PORTAL HYPERTENSION IN CHILDHOOD*

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Portal hypertension in children, as in adults, is usually subdivided into extrahepatic (prehepatic), intrahepatic and suprahepatic. Since the introduction of techniques of estimating intrasplenic pulp pressure and wedged hepatic vein pressure, further classification into presinusoidal and postsinusoidal obstruction has been possible. In presinusoidal obstruction the intrasplenic pressure is raised, while the wedged hepatic vein pressure is normal. In postsinusoidal obstruction both pressures are raised.

AETIOLOGY

During the past 8 years we have admitted 35 children suffering from portal hypertension with proven varices to the Red Cross War Memorial Children's Hospital. Extrahepatic obstruction accounted for 7, intrahepatic for 21 and suprahepatic for 7. This distribution is contrary to experience in most surgical clinics (Table I) which report a marked preponderance of extrahepatic obstructions and, indeed, in our own series 7 of 12 cases referred to the surgical department suffered from extrahepatic portal obstruction. On the other hand, comparison of our figures with data from Sheilla Sherlock's publications[20-22] show a close correlation, i.e. extrahepatic obstruction is responsible for about one-fifth of the cases. It is clear that most surgical series represent selected cases.

TABLE I. 'SURGICAL' SERIES

	Extrahepatic	*Intrahepatic*
Schumaker and King (1952)[18] ..	18	3
Milnes Walker (1962)[24]	28	22
Boles and Clatworthy (1962)[1] ..	12	23
Foster *et al.* (1964)[7]	17	6
Lynn (1964)[14]	31	9
Present series (1965)	7	5
Total	113	68

Table II reflects the various causes of portal hypertension in our series and these are compared with data from Sherlock's writings.

Presinusoidal Obstructions

Cavernomatous transformation of the portal vein is clearly a frequent cause of portal hypertension in childhood. It is a sequel of portal phlebitis and there was a history of umbilical sepsis in 3 of our 6 cases and in 9 of Sherlock's 16 cases.[20] Two of Sherlock's patients had received an exchange transfusion and others have also recorded portal vein thrombosis complicating this procedure, but we have not yet had such a case. In one of our patients appendicitis was probably responsible and in two no definite cause could be ascertained. The ages of the children ranged between 2 and 15 years.

Helminthic infection is an important cause of portal hypertension in Africa. The responsible parasite is usually *Schistosoma mansoni*,[23] which is not encountered in the Western Cape, and our case was due to *Fasciola hepatica.*

Congenital hepatic fibrosis is a condition which has been recognized with increasing frequency in recent years. It is of particular importance to surgeons because it presents with portal hypertension, well-preserved hepatic function and a patent portal vein. Our patients were aged 3 years and 11 years. The ages of Sherlock's patients ranged from 2½ to 16 years,[21] and Boley *et al.*[2] have found a familial history in a third of the cases reported.

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TABLE II. AETIOLOGY OF PORTAL HYPERTENSION

	Present series (35 cases)		*Data from Sherlock (112 cases)*	
	Total	*Haemorrhage*	*Total*	*Haemorrhage*
Presinusoidal obstruction				
Extrahepatic				
Traumatic ..	1	–	–	–
Cavernomatous transformation	6	6	16	14
Intrahepatic				
Congenital hepatic fibrosis.. ..	2	1	15	5
Leukaemia ..	–	–	?	?
Helminthic infection	1	1	–	–
Totals ..	10	8	31	19
Postsinusoidal obstruction				
Intrahepatic				
Post-hepatitis ..	6	4	11	1
Neonatal hepatitis	4	–	5	–
Lupoid	–	–	33	8
Wilson's disease..	–	–	7	3
Biliary atresia ..	2	–	5	–
Galactosaemia ..	1	–	–	–
Mucoviscidosis ..	1	–	1	1
Cryptogenic cirrhosis	4	1	16	3
Suprahepatic				
Veno-occlusive disease.. ..	5	1	–	–
Vena caval thrombosis	2	–	–	–
Constrictive pericarditis ..	–	–	3	–
Totals ..	25	6	81	16

Postsinusoidal Obstruction

Of the numerous cases of cirrhosis in childhood, acute viral hepatitis was most commonly responsible for cirrhosis with varices in our series. This included hepatitis in older children as well as neonatal giant-cell hepatitis. In the latter the portal hypertension did not become evident until the age of 3 years or over. Although we have had cases of *lupoid cirrhosis* and *Wilson's disease* with portal hypertension in adolescents, we have not encountered a case in childhood. *Biliary atresia* is a fairly common cause of portal hypertension, but most of our patients succumbed at home before there was evidence of varices, and therefore only two have been included in the series.

