Portal Diversion for Portal Hypertension in Children

The First Ninety Patients

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Ninety children with portal hypertension were treated by portal diversion. Fifty-two had cavernous transformation of the portal vein and 38 had an intrahepatic block from various causes. There were 59 central splenorenal shunts, 19 mesocaval, 11 portacaval and one distal splenorenal. In 61 peripheral shunts the veins used for the anastomosis were less than 10 mm in diameter. There was no operative mortality in children with extrahepatic block. One child with cystic fibrosis died postoperatively. Thrombosis of the shunt occurred in five children (5.6 per cent) and was responsible for recurrent bleeding in two. Four children with a thrombosed shunt underwent successful reoperation and one is awaiting another anastomosis. No late complications occurred in the 52 children with extrahepatic block, while encephalopathy developed in four children with intrahepatic block. These figures confirm our earlier results in the management of portal hypertension in childhood and suggest that portal diversion is the treatment of choice. Several precautions have permitted lowering of the rate of thrombosis whichever shunt is performed. Portal diversion should be indicated following the first episode of hemorrhage in children with extrahepatic block. In patients with intrahepatic block, congenital hepatic fibrosis and cystic fibrosis are good indications as are in general the hepatic diseases with no or mild activity.

THE TREATMENT OF PORTAL hypertension in children continues to be a challenge. In 1976 we reported our experience with operations for portal diversion in very young children, under six years of age, and showed that operation was feasible and often successful despite the small calibre of the vessels.[2] Since then, our experience has expanded to 100 children under 15 with portal hypertension and variceal bleeding. Ten have undergone variceal ligation only, indicated when no vein over 3 mm in size is available. Ninety children have had various procedures for portal diversion. This communication reports our experience with shunt operations in these children.

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Patients and Methods

Ninety children ranging in age from 18 months to 15 years have undergone various types of portal diversion procedures in our service since 1970 (Fig. 1). Forty-one were under six. The cause of portal hypertension was extrahepatic in 52 and intrahepatic in 38 (Table 1). All patients with cirrhosis were type A according to Child's classification. Thirty-one patients were operated on after the first bleeding episode. Four patients with extrahepatic portal hypertension underwent an emergency shunt (three for recurrent massive bleeding following prior variceal ligation).

Preoperative angiography (selective arteriography of the celiac axis and superior mesenteric artery and/or splenoportography) was obtained in every patient to assess the patency and the size of portal, splenic and mesenteric veins. In patients with intrahepatic portal hypertension, all veins were patent. In patients with extrahepatic portal hypertension, cavernous transformation extended to the splenic vein in two patients and the mesenteric vein in eight patients. The sizes of the veins used for peripheral shunts as measured on angiograms are shown in Figure 2. In the 42 patients with an extrahepatic block having both peripheral veins patent, the mesenteric vein was larger than the splenic vein in 21 patients, smaller in 15 patients and equal in six patients.

The anastomoses performed were central splenorenal in 59 patients, mesocaval in 19 patients, portacaval in 11 patients, and distal splenorenal in one patient. Central splenorenal shunt was performed according to the technique of Clatworthy and Boles,[4] and mesocaval shunt as described by Farge and Auvert.[6] Distal splenorenal shunt was performed according to

