Collateral Pathways in Portal Hypertension *

THOMAS N. P. JOHNS, M.D., BLACKWELL B. EVANS, M.D.

From the Department of Surgery, the Johnston-Willis Hospital, Richmond, Virginia

THE ROLE of operative surgery in the management of patients with portal hypertension is reasonably well established. It consists of operations which divert blood from the portal into the caval lakes by various venous shunting procedures; the aim is to control hemorrhage from gastro-esophageal varices in the majority of cases, to control ascites in a minority.

The success of shunt operations, portal-caval and spleno-renal vein anastomoses being the most used, depends upon the presence of a functioning portal or splenic vein which can be used for a shunt. When such a vessel is not available, as for example in cavernomatous transformation of the portal vein, portal-to-systemic vein anastomosis usually is impossible and the patient becomes a surgical challenge of considerable magnitude.

The three procedures designed to meet this challenge have had some limited success: 1) Ligation of the hepatic and other celiac arteries, as advocated for the relief of ascites by Rienhoff and Woods [25] may reduce the vascular load on the portal venous system and inhibit bleeding; 2) A direct attack on the bleeding either by the ablative or interruptive operations such as esophagogastrectomy or by direct suturing of bleeding varices as an emergency procedure sometimes proves to have lasting benefit; and 3) Operations which attempt to create networks of new collateral channels have theoretical possibilities. To the early operations such as the Talma-Morrison omentopexy [18, 19, 33] have been added the more recent and more sophisticated intra-thoracic transplantation of the spleen, as reported by Nylander and Turunen [20, 35] of Helsinki and later by Foster and Scott.[5]

The purpose of this report is to call attention to the naturally-occurring portal-to-systemic collateral shunts in portal hypertension and to record by way of illustration a case of cavernomatous transformation of the portal vein in which such a naturally-occcurring shunt could be turned to some surgical advantage.

Natural Portal Collaterals

The portal system collaterals are generally thought to be five in number: [11] 1) The esophageal plexus allegedly connects the portal with the azygos system of veins. However, it is more likely that in portal hypertension these vessels are blind ends of dilated veins [34] since the azygos system itself is not usually distended in the presence of varices. 2) The hemorrhoidal plexus is also implicated as a collateral pathway. However, although bleeding piles are supposed to occur frequently in patients with cirrhosis, they are not, at least in our experience, troublesome stigmata of portal hypertension. 3) The peri-umbilical plexus or in its advanced form the caput medusae [3] is evidence of regurgitation of portal blood through the veins in the round ligament of the liver into those of the anterior abdominal wall. Although prominent veins in the abdominal wall are commonly seen in portal hypertension, a true

* Presented before the Southern Surgical Association, Hot Springs, Virginia, December 5–7, 1961.

caput medusae is a late and rare finding. The diagnostic significance of this collateral pathway is greater than its physiological benefit to the patient. 4) The veins of Retzius[17] drain from the intestine to the inferior vena cava. These veins form a wide-meshed retro-peritoneal plexus connecting the posterior duodenal, pancreatic, splenic, colic and portal veins with the phrenic and azygos systemic veins and are found in the retro-peritoneal spaces such as the retro-duodenal space and the diaphragmatic attachments of the spleen. Dilated veins of Retzius are regularly encountered at operations for portal hypertension and the sometimes troublesome oozing from them suggests that they convey a considerable amount of blood. 5) The veins of Sappey[28-30] are accessory portal veins entering the liver capsule from various abdominal organs. These include cystic veins, veins in the gastrohepatic omentum, diaphragmatic veins and veins of the suspensory ligament of the liver. These vessels are the only collateral pathway capable of transporting portal blood into the liver in cases of extra-hepatic portal obstruction and are also regularly seen to be dilated at operations for portal hypertension.

It is probable that of these five collateral pathways, the first three do not shunt significant amounts of portal blood into systemic veins in portal hypertension. In the case of the latter two, *i.e.*, the veins of Retzius and the accessory portal veins of Sappey, observations suggest that they do transmit some collateral flow in portal hypertension. However, it should be pointed out that, according to Poiseuille's law, the entire collateral bed would have to be enormous in order to achieve the functioning capacity of a normal portal vein.

