Portal Hypertension in Childhood

JOE A. PINKERTON, M.D., GEORGE W. HOLCOMB, JR., M.D., JOHN H. FOSTER, M.D.

From the Department of Surgery, Vanderbilt University Medical Center, Nashville, Tennessee 37232

MASSIVE GASTROINTESTINAL BLEEDING in children presents a very disturbing problem and is frequently the first manifestation of portal hypertension. Evaluation of children with portal hypertension using current technics provides a safe and accurate localization of the obstruction of portal flow to the intrahepatic or extrahepatic areas. In the majority of instances portal hypertension occurs secondary to extrahepatic block caused by portal vein thrombosis while cirrhosis causing an intrahepatic block is seen less frequently. The management, operative procedures and overall prognosis differs in children with portal thrombosis and those with cirrhosis. Portal decompression is the objective of operative management of these patients but the complex course frequently demands prolonged non-operative management and a variety of ancillary surgical procedures which do not directly reduce portal venous pressure.

This report reviews the course and management of 33 children under the age of 15 years treated in the Vanderbilt Medical Center with portal hypertension. It includes a long-term follow-up study of 22 patients reported in 1963 [6] and the addition of 11 new patients treated since 1963. All surviving patients were recalled and reevaluated as to current status and shunt patency. One patient was lost to follow-up providing a 97% follow-up.

Presented at the Southern Surgical Association Meeting held at Hot Springs, Virginia, December 6–8, 1971.

Supported in part by USPHS Grant #5M01 RR-95.

Presentation

There were 33 patients who ranged in age from 2 months to 14 years at the first manifestation of portal hypertension. The group included 14 females evenly divided in the cirrhosis and portal vein thrombosis groups and 19 males only 3 of which had cirrhosis. Portal vein thrombosis was the etiology in 23 patients and cirrhosis in 10 patients (Figure 1). The first manifestation of portal hypertension included hemorrhage in 21 patient's, splenomegaly in four, fever in two and ascites in one and hepatomegaly in one patient (Table 1). In seven patients the initial hemorrhage presented as blood per rectum in two of which there was bright red rectal bleeding. The initial findings on physical examination included splenomegaly in 32 patients, hepatomegaly in 18, ascites in seven, jaundice in one and cutaneous spiders in one. Initial hemorrhage as well as recurrent episodes were preceded by a febrile episode with or without an upper respiratory infection in over one half of the bleeding episodes.

Evaluation

Evaluation consisted of complete blood counts and liver function tests, esophagogram and splenoportography with measurement of splenic pulp pressure. The latter test became a routine part of the evaluation in the mid 1950's.

All patients with the portal vein thrombosis had normal liver function tests while eight of 10 patients with cirrhosis had nor-

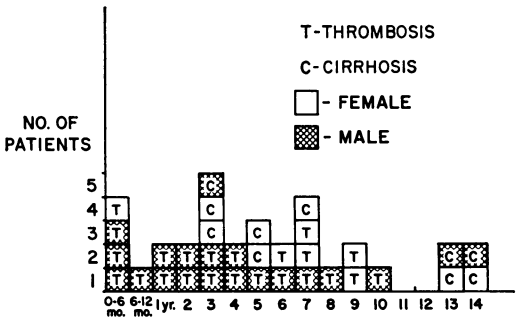

FIG. 1. Age at first manifestation of portal hypertension.

mal liver function. Hypersplenism was reflected in the hemogram in seven patients with portal vein thrombosis and eight patients with cirrhosis. Esophagogram was done in 30 patients. Varices were seen in seven of 10 patients with cirrhosis and in 16 of the 20 patients with portal vein thrombosis.

Splenoportography was done in nine of the 10 patients with cirrhosis and 13 of the 23 patients with portal vein thrombosis. The 10 patients with portal vein thrombosis who did not undergo splenoportography were treated prior to 1953. The splenoportogram has allowed preoperative differentiation between cirrhosis and portal hypertension (Fig. 2) and has been used postoperatively to evaluate shunt patency. Splenic pulp pressure has ranged from 195–600 mm. saline. More than 60 splenoportograms have been done in 22 of the children in this study without complication. One patient was thought to have a subphrenic abscess after the splenoportogram but exploration failed to confirm this diagnosis. Prior to 3 years ago we used general anesthesia for splenoportography, since that time sedation with Ketamine has sufficed. The needle is inserted through the 9th intercostal space and following the procedure the patient lies with the left side down for 2 hours.

TABLE 1. *First Manifestation of Portal Hypertension*

	Cirrhosis	Thrombosis
Hemorrhage	4	17 (7)*
Splenomegaly	4	4
Fever of unknown origin	1	1
Ascites	0	1
Hepatomegaly	1	0

* Blood per rectum.

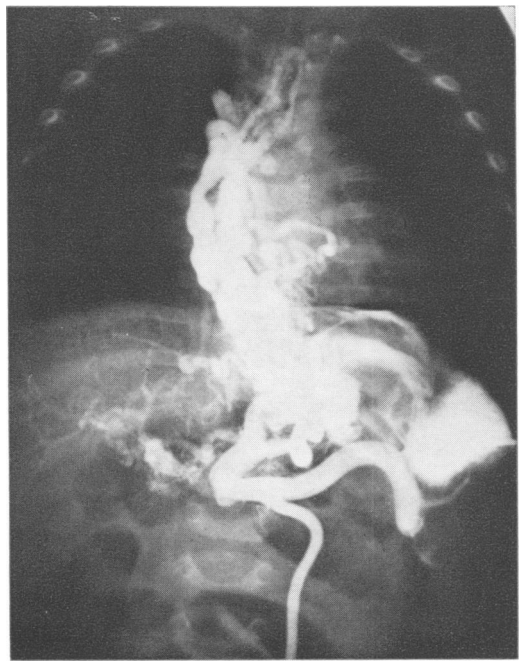

FIG. 2A. Splenoportogram in an 8-month-old boy showing the typical venous pattern of portal vein thrombosis with extensive perigastric and periesophageal collaterals and very little portal flow to the liver.

