Results of Surgical Treatment of Portal Hypertension in Children *

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PORTAL HYPERTENSION in the child and adult differs in several important respects. Cirrhosis of the liver is the most common etiology in the adult while thrombosis of the portal vein more frequently causes portal hypertension in the child. Hepatic insufficiency following an esophageal hemorrhage or a major operation accounts for most of the morbidity and mortality in adult portal hypertension. The child with portal vein thrombosis ordinarily has a normal liver; such a child tolerates a massive hemorrhage or an operative procedure much better than the adult with cirrhosis. Furthermore, cirrhosis of the liver seems to have a better prognosis in the child than in the adult. Cirrhosis in the adult is most commonly related to excessive alcohol intake which all too frequently remains a problem in the management of the patient after an esophageal hemorrhage or surgical treatment. Cirrhosis of the liver in children may follow an episode of hepatitis or is of obscure origin. The management of portal hypertension in the adult has been the subject of many reports; relatively little has been recorded about childhood portal hypertension. A recent review of our experience with the surgical management of portal hypertension in 22 children prompts this report.

The records of all children under 15 years of age who have had surgical treatment for portal hypertension were carefully analyzed; none were excluded. Surviving patients were recalled for a careful evaluation of their current status; this included history, physical examination, hemogram, esophagram, liver function studies and contrast angiographic demonstration of their portal-systemic shunt when appropriate. A 100 per cent follow up was accomplished.

Presentation

The 22 children ranged in age from two to 14 years at the time of their first operation. Sixteen had portal vein thrombosis and six had cirrhosis of the liver. In Figure 1 the sex, age and etiology of the portal hypertension is designated for each patient. Most of the children with thrombosis of the portal vein were under ten years of age, and almost half were under six. In contrast the patients with cirrhosis seemed to have been randomly scattered between the ages three to 14. Boys predominated in the thrombosis group: 11 of 16 patients. This was even more striking in the patients under six years of age where seven of eight were boys. Girls outnumbered boys four to two in the cirrhosis grouping.

The etiology of the portal vein thrombosis or cirrhosis of the liver was obscure in most instances. Only four of 16 patients in the thrombosis group had a history of neonatal omphalitis; two other patients in this group had a past history of jaundice. In the cirrhosis group two patients had a

* Presented before the Southern Surgical Association, Boca Raton, Florida, December 4–6, 1962.

Supported in part by USPHS research grant HE-05286-03(C2), National Heart Institute.

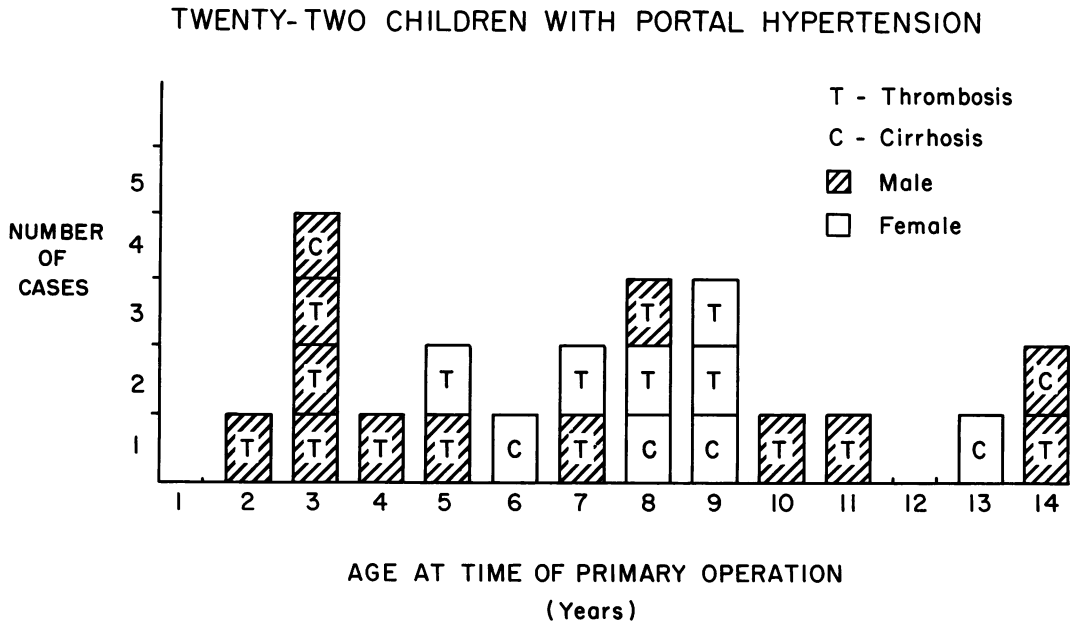

FIG. 1. Sex, age at the time of initial operation and etiology of the portal hypertension in each patient.

past history of jaundice. One patient (M. M.) had been jaundiced at birth and intermittently thereafter; another child (J. S.) had hepatitis six months prior to the surgical admission.

The first manifestation of portal hypertension in the thrombosis group was hemorrhage in 11 cases, splenomegaly in four, and fever of unknown origin in the final patient (Fig. 2). In the cirrhosis group, the initial manifestation was splenomegaly and ascites in three cases, hemorrhage in two, and fever of unknown origin in one. The initial hemorrhage in the patients of the thrombosis group presented as bright red blood per rectum in seven instances.

A striking feature in these children was the frequency of an upper respiratory infection, an infectious disease, or nonspecific temperature elevation in the days immediately preceding a massive upper gastrointestinal hemorrhage. This has been documented in the hospital records of over half of the bleeding episodes sustained by the children in the current study.