An additional collateral pathway has been found between the gastric and splenic veins and the left renal and adrenal veins. First described by Lejars,[14, 15] in 1888, these pathways have been found in numerous instances of portal vein obstruction. These veins are present in normal individuals and may be as large as four millimeters in diameter. They are apparently capable of dilating further to accommodate a considerable blood flow in case of portal vein obstruction.

Case Report

G. B. 52-7126. An 18-year-old white man was admitted May 3, 1949, because of jaundice and fever. There was a history of operation for liver abscess eight years previously. The present illness was of short duration and was characterized by right upper quadrant pain, fever and jaundice. On admission there was tenderness in the right upper quadrant, a moderate degree of jaundice and a small scar above the left costal margin. He improved on symptomatic therapy and was discharged from the hospital in 10 days.

Two months later he was re-admitted with a diagnosis of congenital hemolytic icterus. He had repeated episodes of abdominal pain, anorexia, weakness and jaundice. On admission he had anemia and splenomegaly. A splenectomy was performed. The postoperative course was complicated by a left subphrenic abscess, which was drained, and repeated upper gastro-intestinal massive bleeding requiring 23 pints of blood during the postoperative period. He was discharged after 53 days in the hospital.

His next two admissions during the following year were for symptoms of gallstones. An elective cholecystectomy was planned, but at operation this was abandoned because of too much bleeding. A cholelithotomy and cholecystostomy were carried out instead. He was discharged after 14 days. A right sub-phrenic abscess ensued requiring a fifth admission.

His next seven hospital admissions from July, 1952, to June, 1956, were for continuing upper gastro-intestinal hemorrhage. He had bled approximately 36 times, spent 84 days in the hospital, was treated with the Blakemore bag three times and received 27 pints of blood. A diagnosis of esophageal varices and portal hypertension was confirmed by esophagoscopy.

His thirteenth admission, November 9, 1956, was due to hematemesis and loss of over one liter of blood. He had continued to bleed during the six months since the previous admission. At this time he was admitted in shock and was transfused with blood. On November 21, 1956,

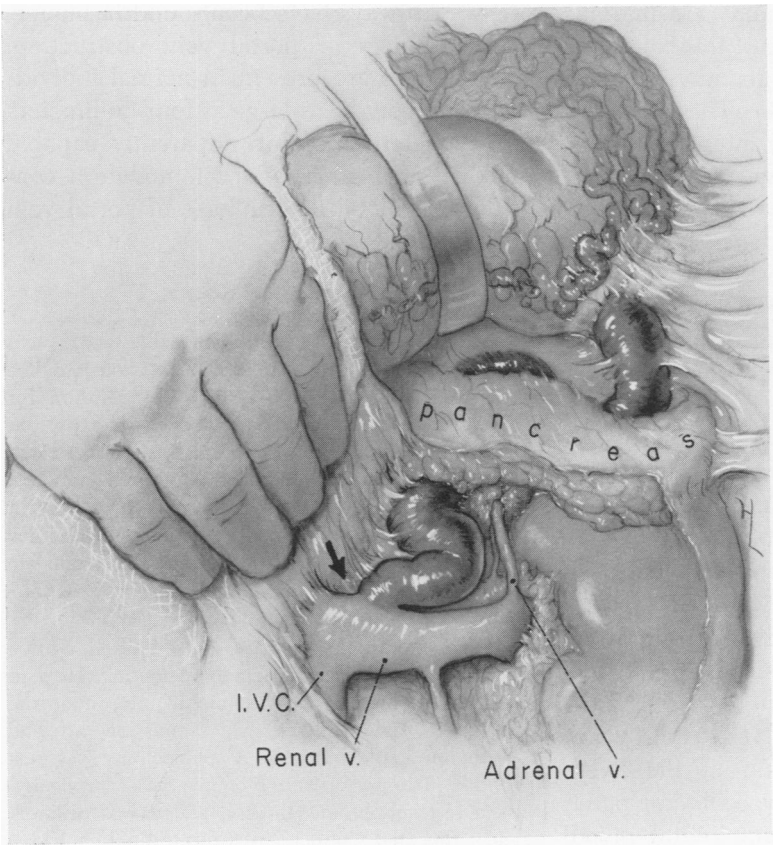

FIG. 1. Operative findings in patient with cavernous transformation of the portal vein. A left thoracoabdominal incision was made for intended esophagogastric resection. A large venous channel was found posterior to stomach and pancreas. This was followed from its connection with the coronary veins above to entrance to left renal vein.

