Portal Hypertension in Children: Three Case Reports and Review of Recent Progress in Treatment

Philip J. Howard, MD

The diagnosis and surgical management of extrahepatic portal hypertension in infants and children have improved dramatically in the last 50 years, as evidenced by the reported mortalities of 46% in 1928-1935 and only 12% in 1974. The diagnosis is often evident from the first sudden gastrointestinal bleeding incident and can be verified quickly with fiberoptic esophagoscopy and x-rays. The surgeon has a choice of multiple shunt procedures to control portal pressure.

Portal hypertension in children that involves an enlarged spleen and esophageal varices is a threatening condition. Quite often the first sign of disease is a sudden gush of blood from the mouth which may indicate ruptured esophageal varices. Primary portal system hypertension without liver disease is usually from thrombosis, phlebitis or deformity of the portal vein or tributaries. The deformity may be smallness of the vein, multiple veins or enlarged areas leading to pooling of blood. Secondary hypertension may be from congenital cystic hepatic disease, cirrhosis or fibrosis after hepatitis from viral, bacterial or other infections, all of which disturb liver function and structure. These are subject to test and examination by biopsy. These conditions with liver disease account for more than half the cases of portal hypertension, while primary cases free of liver disease constitute less than half.1

Clinical study will show the source and severity of bleeding and whether there is liver enlargement. Cases with liver enlargement have poor prognosis, depending on the underlying disease, such as Wilson's disease (disturbed copper metabolism), cystic fibrosis, or mucopolysaccharidoses.

Primary hypertension cases can usually be supported conservatively with sedation, transfusion, and measures to control esophageal varices with local pressure by the Sengstaken-Blakemore tube until the child's veins are large enough for the distal splenorenal or other indicated shunts. If possible, operation should be delayed until the child is 8 years old.

Case Reports

Case 1

A white female, born November 15, 1950, was admitted to Henry Ford Hospital on September 12, 1957 for an enlarged spleen present since the age of 7 months. There had been no interference with growth, development or schooling. At age 23 months her spleen had become enlarged 2 cm below the costal margin, and her liver was felt at the costal margin. Her blood count, urinalysis,
A female infant, born August 31, 1953, was admitted on January 23, 1956 for abdominal pain and vomiting of two days' duration.

On admission at 7 years of age she weighed 50 lbs and was 48 inches in height. Blood pressure was normal. There were bilaterally dilated veins over the abdomen, the spleen was enlarged at 6 cm below the costal margin, and the liver was considered normal in size. Splenectomy was performed because of splenic size. At operation the splenic veins were searched for, but no true splenic vein was found. One or two small veins were seen to run in an aberrant direction. The spleen weighed 200 gms (normal weight, 58 gms). Her postoperative course was uneventful.

As of March, 1978, the patient was reported* to be in excellent health.

In cases of this type it is now surgical practice to perform a distal splenorenal shunt that preserves the spleen.

Case 2
A male born on November 17, 1917 with splenic vein anomalies who grew up without symptoms gradually developed an enlarged spleen and varices with esophageal hemorrhage.

He was first seen on June 5, 1959, when he was 42 years old, with complaints of upper abdominal pain, some weakness and an enlarged spleen of five years' duration. He had no history of either hematemesis or intestinal bleeding. His past history was negative except for jaundice at 11 years of age. His examination showed normal height and weight, good general development, in addition to an enlarged spleen, varices of the esophagus, and anemia. He was treated for anemia for four weeks, and surgery was performed on July 11. At surgery several very large ramifications of the splenic vein were noted, and the surgeon was able to trace one main branch which led into a somewhat thickened but normal caliber splenic vein. Pressure in the splenic vein was 320 to 330 cm of saline. This was then divided, the spleen removed, and a splenorenal shunt completed.

The patient was followed for nine months without complications. He then had a serious hemorrhage from esophageal veins in April, 1960, which healed during rest therapy. He was re-examined on June 2, 1967. Physical examination and gastrointestinal series were normal. He had been seven years without pain or hemorrhage.

This case would probably now have a distal splenorenal shunt without spleen removal.

Case 3
A child treated for six years had three major operations, seventeen admissions for injection of varices and transfusions, and two for orthopedic correction. Her diagnosis was venous malformation at the hilus of the liver and venous enlargement in the central portion of liver substances. Liver function was normal.

