Management of Extrahepatic Portal Hypertension in Children

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Among 69 patients with PVT, 338 variceal bleeding episodes occurred. Only two patients died from bleeding, and both lived in remote communities and were inaccessible to medical care. Fifty-three children underwent 164 operations for the management of PVT. Once operative management was undertaken, subsequent operations frequently were necessary. Nonoperative measures controlled acute variceal hemorrhage in most instances during the past 10 years. Almost all patients who underwent splenectomy alone, variceal ligation, gastric division, splenic transposition, or makeshift shunts subsequently rebled. These operations are rarely indicated in the current management of children with PVT. Portal venography is essential to define the portal venous circulation before a shunt operation is attempted. Cavomesenteric or central splenorenal shunts prevented further bleeding in eight of 15 patients and are the most reliable operations to control bleeding in patients with PVT. Emergency operation is rarely necessary to control bleeding. Sixteen patients (average age 14.6 years) with PVT did not undergo any operations, and are alive. Each of the six patients with PVT who died from complications of portal hypertension did so within nine months of an operation. Four of these patients had previous splenectomy and died with sepsis as one of the major factors. Bleeding episodes became less frequent as the patients increased in age. Patients who underwent shunts under unfavorable circumstances or who received various other operations to treat portal hypertension appeared to have a higher risk of morbidity and mortality than those managed nonoperatively.

EXTRAHEPATIC portal venous thrombosis (PVT) has been recognized as the most frequent cause of portal hypertension in children, yet scant information has been recorded to indicate the long-term prognosis for patients with this condition. Since Clatworthy and associates[2-4] summarized the manifestations of PVT and recommended a logical course of management, most reports on this subject have emphasized clinical experience with various operations to decompress the portal venous circulation or to control variceal hemorrhage, with follow-up periods usually being short or unspecified.[6-9] The frequency of postoperative rebleeding and the increasing morbidity and mortality encountered with subsequent operations suggest that portal venous decompression may not be as straightforward an approach as initially believed. On the other hand, surprisingly little data have been published to indicate the clinical course of patients with PVT who have been treated by carefully planned nonoperative therapy. The optimal course of management for children with this condition, with a particular view toward long-term evaluation into adulthood, has therefore not been clearly defined.

The combined experience of the UCLA Hospital and the Royal Children's Hospital of Melbourne, Australia, in the management of 69 children with PVT is summarized in this report. On the basis of this clinical experience, including several patients who were available for long-term followup observations, certain recommendations are made regarding the management of extrahepatic portal hypertension in children.

Clinical Material

During the 25-year period from 1948 to 1973, 15 children from the UCLA Hospital and 54 patients from the Royal Children's Hospital were treated for PVT. Thirty-eight of the children were male (55%) and 31 were female. Portal hypertension was first diagnosed under six years of age in 55 patients (80%) (Table 1). Fifteen

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TABLE 1. *Age at Which Extrahepatic Portal Hypertension First Diagnosed*

Age (Yrs.)	No. of Patients	
0–1	14	
1–3	23	80%
3–6	18	
6–10	6	
10–14	8	
Total	69	

children (22%) had a definite history of neonatal infection, most commonly peri-umbilical. No apparent association could be established between cannulation of the umbilical vein during the neonatal period and portal vein thrombosis. An episode of dehydration during early infancy occurred in several cases.

The most frequent initial clinical manifestation of portal hypertension was variceal bleeding, which occurred in 39 children (57%). Twenty-three of the remaining 30 patients in the study experienced variceal hemorrhage later in the course of their disease, most on more than one occasion. Seven children with PVT have never bled. Twenty-four patients had less than three bleeding episodes by the age of 16 years. None of the children died during the first episode of variceal hemorrhage. The average total number of bleeds per patient was 4.9; one patient experienced 24 separate episodes of hemorrhage. Transfusions were given during approximately one-third of the bleeding episodes, although it was rarely necessary to give more than one unit.

Symptomless splenomegaly was the second most common initial manifestation of PVT and occurred in 26 patients. Percutaneous splenoportography was performed on each of these patients to confirm the diagnosis and to determine the portal pressure. The liver was not enlarged, and the spleen was at least 2 cm below the costal margin in each of these patients. More than one-half of the patients had radiographically demonstrated esophageal varices; in 19 of these, variceal bleeding subsequently developed.

Ascites occurred in two young children with PVT; however, in both it was of mild degree, did not require specific therapy, and disappeared before the age of four years. Ascites was not used as an indication for operation in any of the patients. Mild ascites occasionally was observed as a postoperative complication after portal decompression. Anemia was the first manifestation of PVT in two infants.