a right thoraco-abdominal incision was made in an attempt to carry out a portacaval anastomosis. Cavernomatous transformation of the portal vein was found; the operation lasted six hours and 10 pints of blood were given. The cavernous masses were dissected into the liver where a fibrous remnant of the portal vein was found. Following operation, the patient developed obstructive jaundice and on December 13, 1956, a Roux-Y cholecystenterostomy was performed followed by clearing of the jaundice. Liver biopsy revealed healthy looking liver with minimal bile duct proliferation and no biliary stasis. During this admission, the patient received 29 pints of blood. He was discharged after 48 days in the hospital free of jaundice.

His next three hospitalizations from September 1958 to May 1960 were for repeated upper gastrointestinal hemorrhage requiring tamponade, transfusions and a total of 42 days in the hospital.

His seventeenth and most recent admission was on October 21, 1960, again because of upper intestinal bleeding. He had an estimated blood loss of five liters and was admitted in shock. He received four liters of blood, was again treated with a Blakemore-Sengstaken bag and improved. On November 3, 1960, a left thoraco-abdominal incision was made with the plan of performing an esophagogastric resection. At operation a large venous channel was found extending from the posterior surface of the stomach behind the pancreas in the direction of the kidney (Fig. 1). On dissection this vein entered the left renal vein just central to the entrance of the adrenal vein. Its largest diameter was 1.5 cm. and its smallest visible diameter at its entrance into the renal vein was 8.0 mm. Uncorrected pressures in the gastric vein tributary and in the renal vein were 185 and 130 mm. of saline, respectively. After cross-clamping the renal vein, the gastric vein pressure rose to 265 mm. of saline. A venoplastic enlargement of the existing splenorenal venous shunt was then carried out by a technic similar to the Finney pyloroplasty (Fig. 2). This resulted in enlarging the existing shunt to an opening of approximately 3.0 cm. in its greatest diameter. Following completion of the venoplasty, the splenic pressure was 165 mm. of saline.

The patient had a satisfactory postoperative recovery and left the hospital on the 48th hospital day. He is well and working as a school-bus driver in December, 1961, 13 months following operation. There has been no further bleeding to date. His peripheral blood findings are normal. Recent esophogram, however, reveals shadows identified as esophageal varices.

In summary, this patient was admitted to the hospital at the age of 18 years with a remote history of liver abscess. He was found to have jaundice and gallbladder disease, and was treated with splenectomy and removal of gallstones. After two years he began a series of approximately 40 upper intestinal bleeding episodes which lasted for the following eight years. He was hospitalized 17 times for a total of 308 days, and he bled an estimated 72 liters of blood during this time. He received 49 liters of blood and was operated upon eight times. He was found to have a cavernomatous transformation of the portal vein and ultimately to have an existing functioning shunt between the gastrosplenic venous system and the left renal vein. Following surgical enlargement of the existing shunt, the patient has been well for approximately one year, with persistent esophageal varices.

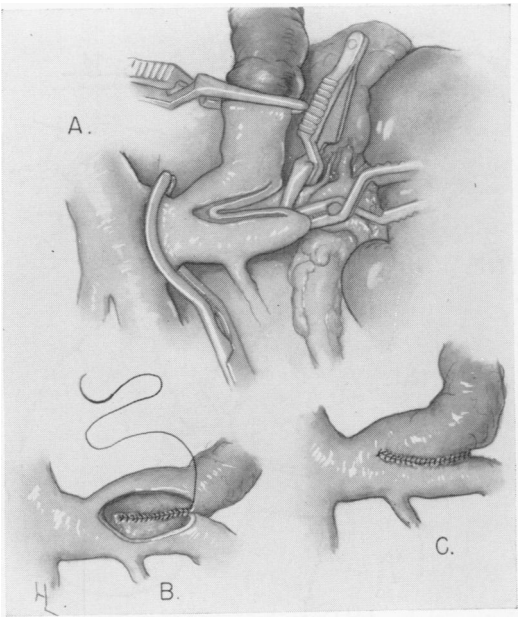

FIG. 2. Technic of enlargement of existing gastric to renal venous shunt. This is similar in principle to Finney's pyloroplasty.

