I will present a personal approach to the problem of portal hypertension, without attempting to cover the subject exhaustively, but emphasizing what I feel to be the important points for consideration in relation to the management of patients.

Causes of portal hypertension

The most important cause of portal hypertension is chronic liver disease which usually implies cirrhosis of the liver.

The second cause of portal hypertension to be considered is obstruction to the portal vein before its entry to the liver, or in some cases obstruction to the splenic vein producing hypertension in only the splenic portion of the portal venous system.

A third and less important cause of portal hypertension is obstruction to the hepatic vein, either to the major hepatic veins as in Chiari's syndrome, or obstruction to the intrahepatic tributaries of the hepatic veins as in the veno-occlusive disease found in some tropical areas. A similar variety of portal hypertension is caused by obstruction to the flow of blood from a patent hepatic vein as is produced by occlusion of the inferior vena cava, or by constrictive pericarditis or by disease of the tricuspid valve.

These three causes of portal hypertension can be looked at in another way. Cirrhosis may be regarded as portal hypertension due to a lesion within the liver, portal vein obstruction may be regarded as portal hypertension due to a pre-hepatic cause, and obstruction to the hepatic vein may be regarded as post-hepatic portal hypertension.

Table 1 lists the mechanical causes of portal hypertension just described, and for the sake of completeness two other causes are added.

The fourth cause of portal hypertension listed is idiopathic. This implies that chronic liver disease is not present, that there is no block in the extra-hepatic portal venous system and no block to outflow from the hepatic veins. A number of possible mechanisms for the portal hypertension have been postulated, including portal-vein spasm, an increase in portal venous blood-flow due for example to abnormal vessels in the stomach, or abnormal communications between branches of the hepatic artery and the portal vein within the liver. All these are difficult to substantiate and would be even more difficult to treat. However, one rare cause of portal hypertension which is more easy to substantiate and to treat is an arterio-venous aneurysm between major arteries and veins in the area, for example between the splenic artery and the splenic vein.

The final cause of portal hypertension listed is secondary, just as a reminder that splenomegaly, with an increase in the portal venous pressure, does occur in a variety of conditions unrelated to primary disease of the liver or portal venous system, for example in the splenomegaly of the myeloproliferative disorders the portal venous pressure is raised. This is included only to emphasize that it should not be allowed to cause confusion in diagnosis.

Presentations of portal hypertension

Table 2 shows the ways in which a patient with portal hypertension may present to the clinician.

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Presentations of portal hypertension</th>
</tr>
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<tbody>
<tr>
<td>Common</td>
<td>Bleeding oesophageal varices</td>
</tr>
<tr>
<td>Common</td>
<td>Splenomegaly</td>
</tr>
<tr>
<td>Rare</td>
<td>Septicaemia</td>
</tr>
<tr>
<td>Rare</td>
<td>Cyanosis</td>
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<tr>
<td>Rare</td>
<td>High output state</td>
</tr>
</tbody>
</table>

Probably the commonest presentation, and the most difficult to manage, is bleeding from oesophageal varices.
A common clinical presentation is the patient with splenomegaly, discovered at abdominal examination for some other reason, and with no obvious cause for the enlargement of the spleen.

Also in this group is the patient having ascites, without any obvious cause for the condition.

A further common clinical presentation is with one of the associations of hypersplenism, whether related to the anaemia, the low white blood count as manifested by infections, or the low platelet count as manifested by haemorrhagic phenomena.

Rarely the patient with portal hypertension might present with cyanosis, with septicaemia often due to a Gram-negative organism, or with a high cardiac-output state.

**Investigation of the patient**

A detailed history and physical examination are necessary and the symptoms and signs will, of course, depend to some extent upon the cause of the portal hypertension. This aspect will not be considered further.

The next problem to consider is whether it is necessary to make a precise diagnosis of the presence of portal hypertension and its cause in patients who present in the ways described. Accurate diagnosis is the basis of management in these patients, but it is possible to give more precise reasons for making a diagnosis, if these are thought necessary.

In the first place it is necessary to know of the existence of portal hypertension in the management of a patient with haematemesis. Should the haematemesis be due to bleeding from oesophageal varices, the possibilities of balloon tamponade, intravenous Pitressin and portacaval anastomosis must be raised. This is, of course, quite different from the management of the patient who is bleeding from a peptic ulcer. Even if the bleeding is so severe that, whatever the cause, surgery is necessary, the surgical approach may well be different according to the aetiology of the haematemesis. In considering a patient with ascites, clearly the management depends on a knowledge of the cause of the condition. In considering a patient with hypersplenism, if the haematological disorder is sufficiently severe, splenectomy may well be contemplated. This however, might be undesirable in a patient with portal hypertension, in whom other procedures may be preferred.

It is therefore necessary to decide whether portal hypertension is present and if so what is its cause, and further investigations will usually be necessary to achieve this.

In the first place, liver function tests and a barium swallow should be performed. Both are innocuous, and a barium swallow can even be performed in the ward if necessary, in a patient with gastro-intestinal bleeding. Oesophagoscopy and liver biopsy will need to be considered, but each carries a morbidity and mortality and the decision thus becomes a matter of judgement and personal preference. Having established the likelihood that the patient has portal hypertension, it may then be necessary to measure the portal venous pressure and to outline the portal venous system, in order to decide upon therapy.