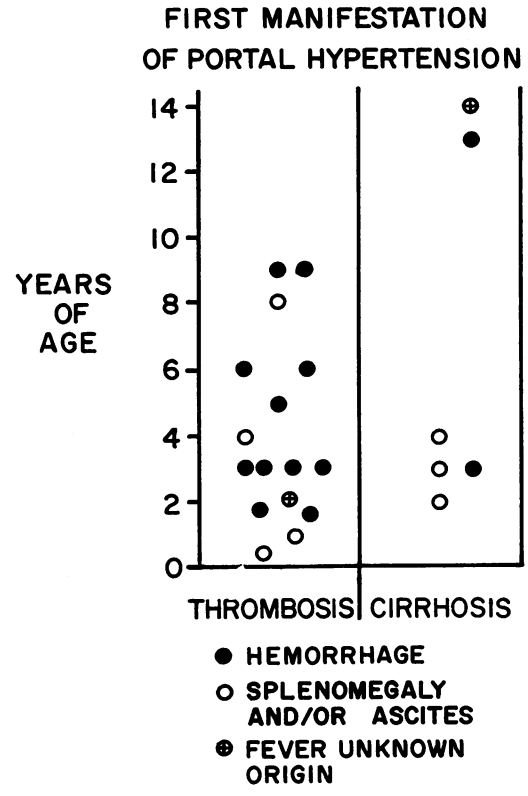

FIG. 2. The first manifestation of portal hypertension and age at onset is depicted for each patient.

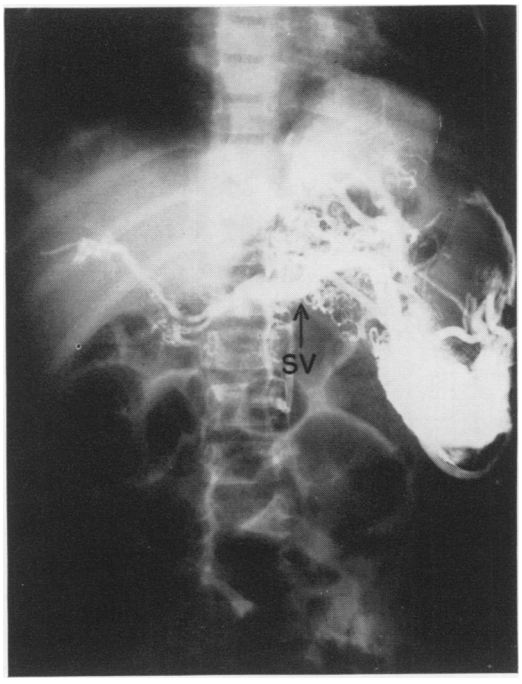

FIG. 3. Splenoportogram in a four-year-old boy (W. G.) showing thrombosis of portal vein, collateral network of perigastric and periesophageal veins and some flow into the liver via collaterals traversing the porta hepatis.

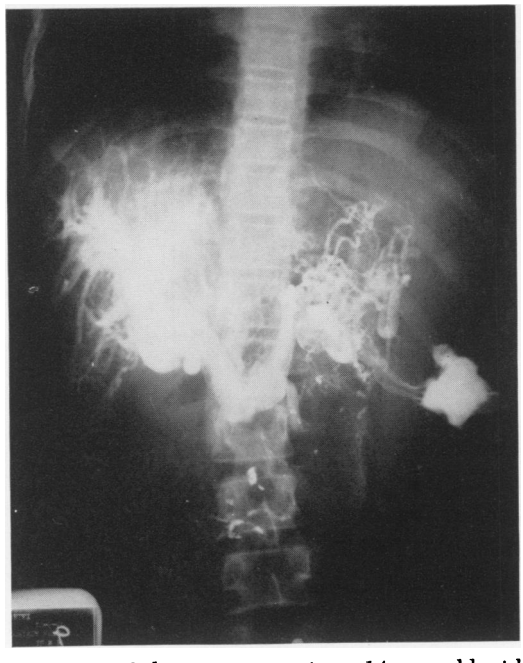

FIG. 4. Splenoportogram in a 14-year-old girl (J. H.) with cirrhosis of the liver. Dilated and tortuous splenic and portal vein, as well as perigastric collaterals are striking.

Diagnostic Studies

In the thrombosis group, 13 of 16 children had an esophagram, and varices were demonstrated in nine instances. Esophagram was done in all six children of the cirrhosis group but revealed varices in only three patients; in two of the three patients with demonstrable varices the primary problem was hypersplenism and ascites.

Liver function studies were performed preoperatively in 12 of the patients with thrombosis and were normal in each instance. Abnormal liver function was present in the six patients with cirrhosis; however, the abnormalities were not severe in any of the children. Hypersplenism was present in four patients from each group; the pancytopenia was most marked in the cirrhosis group.

Splenoportography became a part of the study of patients with portal hypertension in about 1953. Ten of the patients in this study presented after that time. The splenoportogram revealed filling of collaterals in each case and allowed differentiation between the cirrhosis (4 cases) and thrombosis (6 cases) groups (Fig. 3, 4). The splenic pulp pressures ranged from 280 to 530 mm. of saline. Differentiation of thrombosis and cirrhosis prior to the advent of splenoportography was based on dissection of the portal vein at the time of operation, on the results of liver biopsy, or on autopsy findings.

Management

The interval from first hemorrhage to operative treatment was 0–7 years and averaged 2.0 years in the thrombosis group (Fig. 5). Three children had surgical treatment at the initial hemorrhage, six at the second hemorrhage, and the other seven had three to seven episodes of bleeding prior to surgical intervention. The 16 children with thrombosis of the portal vein had 46 episodes of hemorrhage prior to the initial operation. Between bleeding epi-

sodes these children remained amazingly well.

Hemorrhage was the indication for surgical intervention in every patient in the thrombosis group. In the cirrhosis group hemorrhage was the indication for surgery in four cases: two at the initial episode and two after three or four bleeding episodes (Table 1). Hypersplenism and ascites was the indication for operation in the other two patients; both of these patients had varices but had never bled.