Discussion

Nature of Cavernomatous Transformation of the Portal Vein. As in this case, cavernous or cavernomatous transformation of the portal vein is the replacement of the portal vein, and sometimes its major tributaries by angiomatous masses of tortuous distended veins situated in the free margin of the gastrohepatic omentum and enveloping the structures of the porta hepatis. Since these channels have characteristically thin walls and contain portal blood under elevated pressure, cavernomatous transformation may present technical surgical difficulties in controlling hemorrhage. Furthermore, none of these enlarged channels can be used satisfactorily for venous shunt as their angiomatous nature makes them impossible to isolate and manipulate.

Clearly a preoperative delineation of the portal system by splenoportography is most helpful in detecting this condition. In the absence of the spleen, as in this case, visualization of the portal vein can be obtained by cannulating and injecting a small mesenteric vein at the operating table. In retrospect, had this been done in the present case, in which obliteration of the portal vein was suspected, the dissection of the porta hepatis could probably have been avoided.

The pathogenesis of the obliteration of the portal vein and its transformation into angiomatous masses remains a matter of speculation. Since first described by Balfour and Stewart,[1] in 1868–69 and named by Klemperer,[9] in 1928, various theories of the nature of the condition have been proposed.[21, 26]

The original idea of Pick[24] that the lesion is a hamartoma or even a true neoplasm now seems unlikely in the view of the careful pathological observations of Gibson and Richards.[6]

Its occurrence in infants and young children[10, 38] suggests that the disease has a congenital or neonatal origin. The normal process by which the umbilical vein and ductus venosus are obliterated at birth

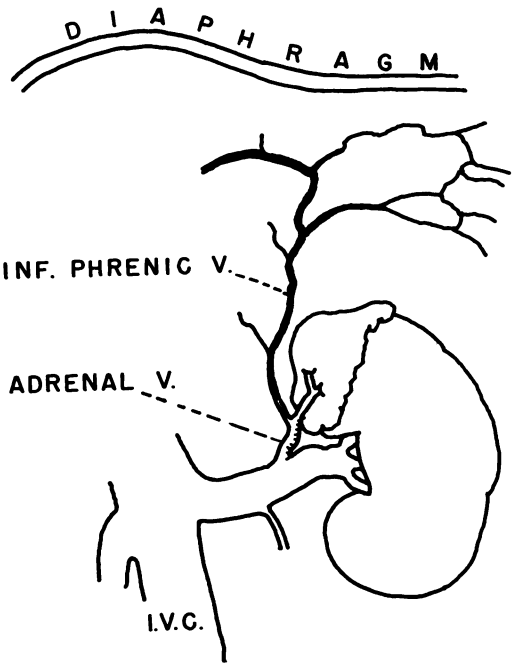

FIG. 3. One of the normal patterns of venous drainage into the left renal vein. There are no connections with the gastric or splenic veins.

may extend retrograde into the portal vein. A related condition is the Cruveilhier-Baumgarten syndrome[2, 3] described in 1829; the case was also reported by Pegot,[22] in 1833. In this disease the portal vein is obliterated but the umbilical vein remains open. The clinical findings are dilated veins in the abdominal wall, patent umbilical vein, caput medusae and a loud venous murmur at the umbilicus. Recent studies of Leger and Hennrich[8, 12] reveal the feasibility of ompholo-caval shunt to relieve this condition.

Obliteration of the portal vein is alleged also to occur as a result of trauma, infection or thrombotic blood dyscrasia. Infection of the umbilicus in the newborn with ascending thrombophlebitis may be implicated as may pylephlebitis secondary to gastro-intestinal conditions such as appendicitis. In the present case, infectious origin is suspected since a liver abscess had occurred. It is significant that at least ten years elapsed between the disease which may have involved the portal vein and the development of portal hypertension. The cavernomatous change presumably involved a gradual dilatation and proliferation of the accessory portal veins of Sappey, as well as recanalization of the thrombosed portal vein. This is in agreement with the findings of Gibson and Richards indicating that a considerable time may elapse between occlusion of the portal vein and the formation of the cavernous mass of veins. They suggest that cavernous transformation does not follow massive thrombosis of the portal vein but is due rather to a series of small, even sub-clinical thrombotic episodes.