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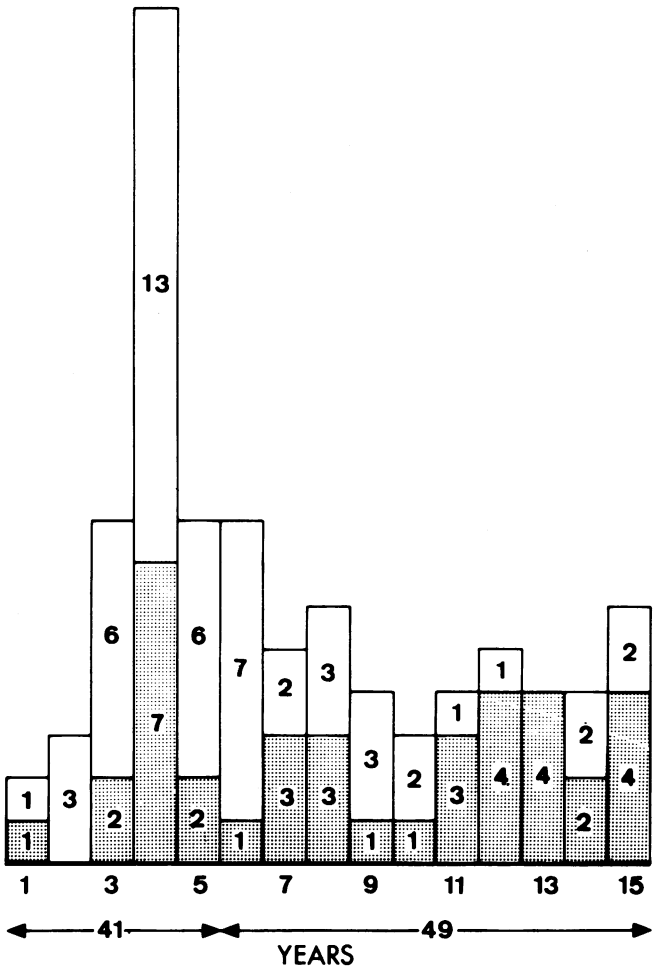

FIG. 1. Ages of 90 children with extrahepatic (open blocks) and intrahepatic (hatched blocks) portal hypertension treated by shunt operations.

the technique of Warren, et al.,[10] without any devascularization procedure. Some technical details should be emphasized: the shunt is performed using interrupted sutures, with 6–0 monofilament nylon. In the splenorenal shunt, the left part of the mesenteric root is opened to approximate the two veins as closely as possible; the renal vein is completely freed and clamped at its two ends in order to obtain a round stoma by excision of a patch from its anterior wall. In mesocaval shunt, the third duodenum is largely freed to avoid compression of the iliac vein turned up towards the mesenteric vein. During completion of the anastomosis, the lumen of both veins is frequently irrigated with heparinized saline. Two children had an associated biliary procedure for congenital stenosis of the bile ducts. A 4-year-old boy first underwent a portacaval shunt for biliary cirrhosis, then two months later repair of a congenital stenosis at the convergence of the bile duct; an 11-year-old girl had simultaneous central splenorenal shunt and hepaticojejunostomy for congenital diaphragm with intrahepatic stones. Two other patients had had previous Kasai procedures for atresia.

TABLE 1. *Etiologic Factors of Portal Hypertension in 90 Children Treated by Portal Diversion*

Extrahepatic block		52
cavernous transformation	34	
iatrogenic portal thrombosis (umbilical vein catheter)	18	
Intrahepatic block		38
cirrhosis	28	
secondary biliary cirrhosis	5	
deficit α 1-antitrypsin	4	
postnecrotic	3	
cystic fibrosis	2	
Wilson's disease	1	
Byler (?)	1	
unknown origin	12	
congenital hepatic fibrosis	7	
miscellaneous	3	