A female infant, born August 31, 1953, was admitted on January 23, 1956 for abdominal pain and vomiting of two days' duration.

The patient was considered to be in constant danger of uncontrollable bleeding and a reoperation to perform a mesocaval shunt was recommended.
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Admission for the third major operation was on February 4, 1962, when the patient was 8 years old. She weighed 32 lbs and was 44 inches in height. Hemoglobin was 8.2 gm%, WBC 4450 mm³, urea nitrogen 9 mg/dl, total protein 5.9 g/dl. The third operation was performed on February 8, 1962. Pre- and postshunt superior mesenteric venography was done with preshunt measure of portovenous pressure at 500 mm water, and postshunt of 240 mm water. The inferior vena cava was divided just proximal to its bifurcation. A surgical path was developed and the proximal inferior vena cava was swung and brought in proximity to the right border of the superior mesenteric vein. An end-to-side anastomosis was made between the end of the inferior vena cava and the side of the superior mesenteric vein. The portal pressure was relieved by approximately 50%.

The child was examined in 1963 and again in 1970, when her weight was 58 lbs and her height 58 inches. She had been relieved of any tendency to bleeding. She continues to be under medical care for orthopedic and urologic difficulties derived from the birth injury.

Discussion

In cases like these it is a matter of surgical judgment whether to do a splenorenal shunt or a distal shunt.

Operations in small children are delayed as long as possible to allow the veins to develop so that distal splenorenal shunting to preserve splenic function can be performed. Portal vein system obstruction in children, which has many causes, can usually be clearly distinguished from adult cases secondary to cirrhosis of the liver. Portal vein congestion is the type commonly seen in infants and children. It may be caused by phlebitis with partial obstruction or by thrombosis of the regional veins, mainly splenic or portal, but involving the tributary collateral veins in increasing pressure. Any type of infection leading to sepsis is a danger. Other main causes are congenital deformities of the splenic and portal veins such as constrictions, small size, extra valves, or when many small veins replace the main vein. There may be complete thrombosis later recanalized, and there may be a variety of vascular dilations of the portal vein or of its hepatic branches. The most severe complication is acute bleeding from esophageal varices by way of increased pressure in the splenic vein and collateral esophageal veins. Many types of intrahepatic disease cause congestion with splenic enlargement and esophageal varices. Some of these are infective hepatitis, congenital cystic disease, metabolic disease, or cystic fibrosis. Conditions of this type are not so clearly surgical problems. They may require surgery, but have differing medical needs. The indication for operation is still uncontrolled bleeding from esophageal varices.¹

The management of children with esophageal hemorrhage from varices was inadequate as long as splenectomy alone was considered the most effective treatment. In the 1927 and 1935 reports by Smith and Howard² and by Smith and Farber,³ 7 of 15 children treated by splenectomy died. Their average age was 9 years. Although their vein structure was large enough for shunt procedure in all but one who was 4 years old, shunt operations were not in use. Such high mortality was convincing evidence that better operations were needed. In 1941 Ladd and Gross⁴ concluded that the main causes of portal obstruction were thrombophlebitis from omphalitis or other sources of thrombi and persistent embryonic valves. They advised splenectomy in early cases to reduce portal engorgement and believed that this procedure reduced the portal load by 20%.

In their more recent cases they used division of esophageal branches of the coronary vein and omentopexy to provide additional collateral channels to reduce esophageal varices.

Between 1937 and 1945 great changes in surgery for relief of portal hypertension were undertaken, and new shunt procedures were tried by Whipple and associates⁵,⁶ and by Blakemore and Lord.⁷ Subsequently, great skill was acquired in blood vessel surgery of the delicate tissue involved in infants and small children. In 1952 Schumacher⁸ reported that 7 of 21 children under the age of 15 years had shunt procedures, with only one poor result. Since then the mortality in this critical group has improved. Gellis reported six children who had splenorenal shunts, each of whom was well at ages ranging between 8 and 12 years.

Thomas and Sherlock⁹ reported on the etiology of portal vein thrombosis in 12 cases aged 4 months to 20 years; 8 had neonatal sepsis, and 4 had sepsis between two and 20 years of age. They then reported 493 infants who had umbilical vein catheterization usually with exchange transfusion. In a ten-year follow-up study on 470 of these children, 80 had general sepsis, including three with umbilical infections at birth. All were treated with prophylactic antibiotics and no portal sepsis occurred. In emergency care of high risk and premature newborn infants, constant use of intravenous and intra-arterial catheters may be needed. Phlebitis, arteritis and omphalitis are rare complications, but when these occur, vigorous antibiotics are indicated.