Growth and development were normal in almost all the children with PVT, and most participated in physical activities to an equal extent as their peers between bleeding episodes. Splenomegaly was invariably present, although the size of the spleen varied considerably and, in general, enlarged as the patient increased in age with no relationship to the occurrence of variceal hemorrhage. Thirty-three children had evidence of hypersplenism as manifested by a leukocyte count of less than 2,000/mm^3 and a platelet count of less than 100,000/mm^3. In two patients from our early experience, the manifestations of hypersplenism were considered sufficiently severe to warrant a splenectomy in the absence of previous variceal bleeding. Both patients subsequently bled on many occasions.

Hepatomegaly did not occur in any of the patients in this study, and liver function tests were almost always within normal range. Liver biopsies were performed on the majority of the children, and the liver was histologically normal in each.

An esophagram was performed on almost all patients, and in approximately two-thirds, varices were demonstrated. This study was less helpful in children under four years of age. Diagnostic esophagoscopy was not performed on any of the patients. Percutaneous splenic portovenography under light general anesthesia was performed on 45 children. Operative mesenteric venography was performed on 16 children and provided good visualization of the portal venous system. This study was of particular value in delineating the location of veins suitable for shunting in patients who had previously undergone splenectomy. Indirect portography by means of selective celiac arterial contrast injection provided satisfactory visualization of the splenic and portal veins in several patients but, in general, was less satisfactory in defining the anatomy of the mesenteric venous system than was direct portography. Portal venous pressures were measured in all patients and values ranged 180–510 mm Hg (Table 2).

Treatment

Fifty-three children underwent 164 operations for the treatment of PVT; the average was 3.1. A variety of methods was used (Table 3). The average interval between the first hemorrhage and initial operative management was 1.7 years. Although 14 patients had only one operation, 12 had four or more operations, and one patient underwent 25 operations for the management of portal hypertension. The average number of bleeding episodes prior to operation was 3.6, and all but two

TABLE 2. *Portal Venous Pressure Measurements*

Pressure (mm H_2O)	No. of Patients
Under 200	7
200–300	27
300–400	22
400–500	9
Over 500	4
Total	69

TABLE 3. *Operations for 53 Patients with PVT*

Operation	No. of procedures*	No. Rebled	No. Reoperated	No. Died
Splenectomy alone	20	19	17	1
Variceal ligation	22	22	20	2
Variceal injection	1	1	1	0
Gastric division	17	15	10	3
Gastrectomy with colon interposition	2	1	1	0
Cavomesenteric shunt	9	4	3	0
Splenorenal shunt	6	3	1	0
Makeshift shunt	6	6	4	0
Splenic transposition	3	2	1	0
	86	73	58	6

* Several patients had more than one of the operations listed. Seventy-eight additional operations were performed on the 53 patients, including lysis of adhesions, revision of anastomosis, gastrostomy, drainage of abscess, etc.

patients had experienced at least one hemorrhage. Once operative management was undertaken, subsequent operations were frequently necessary (average 2.1 per patient), and the morbidity was high. Esophageal balloon tamponade was employed in several children, as was systemic vasopressin. The data suggest that tamponade has been of considerable value, but there does not appear to be any convincing evidence of the value of vasopressin in this series. Nonoperative measures, including hospitalization, bed rest, sedation, vitamin K administration, oral antacids, and withholding of oral feedings were successful in stopping the acute variceal hemorrhage, within 48 hours in most instances. Bleeding episodes frequently followed the onset of an upper respiratory tract infection.

Nineteen of 20 children who underwent splenectomy alone as the initial operation rebled, although the next bleeding episode was delayed an average of 3.4 years. Seventeen of these patients underwent further operations, and one died. Many additional patients underwent splenectomy at the time of other operative procedures.

Twenty-two patients underwent variceal ligation for acute bleeding, but subsequent bleeding occurred in all, and two patients died.

Esophagoscopy with transesophageal injection of the varices with sclerosing solution was performed on one child during an acute bleeding episode; however, further hemorrhage occurred one month later.

Porta-azygous disconnection and gastric division (Tanner procedure) was performed on 17 children; rebleeding occurred in 15, three of whom died.

Two patients underwent esophagogastrectomy; one experienced subsequent rebleeding, and the other underwent multiple operations to relieve obstruction of the interposed colon segment. Neither of these patients died after esophagogastrectomy. One patient who underwent vagotomy and pyloroplasty after four previous operations developed recurrent bleeding and has experienced symptoms compatible with the dumping syndrome.