As a rule, cavernomatous transformation of the portal vein is not associated with primary liver disease; in most cases, as in this one, the liver is normal. However, thrombosis of the portal vein is well known in cirrhosis of the liver. This sometimes occurs as a massive occlusion and produces varices in a matter of days or weeks. Under these circumstances, dilatation of the accessory portal system occurs but cavernomatous transformation is not found.

Our present conclusion is that cavernous transformation is secondary to obliteration of the portal vein, a gradual process which has more than one possible primary cause. The consequent angiomatous changes are due to over-abundant compensatory enlargement of the rich accessory portal system of Sappey.

Significance of the Gastro-Splenic to Renal Vein Pathways. Communications between the portal system and the left renal vein have been reported in isolated instances[7, 23, 31, 37] since described by Lejars,[14, 15] in 1888 and Tuffier and Lejars,[36] in 1891. In Simonds'[32] review of 32 cases of cavernous transformation in 1930, seven had these large shunts between portal tributaries and the renal or adrenal veins. In the studies of Madden[16] *et al.*, in 1954, injection of the portal system in cadavers

with normal livers revealed demonstrable portacaval shunts in four of eight specimens; in two of these the shunts were 3.0 and 4.0 mm. at their narrowest diameters. Detrie and Martini,[4] in 1960, presented six cases of spontaneous portacaval shunts in patients. Our dissections of 12 cadavers reveal that the left adrenal and renal veins regularly receive contributions from the inferior phrenic veins draining the left half of the diaphragm (Fig. 3). Communications from the gastric veins were present in two instances (Fig. 4).

It seems likely that the large venous communications found in this patient, as well as those reported in the literature, represent dilatation of such pre-existing channels. As most shunts have been reported in patients with cavernous transformation of the portal vein, splenic portography should be helpful whenever this severe form of portal vein obstruction is present.

Whether naturally existing shunts of this type are of functional benefit to the patients seems questionable. The long and tortuous course which the portal blood has to pursue through these veins together with their small size indicates that the blood flow through them is not of great consequence. Rousselot [27] who has observed many collateral portacaval shunts in his experience with splenic portography believes these channels are not of great functional significance and therefore do not benefit the patient. In the case recorded here, the channel was 8.0 mm. in diameter at its narrowest visible point and pressure measurements before and after cross-clamping it indicated that the flow was in the direction of the renal vein; however, the pressure differentials were not so great as to suggest high flow-rates and direct flow measurements were not made.

Obviously, it is not known whether enlargement of this shunt will prove of lasting relief to the patient; he still has radiologic evidence of esophageal varices and presumably portal hypertension. However, temporary freedom from hemorrhage for 13 months is the longest such period he has had in nine years. It is presumed wiser, therefore, to have preserved this shunt and enlarged it rather than to have carried out an ablative procedure which would have destroyed it.

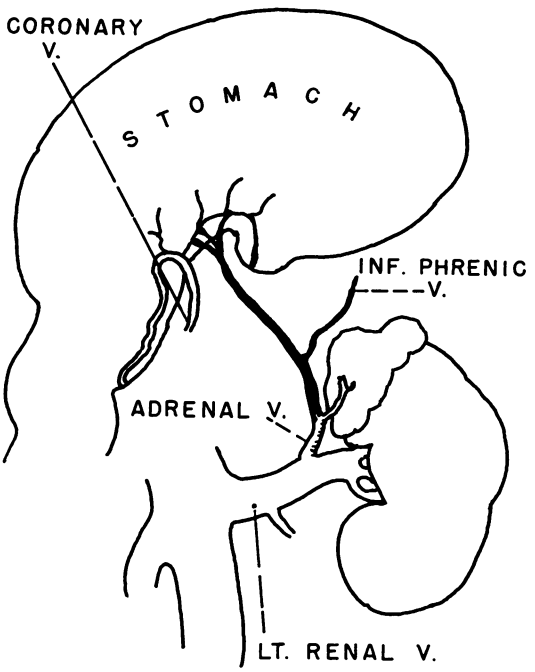

FIG. 4. One type of communication between gastric and renal venous systems. This is an autopsy finding in a patient with normal portal vein.