Cirrhosis

Cirrhosis was the cause of the portal hypertension in 10 patients. There was a history of jaundice or a febrile episode which may have been hepatitis in four patients. The other six patients had no history of antecedent hepatic disease.

Hemorrhage was the primary indication for operation in six patients (Table 2). The mean time interval from the first hemorrhage to operation was 1.5 years with a range of 0–6 years. Two patients were shunted after the first hemorrhage and four after repeated bouts of hemorrhage. The six

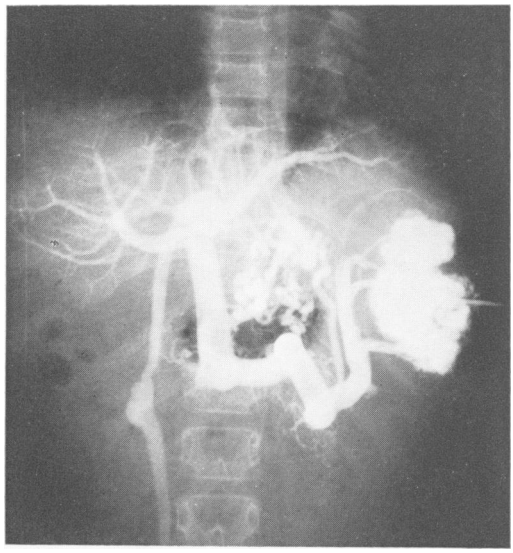

FIG. 2B. Splenoportogram in a 7-year-old girl showing the typical pattern of cirrhosis with a patent portal vein and a very small liver. There is also retrograde filling of the umbilical vein.

patients had an average of three bleeding episodes prior to operation with a range of 1–6 hemorrhages. Hypersplenism was the indication for operation in four patients (Table 2). All four patients had a splenorenal shunt.

Non-Operative Management

Patients who hemorrhaged were managed non-operatively if possible to allow optimal preparation for an elective shunting procedure. Non-operative management consisted of bed rest, sedation, and blood transfusion. Intravenous pitressin was used in one patient and a Blakemore Sengstaken tube in two patients. Ascites was managed with diuretics, sodium restriction and administration of serum albumin.

TABLE 2. *Indication for Operation*

	Cirrhosis	Thrombosis
Hemorrhage	6	21
Initial	2	5
Repeated	4	16
Hypersplenism	4	0

TABLE 3. *Results of Operative Treatment in Ten Patients with Cirrhosis*

Operation	No. Patients	Shunt Patent	Operative Deaths	Late Deaths
Portacaval shunt	4	4	1	1
Splenorenal shunt	6	5*	0	3
Total	10	9	1	4

* The one thrombosis was discovered at autopsy 9 years after the operation.

Operative Management

Patients with cirrhosis have required 12 operative procedures to correct or control the complications of portal hypertension. These included four portacaval shunts, six splenorenal shunts and transthoracic ligation of varices in two patients (Table 3).

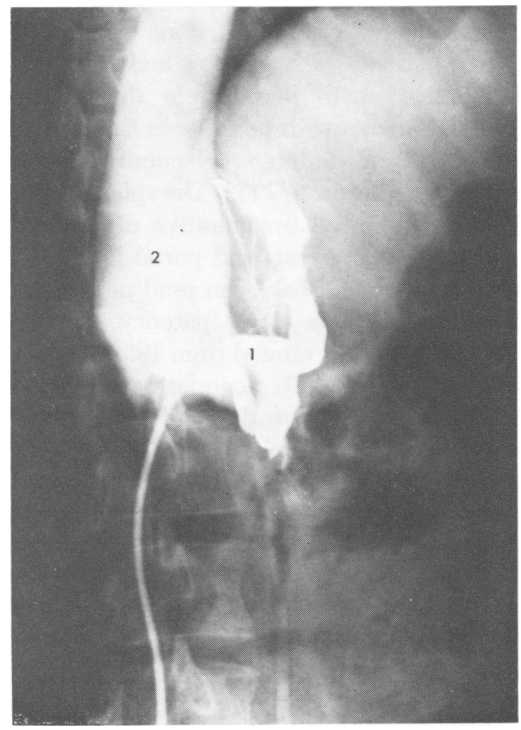

FIG. 3. Demonstration of patency of H-type Teflon graft portacaval shunt 3 years postoperative in a 17-year-old young lady. Catheter is inserted via right saphenous vein and shows patent Teflon graft (1) and flow into portal vein and inferior vena cana (2).

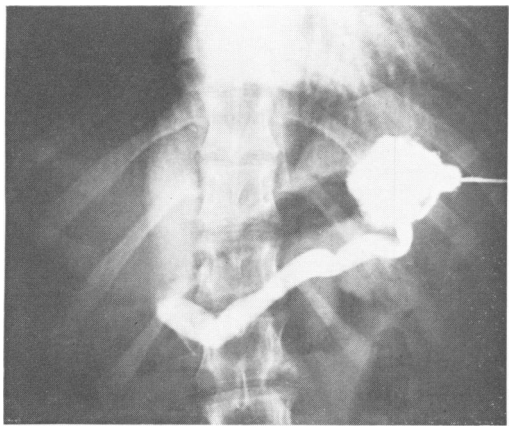

FIG. 4. Splenoportogram in a 10-year-old girl with cirrhosis 4 years after portacaval shunt.