Conservative measures consisting of hospitalization, bed rest, sedation, blood transfusion and vitamin K therapy controlled hemorrhage in the vast majority of instances. The 22 children sustained a total of 112 episodes of hemorrhage: 57 prior to the initial operation and 55 in their subsequent course (Fig. 5). Uncontrollable, continued bleeding was encountered on seven occasions: twice prior to the initial operation and subsequently in five cases. Transesophageal ligation of esophagogastric varices was performed in five of these patients and successfully controlled the bleeding in each instance.

TABLE 1. *Indication for Primary Operation*

	Thrombosis Group No. Patients	Cirrhosis Group No. Patients
First hemorrhage	3	2*
Recurrent hemorrhage	13*	2
Hypersplenism + ascites	0	2

* One patient had uncontrollable bleeding, remainder had interval operation.

Operative Treatment. In the 16 patients of the thrombosis group the initial operation was splenectomy in six, spleno-renal shunt in eight, transesophageal ligation of varices in one and laparotomy in one. Eight of these patients eventually had 13 more operative procedures: make-shift shunt in two, cavosuperior mesenteric shunt in three, esophagogastrectomy in four, trans-

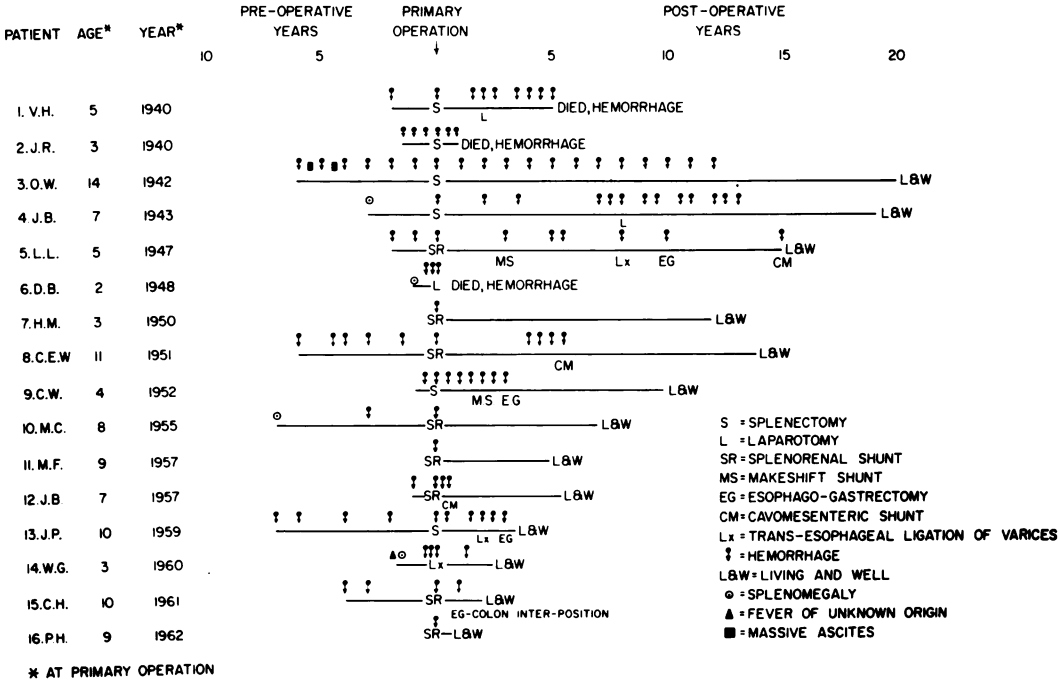

FIG. 5. Graphic presentation of the course of each patient in the thrombosis group.

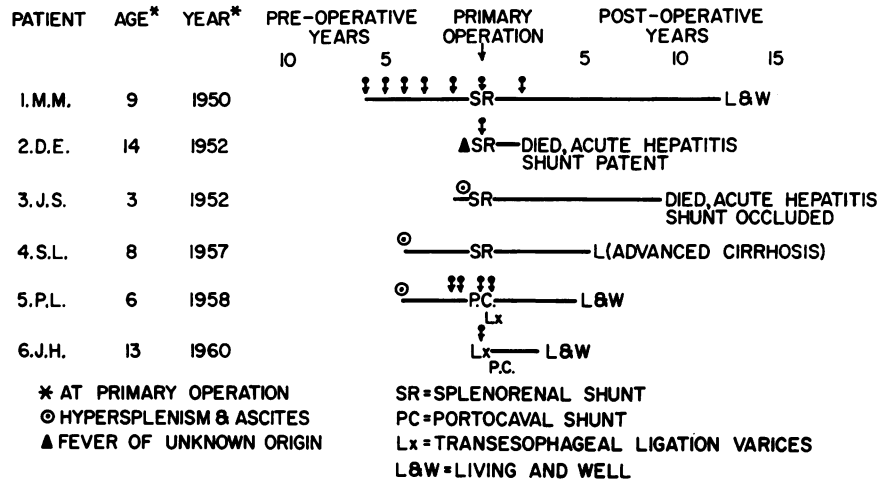

FIG. 6. Graphic presentation of the course of each patient with cirrhosis of the liver.

esophageal ligations of varices in two and laparotomy in two. Make-shift shunt refers to the anastomosis of a nonspecific branch of the portal system to the inferior vena cava. Figures 5 and 6 show the timing of each of the operative procedures in both groups. In the cirrhosis group the initial operation was splenorenal shunt in four, portocaval shunt in one, and transesophageal ligation of varices in one. Two patients had an additional operation.