Summary and Conclusions

1. The five classical collateral pathways of the portal circulation are probably of little physiological benefit to patients with portal hypertension.

2. Naturally occurring shunts between the gastric and splenic veins and the left renal and adrenal veins are an important part of the portal collateral circulation. These shunts occur in approximately 20 per cent of individuals and appear to be capable of dilating in the presence of portal hypertension.

3. A case of cavernomatous transformation of the portal vein is reported in which a large gastrorenal vein shunt was found and enlarged.

Bibliography

1. Balfour, G. W. and T. G. Stewart: Case of Enlarged Spleen Complicated with Ascites, Both Depending upon Varicose Dilatation and Thrombosis of the Portal Vein. Edin. Med. J., **14**:589, 1868–69.
2. Baumgarten, P. C. von: Persistent Patency of the Umbilical Vein together with a Contribution to the Problem of Banti's Disease. Arb. a. d. Geb. d. path. Anat. Inst. zu Tubing, Leipzig, **6**:93, 1907.
3. Cruveilhier, J.: Maladies des Veines. Anatomie Pathologique du Corps Humain. Vol. 1, Book 16, Pl. 6, Paris, Bailliere, 1829–35.
4. Detrie, P. and R. Martini: Spontaneous Portocaval Shunts. J. Chir. (Paris), **80**:452, 1960.
5. Foster, J. H., W. S. Stoney and H. W. Scott, Jr.: An Experimental Study of Thoracic Transposition of the Spleen: a Method of Portal Decompression. Surgery, **49**:223, 1961.
6. Gibson, J. B. and R. L. Richards: Cavernous Transformation of the Portal Vein. J. Path. and Bacter., **70**:81, 1955.
7. Gross, H.: Two Cases of Chronic Obsruction of the Portal Vein. Frankfurt Ztschr. f. Path., **34**:71, 1926.
8. Hennrich, G.: Cruveilhier-Baumgarten Disease and its Surgical Treatment with a Contribution to the Origin of Intrahepatic Portal Hypertension. Acta Hepatosplen. (Stuttgart), **8**:1, Jan.–Feb. 1961.
9. Klemperer, P.: Cavernomatous Transformation of the Portal Vein. Arch. Path., **6**:353, 1928.
10. Lamy, M., L. Jammet, C. Nezelof and C. Guibert: Splenoportal Thrombosis in Children. Arch. Franc. Pediat., **18**:182, 1961.
11. Learmonth, J. R.: On Certain Aspects of Portal Hypertension. Edin. Med. J., **58**:1, 1951.
12. Leger, L., P. Detrie and Rousset-Guyet: Omphalo-caval Anastomosis in Cruveilheir-Baumgarten Syndrome. Presse Med., **68**: 1581, Oct. 1960.
13. Leger, L. and P. E. Holmes: Cavernous Transformation of the Portal Vein. Brit. J. Surg., **48**:190, sup. 1960.
14. Lejars: Collateral Channels of the Renal Vein. Bull. Soc. Anat. de Paris, 2:504, 1888.
15. Lejars: A Case of Supplement to the Portal Circulation by the Left Renal Vein and the Vena Cava. Progress Med., 7:479, 1888.
16. Madden, J. L., J. M. Lore, Jr., F. P. Gerold and J. M. Ravid: The Pathogenesis of Ascites and a Consideration of its Treatment. Surg., Gynec. & Obst., **99**:385, 1954.
17. Mariau, A.: Anatomical Research on the Portal Vein and Particularly on the Anastomoses with the General Venous System. These de Lyon, 1893.
18. Morrison, R.: Operative Cure of Ascites due to Liver Cirrhosis (Talma-Morrison Operation). Proc. Roy. Soc. Med., **5**:37, 1912.
19. Morrison, R.: A Case of Ascites due to Liver Cirrhosis Treated by Operation. Ann. Surg., **38**:361, 1903.
20. Nylander, P. E. A. and M. Turunen: Transposition of the Spleen into the Thoracic Cavity in Cases of Portal Hypertension. Ann. Surg., **142**:954, 1955.
21. Parker, R. A. and R. M. E. Seal: Cavernous Transformation of the Portal Vein. J. Path. and Bacter., **70**:97, 1955.
22. Pegot: Varicose Tumor with Venous System Anomaly, Persistance of the Umbilical Vein and Development of Sub-cutaneous Abdominal Veins. Bull. Soc. Anat. de Paris, **8**:49, 1833.
23. Pensa: A Case of Anastomosis of the Splenic Vein to the Left Renal Vein. Boll. d. Soc. med.-chir. di Pavia, **22**:100, 1908.
24. Pick, L.: Total Hemangiomatous Obliteration of the Portal Vein. Virchows Arch. f. Path. Anat., **197**:490, 1909.
25. Rienhoff, W. F., Jr., and A. C. Woods, Jr.: Ligation of Hepatic and Splenic Arteries in Treatment of Cirrhosis with Ascites. J. A. M. A., **152**:687, 1953.
26. Ritzer, A. A., and W. B. Leach and D. M. Whitelaw: Cavernous Transformation of the Portal Vein in Polycythemia Vera. Canad. M. A. J., **81**:184, 1959.
27. Rousselot, L. M., A. Moreno and W. Panke: The Clinical and Physiopathological Significance of Self-Established (non-surgical) Portal Systemic Venous Shunts. Ann. Surg., **150**:384, 1959.
28. Sappey, C.: A Point of Anatomy Relative to the History of Cirrhosis. Mem. Acad. de Med., **23**:269, 1859.
29. Sappey, C.: Research on some Accessory Portal Veins. Compt. Rend. Soc. de Biol., **1**:3, 1860.
30. Sappey, C.: On Accessory Portal Veins. J. de L'Anat. et Physiol., **19**:517, 1883.
31. Saxer, F.: Contribution to the Pathology of the Portal Circulation. Centralbl. allg. Path., **13**:577, 1902.