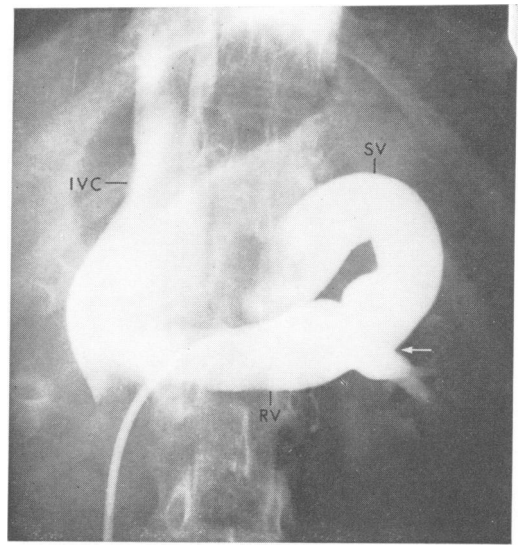

FIG. 5. Demonstration of patent splenorenal shunt (arrow) in a 13-year-old girl with cirrhosis 5 years postoperative. Catheter is inserted via the right saphenous vein. SV—Splenic Vein, RV—Renal Vein and IVC—Inferior Vena Cana.

Portacaval Shunt

Portacaval shunt was done in four patients whose ages were 5, 6, 7, and 14 years. One patient had undergone gastric transection and suture of varices 3 months previously. Side-to-side shunt was used in one patient with repeated hemorrhages who had severe cirrhosis and ascites. In one patient (age 14 years) portacaval shunt was done using woven crimped Teflon graft, 2 cm. in length and 14 mm. in diameter, to create a H type shunt (Fig. 3). This shunt has now been patent for 11 years. End-to-side portacaval shunt was used in the other two patients. One patient bled postoperatively despite a patent shunt demonstrated radiographically (Fig. 4). After balloon tamponade failed to control the hemorrhage, transesophageal ligation of the varices was used. There were no subsequent bleeding episodes. Two shunts are currently patent and the other two were patent at autopsy in patients who died 3 weeks and 12 years postoperatively (Table 4).

TABLE 4. *Causes of Death in Five Patients with Cirrhosis*

Cause of Death	No. Patients	Postoperative
Hepatic failure	2	3 weeks, 6 years
Acute hepatitis and hepatic failure	2	13 mo., 9 years
Tuboovarian abscesses	1	12 years

Splenorenal Shunt

Splenorenal shunt was used in six patients, they ranged in age from 4 to 14 years (average 12.4 years) with only one child under 9 years of age. Four were operated on for hypersplenism and two for hemorrhage. End-to-side splenorenal anastomosis after splenectomy was used in each instance (Fig. 5). Hemorrhage recurred 2 years postoperatively in one patient who survives now 22 years after operative with a patent shunt and no further hemorrhage. One patient died of hepatic failure 9 years postoperatively and was found to have an occluded shunt. This is the patient who was 4 years old at the time of shunting. He had no varices and had not bled after splenorenal shunt. All other splenorenal shunts are open currently or were open at autopsy in the two patients who died 1 and 6 years postoperatively.

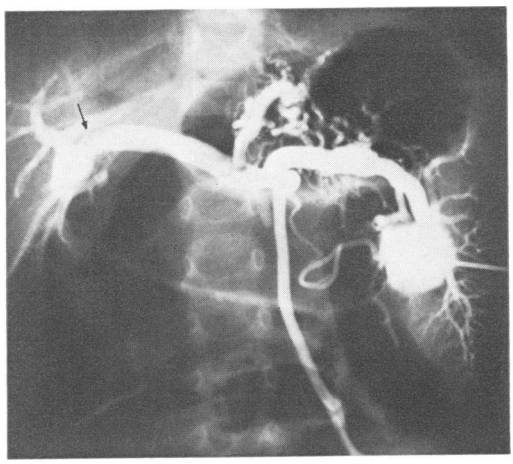

FIG. 6A. Splenoportogram in a 15-month-old boy showing an intraluminal defect (arrow) in the portal vein suggestive of a clot. Splenic pulp pressure at this time was 300 mm. saline.

Course-Early

There was one postoperative death in a 7-year-old girl with severe cirrhosis in whom preoperative laboratory studies included BSP retention 18%, Albumin 2.0 Gm./100 ml. Cephalin flocculation 4 plus, thymol turbidity 7.0, and bilirubin of 1.5 mg./100 ml. Death resulted from hepatic failure 21 days after a side-to-side portacaval shunt. Both patients who required use of the Blakemore-Sengstaken tube and ligation of the varices also required tracheostomy for aspiration pneumonitis.

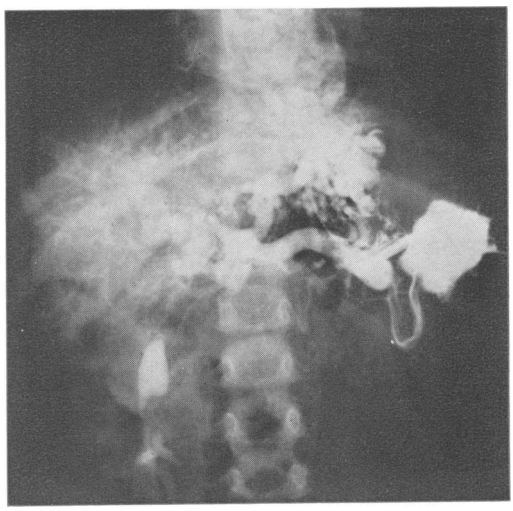

FIG. 6B. Splenoportogram in the same patient at 4½ years of age showing thrombosis of the portal vein with decrease in hepatic flow and development of periesophageal collaterals. Splenic pulp pressure at this time was 450 mm. saline.

Late

There have been five deaths in the 10 patients with cirrhosis (Table 4). Four resulted from hepatic failure. Death occurred 6 years and 9 years after operation as a result of acute hepatitis superimposed on pre-existing cirrhosis in two patients. Death in one patient was unrelated to hepatic disease, but resulted from complications of bilateral tuboovarian abscesses.

The five survivors with cirrhosis are now 1, 2, 7, 12 and 22 years after operation. All patients have patent portosystemic shunts, are free of varices on esophagram, and are leading normal, active lives.