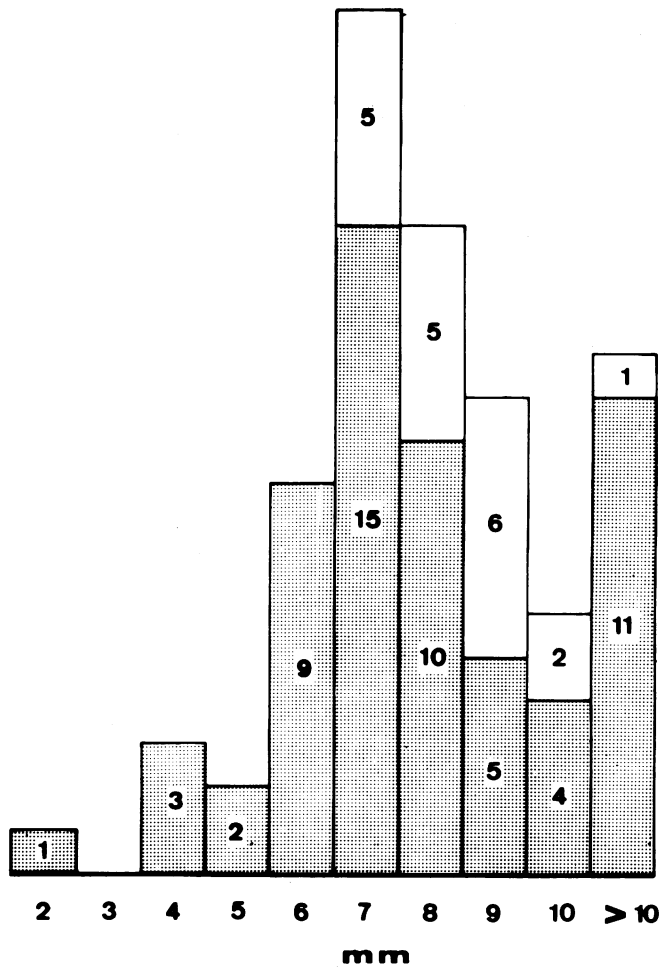

FIG. 2. Size of veins used for peripheral portal diversion: splenic veins (hatched blocks) in 59 central splenorenal and one distal splenorenal anastomoses and mesenteric veins (open blocks) in 19 mesocaval anastomoses. In patients receiving direct portacaval shunt, the portal vein was above 10 mm in diameter.

TABLE 2. *Results of Portal-systemic Shunts in 90 Children with Portal Hypertension*

	Extra-hepatic block (52)	Intra-hepatic block (38)	Total (90)
Postoperative complications			
mortality	0	1	1
morbidity			
intra-abdominal hemorrhage	2	1	3
encephalopathy	0	2	2
Late results			
thrombosis of the shunt	3	2	5
recurrent bleeding	1	1	2
encephalopathy	0	4	4
mortality	0	6	6

Evaluation of the shunt was done at operation by measurement of flow through the anastomosis. In the last 64 patients intraoperative ileoportography was done, leading to reconstruction of the shunt in two cases. Heparin (100 units/kg/day) was infused intraoperatively via peripheral vein as soon as the vessels were clamped and continued for ten days. In case of mesocaval shunt, in addition of anticoagulant therapy, special care was taken against venous complications of the lower limbs, including elevation of the feet and physiotherapy.

Until recently, patency of the shunts was tested by selective arteriography of the superior mesenteric artery one to six months postoperatively. Since the disappearance of esophageal varices seemed to correlate closely with patency of the anastomosis, we then decided to check the presence of esophageal varices by endoscopy or x-ray on the sixth postoperative month; if varices have disappeared, no further investigation is done; if they persist, angiography is then performed. Overall, 61 patients had angiographic control. In the 29 other children, varices were absent on barium swallow (five patients) or at esophagoscopy (24 patients). Mean follow-up averaged four years and two months. The status of all but two patients has been ascertained in the year preceeding preparation of this paper. The two lost to follow-up (with extrahepatic portal block) had been well at 18 months postoperatively without evidence of esophageal varices.

Results (Table 2)

Operative Mortality (60 Postoperative Days)

There was one operative death (1.8%). This occured in a 12-year-old boy with cystic fibrosis who died from respiratory failure, on the fifteenth postoperative day, complicating an episode of acute encephalopathy. No operative fatality occured in patients with extrahepatic block.

Postoperative Complications

Intraperitoneal hemorrhage occurred in three children. In two patients with portacaval vein thrombosis and mesocaval shunt, intra-abdominal bleeding was due to an overdose of heparin and necessitated emergency laparotomy. The rest of the postoperative course was uneventful. In an 11-year-old boy with cirrhosis, intraperitoneal bleeding was due to disruption of the ligature of the distal end of the splenic vein following central splenorenal shunt and necessitated an emergency procedure for religation of the vein. The postoperative course was then uneventful.

