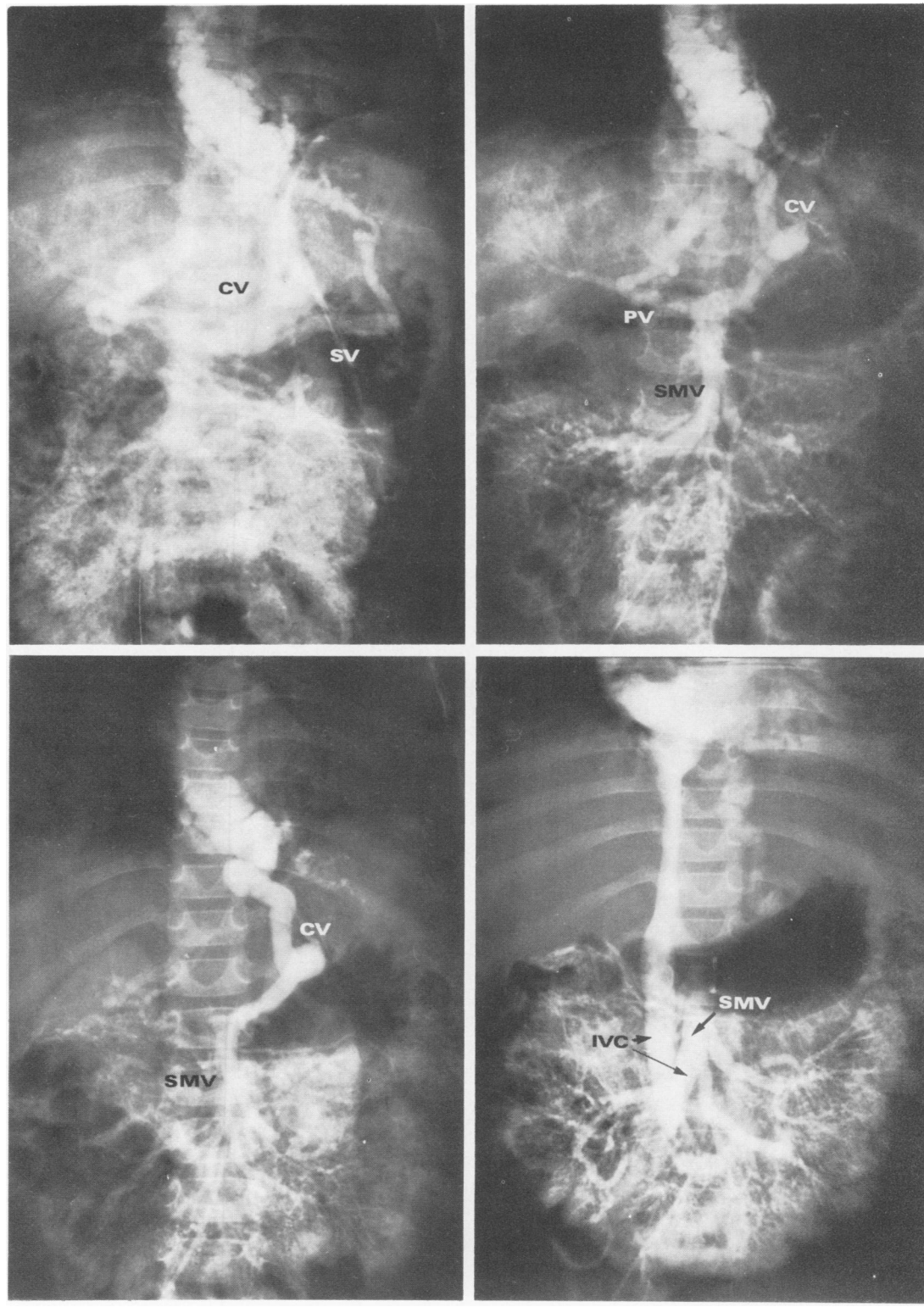


FIG. 7. Repetitive thrombosis of shunts in a 4 year, 5-month-old boy with liver cirrhosis (Case 1, Table 2). (upper left) Preoperative venous phase of superior mesenteric arteriography showing a large coronary vein (CV) and large esophageal varices. Splenic vein (SV) measured 7 mm. (upper right) Control angiogram two months after splenorenal shunt, following a recurrent hemorrhage: the shunt was not functioning and collateral channels were still opacified. A side-to-side portacaval shunt was then constructed. (lower left) Control angiogram two months after the new shunt, following a second episode of recurrent bleeding, showing obstruction of the shunt. The contrast medium opacifies the superior mesenteric vein (SMV) flowing into the coronary vein (CV). (lower right) A mesocaval shunt was finally performed and proven to be patent on a last angiogram. The dye opacifies the superior mesenteric vein (SMV) and the inferior vena cava (IVC).

