Radiological Intervention and the Budd-Chiari Syndrome

Introduction

Budd-Chiari syndrome (BCS) is characterised by obstruction of hepatic venous outflow and comprises a range of different clinical presentations, venous abnormalities, therapeutic options and clinical outcomes. Venous obstruction may be in the small centrilobular hepatic veins, in the main hepatic veins or in the distal IVC.

Clinical presentation

The presenting symptoms depend on the dominant site of obstruction. If this is the hepatic veins, patients usually present acutely with progressive hepatic engorgement, hepatic impairment and portal hypertension culminating in hepatic encephalopathy. Conversely, if the dominant site of obstruction is the IVC, patients present with a chronic history of lower limb swelling dating back many years.

Aetiology

Aetiology also varies according to the principal site of obstruction. In the West, most patients with BCS have an obstruction to their hepatic veins (either centrilobular or main). In about one-third, the cause is unknown, while most of the remainder have an underlying myeloproliferative disorder. In the East, the dominant site of obstruction is usually the IVC and only a minority have an underlying myeloproliferative disorder. IVC obstruction may be a sequel of a congenital venous web anomaly frequently complicated by thrombosis. It is classified as membranous (i.e. "caval web"), if it extends over 1-2 mm, or segmental if it extends over a few centimetres. IVC obstruction in BCS may also be due to caudate lobe hypertrophy, direct tumour invasion or extrinsic compression by hepatic tumour.

Diagnosis

The diagnosis ultimately rests on the demonstration of hepatic vein obstruction. We now rarely perform hepatic scintigraphy preferring to rely on ultrasound (Figures 1a, b). Ultrasound evaluates hepatic and portal vein patency, liver morphology...
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(e.g., caudate lobe hypertrophy) and features of portal hypertension. The hepatic veins may be engorged, irregular, filled with thrombus or absent. Triphasic flow may be dampened (Figure 1b) or reversed. Collaterals (intrahepatic, subcapsular) with a characteristic curved or "hockey stick" appearance may be visible. The IVC may be narrowed, displaced by caudate hypertrophy or contain intraluminal thrombus.12-14

Venography remains the gold standard diagnostic tool (Figures 2a, 5a and 6a, b).

Inferior venography is initially performed to assess intrinsic IVC obstruction or significant extrinsic compression which may be present in sixty-five percent.3 The exact length, location and direction of obstruction can be assessed by simultaneous injections of catheter placed on either side of the obstruction.15 Pressure measurements are obtained from the infrahepatic, intrahepatic and suprarehepatic IVC as an adjunct to surgical planning.

A transjugular approach allows easiest access to the hepatic veins. The walls of the IVC should be gently probed with a suitably shaped catheter to locate the hepatic venous ostia. Flush inferior vena cavaography alone cannot be relied upon to opacify severely stenosed venous ostia. Once cannulated, free and wedged venous measurements are obtained to give an assessment of portal hypertension, the hepatic wedged venous pressure correlating roughly with the degree of portal hypertension. Hepatic venography allows assessment of venous calibre, stenoses and thrombosis of either the large or small veins (the latter evidenced by a "spider's web" appearance which reflects partially recanalised and collaterised third and fourth order venules). A liver biopsy may be taken at the same time. This allows assessment of hepatic fibrosis and differentiates BCS-type changes ("peri-sinusoidal congestion") from hepatic venocclusive-type change where the terminal hepatic venules are obliterated.

Venographic abnormalities at presentation

In patients with dominant hepatic venous obstruction, the degree of limitation of hepatic venous flow at presentation is striking. The most venographic patterns evident at presentation are (i) a "spider's web" appearance alone without obstruction to the main hepatic veins, (ii) complete occlusion of all three veins or (iii) an occlusion of two out of three veins with a severe stenosis of the remaining vein. A critical level of hepatic vein occlusion must be reached before symptoms develop. This is supported by a prevalence of asymptomatic hepatic venous obstruction present in postmortems of 0.183%.16 Intrahepatic collaterals will usually be established at presentation. Similarly, with dominant IVC obstructions, a short ("membranous") or long segment ("segmental") stenosis/occlusion with established aygous and hemiazygous collaterals is typical, accompanied by varying degrees of obstruction to the main hepatic veins.

Treatment options

This may be supportive medical treatment, surgical or radiological.

Medical management

Medical management involves treating underlying myeloproliferative disorders, anticoagulation of hypercoagulable patients, control of ascites and lactulose/dietary restriction to treat hepatic encephalopathy.