Recurrent variceal bleeding occured following portacaval shunt in a 12-year-old girl with cirrhosis and was responsible for severe encephalopathy with coma. Recurrent hemorrhage was secondary to thrombosis of the anastomosis and treated by emergency mesocaval shunt on the fifth postoperative day. No complication was observed at the level of the legs following mesocaval shunt.

Patency of the Shunt

Eighty-five shunts were proven to be patent (including the postoperative death): 55 central splenorenal, 19 mesocaval, 10 portacaval and one distal splenorenal. None of these 85 children experienced recurrent hemorrhage. Late postoperative angiographic controls showed growth of the shunts (Fig. 3).

Thrombosis was observed in four splenorenal shunts and one portacaval shunt (5.6%). In two patients, thrombosis was revealed by recurrent bleeding and demonstrated by subsequent angiogram. In three patients, thrombosis of the anastomosis was discovered by systematic control of varices and confirmed by postoperative angiography. Detailed observations concerning these five children are given in Table 3. Four occurred among our first 20 patients. The cause of the thrombosis was recognized in four patients. In one patient, the vein used for the shunt was very small (2 mm) and heparin was not used because of the emergency of portal diversion performed for massive variceal bleeding. In three patients, there was a technical error. Four of these children had a successful subsequent mesocaval shunt and one is awaiting such a procedure.

Recurrent Bleeding

Two of the five patients with thrombosed shunts rebled (cases 1 and 5, Table 3). None of other patients has had recurrent bleeding.