Portal pressure at the time of operation was measured in 20 patients and ranged from 220 to 500 mm. saline; in 18 instances the pressure exceeded 300 mm. of saline. Pressures were measured before and after a portal-systemic venous shunt in nine patients; these are recorded in Figure 7. Two of these shunts subsequently thrombosed (C. H. and C. E. W.); both were splenorenal anastomoses and were attended by falls in pressure of 223 and 100 mm. of saline, respectively, following establishment of the shunt.

The diagnosis of cirrhosis of the liver was confirmed by wedge biopsy in each instance. In one patient (J. H.) this was an unsuspected finding, was not recognized at the time of operation, and became apparent on microscopic study of the liver. On histologic study three cases were classified as nonspecific portal cirrhosis of mild to moderate degree, two were considered postnecrotic cirrhosis, and the last case was designated "nutritional cirrhosis." One of the patients with postnecrotic cirrhosis had a history of jaundice; the other had never been jaundiced. The second patient in the series with a history of jaundice proved to have nonspecific portal cirrhosis.

Two patients with thrombosis of the portal vein also had mild scarring of the liver compatible with *early cirrhosis*. In one of these (J. B.) the hepatic scarring was noted at the time of splenectomy in 1943; this patient is living and well 19 years later and has no evidence of abnormal liver function. In the other patient (V. H.) the cirrhotic process was noted at autopsy five years after splenectomy; the liver had been normal at the time of removal of the spleen. It should be emphasized that both patients had proven thrombosis of the portal vein; one on the basis of extensive dissection of the porta hepatis and the other at autopsy.

Results as Related to Operative Procedure

Splenectomy. Splenectomy alone was performed in six patients with portal hypertension secondary to portal vein throm-

bosis. Every patient treated by splenectomy bled again and bled repeatedly. A total of 43 episodes of hemorrhage occurred in these six patients; two patients bled to death one and five years following removal of the spleen. Four patients had an additional operative procedure; in two instances an attempt to find a suitable vein for portal systemic shunting failed and paragastric veins were ligated. In one a make-shift shunt was constructed but failed to prevent recurrent hemorrhage, and finally esophagogastrectomy has resulted in freedom from recurrent hemorrhage for 12 years (C. W.). In another patient (J. P.) the secondary operation was transesophageal ligation of varices. This patient bled again repeatedly and esophagogastrectomy was then performed. Thus, two patients died; two ultimately came to esophagogastrectomy; and two have survived for a long time with splenectomy as essentially the only treatment. These latter two patients

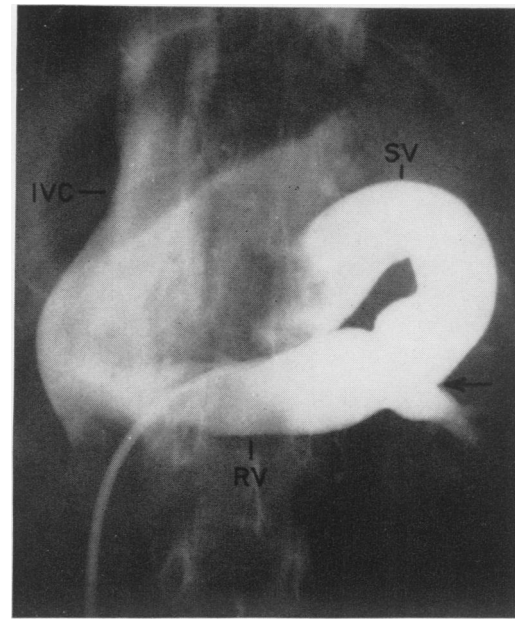

FIG. 8. Demonstration of a patent splenorenal shunt after five years (S. L.). Catheter inserted via right saphenous vein. Subsequent films showed prompt flow of medium back through anastomosis and into inferior vena cava. Anastomosis was 11 mm. in diameter at time of construction.

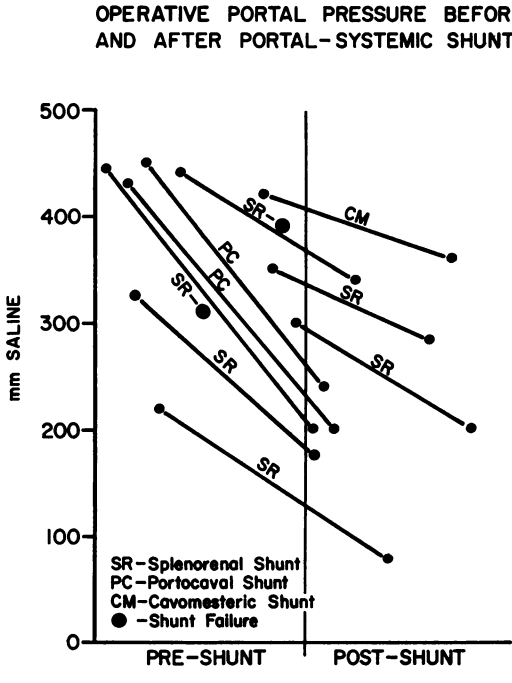

FIG. 7. Pre- and post-shunt operative portal pressures in seven patients from the thrombosis group and two patients with cirrhosis.

are probably the most interesting in the group. They were both boys (O. W. and J. B.); both bled repeatedly for 12 to 13 years, usually every year and often requiring transfusions, until they were over 20 years of age and since then have not bled during the past six to eight years. Both are now living and well, married and have children.