We have had an unusually large proportion of cases caused by *veno-occlusive disease*. These children have come from wheat farms, and the disease presumably resulted from phlebitis of the hepatic vein radicles owing to senecio poisoning, although in one case a congenital membrane of the hepatic vein was apparently present. *Vena caval thrombosis* was responsible for two of our cases, but we have not had any cases of constrictive pericarditis presenting with varices.

CLINICAL PRESENTATION

Whatever the site and cause of portal obstruction, the principal effect is back-pressure on the various tributaries of the portal venous system with congestion of the spleen and enlargement of collateral venous channels at the sites of communication between portal and systemic venous systems, particularly in the lower oesophagus and upper stomach. In addition the patients are liable to septicaemia by Gram-negative organisms and to neuropsychiatric changes, and suffer from the effects of the primary disorder, which is of obvious importance in patients with cirrhosis or veno-occlusive disease.

The clinical presentation depends upon the complications of these effects (Table III):

1. Bleeding from oesophageal or gastric varices.
2. Anaemia or cytopenia owing to the enlarged and overactive spleen.
3. The development of ascites.
4. Liver failure and failure in general health.
5. Jaundice from the primary cause (e.g. biliary atresia) or secondary to hepatic necrosis.

Any one of these complications may dominate the clinical picture, but often several are present in a particular case.

TABLE III. PRESENTATION

	Presinusoidal obstruction		*Postsinusoidal obstruction*		*Total*
	Extrahepatic	*Intrahepatic*	*Intrahepatic*	*Suprahepatic*	
Total cases	7	3	18	7	35
Bleeding varices ..	5	2	1	1	9
Hypersplenism ..	1	1	4	–	6
Ascites	1	–	6	5	12
Liver failure ..	–	–	4	1	5
Jaundice	–	–	3	–	3

Haemorrhage is much more common in cases with presinusoidal obstruction. In our series 7 of the 9 patients who presented with bleeding varices suffered from presinusoidal obstruction. Children with postsinusoidal obstructions usually presented with ascites and/or evidence of impaired liver function, and when bleeding did occur it often aggravated the ascites or precipitated liver failure.

Further analysis of our cases shows that bleeding at some time during the course of the child's illness occurred in 80% of cases with presinusoidal obstruction and only in 20% of those with postsinusoidal obstruction (Table II). The same applies to Sherlock's cases, and from this it will be seen that if a child bleeds from varices the chances of presinusoidal and postsinusoidal obstruction are about even, despite the fact that the latter is a much more frequent cause of portal hypertension. Furthermore, the child with postsinusoidal obstruction often has such severe impairment of liver function that surgical aid is not sought. Indeed, in our series 11 of the 25 patients with postsinusoidal obstruction have died without surgical intervention. It is therefore not surprising that reports from surgical clinics reflect a preponderance of extrahepatic obstructions.

DIAGNOSIS

The diagnosis of portal hypertension may be confirmed by the following:

1. Enlargement of collaterals. Distended superficial abdominal veins are the commonest and are best demonstrated with infra-red photography. A *caput medusae*, if present, is obvious but does not occur in cavernomatous transformation of the portal vein because the obstruction is distal to the entry of the left umbilical vein. A large azygos vein may be visible on plain chest radiographs and so may aneurysmal dilatation of the hemiazygos.[22]

2. The presence of oesophageal and/or gastric varices (Fig. 1). These may be demonstrated by radiography, but are best detected on oesophagoscopy[22] which, however, is dangerous in small children.

3. Congestive splenomegaly with raised intrasplenic pulp pressure on manometry. The normal intrasplenic pressure is 10 ± 2 mm.Hg and the mean pressure in children with portal hypertension varies from 14 to 23 mm.Hg.[3,19,20] It tends to be somewhat higher in extrahepatic than in intrahepatic obstructions. Splenic puncture should not be performed in jaundiced patients nor when the prothrombin index is below 60% and the platelet count below 100,000/cu.mm.[20,22]

4. Splenic venography (Figs. 2 and 3). This is usually performed at the time when the splenic pulp pressure is measured. In portal hypertension the venograms demonstrate enlargement of the portal radicles and the presence of collaterals including oesophageal varices and, in addition, the site of obstruction[19] (see below).