Surgical treatment

Surgical management consists of peritoneovenous shunts, dorsocranial liver resection with venous reconstruction, portosystemic shunting, direct venous repair and orthotopic liver transplantation.3 Mesocaval shunts (which involve interposition of an autologous or dacron graft between the IVC and the superior mesenteric vein) are unsuitable for patients with high caval pressures. Mesoastrial shunts (preferred if the IVC pressure is high) involve interposition of a long narrow polyethylene prosthesis between the superior mesenteric vein and the
right atrium over the liver surface. These grafts are prone to occlusion due to low flow states and graft thrombogenicity. Nevertheless, the 5-year survival for patients undergoing mesocaval or mesoaortic shunts is 60-75%. Hepatic transplantation is reserved for those patients with advanced hepatic fibrosis.

Radiological intervention

Since percutaneous angioplasty of an IVC web was described in 1974, there have been many reports heralding radiological intervention as a successful treatment for BCS. The diversity of radiological options includes standard angiographic dilatation; I,2,6,15,17,20,22,24,26,29,30,31,38 IV C recanalisation utilising a straight transeptal needle,6 endomyocardial biopsy device,18 stiff guide wire24 or laser,27 hepatic vein recanalisations utilising a transhepatic approach,1,20,37 percutaneous stent insertion,1,19,32,33,37 thrombolysis,23,25 transluminal intrahepatic portosystemic shunting (TIPS)34-37 and portosystemic shunt dilatation.38

Patient suitability for radiological intervention

We reviewed 47 patients admitted over an eight year period with BCS.1 Of these 17 (37%) who were suitable for radiological intervention, two had tumour-related BCS and one underwent repeated dilatations of a surgically placed mesocaval shunt. Proper patient selection is important, the decision to proceed with radiological intervention being made following assessment of hepatic biopsy, ultrasound and venography findings. Patients unsuitable for venoplasty are those with (i) portal vein thrombosis, (ii) established hepatic fibrosis, (iii) a "spider’s web" appearance alone on venography without stenoses of the main hepatic veins, and (iv) complete thrombosis of all hepatic veins. Thrombolysis is ineffective in the last group as thrombosis typically extends out into the fifth order hepatic vein radicals and these smaller veins are difficult to clear effectively due to hepatofugal venous flow. Therefore, if the main veins are cleared, there is usually insufficient inflow to maintain patency. Patients in groups (iii) and (iv) may be suitable for TIPS.

Standard radiological interventional techniques

Patient preparation includes drainage of ascites, correction of clotting factors and omission of anticoagulant therapy (prior to percutaneous transhepatic procedures). Intravenous antibiotics are routinely administered. Procedures are tolerated well using intravenous sedation/analgesia with appropriate monitoring. The standard radiological techniques are hepatic and IVC venoplasty and/or recanalisation usually performed through a jugular and femoral approach respectively (Figures 3a-d). For hepatic vein dilatations, we generally use a 6F right internal jugular sheath, a general purpose angled catheter to cannulate and a standard angioplasty catheter to dilate the vein (to 12 mm) until waisting is abolished. The usual stricture encountered is at the venous ostium, is short, hard to dilate and recoils following deflation of the balloon (Figures 2a,b). Nevertheless, although there may not be much visible widening of the stricture following repeated dilatations (Figure 2b), a reduced pressure gradient, increased blood flow and symptomatic improvement does follow. For IVC obstructions, either one large balloon or up to four smaller balloons placed side to side (introduced via the femoral veins) can be used (Figures 3c,d). Thrombus, if present, in either the IVC or the hepatic veins should be cleared by thrombolysis to dilatation to...
reduce the risk of thromboembolism. All patients are maintained on long-term anticoagulation though the potential benefits of this are only anecdotal.

Dealing with completely occluded hepatic ostia

When the main hepatic venous ostia are occluded a transhepatic approach may be used (Figures 4a-d and 5a,b). Ultrasound allows identification of a suitable patent peripheral hepatic vein which is cannulated percutaneously (Figures 4a, 5a). Once the obstructed segment is crossed with a guide wire, the wire can be snared within the RA or IVC and brought out through a jugular sheath (Figures 4b,c). Anchoring both ends of the wire allows dilatation of the recanalised segment (and, if necessary, stent placement) via the jugular vein (Figures 4d, 5b). This combined approach leads to an improved recanalisation success rate and reduced the need for large transhepatic tracks thus lowering the risk of bleeding and intrinsic liver damage. It also allows one to steer, under biplane fluoroscopic screening, to a catheter (or inflated balloon) placed in the IVC or HV when attempting a recanalisation from the HV or IVC respectively. A similar technique can be used for a combined femoral-jugular vein approach (Figures 6a-d).