Portal Vein Thrombosis

Thrombosis of the portal vein was the cause of portal hypertension in 23 patients. A history of neonatal omphalitis was obtained in six patients. Two patients had been treated for neonatal respiratory distress syndrome with umbilical vein catheterization for monitoring purposes. One child had undergone cystojejunostomy for a choledochal cyst in infancy.

One patient, with no antecedent disease to explain the portal vein thrombosis, developed gastrointestinal hemorrhage at 15 months of age and on splenoportogram was found to have a pulp pressure of 300 mm. saline. The portal vein was patent but there was a constant filling defect in the portal vein just before it bifurcated. Repeat splenoportogram at age 4½ years revealed complete occlusion of the portal vein with massive venous collaterals and a splenic pulp pressure of 450 mm. saline. This patient has now had 15 bleeding episodes and is being managed by non-operative means until he reaches adequate size for a successful shunting procedure (Fig. 6).

TABLE 5. *Results of 18 Portal-Systemic Shunts in 13 Patients with Portal Vein Thrombosis*

Type of Shunt	No. of Shunts	Shunt Thrombosis	Shunt Patent	Operative Deaths	Late Deaths
Splenorenal	10	7*	3	0	0
Mesocaval	6	2**	4	0	0
Make shift	2	2***	0	0	0

* Three then had mesocaval shunt.
** One then had splenorenal shunt.
*** One then had a mesocaval shunt.

Variceal hemorrhage was the indication for operative treatment in all patients (Table 2). Operation was performed after initial hemorrhage in four patients and after repeated hemorrhages in 16 patients. There was a mean interval of 2.3 years from the first hemorrhage to initial operation. The bleeding episodes ranged from 1 to 9 prior to initial operation with a mean of 3.2 episodes. Two patients have had no operation thus far and are managed by non-operative measures until they reach adequate size for successful shunt.

Non-Operative Management

The management of hemorrhage in children with portal vein thrombosis is somewhat more refined than that used in instances early in this series. The elements of current management consist of careful monitoring and observation with bed rest, sedation, blood transfusion and intravenous pitressin. Hemorrhage has been of limited duration with this approach and valuable time has been gained to allow growth and increase in size of the portal venous system. Liver function studies have been normal and hepatic failure and ammonia intoxication have not occurred with bleeding episodes in these patients. Four patients, with a total of 27 hemorrhages, are currently being followed until they are large enough for shunting operations. Two patients have had no operations thus far while ligation of varices and thoracic transposition of the spleen have been used as temporizing measures in the other two patients.

Operative Management

Surgical procedures directed at correction or control of the complications of portal hypertension have been necessary in 21 of the 23 patients with portal vein thrombosis. There have been 38 surgical procedures used in the 21 patients.

Splenorenal Shunts

Ages ranged from 3 to 13 years at the time of operation. Three of the 10 shunts are currently patent. The three patients with patent shunts have no evidence of varices on esophagogram (Table 5). These shunts were constructed at ages 7, 9, and 13 years (average 9.7 years). The seven patients in whom the shunt thrombosed ranged in age from 3 to 11 years and averaged 7.7 years. Splenorenal shunt was the initial operative procedure in nine patients and was done after failure of a cavomesenteric shunt in one patient. Recurrent hemorrhage led to subsequent operations in four (three had mesocaval shunts and one an esophagogastrectomy) of the seven patients with occluded shunts. In all three of the patients with occluded shunts who have not been operated upon again, the occluded shunt was found when we recalled them for study and not because they had rebled or were having any trouble. Two have had no

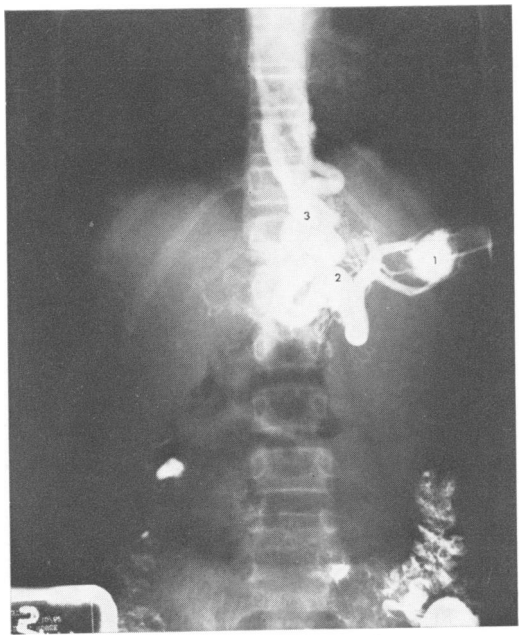

FIG. 7A. Splenoportogram in a 9-year-old girl with portal vein thrombosis. Injection of dye into the spleen (1) is followed by filling of the splenic vein (2) and periesophageal collaterals (3). There is minimal hepatic portal venous flow.

further hemorrhage. One patient has had an isolated hemorrhage 18 years after operation and now survives 22 years after surgical

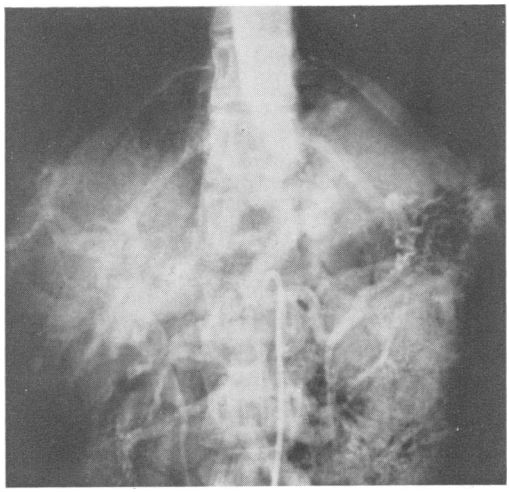

FIG. 7B. Portal venogram in the same patient at 18 years of age. The study was obtained by selective injection of dye into the superior mesenteric artery. There is no evidence of patency of the splenoral shunt done at 9 years of age. There is a striking increase in hepatic portal venous flow and large perigastric and periesophageal collaterals.

treatment and has had no further hemorrhage. All three have varices on esophagram and a striking network of venous collaterals from the portal system (Fig. 7). No shunts have been performed in patients with portal vein thrombosis less than 9 years of age in the past 15 years.