The diagnosis of the cause and site of the portal hypertension is made on the following grounds:

1. *The symptoms and signs.* A careful history and meticulous clinical examination combined with laboratory tests of liver function will often be sufficient to determine the cause of portal hypertension. Particular attention should be paid to enlargement of the liver and spleen, the presence of ascites and/or jaundice, evidence of hypersplenism and impairment of liver function.

A history of umbilical or intra-abdominal sepsis, splenomegaly without hepatic enlargement or impairment of liver function, suggests extrahepatic obstruction owing to portal vein thrombosis.

Hepatosplenomegaly suggests hepatic disease. If there is no derangement of liver function, congenital hepatic fibrosis should be suspected. If there is impairment of liver function the most probable cause is cirrhosis and the cause may be determined from the history and a battery of liver-function tests.

Oedema of the lower limbs, enlarged superficial abdominal veins and gross ascites in addition to hepatomegaly, clearly raise the suspicion of veno-occlusive disease.

2. *Liver biopsy.* A normal histological picture usually rules out cirrhosis as a cause, while various patterns of liver-cell damage, fibrosis and cellular infiltration will often give the clue to the cause of intrahepatic obstructions. Only slight increase in portal connective tissue with venous congestion suggests veno-occlusive disease.

3. *Splenic venography.* This investigation is most useful for differentiating between extra- and intrahepatic obstructions (Figs. 2 and 3). However, it cannot distinguish primary from secondary portal vein thrombosis, nor presinusoidal from postsinusoidal intrahepatic obstruction nor, indeed, intrahepatic from suprahepatic obstruction.

4. *Wedged hepatic vein pressure.* This investigation will determine whether the obstruction is presinusoidal or postsinusoidal. The normal wedged pressure is 7± 2 mm.Hg with a gradient of 3·6 ± 1 mm.Hg between free and wedged pressures.[3,19-21] In postsinusoidal obstructions it is raised up to 32 mm.Hg with gradients up to 23.[3] In presinusoidal obstructions it approximates the normal. In children the investigation is of particular value for the following:

(*a*) Diagnosis of the site of obstruction in splenectomized patients.

(*b*) Differentiation of congenital hepatic fibrosis from cirrhosis.

(*c*) Diagnosis of secondary portal vein thrombosis in patients with cirrhosis.

5. *Inferior vena caval venography* is of value in cases of suprahepatic obstruction to detect the site of the block and the extent of collaterals (Fig. 4).

INDICATIONS FOR SURGERY

1. *Haemorrhage from varices* is accepted as the principal indication and some would regard it as the only indication for surgery. Children who *present* with haemorrhage are, therefore, usually referred directly to surgical clinics. In our series haemorrhage was the indication for surgery in 7 cases and 5 of them had extrahepatic obstruction.

2. *Hypersplenism.* Splenomegaly with neutropenia, thrombocytopenia, or any important degree of anaemia is considered by some as a clear indication for surgery. We feel that the risks of surgery outweigh the risks of hypersplenism *per se* and we recommend surgery only in cases with associated proven varices or evidence of occult haemorrhage.

3. *Large varices.* A case may be made for surgery in patients with portal hypertension in whom large varices are demonstrable, but who have not yet bled. This applies particularly to patients with cirrhosis and marginal liver function in whom the risks of surgery appear to be less than that of a single episode of haemorrhage which may precipitate hepatic coma and death. In patients with presinusoidal obstruction or mild cirrhosis, on the other hand, it is probably better to await events because the risks of haemorrhage are not great if transfusion facilities are available.

4. *Nutritional failure.* Failure to thrive *per se* can hardly be regarded as an indication for surgery. However, if