played a role in that case, failure of this anastomosis has rendered us reluctant at the present time to perform shunts on veins of less than 4 mm in diameter. In the 2 other thromboses, the splenic veins both measured 7 mm and shunt failure was due to thrombocythemia in one case and mechanical compression in the other. Preoperative treatment with antiplatelet drugs appears to be advisable in patients with marked thrombocythemia which, however, is a rare event in portal hypertension before splenectomy. In order to detect any obstruction or stric-

TABLE 2. *Patients with Thrombosis of Splenorenal Shunt*

Case	Sex, Age	Cause of portal hypertension	Size of splenic vein	Cause of thrombosis	Reoperation	Status at present time
1	M, 4y 5m	cirrhosis of unknown origin	7 mm	thrombocytosis	1st rebleeding: side-to-side portacaval shunt 2nd rebleeding mesocaval shunt	no recurrent bleeding patent mesocaval shunt
2	M, 4y	cavernomatous transformation	7 mm	stricture of the anastomosis by the root of transverse colon	Unsuccessful attempt to reopen the shunt	no recurrent bleeding awaiting mesocaval shunt
3	F, 1y 6 m	cavernomatous transformation	2 mm	small size of the splenic vein	none	no recurrent bleeding awaiting mesocaval shunt

ture of the anastomosis, we now control the patency of the shunt in each instance by intraoperative portography. In an older child, not included in this series, operative angiography demonstrated a kinking of the splenic vein which led to reconstruction of the shunt with success.

Portal pressure often remained high at the end of the procedure, although less than preshunt figures. This, we think, is due to the small volume of flow through the diminutive splenic vein. Thus intraoperative portography appeared of greater value than pressure measurements to control the patency of the shunt. That early flow through the shunt was low as seen in the intraoperative portography by the small quantity of dye passing into the vena cava and the persistence, commonly observed, of collateral channels (Figs. 8 and 9). Postoperative angiograms performed one to 6 months later exhibited definite changes: enlargement of the splenic and left renal veins; decrease or disappearance of the collateral circulation, particularly the coronary vein and esophageal varices. That function of the shunt improves as time goes by was