Splenorenal Shunt. Twelve children, ranging in age from three to 14 years of age, had a splenorenal anastomosis, eight from the thrombosis group and four from the cirrhosis group. Seven of the shunts seemed to have remained patent; this has been proven by contrast angiography in three patients (S. L., M. M., and M. F.—Fig. 8) and was confirmed at autopsy in one patient with cirrhosis who died of acute hepatitis (D. E.). The other three patients with apparently patent shunts, all from the thrombosis group, have remained free of hemorrhage for one, 12, and 15

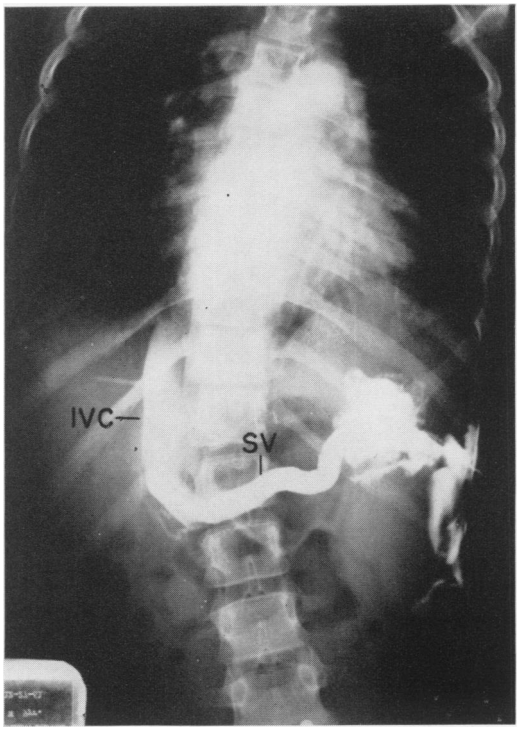

FIG. 9. Splenoportogram in Patient P. L. four years after portocaval shunt.

years (P. H., M. C., and H. M.); none of the three have varices detectable on esophagram. Five of the splenorenal shunts apparently thrombosed. In two instances this could have been predicted at the time of the anastomosis; in the first patient (L. L.), the shunt was constructed with the aid of a vitallium cuff, and in the second patient (J. B.) the splenic vein was 4.0 mm. in diameter. The other three spleno-renal failures occurred in children three, ten, and 11 years old. The three-year-old (J. S.) had cirrhosis; he was well for nine years and then died of an acute fulminate hepatitis. At autopsy the shunt was occluded; it was not possible to determine when the shunt had closed. The ten-year-old (C. H.) had two recurrent massive hemorrhages within less than a year following splenorenal shunt. He then had an esophagogastrectomy with colon interposition and has not bled during the past year. The 11-year-old (C. E. W.) remained free of hemorrhage for four years then bled four times during the next two years, had a cavomesenteric shunt and has remained well for over eight years.

Portocaval Shunt. Two children with cirrhosis had a portocaval anastomosis; both had bled massively from esophageal varices. The first (P. L.) bled again nine days after the shunt was created; a splenoportogram revealed the shunt was patent but conservative measures including balloon tamponade failed to control the bleeding; transesophageal ligation of the varices was effective. The patient has been completely well for over four years and the shunt is patent (Fig. 9). The second patient (J. H.) had a portocaval anastomosis shortly after the ligation of varices; the shunt is patent and the patient well for over two years. In both patients the cirrhosis seems mild and has caused no significant problem; varices have disappeared in both patients.

Cavomesenteric Shunt. In these children (J. B., L. L., and C. E. W.) splenorenal shunt failed and portal flow was diverted via the superior mesenteric vein into the inferior vena cava. In one (J. B.) the inferior vena cava was divided and anastomosed to the side of the superior mesenteric vein; this shunt is patent over four years later and the patient has remained well (Fig. 10). In the second patient (C. E. W.) a large branch of the superior mesenteric vein was divided and the hepatic end of the vein anastomosed to the side of the vena cava; this shunt is patent and the patient has remained well for eight years. In the third patient esophagogastrectomy also failed to control the recurrent hemorrhage and just recently the end of the vena cava was anastomosed to the side of the superior mesenteric vein.

Transesophageal Ligation of Varices. Five patients had suture ligation of esophagogastric varices. The two from the cir-

rhosis group have not bled since the ligation; however, they both have patent portocaval shunts. In one patient (J. H.) who had the ligation as the primary operation, the procedure effectively controlled an unrelenting hemorrhage; however, four months later at the time of portocaval shunting, varices were present on esophagram. The three patients in the thrombosis group who had varix ligation bled again at six, 14, and 24 months following the procedure. Two of these subsequently had an esophagogastrectomy. The third was a three-year-old boy (W. G.) who bled three times prior to varix ligation, bled once 14 months later and has not bled for the past 12 months. The objective in this child has been to gain time until he attains sufficient size so that a shunt may remain patent. He has varices on esophagram at present.

Make-Shift Shunt. Frequently there is a vein in the porta hepatis 4.8 to 8.0 mm. in diameter; on two occasions such a vein has been used for an end-to-side anastomosis into the inferior vena cava. This has usually been a somewhat desperate effort to decompress the portal system when splenic or portal vein was not available. One patient remained free of hemorrhage for two years and one for less than six months; both subsequently had esophagogastrectomy. Make-shift shunt was tried on two other occasions, but the procedure was abandoned because a satisfactory anastomosis could not be accomplished.