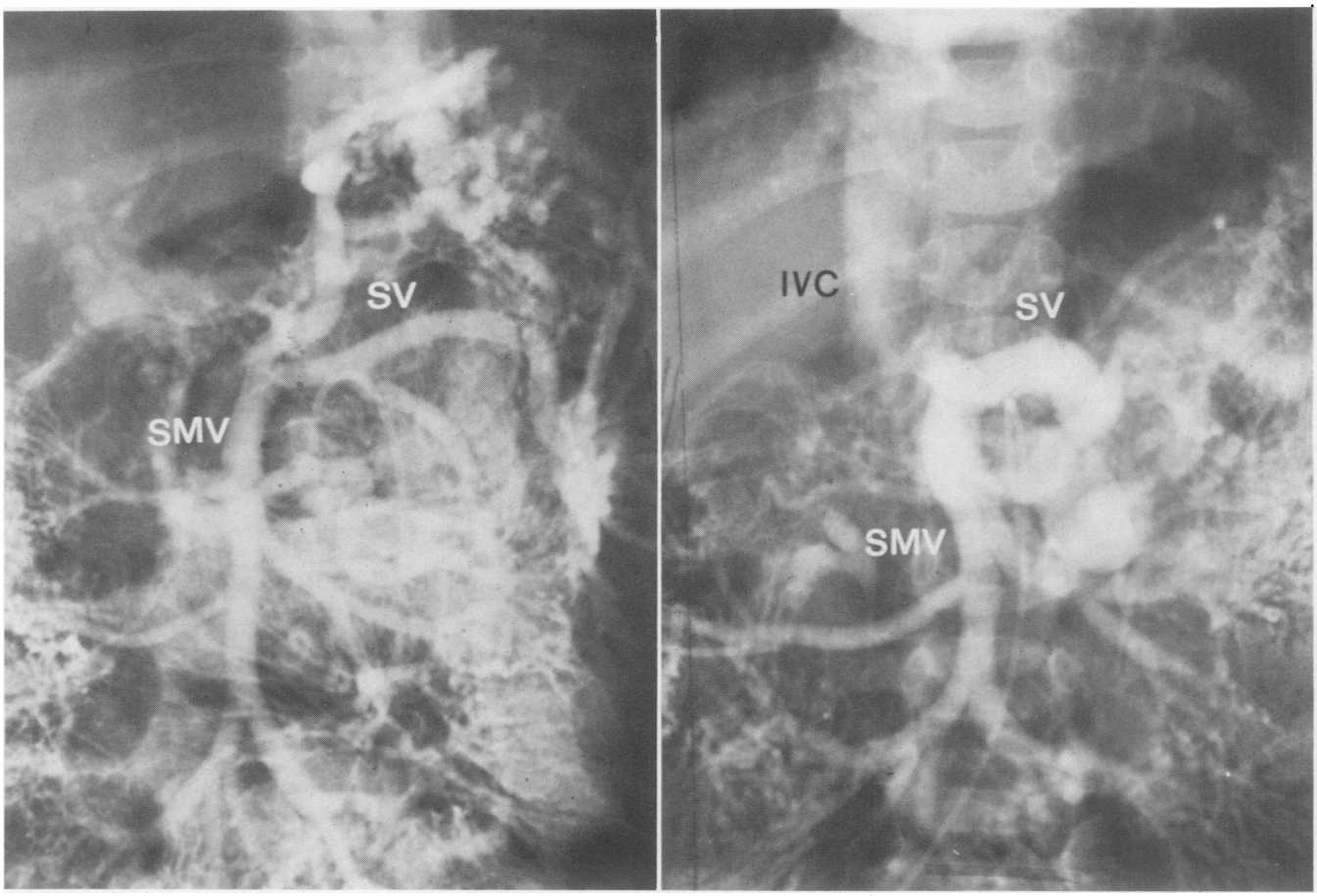

FIG. 3. A. Preoperative angiography of superior mesenteric artery in a 4-year-old boy with cavernous transformation of the portal vein secondary to catheterization of the umbilical vein at birth. Splenic vein (SV) is narrow, 5 mm in diameter. Superior mesenteric vein (SMV) is slightly wider (6.5 mm). Central splenorenal shunt was performed and splenic vein was found to measure 6 mm at operation. B. Postoperative (6 months) angiographic control of the splenorenal shunt in the same child. The shunt is patent as shown by passage of contrast medium into inferior vena cava (ICV). Due to the growth of the child, all veins are a little wider than preoperatively (SMC = 7 mm). Yet, the splenic vein has markedly enlarged and measures 12 mm in diameter.

Encephalopathy

None of the children with extrahepatic block has demonstrated any sign of postoperative encepalopathy. Among the 38 patients with intrahepatic block, apart from the two instances of acute postoperative encephalopathy already described, four children had chronic encephalopathy following splenorenal (two patients) or portacaval shunt (two patients). One patient had Wilson's disease, one patient had a homozygotous deficit in α1-antitrypsin and two a cirrhosis of unknown origin. Neuropsychiatric status progressively deteriorated as well as hepatic function until death in the four.

Late Survival

There were no late deaths in the children with extrahepatic portal hypertension. Six children with intrahepatic portal hypertension died. Four died (including the four above) 5, 10, 15 and 48 months after portal operation from progressive deterioration of hepatic function. A 4-year-old girl with biliary atresia and a Kasai syndrome, operated on by splenorenal shunt, had several bouts of cholangitis and died from fulminant staphylococcus septicemia in the twenty-sixth postoperative month. A 5-year-old girl with histiocytosis died from gram negative septicemia in the twenty-fifth postoperative month while under chemotherapy.

Discussion

In 1976 we described our experience with portal diversion in small children under the age of 6, concluding that such operations were successful in preventing recurrent bleeding.[2] Our continuing experience with this larger group confirms that conclusion: only five of 90 shunts in children thrombosed (5.5%). Infancy or small size of the vessels does not seem to