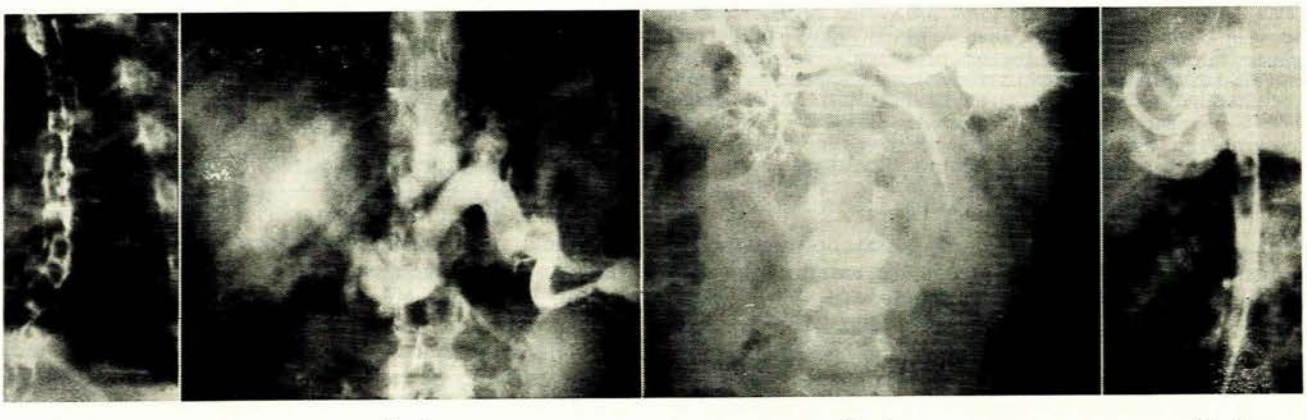

Fig. 1 *Fig. 2* *Fig. 3* *Fig. 4*

Fig. 1. Oesophageal varices in a case of extrahepatic portal obstruction. *Fig. 2.* Splenic venogram showing cavernomatous transformation of the portal vein. Note the gastric varices. *Fig. 3.* Splenic venogram showing patent portal vein and large collaterals. *Fig. 4.* Venogram showing obstruction of the inferior vena cava at the point of entry of the hepatic vein.

varices are present and there is evidence of occult haemorrhage, surgery may be justified.

5. *Ascites*. The earliest attempts at surgical treatment in portal hypertension were directed at the relief of ascites. Then the pendulum swung to the opposite extreme and ascites was regarded as a contraindication to surgery. There are some patients, however, in whom ascites persists in spite of improvement in liver function, and surgery may be beneficial in these cases.

TIMING OF SURGERY

The decision to operate depends upon a number of factors:

1. *The condition of the liver*. Clinical evidence of liver failure, gross ascites and biochemical evidence of advanced liver disease are obvious contraindications to surgical therapy. It is generally agreed that a reversal of the serum albumin-globulin ratio, a serum albumin level below 3 G/100 ml. a serum bilirubin level above 3 mg./100 ml. and a prothrombin index below 70% are contraindications to immediate surgery.[22] On the other hand, marginal liver function in a child with large varices may justify prophylactic surgery because the risks of surgery are less than those of a large haemorrhage which may precipitate liver failure.

2. *The age of the child*. Surgical procedures in children under the age of 4 years are mostly doomed to failure and should be avoided if at all possible. In children between the ages of 4 and 10 years there is still a good chance of failure, and surgery should be undertaken only if haemorrhage is liable to precipitate liver failure or becomes uncontrollable. Shaldon and Sherlock[20] point out that the threat of exsanguinating haemorrhage in cases of extrahepatic obstruction is over-emphasized. Episodes of bleeding may become less frequent as time allows for opening up of collateral vessels to renal and lumbar veins with a drop in portal pressure. Furthermore, the most striking feature of haemorrhage secondary to portal hypertension in a child is the frequency with which it can be controlled by conservative measures.[7] Sengstaken tubes are only very rarely required and emergency surgery is hardly ever necessary.[1,5,7,20] Only 1 of our cases, a boy of 14 years with suprahepatic obstruction, required ligation of varices for uncontrollable haemorrhage.

3. *The site of the obstruction*. The absence of a portal vein in patients with extrahepatic obstruction renders surgery more difficult and less successful. At the same time preservation of good liver function enables the child to withstand a large haemorrhage very well. It is therefore wise to postpone surgical intervention for as long as possible and certainly beyond the age of 10 years.

In intrahepatic obstructions a large portal vein is usually present and therefore surgery can be undertaken in younger patients, i.e. from 4 to 5 years onwards. In suprahepatic obstructions surgery is difficult and dangerous and should be undertaken only for uncontrollable exsanguinating haemorrhage.

SURGICAL PROCEDURES AND RESULTS

The procedures may be divided into 3 categories, viz., decompression of the portal bed, obliteration of the varices, and elimination of the varices. These are shown in Table IV, but only those procedures which are regarded as worth while will be discussed (Table V).