Shunting operations were performed in 21 children and were the most successful of the operations performed to prevent bleeding. End-to-side cavomesenteric anastomoses were performed in nine patients, and four have experienced further hemorrhage. Six patients underwent central splenorenal anastomosis, and three patients rebled. In one of these patients the shunt thrombosis produced complete occlusion of the renal vein as well. Makeshift shunts were performed on six children, and each patient experienced recurrent bleeding shortly thereafter. Cavomesenteric shunts produced the greatest decreases in portal pressure. Each of the shunts that did not decrease the portal pressure more than 120 mm H_2O subsequently became thrombosed, and recurrent bleeding ensued.

Surgical transposition of the spleen into the left chest was performed in three children, two of whom have experienced subsequent variceal hemorrhage.

Sixteen (23%) of the children in the present study did not undergo any operations for treatment of portal hypertension, and all are still alive except for one child who was killed in a railroad accident of causes unrelated to the portal hypertension. An average of 2.4 episodes of variceal hemorrhage occurred in these patients. Portal pressures were measured in all but one of the 16 patients, and the average pressure was 317 mm H_2O. In comparison, the average portal pressure recorded in the 21 patients at the time of portal-to-systemic shunting was 303 mm H_2O. The current average age of this group of nonoperated patients is 14.9 years, ranging 4–23 years, and is only six years younger than that for patients who have undergone operations (21.2 years).

Current followup information is available on 51 patients whose ages range 3–58 years (average 21.6 years). Recent information was not available for 11 patients; however, the average followup available for this group of patients beyond the time when portal hypertension was first diagnosed is 12.9 years. Bleeding episodes became much less frequent after the age of 16 years; only

seven of 29 patients have bled beyond this age, and four of these have required operative treatment.

Only seven patients in this study died, and of these one was killed in a railroad accident (Table 4). The six deaths all occurred in the group of 53 children who underwent operations: one of 20 children died after splenectomy alone (5%); two of 22 died after variceal ligation (9%); and three of 17 died after porta-azygous disconnection (18%). No child died after a shunt operation. All 6 children died within 9 months of an operation.

Discussion

The manifestations of portal hypertension attributable to extrahepatic venous obstruction in children have been reviewed previously,[2-4] and the common findings of variceal bleeding, splenomegaly, anemia, melena, and varying degrees of hypersplenism were observed in almost all patients in the present study. In contrast to the experience of Clatworthy and Boles,[3] ascites was rarely seen. None of the patients had concomitant peptic ulcers.

Portal vein thrombosis is primarily a mechanical problem, with hemorrhage as the only major risk to the patient. In contrast to the high mortality for patients with intrahepatic disease who experience variceal bleeding, the mortality rate is extremely low for patients with PVT, as observed by other authors.[1,11] As noted previously,[10] no direct correlation could be established between the portal venous pressure and the frequency or severity of bleeding episodes. There were 338 episodes of bleeding among the 69 patients in this study. Only two patients died as a direct result of bleeding, and both these patients lived in remote communities and did not receive acute medical care while bleeding. Bleeding episodes were heralded by an upper respiratory tract infection more than 70% of the time. Parents were advised to be particularly observant for melena during such infections. One child experienced 16 bleeding episodes before the age of five years, each related to a recurrence of pneumonitis associated with a bronchopleural fistula following pulmonary lobectomy for lobar emphysema; bleeding episodes became sparse after the fistula closed. Several patients experienced bleeding episodes after ingestion of aspirin, some of which were the most severe bleeds encountered; thus, parents have been advised to refrain from giving this medication. Bleeding episodes were generally more severe when they occurred following operations to relieve portal hypertension than in patients who had not been operated upon.

Emergency operations for bleeding from PVT have been performed rarely during the past 10 years. Most variceal ligation procedures were performed prior to that time, and the reason for operation in many of these patients was not apparent. This procedure was rarely done as the initial operation, and in no patient was it performed for less than three bleeds. In general, we agree with the view of Shaldon and Sherlock[11] that the variceal hemorrhage can be controlled by nonoperative methods; however, a few children have bleeding that is so persistent and catastrophic as to require balloon tamponade, intra-arterial pitressin infusion, variceal ligation, or gastric division, despite the fact that further bleeding has followed all such operations in our experience. No emergency shunts were performed in this series.