**Measurement of portal venous pressure**

There are three commonly used methods of measurement of portal pressure; intrasplenic, intrahepatic and wedged hepatic vein pressure.

An estimate of the pressure within the portal vein can be obtained by measurement of the pressure within the spleen by insertion of a needle percutaneously, and the pressure recorded is closely related to the pressure within the portal vein.

Another commonly used means of estimating portal venous pressure is by measurement of the wedged hepatic-vein pressure. If a catheter is passed into a tributary of a major hepatic vein until the catheter is wedged tight within a small vessel, then the column of fluid within the catheter is in direct communication with the hepatic sinusoids and so the pressure recorded represents hepatic sinusoidal pressure, which is closely related to portal venous pressure, provided that there is no block to the vascular channels between the portal venous drainage area and the hepatic sinusoids. A more recently described method of estimating portal pressure is by measurement of the pressure within the hepatic parenchyma. This is performed by placing a needle or plastic cannula into the liver substance, injecting a small amount of saline, and measuring the pressure recorded. This probably represents the pressure within a small haematoma which is in direct communication with hepatic sinusoids. The pressure measured in this way has been shown to be closely related to the portal venous pressure.

There are several other ways of obtaining an estimate of portal venous pressure, less often used. For example pressure has been measured in oesophageal varices as seen at oesophagoscopy, pressure has been measured in collateral venous channels leading to the portal vein from the anterior abdominal wall, and pressure can be measured within the portal vein by threading a cannula from the umbilical vein.
Outlining the portal venous system

Measurement of portal venous pressure gives an indication of whether or not there is portal hypertension, but in itself gives no indication of the cause of the portal hypertension. A reasonable next step is to attempt to outline radiologically the branches and tributaries of the portal venous system. Four of the ways of outlining the portal venous system are splenoportography, arteriography, umbilical venography and mesenteric venography.

The most common is splenoportography, using a percutaneous needle within the spleen, as for measuring splenic pressure. Contrast medium is injected into the spleen and films are taken at intervals thereafter, showing the progress of contrast medium along the splenic vein into the portal vein and into the liver.

Recently, demonstration of the portal vein radiologically by arteriography has become possible. After injection of contrast medium into the coeliac axis or the superior mesenteric artery, the medium finds its way into the portal vein some seconds later, and outlines the portal vein and sometimes its intrahepatic branches. The contrast obtained is not as good as with splenoportography, but is sufficient to demonstrate whether the portal vein is patent, for example after a splenorenal anastomosis or after a splenectomy.

The umbilical route has been utilized for injection of contrast medium into the portal vein by cannulation of the umbilical vein and passing the cannula down into or near the portal vein. This method can provide pictures with contrast as good as those after splenoportography, and can be used in a patient who has had a splenectomy. It seems possible that umbilical portography and arteriography could be used when there is a haemorrhagic tendency which might preclude splenoportography.

Finally mesenteric venography involves demonstration of the portal vein by injection of contrast medium into one of the mesenteric tributaries of the portal vein either at the time of definitive surgery or through a small incision on a prior occasion, so as to plan a further surgical approach.

Indications for splenoportography

Splenoportography is the most commonly used of the procedures for outlining the portal venous system, and some of the indications will now be considered in terms of the clinical problems involved in patients with portal hypertension.

Splenoportography is a procedure with a definite risk, and mortalities have been described. Thus the information to be obtained must be necessary for planning the management of the patient.

For example splenoportography may justifiably be utilized in the investigation of a patient who has unexplained splenomegaly or who is known to have portal hypertension either because of the demonstration of oesophageal varices or because of the demonstration of an elevated intrasplenic pressure.

Splenoportography may be used in the investigation of a porto-caval shunt, to demonstrate whether or not this is still patent, for example if there is recurrent bleeding or persisting evidence of portal hypertension.

Splenoportography is extremely useful in the diagnosis of extrahepatic portal venous obstruction, in which a block to passage of contrast medium is seen in the portal vein. It should, however, be remembered that contrast medium may fail to travel into a patent portal vein if there are sufficiently large collateral vessels channelling the blood away or if the portal vein is acting as an outflow tract from the liver. If extra-hepatic portal venous obstruction is suspected this may be confirmed by the demonstration of an elevated intrasplenic pressure associated with a low or normal intrahepatic sinusoidal pressure as demonstrated by wedged hepatic-vein pressure measurements, or alternatively by attempting to outline the portal vein by mesenteric venography.

Splenoportography may also be used in the diagnosis of tumours involving the splenic or the portal vein or space-occupying lesions within the liver particularly those near the hilum, although in this latter group arteriography will probably supplant splenoportography.

Occasionally splenoportography may be of help in the evaluation of patients with obscure cerebral or neurological syndromes in whom the absence of collaterals may be taken effectivley to exclude chronic portal-systemic encephalopathy from the diagnostic arena.

Finally the most important use for splenoportography is in preoperative evaluation of a patient in whom a portacaval shunt is desirable, to ensure that the portal vein is patent.

Conclusion

Investigation of the portal venous system must be interpreted in the light of the condition of the patient, the clinical picture and the results of other investigations, in deciding upon the precise diagnosis and need for therapy in the individual patient.
Investigation of portal hypertension.

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