TABLE IV. SURGICAL PROCEDURES IN PORTAL HYPERTENSION
(Present series: 15 operations in 12 patients)

Decompression of varices
- *Reduction of Splenic Circulation:*
  - Ligation of splenic artery
  - Splenectomy (6)
- *Direct Shunts:*
  - Portacaval (2)
  - Spleno-renal (2)
  - Mesenterico-caval
  - Makeshift (2)
- *Indirect Shunts:*
  - Supradiaphragmatic transposition of spleen
  - Omentocavopexy
  - Thoracic subcutaneous ileopexy
  - Poudrage

Obliteration of varices
- Ligation of varices (1)
- Gastric transection
- Oesophageal transection (1)

Elimination of varices
- *Oesophago-gastrectomy with direct anastomosis:*
  - Oesophago-gastrostomy
  - Oesophago-jejunostomy
- *Oesophago-gastrectomy with interposition:*
  - Jejunum
  - Colon (1)

TABLE V. RESULTS OF 15 OPERATIONS ON 12 PATIENTS

Operation	*Type of lesion*	*Indication*	*Age at operation (years)*	*Follow-up*	*Result*
Splenectomy	Extrahepatic	Haemorrhage	4	6 months	Recurrent bleeding. Re-operation
	Extrahepatic	Haemorrhage	2	6 months	Recurrent bleeding. Re-operation
	Extrahepatic	Hypersplenism	14	6 months	Well
	Cirrhosis	Haemorrhage	14	2 months	Recurrent bleeding. Re-operation
	Cirrhosis	Hypersplenism (large varices)	15	3 years	Well
	Cirrhosis	Large varices (hypersplenism)	10	4 years	Well
Portacaval shunt	Primary hepatic fibrosis	Hypersplenism (large varices)	11	2 years	Well
	Cirrhosis	Haemorrhage after splenectomy	14½	6 years	Well
Spleno-renal shunt	Extrahepatic	Haemorrhage	9	3 years	Well
	Extrahepatic	Haemorrhage	13	8 years	Well
Makeshift shunts	Extrahepatic	Haemorrhage	15	4 years	Recurrent bleeding.
	Supra-hepatic	Nutritional failure	2	6 months	Died
Ligation of varices	Supra-hepatic	Haemorrhage	15	3 years	No bleeding
Oesophageal transection	Extrahepatic	Haemorrhage after splenectomy	2½	2 years	Well
Oesophago-gastrectomy and colonic interposition	Extrahepatic	Haemorrhage after splenectomy	4½	6 months	Well

Decompression of the Portal Bed

A. Splenectomy. It has been estimated that splenectomy reduces the portal flow by 20 - 35%, and in the past this has been the basis of the procedure in cases of portal hypertension.[7] However, there seems to be little dispute today that splenectomy alone fails to reduce the portal pressure excepting in the rare case of peripheral splenic vein thrombosis with varices.[8] The operation may have a place in cases of severe hypersplenism, but then it should

rather be combined with spleno-renal shunt or at least with ligation of the varices.

In our series splenectomy was performed in 6 patients. In 3 the operation was done for bleeding varices and all of them bled again and required a second operation within 6 months. This is in keeping with the experience of others and it is agreed today that the operation has hardly any place in the treatment of bleeding varices especially if the obstruction is extrahepatic.[7] Indeed, splenectomy alone is contraindicated because in the first place it destroys a large number of natural porta-systemic shunts between the splenic vein and the renal, azygos, caval and other retroperitoneal veins and, secondly, it eliminates the possibility of a subsequent spleno-renal shunt.

In 2 patients with posthepatitic cirrhosis the operation was done for hypersplenism and varices with occult blood in the stools. There have been no untoward symptoms after 3 and 4 years respectively, but it might be argued that these children would have remained well without surgery. Occasional successes have been reported by others when splenectomy was done for hypersplenism in mild or moderate cirrhotics.

Splenectomy was performed in 1 patient who developed peripheral splenic vein thrombosis with varices following trauma. He has remained well for 6 months and a good long-term result can be predicted.

B. Portal-systemic shunts. The best method of decompressing the hypertensive portal system is to shunt the portal blood into the systemic venous system. The small size of the veins in children, however, poses a special problem because of the risk of postoperative thrombosis of the shunt. It is agreed that any type of shunt in children under the age of 4 or 5 years is doomed to failure and therefore contraindicated in this age group.[1,5,20] Indeed, many authors question the efficacy of shunts in children under the ages of 10 - 12 years.[10,20]

A recent report by Meiss[16] suggests that shunting procedures provide adequate protection against recurrent haemorrhage only if combined with ligation of varices. Others, however, claim good results in older children[1,5,6,10] and this is borne out by our limited experience.