In all but two patients, splenectomy alone eventually was followed by recurrent hemorrhage that made difficult the assessment of the portal venous system prior to a later operation. Splenomegaly *per se* has rarely been a problem, and no case of splenic rupture has been encountered, despite a lack of restrictions on physical activity. Of particular note is the fact that four of the seven children who died had undergone a previous splenectomy, and each died with sepsis, one from pneumococcal infection. Our experience would substantiate previous reports that recommend that splenectomy alone for bleeding or for hypersplenism has no place in the management of PVT, particularly in the young child. On the other hand, splenectomy may facilitate construction of a portal-systemic venous shunt in older children and may reduce abdominal swelling, thus enabling return to normal physical activities.

In seven of the 15 patients with cavomesenteric and splenorenal shunts, thrombosis and rebleeding occurred.

TABLE 4. *Cause of Death in Patients with PVT*

	Operations	Complications	Age at Death*
1.	Variceal ligation and Splenectomy	Gastric perforation; sepsis	9 yrs.
2.	Splenectomy alone	Pneumococcal meningitis; sepsis; variceal hemorrhage	14 mos.
3.	Gastric division (Tanner procedure)	Died from hemorrhage in remote community	18 mos.
4.	Gastric division and Splenectomy	Gastric fistula; sepsis	23 yrs.
5.	(1) Splenectomy alone (2) Variceal ligation	Subphrenic abscess; pneumonia; sepsis; jaundice	8½ yrs.
6.	Gastric division and Splenectomy	Died from hemorrhage in remote community	15 mos.
7.	No operation	Killed in railroad accident; No complications related to PVT	12 yrs.

* Each patient died within 9 months of an operation.

Six additional patients had makeshift shunts, and each has rebled. End-to-side cavomesenteric shunts were the most successful in preventing rebleeding, although the long-term benefits were unreliable, particularly when performed in patients under the age of six years. In several patients, the thrombotic process occluding the portal vein extended into the splenic and mesenteric veins, making surgical decompression difficult. In one such patient the renal vein thrombosed after a splenorenal shunt. We concur with Clatworthy[5] and others that a shunt with a diameter of less than 1 cm is unlikely to remain patent and that shunt procedures should be deferred until the patient is eight years of age. In the rare patient with PVT that extends to the proximal splenic vein, the Warren distal splenorenal shunt may be helpful.

Progressive portal hypertension in children with PVT is usually associated with an increase in the splanchnic-systemic venous collateral circulation.[12] Variceal hemorrhage is usually the result of an insufficiency of these collaterals,[10] and repeated intra-abdominal operations are likely to disrupt many of these collateral channels. One must, therefore, weigh the probability that a large shunt can be established that will remain patent against the chances that the collateral circulation might be divided in vascular adhesions or that injury to other structures might be produced. A large number of patients in this series have experienced postoperative complications, including ascites, intestinal obstruction, wound infection, intestinal perforation, hemorrhage, and jaundice. One patient sustained an injury to the common bile duct during construction of a shunt at 3 years of age, and severe cirrhosis and jaundice subsequently developed. The condition was relieved several years later with a left hepatico Roux-en-Y jejunostomy. Seventy-eight operations were performed to treat complications of previous operations other than bleeding.

Concern regarding the long-term effects of portal-systemic shunts for PVT has been expressed by Voorhees and associates[13,14] who found that in five of eight patients with patent shunts who were followed beyond the age of 20 years, emotional disorders and abnormal electroencephalograms developed. A few of the shunted patients in the present study were emotionally unstable, but this was attributed to the effects of prolonged hospitalization and repeated operations, and we have no documented evidence of abnormalities of the central nervous system.

The experience from this study suggests that a few children with PVT may be managed very satisfactorily and safely without operation despite recurrent variceal bleeding. Bleeding episodes generally became less frequent as the patients increased in age and became less susceptible to respiratory infections. When bleeding continues to recur with moderate frequency, a planned portal-to-systemic venous shunt would seem justified, provided that a shunt with a diameter of more than 1 cm can be constructed. Portal venography is very helpful in making this decision. Patients who undergo shunts under less favorable circumstances and those who receive various other operations to treat portal hypertension appear to have a higher risk of morbidity and mortality than do those managed nonoperatively. There was no significant difference in the incidence of hemorrhage in nonoperated patients as compared to operated patients in this series, with followup extending into late adolescence.

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Discussion

Dr. H. William Clatworthy, Jr. (Columbus, Ohio): This paper gives us some insight in a reasonable number of cases—sixteen—into the natural history of this disease; and this information really has been very hard to glean from the previous literature.

I feel these children with extrahepatic portal bed block must