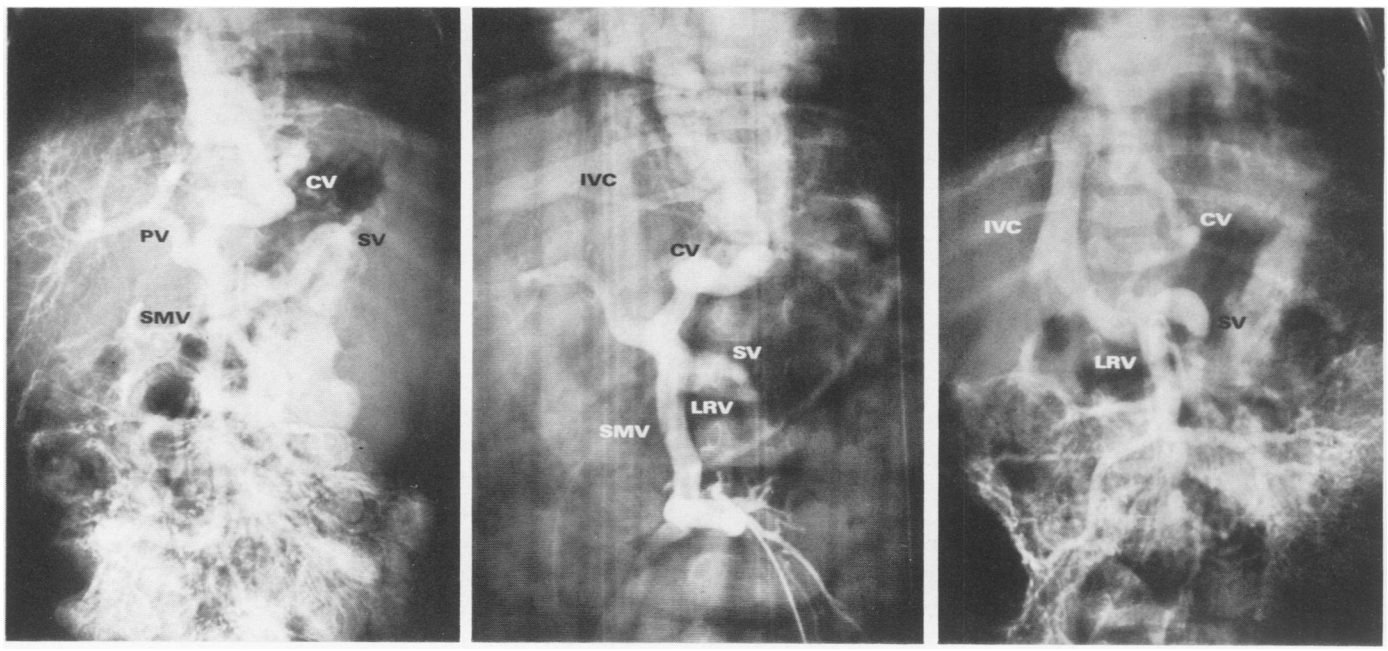


FIG. 8. Pre- and postoperative angiographic studies, in a 23-month-old boy with biliary atresia previously treated by portoenterostomy. (Left) Preoperative venous angiogram: most of the portal blood is diverted through a large coronary vein (CV) towards esophageal varices; the splenic vein (SV) measures 8 mm and is larger than the portal vein (PV) and superior mesenteric vein (SMV). (Center) Preoperative jejuno-portography following the performance of a splenorenal shunt. Although the shunt is patent as demonstrated by the visualization of splenic (SV) and left renal (LRV) veins and of the inferior vena cava (IVC), most of the dye still flows through the coronary vein (CV). Pressure gradient was the same before and after shunting. (Right) Postoperative angiogram taken a month later. The size of the splenic vein (SV) has increased and most of the contrast medium is now flowing through the shunt. The size of the coronary vein (CV) has decreased dramatically. Esophageal varices had almost totally disappeared at that time on esophagoscopic examination.