Esophagogastrectomy. In four children of the thrombosis group, repeated attempts to decompress the portal system failed. Recurrent massive hemorrhage led to the decision to perform esophagogastrectomy, and in one of these colon was interposed in the reconstructive procedure. The patients have done quite well for periods of six months, one, five and 12 years. Just recently a patient (L. L.) who had remained free of hemorrhage for five years bled again; both gastric and esophageal varices were present; a cavomesenteric shunt was per-

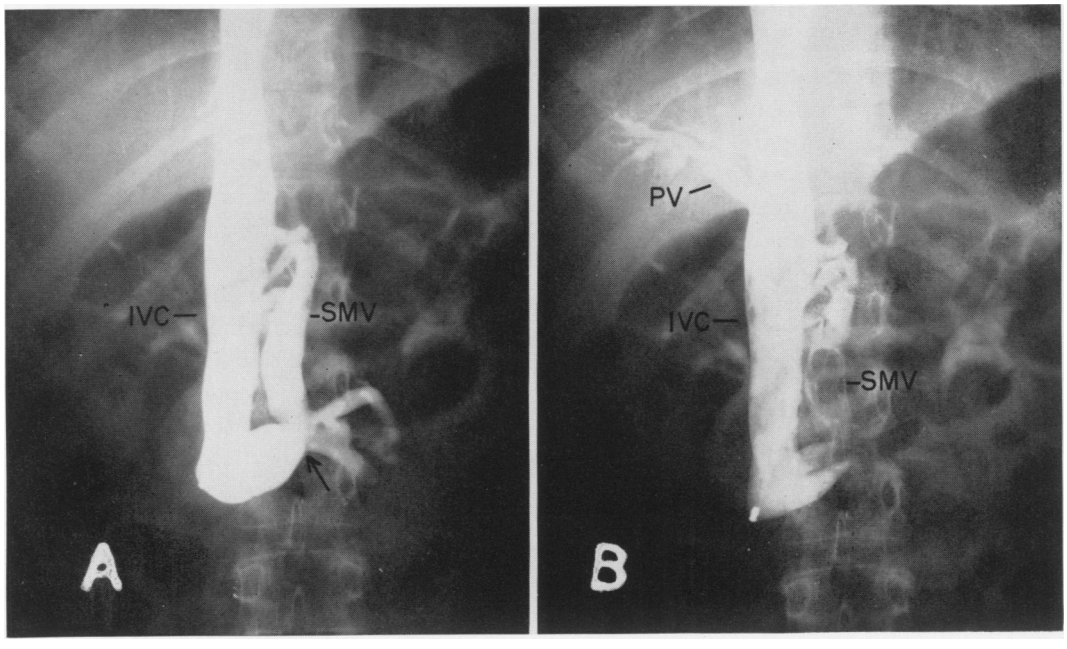

FIG. 10. Demonstration of cavomesenteric shunt after four years (J. B.). Catheter inserted via left antecubital vein. A. Shows patent anastomosis, filling of superior mesenteric vein and beginning opacification of collaterals which circumvent the thrombosis of the portal vein. B. Later film showing intrahepatic portion of portal vein opacified via collaterals.

TABLE 2. *Summary of Experience with 22 Children with Portal Hypertension*

Etiology	No.	No. Operations	Recurrent Hemorrhage Pts.	Recurrent Hemorrhage No.	Deaths Operative	Deaths Late
Portal thrombosis	16	29	11	54	1	2
Cirrhosis	6	9	2	2	0	2

formed. None of the patients have had evidence of esophagitis and nutritional status has been satisfactory. The 12-year follow up was in a boy seven years old at the time of the esophagogastrectomy; he has developed normally and remained well.

To summarize, the 16 patients in the thrombosis group had 29 operative procedures; there was one operative death in a patient in whom nothing was done to stop the bleeding or to decompress the portal system (Table 2). There were two late deaths from bleeding in patients who were treated by splenectomy alone. The other 13 patients can be classified as living and well, although one has varices (W. G.).

In the cirrhosis group the six patients had eight operations (Table 2). There were no operative deaths. There were two late deaths; these were both of the boys of the cirrhosis group and both died of acute hepatitis. The four girls are living 2.5, 4.5, 5.5, and 12 years later. One of these (S. L.) now has advanced cirrhosis, afibrinogenemia and the nephrotic syndrome. The other three girls are well; the patient followed for 12 years is married and has one child. Current liver function studies in these three patients are within normal limits.

Portal systemic venous shunts were performed in 19 patients; two of these were make-shift shunts and two were splenorenal shunts with little or no chance of success. Of the 15 shunts that had a reasonable chance to remain open, 12 have apparently remained patent. This has been proven in eight patients and adjudged so on the basis of the patient's course in the other four. The follow up period in these 12 children with patent shunts ranges from one to 12 years, mean five and one-half years.

Discussion

Portal hypertension in childhood may be first manifest in a variety of ways: hematemesis, melena, bright red blood per rectum, ascites, splenomegaly, hypersplenism, or fever. Clatworthy and Boles have emphasized the frequency of ascites as an early manifestation of portal hypertension secondary to portal thrombosis; in their experience, this most commonly occurred in the first year or two of life.[4] In the present series this did not occur or was not detected. The 16 children with thrombosis first presented as follows: hemorrhage—12, splenomegaly—3; and fever of unknown origin—1.

The etiology of portal vein thrombosis was obscure in most of the patients. Neonatal omphalitis is frequently cited as the most common cause of portal thrombosis.[4, 8] It was found in only four of the patients. However, reliable documentation of the neonatal period is often difficult to obtain. Children with portal thrombosis do not usually have liver disease. In the series of 16 patients reported by Shaldon and Sherlock not one of the patients had ever displayed evidence of hepatic disease.[8] In the present series two of the children with portal thrombosis had a past history of jaundice and two others were eventually proven to have mild *early cirrhosis.*

The frequency of passage of bright red blood per rectum as the first indication of

bleeding in the younger patients with portal vein thrombosis is noteworthy. Bright red blood per rectum can be a misleading sign unless one is aware of this frequent occurrence in young children with portal hypertension.