TABLE 3. *Patients with Thrombosis of the Anastomosis*

Case	Sex, Age	Case of portal hypertension	Type of operation	Size of the vein	Cause of thrombosis	Reoperation	Status at present time
1	M, 4y5m	Cirrhosis of unknown origin	Splenorenal shunt	7 mm	Thrombocytosis	1st rebleeding side-to-side portacaval shunt, 2nd rebleeding mesocaval shunt	No recurrent bleeding patent mesocaval shunt
2	F, 11y	Cavernomatous transformation	Splenorenal shunt	8.5 mm	Unknown	Mesocaval shunt	No recurrent bleeding patent mesocaval shunt
3	M, 4y	Cavernomatous transformation	Splenorenal shunt	7 mm	Stricture of the anastomosis by the root of transverse colon	1st unsuccessful attempt to reopen the shunt 2nd mesocaval shunt	No recurrent bleeding patent mesocaval shunt
4	F, 1y6m	Cavernomatous transformation	Splenorenal shunt	2 mm	Small size of the splenic vein	None	One recurrent bleeding awaiting mesocaval shunt
5	F, 12y	Cirrhosis of unknown origin	Portacaval shunt	12 mm	Stricture of the anastomosis by lymph nodes of the hepatic pedicle	Rebleeding—emergency mesocaval shunt	No recurrent bleeding patent mesocaval shunt

represent a contraindication to portal diversion. Among the 41 children under six years of age, only three thromboses occurred (7.3%). It must be noted that four of the five thromboses observed occurred among the first 20 patients; only one thrombosis has occurred among the last 70 patients (1.4%). The decrease in the incidence of thrombosis of the anastomosis from 13% in 1976 to 5.5% at present is explained by several factors: 1) The gain of technical skill due to greater experience. Analysis of the causes of thrombosis has allowed recognition of several technical pitfalls which are now avoided. 2) The performance of intraoperative portography which proves the patency of the shunt. If a stricture or twisting of the shunt is seen, the anastomosis may be repeated. This was done in two patients. A thick splenic vein twisted during a splenorenal shunt in a 13-year-old girl, and in a 5-year-old girl a low third portion of duodenum compressed an ileomesenteric anastomosis. This was corrected by transection of the duodenum and gastroenterostomy (Fig. 4). 3) Heparin is systematically given early as soon as the vessels are clamped, decreasing the risk of clot formation even during the construction of the shunt. The precocity of thrombosis of the shunt was observed in one patient who had a portacaval anastomosis. In the minutes following completion of the anastomosis, the stoma was obstructed by a platelet clot. Great care must be taken to keep the dosage minimal since short-term overdosage was responsible for postoperative intraperitoneal hemorrhage in two patients. We use continuous iv heparin infusion (100 units/kg/day) during the four first postoperative days, then subcutaneous heparin. The duration of anticoagulant therapy is arbitrarily limited to ten days, in fact, we think that if the shunt works well for the first postoperative hours, the risk of thrombosis is very low. We have not observed late thrombosis when the shunt was patent on the peroperative angiographic control. The high rate of shunt patency is independent of the type of anastomosis performed. It must be noted that, after mesocaval shunt, no leg edema or signs of peripheral thrombosis were detected in any patient even those with cirrhosis. This is in sharp contrast with what is usually reported in adult cirrhotic patients who present with marked leg edema following ligation of both iliac veins. The ability of children to develop new collateral channels is probably responsible for this.

The site of the anastomosis depends on the level of the block. In intrahepatic portal hypertension, there seems to be no difference among the types of portal-systemic shunts for the immediate and late results and we would now recommend choosing the easiest to perform. The first choice, therefore, is portacaval shunt. In case of anticipated technical difficulties, for example previous operation in the right hypocondrium, a peripheral shunt may be preferred. In extrahepatic blocks the type of shunt depends on the extension of the cavernous transformation. In a few cases (one-fifth in our experience), only one vein (generally the splenic vein) is patent and can be used. When both veins are patent and suitable, the choice of the type of anastomosis depends on several factors. We initially preferred central splenorenal shunts because of our large experience with adults. Now we are more selective. We take into account both the risk of splenectomy in young children and the size of the veins. The risk of