1. *Portacaval shunt* provides the maximal stomal size and the most effective portal decompression.[7] Of all the types of shunt it is the least likely to thrombose. Unfortunately the procedure is not possible in the great majority of children who come to surgery because so often the obstruction is extrahepatic and the portal vein unsuitable for anastomosis. However, Hunt[9] reports 3 cases with extrahepatic obstruction in whom clot was scraped from the portal vein and a successful anastomosis made. Shaldon and Sherlock[20] mention that in 5 out of 16 patients with extrahepatic obstruction the thrombus occupied only the hepatic end of the portal vein and a portacaval shunt was thus feasible.

The ideal case for portacaval shunt is the patient with presinusoidal obstruction owing to congenital fibrosis of the liver, because liver function is preserved and the portal vein is patent. In such cases the traditional end-to-side anastomosis is effective.[2] We have performed the operation on one such patient and he has remained well and free from bleeding for 2 years.

The operation is also suitable for cases of posthepatitic cirrhosis, but in most cases the disease of the liver dominates the picture and precludes surgery. A side-to-side anastomosis is preferable because the obstruction is postsinusoidal and the portal vein may be functioning as an avenue of egress for blood entering the liver.[13] A successful shunt will protect the patient from haemorrhage and its disastrous effects on the diseased liver, and in children postoperative encephalopathy is not a problem.[5,24] Therefore prophylactic shunts in patients with mild cirrhosis and varices may be worth while.[1,5]

We have done a portacaval shunt in a boy of 14 years with mild posthepatitic cirrhosis who continued bleeding after splenectomy. He has remained well and free from further haemorrhage for 6 years. Milnes Walker[24] reports on 22 children with intrahepatic obstruction who were referred for surgery. Portacaval shunts were performed on 15 without mortality, but 4 have subsequently died of liver failure. He concludes that the operation should be considered in patients with cirrhosis if liver function is not deteriorating and there is a risk of serious haemorrhage from varices. Clatworthy has found that cases with posthepatitic cirrhosis are more suitable for surgery than those with biliary cirrhosis.[1,5] Bross and Slowikowsky[4] report recurrent bleeding in 20 - 25% of cases.

2. *Spleno-renal shunts* provide a less effective decompression of the portal bed and are liable to thrombose particularly in children under the age of 10 - 12 years.[10] The central type of shunt described by Clatworthy[1,5] seems to have the best chance of remaining patent and he has achieved success in children as young as 4 years. In cases of extrahepatic obstruction this procedure is probably the best that can be offered, and some authors claim long-term patency of the shunts in well over 50% of cases.[8] Clatworthy prefers the procedure also for cases of intrahepatic obstruction owing to cirrhosis, and he claims successful results with no further bleeding in 5 centrally placed spleno-renal shunts in children varying in age from 4 to 11 years and followed-up for 7 months to $3\frac{1}{2}$ years.[1,5] Foster *et al.*[7] report on 12 spleno-renal shunts of which 7 have remained patent for periods of 1 - 12 years.

We have performed the operation in 2 children with extrahepatic obstruction and both have remained well and free from bleeding for long periods. This agrees with Clatworthy's experience, but others have had disappointing results,[9] and it should be remembered that thrombosis of the shunt may lead to loss of the kidney as well.

3. *Mesenterico-caval shunts* have come into vogue in recent years and the initial reports are promising.[6,23] Success of this shunt depends primarily on a normal superior mesenteric vein free of active thrombotic disease. The procedure is simpler than portacaval or spleno-renal shunts because vascular plexuses and collaterals are not so well developed in this region. It provides a wide anastomosis, does not exclude all the portal blood-flow to the liver and does not affect splenic or hepatic function.[6]

Bross and Slowikowsky,[4] who have modified the procedure by using the left common iliac vein, have found that it can be used in children as young as 4 years. They report on 12 cases with extrahepatic portal obstruction treated by this method. One died of postoperative wound

disruption, but the remaining 11 have remained well and free from bleeding for follow-up periods of 4 months to 3½ years. Significantly, none of their patients have been troubled with oedema or functional impairment of the lower limbs, and Voorhees and Blakemore[23] have found that in 26 patients of all ages oedema was only a problem in patients with cirrhosis and hypoproteinaemia. Boles and Clatworthy[1] have had no trouble with oedema in children, but 2 out of 5 cases have bled subsequently. Foster *et al.*[7] have had no rebleeding in 3 children after follow-up periods of 1 - 8 years, but Paltia and Sulamaa[17] report recurrent bleeding in 3 out of 4 children treated by superior mesenteric to vena cava shunts.