The most helpful studies in establishing the diagnosis were the liver function tests, the esophagram, and the splenoportogram, the latter being the definitive diagnostic study. Shaldon and Sherlock have stated that the esophagram may frequently fail to demonstrate varices in young children who cannot co-operate well in the examination.[8] We question this on the basis of recent experience with the use of a small catheter to instill the contrast medium into the distal esophagus for demonstration of varices. Splenoportogram has allowed measurement of portal pressure, demonstration of collaterals and differentiation of the patients with thrombosis and cirrhosis. Other reports have differentiated splenic vein and portal vein thrombosis; we have been unable to make such a sharp distinction.[2] In three patients we have encountered a block in the splenic vein on splenoportography but not with demonstration of collateral filling of a normal caliber portal vein beyond this blockage. Furthermore, as has been previously noted, the splenoportogram is not a completely reliable index to the actual site of obstruction; the portal system may be quite patent beyond the point where the contrast opacification stops, this being dependent on the direction of flow and the pressures involved.[8]

In the management of children with portal hypertension the most striking feature was the number of episodes of bleeding, the frequency with which the hemorrhage was controlled by conservative measures and how well repeated operative procedures were tolerated. In the 22 children there were 112 episodes of hemorrhage. Five children required varix ligation to control the bleeding. Three children bled to death. The first two were patients treated by splenectomy; one died in the hospital (1945), and the other died at home (1941). The third death, in 1948, occurred in a two-year-old child with celiac disease who had bled on three occasions over a six-month period. Liver function studies were normal and varices were demonstrated on esophagram. Exploratory laparotomy was performed to establish a definitive diagnosis; splenoportography had not yet been developed. The portal pressure was found to be 500 mm. of saline, there were many dilated and tense veins in the porta hepatis, and the liver was normal grossly and microscopically. Postoperatively the child bled massively and died 48 hours later. Today such a laparotomy would be unlikely, and if bleeding of this magnitude persisted, transesophageal ligation of the varices would be performed, as would also be done in the splenectomy patient who bled to death in the hospital.

These children can bleed to death. However, it is unlikely with good medical management consisting of sedation, bed rest, blood transfusions, vitamin K therapy if needed, and perhaps balloon tamponade. We do not agree with Arcari and Lynn that emergency operation is unnecessary and unwise in any of these patients.[2] If bleeding continues despite conservative measures, then transesophageal ligation of the varices should be performed. Able has demonstrated the value and safety of this procedure in children with portal hypertension.[1] *Suture ligation of varices is admittedly a temporary expedient, but gaining time is an essential in the management of these children.* In the older and larger child, it allows performance of portal decompression after a short interval on an elective basis. In the young child with bleeding esophageal varices the objective is to stop the bleeding in the most conservative fashion and as often as necessary until the child reaches a size which will allow successful portal decompression.

In the intervals between hemorrhage, the children in the portal thrombosis group were extremely well both mentally and physically. Hypersplenism was manifest in several of these children but did not cause significant difficulty in a single child. This is quite different from the cirrhosis group where hypersplenism was a major indication for operation in two of the six children. Even the child with cirrhosis seems to fare better than his adult counterpart. Three of of the girls have remained well for prolonged periods of time (three to 12 years), and a fourth did quite well for nine years and has just recently developed evidence of advanced cirrhosis. The two boys in the cirrhosis group eventually died of acute hepatitis; however, one of these was well for the preceding nine years.

Choice of Operative Procedure. There seems to be rather widespread agreement that splenectomy has no place in the surgical treatment of portal hypertension. Splenectomy may have a place in the treatment of severe hypersplenism, but even then if portal hypertension is present and the splenic vein is of sufficient size, a splenorenal shunt should be done.

Decompression of the portal system is the only treatment likely to provide permanent benefit; this is especially so in the patient with portal vein thrombosis where a successful shunt is curative. In the present series 12 of 19 shunts remained patent and four of the failures were predictable at the time of construction. In general, the larger the portal-systemic anastomosis, the more likely is prolonged patency. Therefore, when possible, portocaval shunt is preferable to splenorenal shunt. A cavosuperior mesenteric shunt may be preferable to splenorenal anastomosis in many instances; a recent report by Vorhees and Blakemore confirms the value of cavomesenteric anastomosis.[3, 6, 10] Certainly the central type of splenorenal shunt, as described by Clatworthy, is superior to the anastomosis of the more peripheral portion of the splenic vein.[4]

Transesophageal suture ligation of varices as an emergency measure seems to have a definite place in the surgical armamentarium for the treatment of portal hypertension. Esophagogastrectomy should be reserved until all possibilities of a successful portal decompression have been exhausted. In some cases esophagogastrectomy will undoubtedly be required and the results obtained with this procedure in the present series have been generally good. However, viewing the problem at this time one wonders whether cavosuperior mesenteric shunt could have been performed in these patients and a more physiologic result obtained. The three patients in this series treated ultimately by cavomesenteric shunt have done extremely well. In one of these esophagogastrectomy was followed by recurrence of varices and hemorrhage five years later, and cavomesenteric shunt was then accomplished satisfactorily. When esophagogastrectomy is performed a pyloroplasty should be performed and interposition of colon also appears worthwhile.

Several groups have attempted to define the age when a portal-systemic shunt is likely to remain patent. Clathworthy and Boles set age at four years but qualified this by adding, "if the splenic vein is of good size."[4] Arcari and Lynn designated ten years or weight exceeding 60 to 70 pounds.[2] Both of the latter groups focused their attention on the splenorenal shunt. It is interesting that Shaldon and Sherlock believe that unless a portocaval shunt can be done, and this seemed possible in five of their 16 patients with portal thrombosis, they prefer to manage the patient medically.[8] They qualified this by waiting until the child is seven or eight years old. They have been dissatisfied with the results of splenorenal (two cases) and cavomesenteric (two cases) shunts in children. Certainly others have expressed similar dis-

satisfactions.[5] The results of portal-systemic shunting in the present series have been reasonably good. It is difficult to set an age when a shunt is likely to be successful. We currently strive to tide the patient over until he is six to ten years of age, but the more important requirement is that the splenic or portal vein be one centimeter or larger in diameter on splenoportogram. The patients in Shaldon and Sherlock's series with portal thrombosis and yet with a segment of portal vein suitable for portocaval anastomosis would seem to indicate that closer investigation is warranted in an effort to detect similar patients. Even though splenoportography does not reveal a patent segment of portal vein, it might seem worthwhile to perform operative portography in an attempt to definitely delineate the site of blockage.