Cavomesenteric Shunts

Six patients ranging in age from 6 years to 20 years have undergone cavomesenteric shunts. In two patients the shunt thrombosed in the early postoperative period. In one, a 9-year-old girl, a splenorenal shunt was done. It is patent and she has had no further trouble. The other patient with an occluded shunt developed ascites which subsequently cleared and he has had one episode of hemorrhage. He is being managed conservatively and will have a splenorenal shunt later. Four patients have radiographically proven (Fig. 8) patent cavomesenteric shunts which have remained functional for 7, 9, 13, and 17 years respectively (Table 5). There has been no further hemorrhages in any of the four patients and no evidence of varices on esophagogram. Three of these four patients underwent shunt, 3 months, 5 and 15 years previously, which had thrombosed.

Make Shift Shunt

In five patients (four were operated upon before 1955) an attempt was made to find a large collateral vessel suitable for anastomosis to the inferior vena cava. Four patients had previously undergone splenectomy, one for a splenorenal shunt which had failed. The area of the portal vein and superior mesenteric vein was dissected in these patients and in only two was a usable vein found. The make-shift shunts failed in both of these patients (Table 5). Two patients in whom no shunt could be done died of hemorrhage in the postoperative period. The third patient has had recurrent hemorrhage but no further operation.

Splenectomy

Splenectomy alone has not been used in the past decade. Prior to this time period splenectomy alone was used in six patients (only one was done since 1952) whose ages ranged from 3–14 years. Splenectomy was done as the initial operation in all six patients. All patients rebled in 6 months to 2 years after splenectomy. Three patients had subsequent operations. Two of the three patients in whom the only operation was splenectomy died from hemorrhage 8 months and 5 years after operation.

Ligation of Varices

Varices were ligated in four patients ranging in age from 8 months to 12 years. Transgastric technic was used in one patient and transesophageal ligation of varices was used in three patients. This was used as the initial operation in two patients ages 8 months and 4 years in an attempt to defer shunting procedure to an older age. Two patients had ligation of varices after previous operations. There were no postoperative deaths in the four patients who underwent ligation of varices, but all rebled from 6 months to 1 year after operation.

Esophagogastrectomy

Esophagogastrectomy was used in four patients ranging from 7 to 15 years of age. These patients had undergone prior operations all of whom subsequently rebled. Anastomotic stricture, resistant to repeated dilatations, in one patient was managed with substernal colon interposition. Recurrent hemorrhage 3 months postoperative, necessitated complete resection of esophagus and proximal stomach. He subsequently has done well with no further hemorrhage. One other patient who underwent esophagogastrectomy had a colonic segment interposed and survives 11 years after operation with no further hemorrhage.

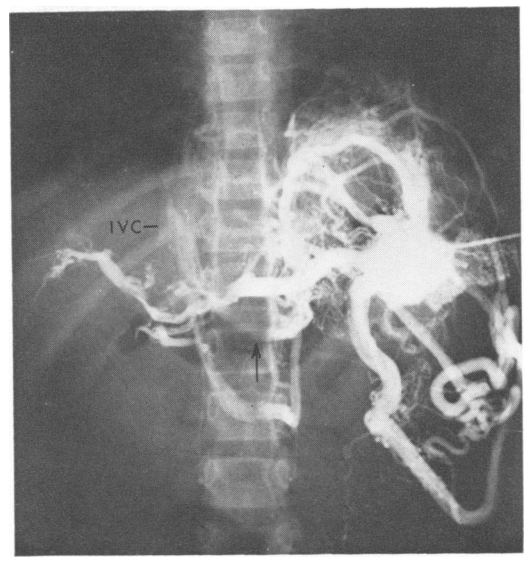

FIG. 8A. Splenoportogram in a 6-year-old boy with portal vein thrombosis demonstrating a striking pattern of venous collaterals. The inferior vena cana (IVC) fills from the left renal vein (arrow) via a naturally occurring splenorenal collateral vessel.

Thoracic Transposition of the Spleen

One patient underwent thoracic transposition of the spleen at age 2 years after re-

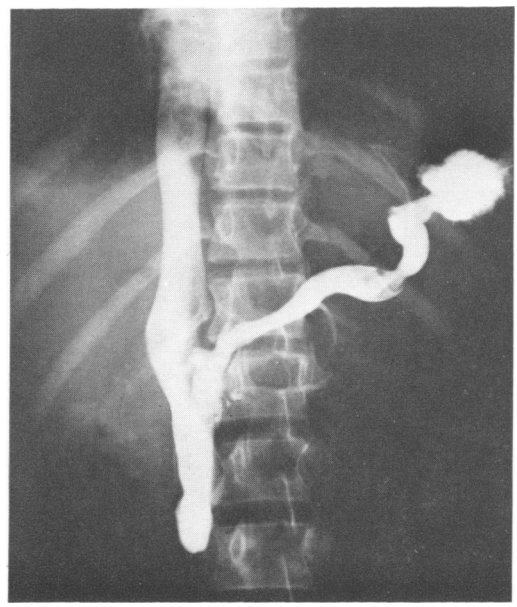

FIG. 8B. Splenoportogram in the same patient at 14 years of age demonstrating a patent cavomesenteric shunt which has remained patent for 8 years. A complete absence of the venous collaterals in A is seen.

