Leading article

Extra hepatic portal venous obstruction

The considerable diagnostic and therapeutic advances in portal hypertension during the last decade have rekindled interest in this subject throughout the world. In Western countries cirrhosis is present in over 90% of adults bleeding from oesophageal varices, but in children and also in the Third World non-cirrhotic causes of portal hypertension are much more common. On a world wide basis hepatic fibrosis as a result of Schistosomiasis is arguably the most common non-cirrhotic disorder while other rarer causes include Budd Chiari Syndrome, veno occlusive disease, agnogenic myeloid metaplasia, congenital hepatic fibrosis, partial nodular transformation and idiopathic portal hypertension as well as a variety of drugs and toxins. To this long list of prehepatic and intrahepatic causes must be added obstruction to the portal venous system.

The most common abnormality consists of a thrombus surrounded by a cavernomatous leach of collateral vessels; early reports suggested that the primary pathological process was an angiomatous malformation but it is now generally accepted that these vessels are collateral veins which form to bypass the block. In other cases surgery or autopsy reveals a hypoplastic fibrous stricture of the portal vein to be the cause of the portal hypertension. Whether this represents the final stage of the body response to a thrombus or a completely different pathological process is uncertain. The causes of such obstruction are numerous (see Table) and the site is important as it may have profound implications with regard to management. The obstruction itself may be confined to a segment of the portal vein or it may involve all the major veins, often with extension into the liver. Abdominal injuries are well recognised causes of both splenic and portal vein thrombosis while the latter can easily result from difficult or careless biliary tract surgery. Isolated occlusion of the splenic vein occasionally arises as a sequel to severe pancreatitis either because of thrombosis alone or pressure from a pancreatic pseudocyst. Although uncommon, this is important to recognise as without proper investigation alcoholics bleeding from oesophageal varices with this condition may be erroneously diagnosed as having cirrhosis. Most patients with portal hypertension caused by extra hepatic venous obstruction have a much more favourable prognosis than those with cirrhosis because overall liver function is essentially normal. Notable exceptions to this occur when tumours encroach upon or occlude the splenic or portal vein. Such events almost invariably lead to rapid onset of ascites, bleeding varices and to downhill progression to death within a few weeks or months. Portal vein thrombosis may arise as a complication of cirrhosis: it is generally considered to be uncommon although a recent report from Italy claims an incidence of 16%. The hepatic veins appear to be more susceptible to thrombotic occlusion secondary to haematological disorders, pregnancy or to the oral contraceptive, but cases involving the portal venous system have also been recorded.

Portal vein occlusion in children has traditionally been considered to arise
as a complication of umbilical sepsis but several observations lead one to question the importance of this factor. First, sepsis caused by omphalitis or intra-abdominal sepsis has been documented in only a minority of cases. Furthermore a prospective study of neonates undergoing umbilical vein catheterisation found no clinical cases of venous occlusion after this procedure. Second, a high incidence of congenital abnormalities has been reported in children with this condition raising the possibility that the portal venous abnormalities might be congenital in origin in some cases.

Another intriguing aspect of portal vein occlusion is its high prevalence in India, where the condition is responsible for 20–30% of all cases of variceal haemorrhage. Although neonatal dehydration and sepsis may play a greater role in the subcontinent compared with the West this is not supported by clinical observations. Whether or not there is a single pathological entity which is distinct from the disorders seen elsewhere in the world is unknown. A possible environmental factor, perhaps akin to that postulated in the same country for non-cirrhotic portal fibrosis may exist; there is clearly scope for carefully detailed research both in India and in other countries in the Third World where we have much less information on the aetiology of portal hypertension in children and young adults.

Acute uncomplicated obstruction of the portal vein presents with sudden onset of ascites, which tends to resolve spontaneously as a collateral circulation develops to bypass the block. After this the patients remain symptom free for a variable period of time until the onset of other features of portal hypertension. In young patients the acute symptoms are often absent and 80% present for the first time with variceal bleeding. Gross splenomegaly is a very common feature and the physical discomfort of this large organ is the only significant clinical complaint. Liver biochemical changes, when present, are trivial but hypersplenism is common and often severe enough to lead to pronounced anaemia. Although ascites was frequently reported in early series it has not been a feature in later reports although it may occur as a sign of decompensation after gastrointestinal bleeding. The presence of oesophageal varices in a patient with normal liver biochemistry and histology should alert the clinician to the possibility of the diagnosis. Quiescent macronodular cirrhosis may produce a similar picture, so confirmation should be established by radiological means. Splenoportovenography or selective coeliac angiography is unquestionably the investigation of choice as both techniques produce images of the portal venous