Conclusions

On the basis of a careful analysis of experience with the surgical management of portal hypertension in 22 children, the following conclusions seem justified.

1. Splenoportography is the most helpful and definitive diagnostic study.

2. Conservative measures will control the vast majority of bleeding episodes.

3. Occasionally, suture ligation of esophagogastric varices is required to control hemorrhage.

4. Control of hemorrhage by conservative measures or varix ligation gains valuable time for growth in the young child and allows interval operation in the older child.

5. Splenectomy alone is not indicated in portal hypertension.

6. A portal-systemic shunt is the only procedure likely to give lasting benefit; this is especially true in portal vein thrombosis where a successful shunt is curative.

7. A centrally placed, splenorenal shunt one centimeter or more in diameter seems to be satisfactory.

8. A portocaval shunt is the best method of portal decompression but is usually not possible in portal vein thrombosis; however, it may be possible more frequently than has been realized.

9. Cavo-superior mesenteric shunts can provide excellent portal decompression; this possibility should be investigated before resorting to esophagogastrectomy or if the splenic vein is too small.

10. Make-shift shunts are unsatisfactory.

11. When a shunt is impossible, esophagogastrectomy plus pyloroplasty has checked bleeding for years and nutritional problems have been minimal. The interposition of colon or a reversed gastric segment would seem a worthwhile addition to the procedure.

Summary

Experience with the surgical management of portal hypertension in 22 children has been carefully analyzed. A 100 per cent follow up evaluation was accomplished.

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Discussion

Dr. James D. Hardy (Jackson): With respect to Dr. Patton's paper, I would like to present a clinical syndrome which was new in our experience and which we have not found reported in the literature. We have looked upon the condition as one of acute hepatic sinusoidal insufficiency.

The patient was a 14-year-old girl admitted to the medical service with acute hepatitis and a serum bilirubin level of 40.3 mg.%. The liver was palpable several centimeters below the right costal margin. She was placed on prednisolone and other therapy, and gradually the liver size diminished (slide) until the organ was no longer palpable. Her condition appeared to improve gradually until one night two months following admission when she developed increasingly severe abdominal pain and swelling, followed by profound shock.

Exploratory laparotomy revealed massive ascites and tremendous boggy edema of all organs drained by the portal system. The spleen was much enlarged but the liver was small and grossly nodular, having declined in mass from the markedly enlarged original size to a very small size. Portal vein thrombosis was suspected, but no thrombus was palpable in this vessel. Emergency portacaval shunt was considered, but inasmuch as the blood pressure had been unobtainable since before the operation had been initiated, it was elected to replace third space fluid loss, and to perform portacaval shunt at a later date, under less hopeless circumstances.

Unfortunately, the patient died and autopsy disclosed no thrombus in the portal or hepatic veins.

It was concluded that the acute and fatal portal hypertension had developed when rapid shrinkage of the liver had critically decreased the capacity of the hepatic vascular bed, in a patient who had not yet developed effective collateral circulation. In other words, there must be a critical point at which there is not enough flow in the sinusoids between the cords of liver cells (slide) to accommodate portal venous drainage.

With respect to Dr. Kirtley's paper, this youngster (slide) represented an instance where it was easier, in fact possible, only to perform a side-to-side portacaval shunt rather than an end-to-side portacaval shunt.

From exchange transfusion maneuvers at birth, he had developed umbilical vein infection and portal vein thrombosis, followed at age 18 months by upper gastro-intestinal hemorrhage, due to esophageal varices.

Sent to us at age four, the patient was operated upon with a view to performing an end-to-side portacaval shunt. However, the portal vein proximal to the visualized thrombus at this point was so delicate, and was bound so densely in scar tissue, that side-to-side shunt proved to be technically expedient (slide).

Actually, of course, this side-to-side shunt was, physiologically, an end-to-side shunt, because of thrombosis of the portal vein in the hilum of the liver.

Dr. J. M. Davis (Jacksonville): We rise to briefly relate our experience with three youngsters with extra hepatic portal vein obstruction who were treated by transthoracic ligation of the varices as described by Crile and as was mentioned by both of the authors this afternoon.

Two of the children, 21 months and 3½ years of age, had undergone splenectomy because of massive gastro-intestinal bleeding. Transgastric ligation of the esophageal varices had been done in addition to splenectomy in the second patient. Both developed recurrent massive hemorrhage three months postop. As an emergency measure each was treated by transthoracic esophagotomy with ligation of the varices. It has now been 6½ and 3½ years since this was done and neither has shown evidence of any further bleeding.

The third patient, a ten-year-old boy, underwent a splenorenal shunt three years ago. Within the past year this child has developed recurrent bouts of hemorrhage and because of this his varices have recently been ligated transthoracically. It has only been a few months since his surgery and it is, of course, too soon to know whether he is going to get a good long term result or not.

We believed these children were of interest in that the bleeding was not controlled by splenectomy alone, which is well recognized, or by splenectomy combined with a shunt, or with transgastric ligation of varices. However, thus far all have shown a most satisfactory response to the transthoracic ligation of the varices. On the basis of this limited experience it would appear that the procedure has definite merit not only in the emergency treatment of this very difficult problem, but also as a definitive measure.

Dr. Denton A. Cooley (Houston): Recently we have demonstrated the feasibility of side-to-side splenorenal anastomosis for relief of portal hypertension. This technic offers several practical as well as physiological advantages over the stand-