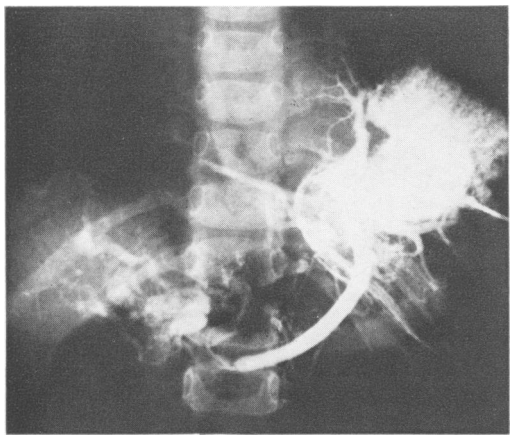

FIG. 9A. Splenoportogram in a 4-year-old boy who had thoracic transposition of the spleen at 2 years of age. Striking collaterals are shown to the intercostal vessels of the thoracic cage.

peated bouts of hemorrhage during 1 year for which he required transfusion of 26 units of whole blood. He had two hemorrhages in the first year after operation but has had no further hemorrhage in the past 8 years. A striking pattern of venous collaterals developed between the spleen and the chest wall, however, with increasing age these have diminished so that at present there are no demonstrable collaterals but instead a network of intra-abdominal collaterals with a greater portal blood flow to the liver which have increased with the age of the patient and include at present varices about the gallbladder (Fig. 9).

Overall Results of Portosystemic Shunts in Childhood Portal Hypertension

Table 6 summarizes the results of all the shunts done for portal hypertension in children. There was one operative death after portacaval shunt in a 7-year-old girl with severe cirrhosis. The best results (100% patency) were obtained with portacaval shunts in children with cirrhosis. The worse results (30% patency) were obtained with splenorenal shunts in children with portal vein thrombosis. The high incidence of shunt thrombosis in this latter group is related to the age and size of these children. Most were under 8 years of age. This is emphasized by the fact that five of six splenorenal shunts in children with cirrhosis remained patent; these children were over 8 years of age and most of them were 10 years of age or older. The one shunt that thrombosed in the cirrhosis group was in a 4-year-old boy. The splenic vein in most children under 8 years of age is less than 1 cm. in diameter.

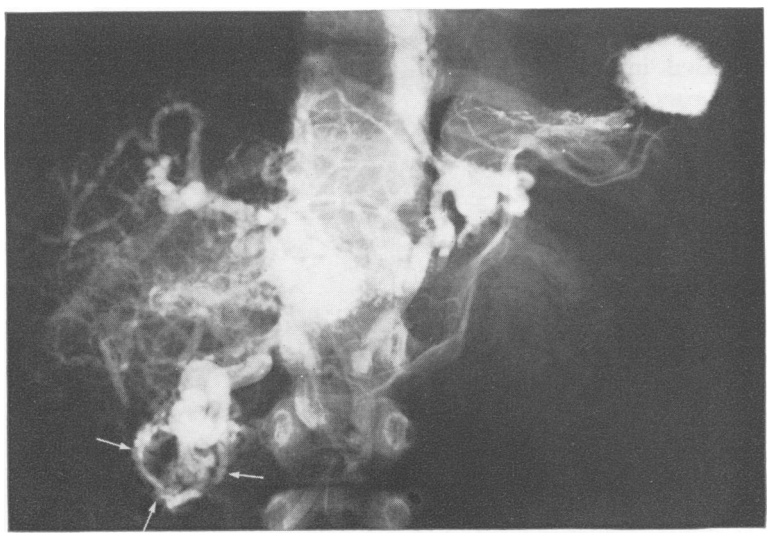

FIG. 9B. Splenoportogram in the same patient at 10 years of age shows loss of intrathoracic collaterals and increase in hepatic portal venous flow and intraabdominal collaterals with varices around the gallbladder (arrow).

TABLE 6. *Portasystemic Shunts for Childhood Portal Hypertension*

Type of Shunt	Thrombosis		Cirrhosis		Combined		Shunt Patency Rate
	Total	Patent	Total	Patent	Total	Patent	
Splenorenal	10	3	6	5	16	8	50%
Cavomesenteric	6	4	—	—	6	4	66%
Portacaval	—	—	4	4	4	4	100%
No. patients shunted	12		10		22		
No. with patent shunt	7		9		16		
% with patent shunt	58%		90%		73%		

However, shunt patency may not be only criteria for a satisfactory result. Three of the children with a thrombosed splenorenal shunt had repeated episodes of hemorrhage preoperatively and were found to have a thrombosed shunt 9, 12 and 16 years later when we recalled them for follow-up study. None had rebled at that time, one has had a single hemorrhage since that time.

Discussion

The nature of portal vein thrombosis and cavernomatous transformation of the portal vein has received various explanations, however, it is currently felt by most authors that it is the end result of thrombosis of the portal vein with the combination of recanalization and enlarging periportal collaterals producing the picture of so-called cavernomatous transformation. Serial splenoportograms in several patients in this study have shown both progressive and regressive thrombotic changes in the portal vein. One 15-month-boy when first seen had mild stenosis in the portal vein just before it bifurcated. Some 41 months later a repeat splenoportogram revealed complete occlusion with massive collaterals in the porto-hepatis (Fig. 6). This is the only patient in this series, or any other that we are aware of, who progressed from mild stenosis to complete occlusion. On the other hand we have seen several patients who when first studied had complete occlusion of the portal vein, virtually no portal collateral flow to the liver and who on later study showed massive portal flow via collaterals (Fig. 7). The patient with the thoracic spleen showed early development of thoracic collaterals which have seemingly disappeared as the collaterals in the porto-hepatis increased (Fig. 9).

Portal vein thrombosis hase been attributed to umbilical infection, intra-abdominal infection, neonatal sepsis and severe dehy dration. However, in the majority of patients there is no history of infection or other disease which might explain the thrombosis. In two patients in this report portal vein thrombosis was attributed to neonatal transhepatic catheterization of the right atrium via the umbilical vein. However, recent reports [12, 14, 15] have indicated that umbilical vein catheterization, done for the purpose of blood sampling, exchange transfusion or physiologic monitoring, is an uncommon cause of portal vein thrombosis.