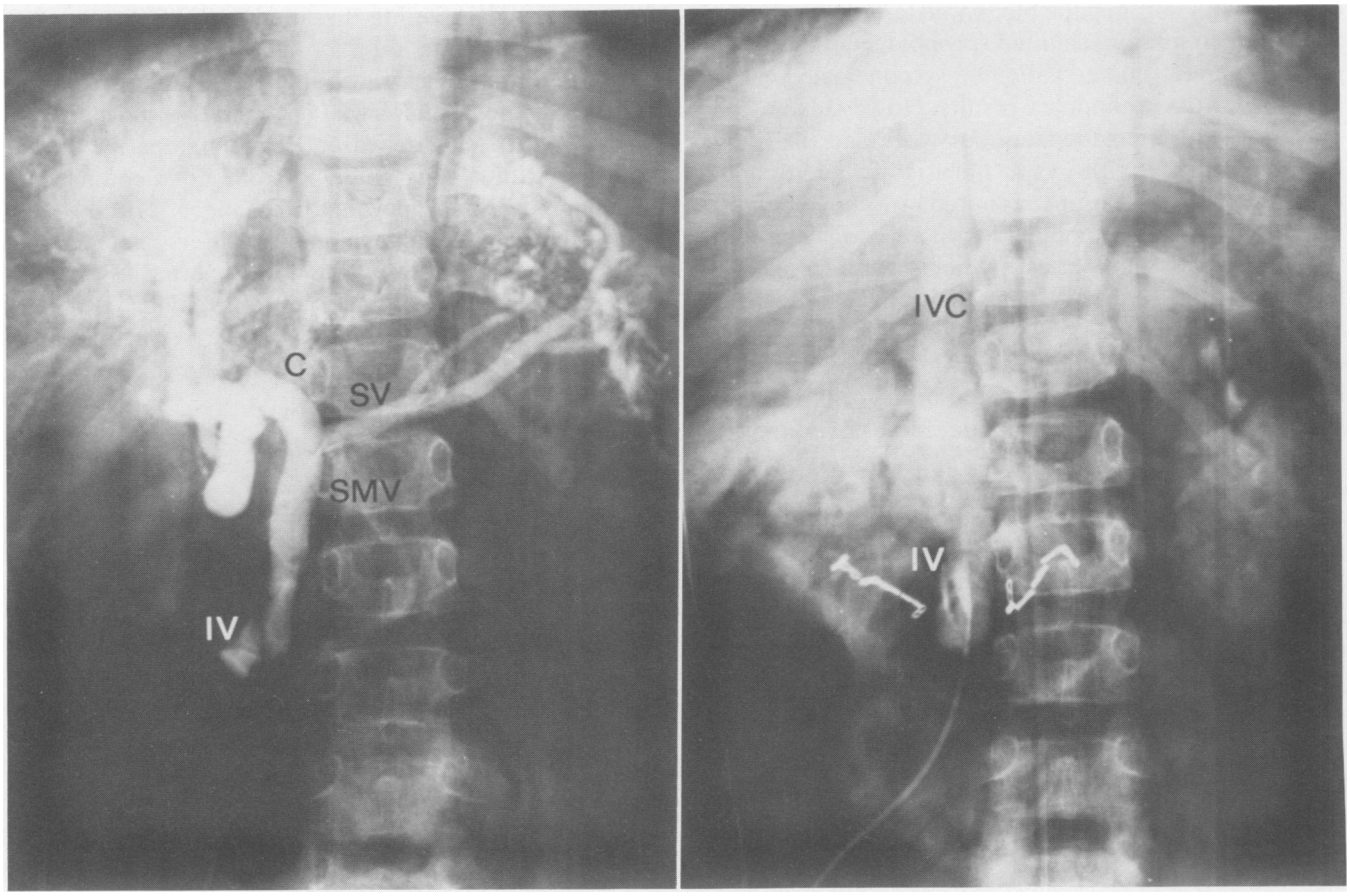

FIG. 4. Peroperative ileoportographies in a 5-year-old girl receiving a mesocaval shunt for extrehepatic portal hypertension. A. First angiographic control: The contrast medium opacifies the superior mesenteric vein (SMV), the splenic vein (SV) and the carvernoma (C). Although the distal part of the right iliac vein (IV) is opacified, almost no flow is visible in the vena cava, suggesting that the shunt is not properly functioning. The cause of the failure of the shunt was a compression by a low third duodenum. The duodenum was then cut using mechanical sutures. B. Second ileoportography after section of the third duodenum. Contrast medium flows directly through the anastomosis into the iliac vein (IV) and inferior vena cava (IVC). Clips on the two ends of the duodenum are seen on each side of the anastomosis.