Recurrence of hepatic stenoses

Restenosis is the rule rather than the exception. Five out of 18 (28%) initial angiographic dilatations in our series failed to provide adequate venous return in the first instance due to a combination of restenosis and thrombosis. Adopting an aggressive attitude to re-intervention during the early treatment period with re-dilatation, use of thrombolytic agents and stent deployment as necessary improved our initial success rate from 72% to 83%.

Regular review is mandatory. HV restenosis occurred in all patients followed up for longer than 10 months although the duration between restenoses may vary. Generally, IVC stenoses recur less frequently than hepatic vein stenoses. The aim of post-intervention surveillance is to re-intervene before critical stenosis or occlusions recur, particularly as they may pre-date clinical or biochemical deterioration. We adopt a policy of regular clinical and ultrasound assessment (varying from three-monthly, six-monthly and yearly once the patient is stabilised) supplemented by hepatic venography annually (or sooner should clinical, biochemical or ultrasonic deterioration occur). Whilst this policy is uniformly well
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Figure 6a-d:

(a) Hepatic inferior venacavography. Renal tumour recurrence in the intrahepatic IVC with diversion into collaterals. It was not possible to enter an hepatic vein from above.

(b) An inferior venous collateral was cannulated (arrowheads).

c) A guide wire was passed to the right atrium and snared.

(d) The ostial obstruction was stented via the jugular vein. This patient who presented with severe hepatic encephalopathy remained well for a further 18 months.

tolerated, in occasional circumstances where frequent repeat dilatation is necessary, stent insertion has been undertaken. Radiological intervention does not address the underlying pathophysiology of hepatic venous outflow obstruction. Thus the disease can never be deemed cured and continued review is essential.

Stenting

IVC stent deployment has been reported more commonly than hepatic vein stenting. We reserve stenting for situations where a) the patient is critically ill and initial dilatations are unsuccessful, b) repeated re-dilatation is required at short intervals on review and c) malignant disease. Where there is significant IVC narrowing, placement of an intrahepatic stent may reduce caval pressure to allow mesocaval shunting rather than more hazardous mesoatrial shunting. As reduction in liver swelling may lead to apparent stent movement, we aim to deploy hepatic vein stents with the tip not protruding much beyond the venous ostium. IVC stents should not extend into the right atrium or the infrahepatic IVC where they will interfere with liver removal (if transplantation is subsequently required).

Safety of radiological intervention

Very few complications following radiological intervention for BCS have been reported. IVC stent migration is the only significant complication reported to date. In 49 angiographic dilatations (including 11 recanalisations), we had three serious complications. In one patient, a small pseudoaneurysm of a right hepatic artery branch presented as haemobilia within days of percutaneous transhepatic intervention. Cephalad migration of two interlaced upper IVC stents into the right atrium by approximately 3 cm occurred in another patient. This cephalad migration may have precipitated an increased frequency of atrial tachyarrhythmias. Myocardial puncture (without haemodynamic compromise) occurred in another patient during recanalisation of an upper IVC occlusion.

Effectiveness of radiological intervention

Overall, the efficacy of radiological intervention compares favourably with surgical shunting. A "marked improvement" (i.e. with no symptoms related to hepatic venous outflow obstruction) following radiological intervention was seen in 10 of the 18 (56%) patients. Some of the patients developed recurrent progressive symptoms in the weeks prior to re-intervention. "Improvement" (i.e. continued mild symptoms of hepatic venous outflow obstruction) was seen in 4 out of 18 (22%) patients again with progressive symptoms prior to re-intervention.

Failure of radiological intervention

As most reports consist of one or two patients, there is little mention of failed radiological intervention in the literature.
Radiological intervention failed in 5 out of 18 of our patients, primarily during the earlier years of the study. Three of these patients failed early: recurrent thrombolyis (n = 1); failure to recanalise IVC occlusion (n = 1); and failure to recanalise hepatic vein occlusion (n = 1). Two late failures occurred (restenosis, n = 2). Of late, better patient selection, use of a combined transhepatic-transjugular route and stenting have led to an improved success rate. Should radiological intervention fail, patients can still proceed to surgical shunting or transplantation. Moreover, a limited response to radiological intervention in a patient with severe hepatic failure, may make subsequent surgery less hazardous.

TIPS

When the main hepatic veins are completely occluded, TIPS have been successfully performed (usually through an occluded RHV sinus) as an alternative to surgical portosystemic shunting to relieve portal hypertension and decrease hepatic congestion. Whilst contraindicated in the presence of a concomitant severe upper IVC stenosis, TIPS, as opposed to mesocaval shunting, can be considered when severe caudate lobe hypertrophy exists and does not interfere with subsequent liver transplantation. As venous drainage through the hepatic veins is not restored, TIPS carry an inherent risk of portal-systemic encephalopathy although the incidence of this, following surgical portosystemic shunting for Budd-Chiari, is low.

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