The etiology of childhood cirrhosis is also often difficult to determine. Many causes have been listed including congenital hepatic fibrosis, the cirrhosis of cystic fibrosis, Wilson's disease, and cirrhosis due to neonatal and juvenile hepatitis.[5, 8, 10, 13] We have excluded infants with biliary atresia and other forms of obstructive cirrhosis from this study. Four of the children had histories of jaundice or prolonged febrile illness in the early years of life and probably had hepatitis. In the other six children there were no histories of antecedent illness suggestive of hepatitis.

Evaluation of the child with suspected portal hypertension is fairly well standardized.[8, 14, 16] It includes determination of liver function studies, complete blood counts, clotting studies, esophagogram, and splenoportogram in addition to complete history and physical examination. However, the definitive study is splenoportography with measurement of splenic pulp pressure. A splenic pulp pressure of 30 cm. saline or greater indicates portal hypertension. A normal splenoportogram should show only the splenic vein and the portal vein; opacification of tributaries of either of these veins indicates obstruction and portal hypertension. Splenoportography also differentiates portal vein thrombosis and obstruction secondary to cirrhosis (Fig. 2). When properly performed splenoportography is a safe and valuable diagnostic study. The use of Ketamine sedation rather than general anesthesia appears to be a step forward in the technic of splenoportography. Operative portography is still advocated by some[14] but is suboptimal in our opinion because (1) it usually consists of a single technic (2) we prefer to study the radiographs preoperatively and (3) because of satisfaction with preoperative splenoportography. Selective superior mesenteric arteriography provides an alternate method of portography and is especially helpful in patients who have had previous splenectomy.

The child with portal vein thrombosis usually presents at an earlier age than the patient with cirrhosis.[14, 18] Ascites is often present early in the course of portal vein thrombosis but is usually transient.[4, 7, 9] Hematemesis is the characteristic mode of presentation of the child with portal vein thrombosis, however, the initial bleeding may present as melena or bright blood per rectum. In this report, as well as others,[1, 8, 9] there is a frequent relation between a febrile episode with an upper respiratory infection and the onset of bleeding. It has been suggested that this is related to the use of aspirin during the febrile episode.[1, 7] Hemorrhage usually occurs before the age of 10 years in portal vein thrombosis.[2, 17] Hypersplenism is a more frequent problem in the patient with cirrhosis though it seldom is a problem until after 4 years of age.[4]

The initial management of children with portal hypertension due to cirrhosis and portal vein thrombosis is usually the same. The usual mode of presentation is upper gastrointestinal hemorrhage from esophageal varices. In both the cirrhotic child and the one with portal vein thrombosis the bleeding can usually be controlled with bed rest, sedation and transfusion of whole blood. If significant bleeding persists balloon tamponade with a Blakemore-Sengstaken tube can be used. In recent years we have been reluctant to use balloon tamponade in children and have come to rely more on the use of intermittent doses of 2–3 units of intravenous pitressin. This has been effective in both the cirrhotic and the child with portal vein thrombosis. If bleeding continues we have resorted to transesophageal ligation of varices with control of hemorrhage which has persisted from 3 months to 2 years in the five children in which it was done.

Once the hemorrhage has been controlled the therapeutic objective is to get the patient in the best possible condition for a portal systemic shunt. Management of the cirrhotic and the child with portal vein thrombosis differs primarily in the timing of the portal systemic shunt. The child with cirrhosis is usually 5 years of age or older. At this age the portal vein is large enough to allow a successful portacaval shunt. Therapy is directed toward getting the child in the best possible nutritional state and then doing the shunt.

In control the child with portal vein thrombosis usually presents with the initial bout of hemorrhage prior to the age of five, the protal vein is thrombosed and not available for a shunt and the splenic and superior mesenteric veins are usually too small for a successful shunt. Furthermore,

control of the hemorrhage is more easily accomplished with conservative measures and the child is well between bleeding episodes. In these children we have adopted a policy of conservative management until they are 8–10 years old before undertaking a shunt. We have four children in this category at the present time. One of them had transesophageal ligation of varices at the age of 9 months, he is now 5 years old and had had three episodes of bleeding in the interim. Another is the child with the thoracic transposition of the spleen who bled twice during the following year but has not bled in the past 8 years. Two others have been managed with completely non-invasive technics, are now 4 and 5 years old and have had a total of 18 bleeding episodes. The major reason for adopting this policy was the high incidence of shunt thrombosis in children under 8 years of age in the 1950's and early 1960's.

Another problem in the post-hemorrhage management of the child with portal vein thrombosis is ascites. For reasons that are poorly understood following the bleeding episode they develop massive ascites. We have seen this repeatedly in the children who we are following until they grow to an adequate size for successful shunting. We have managed this problem with salt restriction, intravenous administration of human serum albumin and diuretics. Our pediatricians look askance on our giving albumin to a child with a serum albumin level of 3.0 Gm./100 ml. or more. However, in each instance the ascites has cleared in 4 to 8 days and has not recurred until the child bleeds again.

The only clear cut indication for operative treatment in both the cirrhotic and the child with portal vein thrombosis is massive variceal hemorrhage. Shunting procedures in the child with cirrhosis, varices, hypersplenism and who has never bled is more controversial. However, if one is going to do a splenectomy for the hypersplenism in such a child we feel splenorenal shunt should be done at the same time.

