Reprinted from THE JOURNAL OF PEDIATRICS, St. Louis
Vol. 103, No. 5, PP. 741-742, November, 1983 (Printed in the U. S. A.)

Comment: Although pediatricians are not called on to make technical surgical decisions, we are frequently consulted by our patients and their families to confirm the advisability of proposed surgical intervention. Because of the complexity and the severity of portal vein obstruction in children, it is especially important that we understand the issues addressed in the two papers in this issue by Alvarez et al. and in the following commentaries. Unusually important are the issues in regard to early intervention and the long-term implications of dangerous hemorrhage and encephalopathy. Readers will quickly detect that perfect agreement has not been reached. Yet we agree with our four reviewers that the series is large, the results impressive, and the implications in reference to diagnosis and management significant. Accordingly, we hope that all physicians will find this timely, comprehensive revisitation as stimulating and as informative as we have.—R.E.M.

Portal vein thrombosis and portal diversion

THE ARTICLES BY ALVAREZ ET AL., in this issue of The Journal advocating portosystemic shunt procedures in children with portal vein thrombosis are certain to be controversial. In the United States and Canada, the consensus developed by pediatric surgeons during the last decade is that such shunt procedures are not often indicated.[1,2] It has been claimed that children with portal vein obstruction rarely bleed to death if given conservative treatment. However, the most important reason for a nonsurgical approach has been a very high incidence of shunt thrombosis in patients younger than 10 years old or if the portal vein is <10 mm in diameter.[1,2]

Articles on the subject may outnumber the patients treated, and the personal experience of some of the authors has apparently been miniscule. The largest American series, reported by Grauer and Schwartz,[3] consisted of 19 patients. It is clear from that experience that portosystemic shunts can be performed in children with little fear of thrombosis. The importance of technical proficiency is well illustrated in the experience of Alvarez et al.: more than 90% of the anastomoses apparently remained patent.

Granting that effective operations are feasible, there are physiologic consequences of portal diversion that have become clear only recently. When a portal vein is gradually occluded in animals[4] or is obstructed by thrombosis in humans,[5] the lost venous inflow to the liver is restored to a significant extent by hepatopetal (liver seeking) collateral veins. An effective portosystemic shunt "steals" this collateral flow, placing the liver in the same state as that produced in an anatomically normal patient by a completely diverting end-to-side portacaval shunt.[5] Warren et al.[5] have described two patients with portal vein thrombosis who developed hepatic encephalopathy long after portosystemic shunts were established; the encephalopathy was relieved when the shunts were taken down.

Completely diverting portacaval shunts cause wide-ranging changes in the liver of rats, dogs, swine, subhuman primates, and humans.[6] The hepatocytes undergo atrophy and hyperplasia, fatty infiltration, deglycogenation, and drastic changes in organelle structure.[6] The most specific change in the organelles is depletion (with disruption) of the rough endoplasmic reticulum. The changes are caused by the diversion around the liver of the so-called hepatotropic factors in the portal venous blood, of which insulin is the most important.[7]

See related articles, pp. 696, 703, and 742.

Because the rough endoplasmic reticulum is the "factory" of the liver cell, it is not surprising that many and probably most hepatic synthetic processes are reduced by complete portal diversion. Depressions have been described in lipid and bile acid synthesis, the Krebs-Henseleit cycle, and the hepatic microsomal mixed-function enzyme system.[8] These discoveries have shown how undesirable portosystemic shunting is, and have emphasized that complete portal diversion procedures should be done only for life-threatening indications. The 12 prophylactic portosystemic shunts in the 76 patients of the Alvarez series were not lifesaving, and may provoke unusual criticism.

Nevertheless, the data of the French group have provided convincing evidence that the benefits of a portosystemic shunt exceed the risks in patients with portal vein thrombosis and life-threatening complications of portal

hypertension. The absence of encephalopathy was striking but in accord with experience in patients in whom the reason for complete diversion was an inborn error of metabolism. The clearest evidence has been in patients with familial hypercholesterolemia. The architecture of the liver is completely normal in this disease. Several dozen patients have been studied for long periods after the creation of completely diverting end-to-side portacaval shunts, and there has been only one equivocal example of encephalopathy,[8] which was effectively treated with a low-protein diet.

The demonstration that portal diversion procedures can be done in children with a high degree of technical success and with a low incidence (zero in this experience) of consequent hepatic failure or encephalopathy may change the excessively conservative attitudes prevalent in North America.

Thomas E. Starzl, M.D., Ph.D.
Department of Surgery
University of Pittsburgh
School of Medicine
1084 Scaife Hall
Pittsburgh, PA 15261

REFERENCES

1. Clatworthy HW Jr: Extrahepatic portal hypertension. *In* Child CG, editor: Portal hypertension. Philadelphia, 1974, WB Saunders, pp 257-261.
2. Fonkalsrud EW, Myers NA, Robinson MJ: Management of extrahepatic portal hypertension in children. Ann Surg **180:**487, 1974.
3. Grauer SE, Schwartz SI: Extrahepatic portal hypertension: A retrospective analysis. Ann Surg **189:**566, 1979.
4. Bollman JL, Flock EV, Grindlay JH, Bickford RG, Lichtenheld FR: Coma with increased amino acids of brain and cerebrospinal fluid in dogs with Eck's fistula: Prevention by portal-systemic collateral circulation. Arch Surg **75:**405, 1957.
5. Warren WD, Millikan WJ Jr, Smith RB III, Rypins EB, Henderson JM, Salam AA, Hersh T, Galambos JT, Faraj BA: Noncirrhotic portal vein thrombosis: Physiology before and after shunts. Ann Surg **192:**341, 1980.
6. Putnam CW, Porter KA, Starzl TE: Hepatic encephalopathy and light and electron micrographic changes of the babboon liver after portal diversion. Ann Surg **184:**155, 1976.
7. Starzl TE, Porter KA, Watanabe K, Putnam CW: The effects of insulin, glucagon and insulin/glucagon infusions upon liver morphology and cell division after complete portacaval shunt in dogs. Lancet **1:**821, 1976.
8. Starzl TE, Porter KA, Francavilla A: The Eck fistula in animals and humans. Curr Probl Surg (In press.)