We have not yet used this procedure in a child and it is our belief that it should be reserved for children in whom the portal vein is occluded and spleno-renal shunts impossible because of the small size of the vessels (i.e. before the age of 10 years) or previous splenectomy.

4. *Makeshift shunts* between smaller radicles of the portal system and the vena cava or its tributaries are almost invariably doomed to failure and are not recommended. Much the same applies to *indirect shunts*, although recent reports suggest that transposition of the spleen into the thoracic cavity may be useful, particularly if combined with ligation of the varices.[14,17] Thoracic subcutaneous ileopexy is still an experimental procedure and omentocavapexy is unlikely to produce long-term relief.[17]

Obliteration of the Varices

A direct attack on the varices by suture ligation or porta-azygos disconnection is a valuable method of dealing with severe bleeding varices that fail to respond to conservative measures. Most authors are agreed that prolonged freedom from bleeding cannot be expected.

(a) *Transoesophageal ligation* of the varices is the simplest method of controlling bleeding varices and is especially valuable as an emergency procedure in children. It is recommended by Clatworthy,[1,5,6] and Foster *et al.*[7] report freedom from recurrent haemorrhage for periods up to 30 months. The operation was done on one of our patients—a boy of 15 years who bled profusely from varices secondary to suprahepatic obstruction. He has remained free from haemorrhage for 3 years.

The procedure has also proved of value as an elective operation for bleeding varices in children who are too small for a shunt[7,20] and in older children with extrahepatic obstruction who have had a previous splenectomy.[1,5] Paltia and Sulamaa[17] report on 23 cases ranging in age from 6 months to 9 years who were treated by ligation of varices, the procedure being repeated several times in 5 patients. Seventeen (75%) remained symptom-free for periods up to 9 years. On the other hand, Meiss[16] has found that ligation alone affords no protection against recurrent haemorrhages.

Ligation of varices is claimed to be of particular value when used in conjunction with some other procedure. Meiss[16] has found that various types of porto-systemic shunts provide adequate protection against haemorrhage only if combined with ligation of varices. Paltia and Sulamaa[17] claim good results with transposition of the spleen into the thorax combined with ligation of varices. Foster *et al.*[7] cite the case of an 8-year-old child who continued bleeding despite a patent portacaval shunt and in whom subsequent ligation of the varices resulted in freedom from bleeding for a follow-up period of 5 years.

(b) *Oesophageal transection and resuture* is an alternative method of obliterating the varices and has at least the theoretical advantage of completely dividing all the varices entering the oesophagus. On the other hand, there is an increased risk of suture leakage. Milnes Walker[24] has performed the operation on 14 children with extrahepatic obstruction with no operative mortality. One died 4 years later of haemorrhage, 4 bled again during a follow-up period of 2 - 10 years and 9 remained free from bleeding during a follow-up period of 1 - 10 years. The procedure was carried out in one of our patients—a boy of 2½ years who had recurrent haemorrhage after a previous splenectomy, and he has remained free from bleeding for 2 years.

(c) *Proximal gastric transection* has been used with success as a method of obliterating varices in adults, and Hunt[9] reports a fair degree of success in children with extrahepatic portal obstruction. He performed the operation in 9 patients and 4 were successful for follow-up periods of 2 - 7½ years although the children were underweight and liable to dyspepsia. One case was a failure because of severe reflux oesophagitis and 4 bled again after periods ranging from 13 to 38 months after surgery. In cirrhotics the operation is often exceptionally difficult because of dense adhesions, and it is not recommended in children.[9]

Elimination of the Varices

(a) *Resection of the varix-bearing area*, viz. lower two-thirds of oesophagus and upper third of the stomach, has been used for control of recurrent haemorrhage in 'unshuntable' patients, but has many disadvantages. Firstly, major gastric resection is not desirable in small children because of the nutritional failure that follows.[12] Secondly, the sacrifice of the oesophago-gastric junction in children with active gastric secretion frequently results in peptic oesophagitis and consequent stricture formation, although Foster *et al.*[7] have found that this is greatly reduced by combining the operation with vagotomy and pyloroplasty. Finally, the operation is followed by a high immediate postoperative mortality and a high rate of recurrent haemorrhage.[15] The operation is therefore not recommended as an elective procedure. Schumaker and King[10,18] have used it as an emergency procedure for uncontrollable haemorrhage, but the mortality has been prohibitive.