The operation of choice for the cirrhotic child is a portacaval shunt which has proven effective in preventing further hemorrhage both in this series and others.[8] Splenomegaly and hyperplenism regresses over varying periods of time after the shunt.[14] If the splenic vein is 1.0 cm. in diameter or greater, a splenorenal shunt may be equally effective. Shunt patency in the cirrhotic children in this series was 90%. The ultimate prognosis of the child with cirrhosis is closely related to the hepatic reserve much as in the adult cirrhotic. However, even the existence of severe cirrhosis in a child does not preclude the possibility of prolonged survival with careful management.[11]

The operation of choice in the child with portal vein thrombosis is not so clear cut. Either a mesocaval shunt or a central splenorenal shunt as described by Clatworthy[3] should be equally effective if the splenic or the superior mesenteric vein are large enough to allow construction of an adequate size shunt. The splenic vein should be at least 1 cm. in diameter or greater. One problem with the mesocaval shunt is compression by the third portion of the duodenum. In order to get an adequate length, one may need to use a common iliac vein for anastomosis to the superior mesenteric vein. Tank and others[14] feel the mesocaval shunt is the operation of choice because it allows successful shunting at an earlier age. The mesocaval shunt has been especially valuable in patients who have had a previous splenectomy or a splenorenal shunt that has thrombosed. The fact that seven of the ten splenorenal shunts in this series thrombosed would seem to support the advocates of the mesocaval shunt. However, most of those splenorenal shunts were done in the 1950's and in children under 8 years of age.

Several other types of operative treatment of portal hypertension due to portal

vein thrombosis have been used in this series and others. Splenectomy alone is to be condemned. Three of the six children so treated in this series bled to death. Most of these patients were treated in the 1940's and early 1950's. Esophagogastrectomy, possibly with colon interposition, still has a place when all other procedures have failed. Two of the four patients who had esophagogastrectomy are doing extremely well, both are in their 20's and leading normal lives. One patient with an esophagogastrectomy bled repeatedly 5 years later, had two explorations but a portal decompression was not done and he died shortly after the second exploration. It would appear that this patient should have had a mesocaval shunt. The fourth patient with an esophagogastrectomy had a recurrence of variceal hemorrhage and did then have a successful mesocaval shunt. It should be noted that these patients with espohagogastrectomy were treated in the 1950's before the mesocaval shunt was widely recognized as a good operation.

Thoracic transposition of the spleen, as used in one patient in this series, has not had widespread application in this country. It was used in a 2-year-old child following repeated bleeding episodes which required transfusion of 26 units of whole blood in one 12-month period. The child has developed normally, has not bled for 8 years but he does have esophageal varices. If he should bleed again a portal systemic shunt will be done.

It is important to keep in mind that the child with portal vein thrombosis has a normal liver and if a successful portal systemic shunt can be constructed, the prognosis is excellent. A conservative approach to the management of bleeding episodes until the child reaches an adequate size increases the likelihood of a successful shunt. If one type of shunt thromboses, the other type of shunt can then be done. Of the 23 patients with portal vein thrombosis 19 are living. The four deaths are the result of treatment as it existed in the 1940's and early 1950's. It is our opinion that had these children been treated by current methods of management they would all be alive. Of the 19 children who are living: seven have a patent shunt and are completely well, three with an occluded shunt are well, two with an esophagogastrectomy are completely well, five are awaiting a definitive shunt and finally two patients treated by splenectomy alone are in their 30's and still have varices and occasional melena but are leading essentially normal lives.

Summary and Conclusions

The presentation, evaluation and management of 33 patients with childhood portal hypertension have been reported. The course and management of those with cirrhosis has been contrasted to the spectrum of problems encountered in the child with portal vein thrombosis. On the basis of this experience the following conclusion seem justified, they have changed little since our 1963 report.

1. Splenoportography is the definitive diagnostic study.
2. Conservative measures will control most bleeding episodes.
3. Suture ligation of the varices is occasionally required.
4. Control of the hemorrhage by conservative measures gains valuable time for growth of the child so that a successful shunt can be constructed.
5. Splenectomy alone is not indicated in portal hypertension.
6. A portal-systemic shunt is the only procedure likely to produce a lasting good result.
7. A centrally placed splenorenal shunt one centimeter or more in diameter is satisfactory.
8. A mesocaval shunt is also an excellent procedure and may be the procedure of choice in many cases.
9. With the above two shunts available it should seldom be necessary to perform esophagogastrectomy.

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Discussion

Dr. J. Philip Sandblom (Lausanne, Switzerland) discussing Papers 26, 27, 28: I had the great fortune about 25 years ago to be along when Dr. Whipple and Dr. Blakemore did their first work in portal hypertension. I immediately returned to Europe, and we had a center in Lund for treatment of portal hypertension, which gave us the opportunity of studying this abundant material.

When we started to perform the shunts, the medical people were a little bit dubious. They told us this was a very unphysiologic procedure; but we answered that pretty soon we found out that our patients preferred an unphysiologic life to a physiologic death.

I think a very important part of Dr. Foster's report is that he has given us such detailed information about these unphysiologic lives and what will be the end. Of course, children should offer very valuable material in this respect because of their long life expectancy.

On the other hand—and I would like to hear Dr. Foster's opinion about this—it might not be exactly fair to compare children to adults, because children might have greater possibility to adapt themselves to new circulation of this kind. We know that children sometimes tolerate procedures and adapt themselves in a much better way than adults do.

In any case it is of great value to see the results of this material, which is so well studied, well followed and well managed.

When the shunt functions well these patients seem to live a perfectly normal life; and as I understand, you have not at all found the same amount of postoperative complications, especially of encephalopathy, that is so commonly reported in adults. I was very pleased to hear this, because this is our experience even with adults. We do not have at all the same amount of late encephalopathy that has been reported especially from the United States. I should like to know if you have any explanation for this difference in the results.

You prefer to defer operation until the child has reached a certain age and size so the shunt can be made fairly large. There is less risk in delay of operation in children with extra-hepatic block because they have healthy livers which eliminates the hazard of hemorrhage as may occur in patients with cirrhosis. The healthy livers in thrombotic patients will not be so damaged by anoxia from the intestines as in cirrhotic patients. So it is very fortunate that portal hypertension in children mostly is caused by portal thrombosis.