32. Simmonds, J. P.: Chronic Occlusion of the Portal Vein. Arch. Surg., 33:397, 1936.
33. Talma, S.: Surgical Opening of a New Channel for the Blood of the Vena Pena Porta. Berlin klin. Wchnschr., 35:833, 1898.
34. Tuccu Tewelde, M.: Anatomy of the Abdominal Esophagus and Physiopathology of Esophageal Varices. Presse Med., 68:1697, 1960.
35. Turunen, M. and H. Laitinen: Collateral Circulation Between a Spleen Transposed into the Thoracic Cavity and the Vena Cava Superior. Ann. Surg., 149:443, 1959.
36. Tuffier, T. and Lejars: Veins of the Fatty Capsule of the Kidney. Arch. de Physiol. Norm. et Path., 3:41, 1891.
37. Umber, F.: A Contribution to Portal Vein Obstruction. Mitt. a. d. Grenzgeb. d. Med. u. Chir., 7:487, 1901.
38. Yi Yung Hsia, D. and S. Gellis: Portal Hypertension in infants and Children. Am. J. Dis. Child., 90:290, 1955.

Discussion

Dr. John Foster (Nashville): Drs. Johns and Evans were ingenious in taking advantage of this naturally occurring portal-systemic shunt which they enlarged by means of a venoplasty.

I think this case emphasizes the value of preoperative splenoportography or, in a case like this in which the spleen has already been removed, of operative portography in helping assess an individual case as regards alternate methods of treatment. Dr. Johns has certainly emphasized this in the manuscript.

This study of the portal collaterals and pathways has been of particular interest to us for several years, especially as regards the propensity of the spleen to develop collateral pathways when it is transposed into the left thoracic cavity. We did this in dogs, taking the lead from the Finnish group who first suggested it, and have been able to very clearly demonstrate the feasibility of shunting the blood from the portal system through the thoraco-splenic collaterals that form promptly after the spleen, with its pedicle intact, is placed in the left chest.

We would not like to give anyone the impression that we think this is a procedure for general application in portal hypertension but one which may find some applicability in selected cases in which it is not possible to perform a shunt procedure. (I.e., patients in whom a vein suitable for a porto-systemic is lacking, small children in whom a shunt has little chance of remaining patent, and possibly in very poor-risk patients who seem unable to tolerate a shunt operation.)