(b) *Oesophago-gastric resection with jejunal or colonic interposition* is a more rational although, perhaps, a more formidable procedure. An interposed jejunal loop provides protection against reflux and exhibits greater resistance to acid pepsin ulceration than either stomach or duodenum, and Habif[8] reports satisfactory results in adults with varices. Our experience with the operation when performed for caustic and peptic oesophageal strictures in children indicates that it is well tolerated and not followed by oesophagitis, but we have not yet used it for varices.

In 1955 Koop and Roddy[12] reported on 5 children with bleeding varices treated by oesophagogastrectomy and colonic interposition. There was no operative mortality and no recurrence of haemorrhage for follow-up periods of 5 months to 1 year. In 1965 Koop and Kavianian[11] reported that all these children were still alive and well during June

1964, extending the follow-up to periods of 7 years and 7 months to 8 years and 1 month. However, only one of the children has not bled at all and one required a subsequent mesenterico-caval shunt. In addition they report on 6 other children with extrahepatic portal hypertension treated by similar procedures. One who had not bled was lost to follow-up after one year. Four others who have been followed-up for periods of 1 year and 8 months to 5 years and 9 months have not bled postoperatively and are all alive and well. One patient died from recurrent haemorrhage. As a result of this experience the authors now use colonic transplants earlier in the course of management of varices, i.e. as the initial treatment in patients with extrahepatic portal hypertension who have had a life-threatening haemorrhage and in whom a spleno-renal shunt is not possible. They have used the same procedure in 2 children with advanced cirrhosis. However, diminished appetite, substernal pressure on eating, a need for small feedings to reduce postprandial discomfort and a failure to gain weight for periods of 4 months to 1 year have continued to be problems. Our own experience with colonic interposition used for oesophageal atresia and caustic and peptic strictures of the oesophagus indicate that this procedure reduces the incidence of postoperative oesophagitis and does not lead to nutritional failure. We have treated one child with bleeding varices by this method—a girl of $4\frac{1}{2}$ years with extrahepatic obstruction—and she has remained well for 6 months. There is obviously still a need for further assessment of the long-term results of colonic transplants, but it appears to be a worth-while addition to the management of bleeding varices where shunt procedures are impossible or have failed especially in small children.

CONCLUSIONS

In the light of our present knowledge and experience of portal hypertension in childhood the role of surgery in the management may be summarized as follows:

Indications for Surgery

Extrahepatic obstructions—haemorrhage from varices

Intrahepatic obstructions—

- Presinusoidal (e.g. primary hepatic fibrosis): haemorrhage from varices
- Postsinusoidal (cirrhosis): liver function satisfactory—haemorrhage from varices
  Liver function marginal—demonstrable varices
- Suprahepatic: uncontrollable haemorrhage only.

Timing of Surgery

Emergency surgery—rarely necessary, but may be required especially for intrahepatic obstruction and occasionally for suprahepatic obstructions

Elective surgery
- Extrahepatic: delay until 10 years
- Intrahepatic: delay until 4 years
- Suprahepatic: emergency only

Choice of Surgical Procedures

Emergency
1. Ligation of varices
2. Oesophageal transection

Elective
- Under 4 years
  1. Ligation of varices
  2. Oesophageal transection
  3. Oesophago-gastrectomy with colonic interposition
- 4 - 10 years
  1. Portacaval shunt
  2. Mesenterico-caval shunt
  3. Oesophago-gastrectomy with colonic interposition
- Over 10 years
  1. Portacaval shunt
  2. Spleno-renal shunt
  3. Mesenterico-caval shunt
  4. Oesophago-gastrectomy with colonic interposition

The problem of portal hypertension in general and particularly in childhood is still far from solved. Much has been achieved by both medical and surgical means, but a great deal of controversy exists mainly because of the unpredictable life history of the disorder. Patients can be found whose varices have never bled, whose varices have bled once and not again, and whose varices have disappeared, but there are others who die as a result of a first haemorrhage and many who keep on bleeding. There are no reliable figures available upon the likelihood of varices bleeding or disappearing and the results of surgical procedures are equally unpredictable in a given case.

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