Esophageal Varices in Children


Banti's disease may be defined as a disease of the spleen, marked by splenomegaly, anemia and leukopenia, and later by cirrhosis of the liver with ascites and jaundice. Banti wrote several articles between 1894 and 1910, describing cases of splenomegaly and anemia which he followed from childhood into adult life.

The surgeon generally considers Banti's syndrome to include the triad of cirrhosis of the liver, splenomegaly and esophageal varices. Chronic portal hypertension is the common denominator leading to the pathological alterations and the clinical picture of the hepatosplenopathies, loosely called "Banti's disease". Some cases, especially those of various types of extrahepatic causes of portal hypertension, are not associated with cirrhosis of the liver. The obstruction is intrahepatic in 80% of cases according to Macpherson, and 70% according to Linton. In only 2% is the obstruction confined to the splenic vein.

The portal system is interposed between two capillary beds. About 75% of the blood to the normal liver is derived from the portal vein; the remaining 25% comes from the hepatic artery. Approximately 40% of the portal blood comes from the spleen.

The portal vein contains a large volume of blood under low pressure; the hepatic artery has a small volume under a high pressure.

Because the portal system lies between two capillary systems and the sinusoids of the liver are embedded in a freely expansile stroma, it follows that during the course of cirrhotic disease there may be an increased resistance to portal venous flow. Particularly in the periportal type of cirrhosis, progressive fibrotic distortion, twisting and obliteration of the finer radicles may occur. Delicate sinusoids may be compressed by regenerating liver cells, and this impedes free exit of blood to the central vein. This process in turn decreases the nutrition available to nearby hepatic cells, augmenting the pathologic process; ultimately, a hepatic arteriole-portal venous shunt may occur, with resultant increase in portal pressure. The average hepatic artery inflow in cirrhotic livers is believed to be increased over that of normal livers.

Extensive collateral circulation develops. This is found in the region of the gastric cardia and lower esophagus, between the superior hemorhoidal veins and the middle and inferior hemorrhoidal veins, between retroperitoneal connections with the veins of Retzius, ligamentous attachments of the liver via the veins of Sappey, and the veins of the falciform ligament which may produce a "caput medusae". Retroperitoneal paravertebral veins coursing behind the diaphragm and joining with the hemiazygos, and thus the superior vena caval system, are added anastomotic channels.

Thrombosis of the portal vein is rarely associated with cirrhosis of the liver. Merendino and Dillard reported that they had never encountered rectal hemorrhage in patients with portal bed block. The major difference in bleeding potential between the esophageal varix and the rectal hemorrhoid is due to the juxtaposition of gastric chyme to the esophageal varix. The large esophageal varices are dilated vascular structures lying between two low pressure systems, the lumen of the esophagus and intrapleural space, so that there is no support for the veins. Such delicate structures can be eroded by trauma from a bolus of food or by acid-pepsin action of gastric juice. Bleeding may be associated with hemorrhagic diathesis due to the hypersplenism which occurs as a result of congestive splenomegaly. Ascites is associated with bleeding...
varices at times, or it may be the only complaint of the patient with portal hypertension. The development of ascites in a patient with portal hypertension may be due to a number of factors, among them intrahepatic cellular edema, protein and electrolytic imbalance, and hormonal disturbances. It is of some importance to realize that the principal causes of portal hypertension in adults and in children are different. Arcari and Lynn studied a collected series of 75 cases of bleeding esophageal varices in children. The site of the obstructive process responsible for the portal hypertension and its sequelae was predominantly extrahepatic and in the portal venous system itself. Sixty-eight of the 75 cases were due to extrahepatic portal obstruction; of these 68 patients, two had ascites.

Many patients with this disorder present during infancy with established portal hypertension due, for example, to an omphalitis in the neonatal period. It is reasonable to assume that a septic thrombophlebitis originates in the umbilical vein and leads to bland or septic thrombosis in the portal venous system. When first seen, the infants may have ascites, growth failure and anemia. After a variable period of time and with appropriate therapy, recanalization of the thrombotic process and the gradual development of collateral venous channels lead to the disappearance of the ascites, and for a period they may grow and develop in near-normal fashion. Later, usually after several years, they present clinical problems because of either massive upper gastrointestinal hemorrhage from esophageal varices or secondary hypersplenism.

In the majority of such cases the cause of portal hypertension with bleeding esophageal varices is difficult to establish. Arcari and Lynn found causes such as cavernous transformation of the portal vein, stenosis of the portal vein, and thrombosis of the splenic vein. Bleeding first occurred before seven years of age in more than 70% of the children in their series; none died during the first bleeding episode. Most of them bled again within two years of the first episode; very few died of the second hemorrhage. All of these considerations allow time for reasonable planning and make it possible to defer surgery until the child has developed further and is older. However, these workers point out that undue delay after the second bleeding episode is accompanied by rise in mortality.

It seems that many of these children present with some degree of hypersplenism, although they have normal liver function studies and/or normal liver biopsies. The commonest single initial operative procedure in the series of Arcari and Lynn was splenectomy. This procedure should not be undertaken lightly because splenectomy alone in the presence of bleeding esophageal varices, whether hypersplenism is present or not, is of little value. In the few instances of isolated thrombosis of the splenic vein, the procedure would be curative. Splenectomy should always be combined with a splenorenal shunt if portal hypertension or esophageal varices have been demonstrated. The high incidence of extrahepatic obstruction, with involvement of the portal vein itself, frequently makes it impossible to perform a portacaval anastomosis to provide definitive treatment for these children.