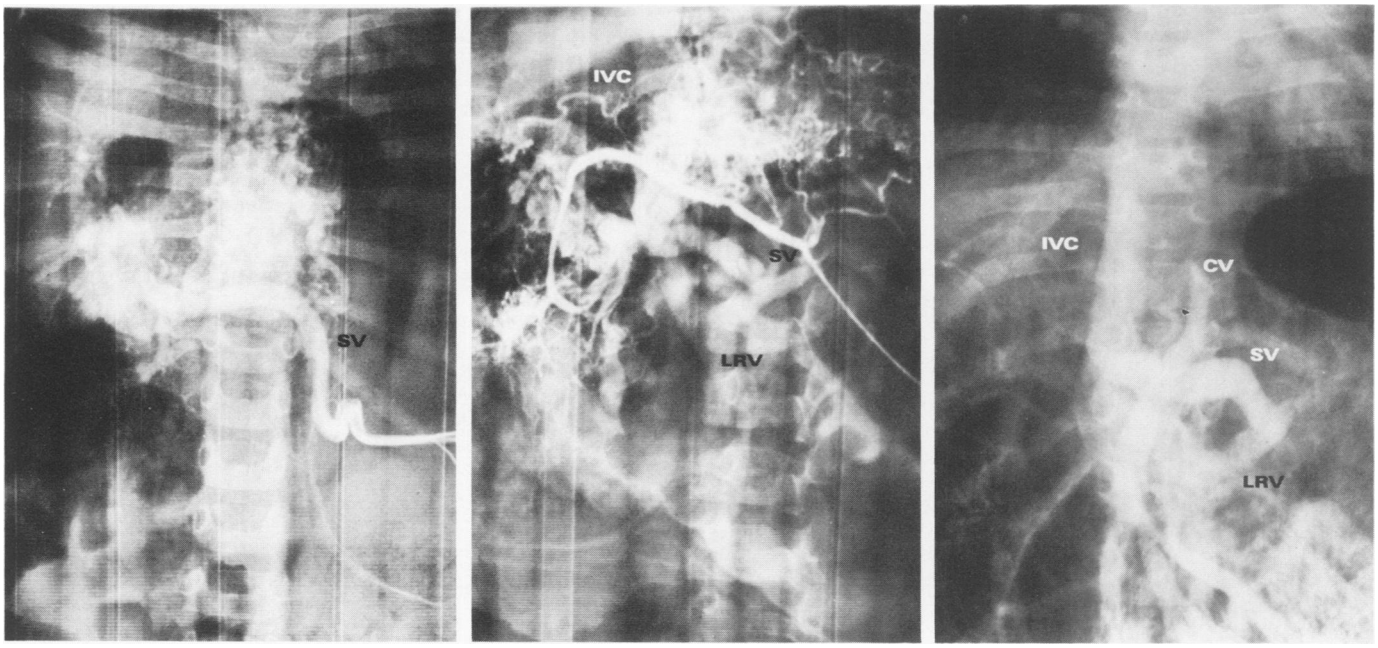


FIG. 9. Pre- and postoperative angiographic studies in a 36-month-old boy with portal thrombosis secondary to catheterization of the umbilical vein at birth. (Left) Preoperative splenoportography showing huge cavernomatous channels; the splenic vein (SV) measured 8 mm. (Center) Preoperative angiographic control by a catheter placed into the right gastric vein. Although the shunt is patent, only a small fraction of dye is diverted through it to the left renal vein (LRV) and inferior vena cava (IVC) and most of the portal blood still runs through collateral veins. (Right) Postoperative angiographic follow-up study a month later: the splenic vein (SV) has enlarged and all the portal blood is diverted towards the inferior vena cava (IVC) through the left renal vein (LRV).

emphasized by endoscopic examinations. In the immediate postoperative period varices often persisted although less dilated than preoperatively, and had disappeared at later visualization.

The success of these techniques argues that portal diversion should be strongly considered in the management of portal hypertension in early childhood. Until now, proposed surgical procedures had unknown effects and considerable risk. Direct attack on varices seemed to bear a high rate of postoperative rebleeding; this occurred in 81% of cases of variceal ligation in the series of Voorhees et al.[18] and 85% in the cases of Mikkelsen.[13] Rebleeding was also frequent following esophago-gastric resection and was observed in 45% of the cases of Koop and Kavianian[9] and 28% of the patients of Voorhees et al.[18] In addition, operative mortality was 21% in this last series. Recurrent bleeding due to thrombosis of the anastomosis was also a frequent complication of portal-systemic shunts in children. Thrombosis of splenorenal shunts was diversely observed in reported series of young children under 8 years of age: 82% for Clatworthy and de Lorimier,[5] 56% for Raffensperger et al.[16] and 50% for Pinkerton et al.[15] These results explain the lack of enthusiasm for operation and seemed to justify the conservative treatment of these children. Another argument for this non-operative approach was the low mortality rate of non-operated patients which was 4% in the series of Voorhees et al.[17] and 3% in the series of Fonkalsrud et al.[7] This led to the attitude recently defined by Clatworthy[6] that "any major surgical intervention including efforts to shunt with small vessels (under 1 cm) would definitely be contraindicated." The same opinion was expressed by Fonkalsrud[7]: "a shunt with a diameter of less than one centimeter is unlikely to remain patent and shunt procedures should be deferred until the patient is 8 years of age."

However, in some series, death associated with hemorrhage appeared more frequent, reaching 13% for Foster et al.[8] 21% for Arcari and Lynn[1] and 31% for Walker.[20] In addition variceal hemorrhage leads to repeated hospitalization, multiple operations and exposes to transfusion hepatitis. Clatworthy,[6] in his excellent review on 438 patients, reported a 5% mortality rate due to variceal hemorrhage, 5% dying from repeated operations and 2% due to transfusion hepatitis; the total figure reached 12% which can be taken as the spontaneous risk of conservative treatment.

Neuro-psychiatric abnormalities were described in 50% of shunted patients by Voorhees et al.[18] If this were really the case, it would darken the prognosis of shunted children and throw doubt on the benefit of shunt operations in young patients. Mental disorders were not observed in our series but the followup may be too short to appreciate the true incidence of this serious complication. The

significance of neurological disorders described by Voorhees et al.[18] is not clear. There is no similarity between these troubles and what is generally described, at least in adults, as post-shunt encephalopathy. Besides, the same authors[19] observed a similar percentage of neurologic disorders in patients with esophago-gastric resection without shunt. Fonkalsrud et al.[7] have drawn attention to the fact that neuropsychic disorders may be due in these children to prolonged hospitalization and multiple operations. In that case, permanent cure of portal hypertension by an early portal-systemic shunt could represent the best prevention against the occurrence of such troubles.

Addendum

Since June 1975, 6 more children under six years of age with portal hypertension, intrahepatic in 2 and extrahepatic in 4, have been operated on and received 4 splenorenal shunts and 2 portacaval shunts. All these shunts were patent on intraoperative angiographic control. In addition, one of the two children with thrombosed shunt (case 2 of Table 2) has had a successful mesocaval shunt.

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