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Table Causes of extrahepatic portal venous obstruction
system of sufficiently high quality to permit identification of the site of occlusion. Despite the thrombocytopenia associated with hypersplenism bleeding problems rarely occur in these patients because the platelets, although few in number, function adequately and a normal bleeding time is arguably a more discriminatory test than a quantitative platelet count. Ultrasound is a reliable non-invasive alternative but may be unreliable in cases with lesser degrees of obstruction.11

The treatment of portal venous obstruction depends very much upon the age of the patient, the site and nature of the obstruction, and the clinical features. Variceal haemorrhage is the major complication requiring treatment. Patients with portal hypertension who have not bled from varices should in general be left well alone as bleeding may not occur for many years and premature interference may do more harm than good. Indeed as documented by Dilawari and Chawla in this issue (page 1198) many patients appear to protect themselves from bleeding by the opening up of spontaneous internal shunts. In general hypersplenism is not an indication for surgical intervention, but in occasional patients symptoms of anaemia or physical discomfort caused by splenomegaly may be sufficiently severe to merit splenectomy.

Once variceal bleeding has occurred some form of therapy should be considered. If the obstruction is confined to the splenic vein (sinistral hypertension12) splenectomy alone is sufficient as this relatively simple procedure is curative and can be carried out in children and adults alike. The problem is not so straightforward when the portal vein is involved. Webb and Sherlock13 in a retrospective analysis of extra hepatic portal obstruction observed a high incidence of rebleeding, encephalopathy, and mortality among patients subjected to portal decompression surgery. On the basis of these results they advised against surgical intervention, but they did not distinguish between children and adults, and a previous study from the same unit14 showed that hepatic function in extra hepatic portal obstruction deteriorates significantly with increasing age of the patient. In addition, most operations were done in peripheral hospitals by surgeons with relatively little experience of the procedure. By contrast surgeons from specialist units in Paris15 and New Delhi16 report a remarkably low incidence of both rebleeding and encephalopathy in children and young adults. The French group claim a postoperative shunt patency of over 90% with no recurrent haemorrhage after a mean follow up of three and a half years. They report successful shunt operations in children as young as 18 months old and claim to be able to anastomose veins of as little as 4mm in diameter.17 Both mesocaval and proximal lienorenal shunt with splenectomy appear equally successful, while the latter also deals with any problems of hypersplenism. Many patients, however, with hypersplenism undergoing successful decompression shunt surgery alone with preservation of the spleen experience significant haematological improvement with regression of spleen size. Clinically significant postoperative encephalopathy is also rare in the French and Indian experience. These patients clearly differ from well compensated cirrhotics because, as the Chandigarh workers show (p. 1198 this issue) they are able to tolerate large spontaneous intrahepatic shunts without difficulty. It should be emphasised, however, that these encouraging results refer to relatively short periods of follow up in young individuals. A similarly favourable short term prognosis has been observed
with non-cirrhotic portal hypertension, but a 30 year follow up has revealed a substantial incidence of late encephalopathy.

The recent widespread use of endoscopic sclerotherapy has added a further therapeutic option. It is clearly the treatment of choice in infants whose vessels are too small for surgical anastomosis and indeed as long ago as 1965 Gibson and colleagues reviewed the published experience of this procedure although at the time it was considered merely as a temporary expedient until the children were big enough for surgery. It is also clearly indicated in those in whom extensive thrombosis of the portal venous system precludes any operative procedure. Elderly patients and those with other major systemic disorders are probably best treated by sclerotherapy while there is usually no alternative for those who rebleed after a failed shunt procedure. There is clearly a case to be made for sclerotherapy in the young otherwise healthy patients although many of them may find the prospect of regular endoscopy with periodic injection for the rest of their lives tedious as well as economically burdensome. Such individuals might prefer a once and for all surgical procedure particularly if they can be guaranteed a good chance of success and no significant longterm sequelae. With our current knowledge, however, it is doubtful whether we are in a position to give such reassurances.

In my view a patient with suspected portal venous obstruction should be fully investigated and the diagnosis of cirrhosis definitively excluded as this has major implications with regard to prognosis. Splenectomy should not be recommended unless it can be clearly shown that the enlarged organ is causing physical symptoms. Oesophageal varices should be left well alone unless and until bleeding occurs. In the event of variceal haemorrhage I would favour endoscopic sclerotherapy irrespective of the age of the patient, reserving shunt surgery for those in whom endoscopic treatment is unacceptable for either technical, administrative or financial reasons. Having opted for surgery, I would choose the surgeon with care as wide experience of this type of surgery is shared by relatively few today. The choice between surgery and sclerotherapy is perhaps more emotional than scientific; those seeking evidence from a prospective controlled trial should remember that the results of such a study are only likely to be realised long after the life of the investigator!

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