acute bacterial infection in young splenectomized children, although low (6%),[5] must be considered since it may lead to death by fulminant bacteremia. In two patients in our series, late mortality was due to septicemia following splenorenal shunt. It must be noted that both children had an underlying disease predisposing them to spontaneous infection (cholangitis from biliary atresia and chemotherapy for histiocytosis) and that the micro-organism was not the usual pneumococcus. However, in view of this risk, we now prefer when there is an underlying disease which may increase the risk of infection and/or when the child is under four years of age, to avoid splenectomy. If a central splenorenal shunt has to be performed (absent or small superior mesenteric vein) antibiotics are given to the child until adulthood. Distal splenorenal shunt (leaving the spleen) seems to be a good alternative for patients when the mesenteric vein is not suitable and when splenectomy is inadvisable. This was the case in the patient in whom we performed a distal splenorenal shunt. The patient was a 4-year-old boy in whom a previous Roux-en-Y biliojejunostomy precluded construction of a mesocaval shunt. Gastric devascularisation is not done with distal splenorenal shunt because conservation of the portal blood flow is not an issue in extrahepatic block. Technical feasability of mesocaval shunt is facilitated by 1) prolongation of the vena cava by the right iliac vein according to the technic described by Farge and Auvert;[6] 2) extensive mobilization of the third duodenum; and 3) the possibility of implanting the iliac vein in the right branch of the mesenteric vein when the formation of the mesenteric trunk is quite proximal.

Indications for Operation Depend Upon the Site of the Block

In children with intrahepatic hypertension, there is no proof in favor of the benefit of portal diversion. To determine the usefulness of shunt operations in

children with intrahepatic block we started a study nine years ago comparing shunted (prophylactic and therapeutic) and nonshunted patients. No conclusion can be drawn yet from preliminary results. However some observations can be made from our work and the literature. 1) Patients with cystic fibrosis are good candidates for shunt operations, as noted by Schuster et al.,[8] who had very few complications and good long-term survival. 2) In our series, patients with congenital hepatic fibrosis equally seem to be adequate candidates; none of our seven patients developed encephalopathy or died. 3) Patients with Wilson's disease, homozygotous (zz) deficit in α1-antitrypsin and perhaps some forms of intrahepatic cholestasis may be bad candidates since they have a continuing liver disease.

In children with extrahepatic portal hypertension, we advocate operation following the first episode of upper gi bleeding, particularly in young children. In our experience when bleeding episodes come early, they tend to repeat with greater frequency than in older children. Among patients operated on before the age of four years 50% had had more than two episodes of hemorrhage as compared with 12% among those operated on at an older age. The nonoperative attitude adopted by several authors developed from an experience with extrahepatic block in adults in whom bleeding episodes are usually few and mild. This does not seem to be the case in children. In 67% of our patients there was more than one bleeding episode. In the series of Foster, et al.[7] and Arcari and Lynn,[1] the mortality associated with hemorrhage reached 13% and 21% respectively. In the larger series of Clatworthy,[3] the overall mortality was 12% either directly or indirectly related to upper gastrointestinal bleeding. In our series, seven patients underwent emergency procedures for massive life-threatening hemorrhage. Our overall good results are a strong argument in favor of early operation in extrahepatic block. Among the 52 operated patients there was no operative mortality or late mortality. There was no encephalopathy. Even children with a long follow-up did not demonstrate any significant neuropsychiatric changes. The absence of postoperative encephalopathy differs from Voorhees, et al.[9] and support the hypothesis already suggested that most of these neuropsychiatric troubles may well have been developed as a result of oversolicitous parents and the patient's own concern for the risk of life-threatening bleeding.

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