By 1965 the splenorenal and selected shunt for relief of portal obstruction were well known but opinion about the necessity of the operation was divided. Lynn¹⁰ reported that in 31 cases in children, 14 had a splenorenal shunt with ligation of varices, 7 had portacaval shunts, and 7 had intestinal interposition, of which 5 were colonic and 2 were jejunal. He commented: "While esophagogastrectomy with interposition of colon seems unphysiologic and cumbersome, it seems the most satisfactory method available at the time of this report."

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In a series of five children operated on for extrahepatic portal obstruction, Koop\textsuperscript{11} reported that all had colonic replacement of distal esophagus and proximal stomach. All were followed for at least seven years. All recovered and only one required a later superior mesenteric to vena cava side-to-side anastomosis. This report represents five children followed seven years with no complications of liver disease or neurologic sequelae.

Echeverria\textsuperscript{12} reported on resection of the esophageal varices in five cases of portal hypertension in children. X-ray views of the varices were obtained by using thick barium swallows. He briefly reviewed injection of sclerosing fluid into the dilated veins, resection of the lower esophagus and upper pole of the stomach, ligature of the splenic vein and splenectomy, gastrectomy, devascularization of the stomach, tampon in the esophagus, or ligation of the hepatic and splenic arteries as "having fallen into disuse." His choice was thoracotomy by resection of the seventh and part of the sixth rib. A longitudinal incision was made into the esophagus with exposure and dissection of the varices, ligating and cutting each; then the esophagus was closed in two layers, as well as the thorax. No time of follow-up was indicated, but all patients recovered.

Turcotte\textsuperscript{13} described 31 portosystemic shunts in children for bleeding varices. The most common etiology was portal vein thrombosis which occurred in 13 cases. Other causes listed in his series are six posthepatitis cirrhoses, four congenital cirrhoses, and one each of mucoviscidosis, biliary atresia, familial biliary stasis, Budd-Chiari syndrome, Gaucher's disease, and congenital stenosis of the portal vein. Other etiologies he recognized were exchange transfusion, infectious mononucleosis, chemotherapy for Wilms' tumor with resulting cirrhosis. Many of his children looked remarkably healthy until they had a sudden gastrointestinal hemorrhage. He advised barium swallow, fiberoptic endoscopy, and retrograde mesenteric angiography. Prognosis depends on maintained hepatic function. The children with liver disease, biliary atresia, biliary stasis, Budd-Chiari syndrome and Gaucher's disease did not survive.

Clatworthy's classification\textsuperscript{14} includes rare possible causes of presinusoidal extrahepatic portal block and thus includes nearly all infants and children. Cavernous malformation of the portal vein is most commonly observed, possibly caused by aberrations in its formation from vascular network, or from antepartum or postpartum thrombosis with subsequent recanalization. Congenital stenosis or atresia have occurred, and omphalitis, general sepsis, and other purulent infections have also been recognized. Intraluminal trauma has been suspected from exchange transfusions. Blunt abdominal trauma with pancreatic injury may lead to local thrombosis, especially of the splenic vein. Similarly, neo-

plastic or cystic disease may involve the liver, kidneys or adrenals with resulting injury to tissue leading to thrombosis of the portal venous system. Arterial venous fistulae of a congenital or traumatic nature may lead to sclerosis or thrombosis or to an increase in portal pressure. The collateral circulation is the usual route to circumvent obstruction, since the gastroesophageal plexus commonly cause trouble. The other collaterals, diaphragmatic, periumbilical, lumbar, splenorenal, spermatic or ovarian, and perirectal are considered advantageous, seldom leading to severe hemorrhage in children. Bleeding esophageal varices are still the main troublesome event in children. Clatworthy\textsuperscript{14} describes four types of splenorenal and four types of mesocaval shunts. A report of 418 cases of extrahepatic portal bed block showed 53 deaths, 23 from hemorrhage, 22 from operation, and 8 from hepatitis. This 12% loss rate and 88% cure rate compare favorably with the 1935 figures of 46% loss and 54% cure with inadequate operations.