Splenorenal shunt procedures are considered of definite value where they are indicated. Of the factors which influence the choice of treatment by means of splenorenal shunt, perhaps age and development outweigh all others. Arcari and Lynn consider that children 60 to 70 lb. in weight and about 10 years of age are most suitable candidates for successful shunt procedures because of the high incidence of portal thrombosis in such children. Clowther and Boles consider patients over 4 years of age, control of bleeding being best accomplished by a splenorenal shunt if the splenic vein is of good size. A centrally placed anastomosis, using a short proximal segment of splenic vein, has definite advantages over the conventional technique. King and Shumacker believe that the procedure should not often be carried out on children less than 11 years of age. Pietri, who examined patients at various times after utilizing splenorenal shunts, found a high incidence of failure.

In summary, although splenorenal shunt is effective in the treatment of portal hypertension due to extrahepatic block, particularly if the patient is a young adult, there are certain instances in which the splenic vein will not be satisfactory for anastomosis because of age or stage of development, or in which the patient will bleed again after a shunt. This procedure is advantageous because it offers an opportunity to treat the hypersplenism at the same time.

In the treatment of post-splenectomy bleeding from esophageal varices, transesophageal ligation of varices is, at least, an effective method of temporizing and, at most, a procedure of long-term value. Mesenteric-caval shunt is a method of dividing the inferior vena cava and anastomosing the proximal end to the side of the superior mesenteric vein. Marion, Bouquet and Yon have reported 11 cases treated in this manner and consider this type of shunt most effective in young adults.

Esophagogastrectomy involves resection of the site of origin of the esophageal varices. Peptic esophagitis can be a morbid aftermath of this procedure and is a very common occurrence. Transection operations for portal hypertension, in patients in whom shunts are not feasible or advisable, may include gastric transection with or without extensive devascularization; complete esophageal transection and mucosal esophageal
transection, carried out at the level of the esophageal hiatus.  

Interposition operations are an answer to some of the problems presented by the patient who has post-splenectomy bleeding or others in whom shunts are not feasible or have failed. The jejunum and colon, usually the transverse colon, are utilized.$^{11,14,18}$ The resections of the esophagus and the stomach need not be extensive; only a segment of the esophagus up to the inferior pulmonary vein, and usually no more than the upper 5 cm. of stomach, need be removed.$^{19}$

**Case Report**

G.S., a 15-year-old boy, was admitted to hospital on August 8, 1960, with massive bleeding from esophageal varices which had begun 24 hours earlier. The first episode of bleeding was at four years of age; this reoccurred four times between the ages of four and eight years.

When he was eight years old, elective splenectomy was carried out, and the pathological diagnosis at that time was Banti's disease. Three years later, he was again admitted with massive bleeding and was transferred to the Hospital for Sick Children, Toronto, where thoracotomy and esophagotomy was performed and the esophageal varices were oversewn. No further bleeding occurred until the present admission.

Physical examination revealed a young man, over 200 lb. in weight, who was bleeding from esophageal varices. Blood replacement was carried out. Liver function tests were essentially normal. Esophageal tamponade, utilizing the Blakemore tube, was carried out intermittently from then until August 25, 1960. On August 13, 1960, an approach through a right thoracoabdominal incision for a planned portacaval shunt, revealed obliteration of the portal vein with evidence of an old infective process.

A jejunal vein was cannulated, and the portal vein pressure was 520 mm. of water (Fig. 1). The gallbladder showed gross evidence of chronic cholecystitis and was removed. The liver was normal.
Following the operation, he soon bled again. Ammonia intoxication did not occur because of the detoxifying ability of the normal liver. On August 25, 1960, through a left thoracoabdominal approach, the lower esophagus and the upper stomach were mobilized (Fig. 2). In addition to the dilatation of the varices making up the varices, some paravertebral veins of tremendous size were seen coming up alongside the diaphragm in tortuous fashion; these were carefully avoided in the subsequent procedures and left intact. The esophagus just distal to the inferior pulmonary vein was transected, as was the upper portion of the stomach, an area approximately 7 cm. in length. The transverse colon, with the middle colic vessels maintained for blood supply, was interposed in isoperistaltic fashion (Fig. 3); the diaphragm was approximated and gastrostomy was carried out. From his admission on August 8, 1960, to the time of operation on August 25, 1960, he received 36,500 c.c. bottles of blood.

His postoperative recovery was slow but without major morbidity. At the time of discharge from hospital on October 9, 1960, he weighed 163 lb. Since discharge, he has had difficulty in maintaining adequate nutrition. At the time of the latest examination on May 29, 1962, he weighed 135 lb. There has been no bleeding since operation.

Summary

The causes of bleeding esophageal varices are different in adults and in children. Portal hypertension and erosive action on the lower esophagus are common factors. The cause of the portal hypertension is intrahepatic in the adult, and often extrahepatic in children. The surgical procedures indicated for adults and children are also different; shunt operations are the procedures of choice, but often are not feasible in children. In such cases, direct attack on the site of the esophageal varices may be the preferred method of treatment, as opposed to indirect attack by means of tension-lowering procedures.

References