Long-term survivals included two children in Linton's report\textsuperscript{15} who lived 11 and 13 years after shunts. Clatworthy\textsuperscript{14} reports two such cases living 11 and 20 years, Hsia and Gellis\textsuperscript{1} five cases who lived 8 to 13 years, Schumaker\textsuperscript{8} 10 cases who lived over 7 years and two who lived 10 and 20 years. These long survivals indicate the excellent liver function that accompanies portal obstruction in children unless they also have liver disease as diagnosed by biopsy.\textsuperscript{16}

On the other hand, in adults with cirrhosis the prognosis depends on the amount of reserve function. In Galambos' study\textsuperscript{17} of adults with advanced cirrhosis, liver function was assessed by rate of urea synthesis under controlled dietary conditions in which the liver functioned at its maximum rate. All those patients with selected shunts had some portal circulation, but those with total shunts had no portal circulation. The resulting onset of encephalopathy was one in 22 for those with some portal circulation, but 10 in 24 for those with no portal circulation. Galambos suggests that every effort be made to select an operation which decreases the flow through the bleeding varices but still, if possible, maintains some positive flow of portal blood through the liver.

Rosch and Dotter\textsuperscript{19} advise that an angiographic examination gives decisive information about the vein and arterial structure of the operative area in portal obstruction and should be done in all cases in addition to blood tests, liver function tests and barium swallow. Contrast material injected into the spleen gives the best information of the splenic area. Angiographic study of the superior mesenteric artery with venous phase pictures shows the portal bed. Inferior mesenteric arteriograms and inferior venocavography may be used. They reported these examinations in a series of 38 children who had 33 splenoportograms, 5
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having had previous splenectomies. Their patients ranged in age from 2 to 18 years and had extrahepatic obstruction.

Leger has studied pressure in the portal system. The portal vein starts from a bed of capillaries and ends in a bed of capillaries. Its flow is normally from splenic, superior and inferior mesenteric veins to its termination in right and left hepatic veins and into the hepatic capillary bed; thence through the hepatic sinusoids to the hepatic vein, inferior vena cava and left auricle. Manometric pressure can be measured in the spleen by needle puncture and is approximately 10 cm of saline, the same as the wedged hepatic vein pressure obtained by venous catheter via inferior vena cava and hepatic vein. Obstruction of the splenic vein at first raises pressure in the spleen only, whereas obstruction in the portal vein raises pressure in all parts of the portal system although the wedged hepatic vein pressure remains normal. Cirrhosis or fibrosis of the liver parenchyma causes pressure on the sinusoidal system, increasing the wedged hepatic vein pressure, which rises proportionally as fibrosis becomes more severe. Under 25 mm Hg is considered mild, 25-30 mm Hg moderate, and above 30 mm Hg severe. These degrees of pressure are seldom present in children, nor are the special pressure conditions of schistosomiasis (intrahepatic presinusoidal pressure). In this latter, the wedged hepatic pressure is normal.

The situation in children where the diagnosis of "idiopathic" is accepted points to the slow development of venous expansion, laking or pooling. It is also recognized that forward pressure in regional arteries may tend to actual arteriovenous shunt or anastomosis.

Summary

The management of sudden bleeding from esophageal varices and splenic enlargement must be considered in stages and with respect to the child's age and size. Diagnosis must be complete as to site of portal obstruction by splenoportogram, and as to varices by esophagoscopy or barium swallow x-rays. The renal veins must also be studied since distal splenorenal shunt depends on size and normalcy of the left renal vein. The normal position of left renal artery should be ascertained as it is frequently out of normal position. The younger or smaller the child, the more conservative the action must be. Because young children tend to stop bleeding with sedation and transfusion, the amount of bleeding must be estimated by hemoglobin determination. Local pressure on the esophageal varices by a Sengstaken tube may be used. At times varices may have to be tied by local operation in order to sustain the child long enough for splenic vein growth to 1 cm in diameter. Uncontrollable hemorrhage requires surgery, and a distal splenorenal shunt is preferred, or other distal shunt which will decompress or relieve the esophageal varices and avoid splenectomy. The distal splenorenal shunt is preferable because it supports some liver circulation and leaves the spleen intact. Even though the liver function of children is superior to that of adults, every improvement in liver circulation should be sought.

Advances made in the last 50 years in better techniques for esophagoscopy, improved portograms, angiography and venacaval studies have significantly decreased the mortality rate among children with portal vein hypertension. Skillful surgery, fluid balance management, and antibiotic care have also shared in reducing the mortality for these difficult patients.
References