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Review

Budd–Chiari syndrome

H. W. TILANUS

Department of Surgery, Erasmus University Hospital 'Dijkzigt', Dr Molewaterplein 40, 3015-GD Rotterdam, The Netherlands
Correspondence to: Mr H. W. Tilanus

Budd–Chiari syndrome is the generic term for different forms of hepatic venous outflow obstruction resulting in a clinical picture of portal hypertension and hepatomegaly. Three levels of venous outflow obstruction may be recognized, affecting respectively the small intrahepatic venules, the large hepatic veins and the inferior vena cava (IVC). Each level of obstruction is related to a different aetiology. Clinical manifestations range from mild symptoms to acute or chronic end-stage liver disease. Treatment is surgical in the great majority of patients. Occlusion of the IVC may be treated by removal of the caval obstruction in selected patients. Hepatic outflow obstruction may be circumvented by different forms of shunting from the portal or upper mesenteric vein to the IVC or right atrium, depending on the level of obstruction and the difference in venous pressure. For the rare patient presenting with acute or chronic end-stage liver failure, hepatic transplantation may be a life-saving procedure.

The syndrome of portal hypertension with hepatomegaly due to hepatic venous outflow obstruction was first described by Budd[1]; the associated histology was first described by Chiari[2] hence the name Budd–Chiari syndrome. The outflow obstruction, caused by occlusion of the smaller or larger hepatic veins or the inferior vena cava (IVC) between the liver and the right atrium, results in right-sided upper abdominal pain, hepatomegaly and (often) massive ascites. For this review the proposal of Ludwig *et al.*[3] is adopted: Budd–Chiari syndrome consists of hepatic venous outflow obstruction and its manifestations, regardless of cause, the obstruction being either within the liver or in the IVC. Functional hepatic outflow obstruction caused by congestive heart failure is not considered to be Budd–Chiari syndrome.

The syndrome may be specified by several characteristics; anatomical, aetiological and morphological. Although the diagnosis is quite straightforward, definitive treatment is often postponed until serious liver damage becomes apparent, and surgical intervention is associated with many complications. Several surgical procedures lay claim to superior results but, in this rare disease, it is difficult to choose the right form of hepatic venous decompression for an individual patient. This is especially so in the light of new treatment options such as liver transplantation and, more recently, transjugular intrahepatic portasystemic shunting. The aim of this review is twofold: to provide guidelines for diagnosis and therapy in general, and to describe an approach for the management of the individual patient.

Aetiological considerations

Traditionally, the level of hepatic venous occlusion in Budd–Chiari syndrome is differentiated into three groups: the small hepatic venules, the major hepatic veins and the IVC.

The first description of occlusion of the small hepatic venules by Chiari[2] used the term 'obliterative endophlebitis'; today this would be regarded as veno-occlusive disease. It may result from ingestion of hepatotoxins such as pyrrolizidine alkaloids[4–7]. Bras *et al.*[8] described the disease in Jamaica and related it to the ingestion of 'bush teas', which contain pyrrolizidine alkaloids such as senecio and crotalaria. The same histopathological picture results from chemotherapy and radiotherapy regimens to prepare patients for bone marrow transplantation[9,10]. It has also been described in relation to conventional chemotherapy and to azathioprine treatment after renal transplantation[11].

Occlusion of the major hepatic veins is, in many instances, secondary to an underlying disease (detected or undetected). Three main causative states may be recognized: a hypercoagulable state, malignancy and a residual group of miscellaneous diseases. A hypercoagulable state may occur in the course of various myeloproliferative syndromes, paroxysmal nocturnal haemoglobinuria, circulating lupus coagulant, Behçet's disease, and deficiencies of antithrombin III, protein C and protein S[12–19]. The use of oral contraceptives is frequently listed under the factors predisposing to Budd–Chiari syndrome[20–23]. In case–control studies, however, the risk of the syndrome from oral contraceptive use seems significant only in the presence of a thrombogenic tendency in patients with an underlying haematological disorder, which, at time of diagnosis of Budd–Chiari syndrome, may still be latent[24–26]. Increased thrombogenicity may play a role in eclampsia and in pregnancy as a cause of the syndrome, but why the major hepatic veins are the target vessels is unclear[27–29]. Crohn's disease and aspergillosis may cause Budd–Chiari syndrome but this is anecdotal[30,31]. Hepatocellular carcinoma, cysts and abscesses within the liver, and tumours outside the liver (such as rhabdomyosarcoma, leiomyosarcoma and leiomyoma) may obstruct the hepatic veins, leading to the syndrome[32–36].

Occlusion of the IVC between the hepatic veins and the right atrium may be caused by a membrane, by thrombosis or by compression from outside[37–39]. The first description of a membranous web is attributed to Osler[40] in 1879. It is a rare cause for Budd–Chiari syndrome in

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the Western world, but accounts for the majority of cases in oriental series and in South Africa[41–43]; it is also noted with greater frequency in areas of the USA inhabited by oriental immigrants[44]. There is increasing evidence that pre-existing cirrhosis and previous hepatitis predispose to a membranous obstruction and in affected patients an unusually high incidence of hepatitis B positivity is found[45–49]. The nature of the web, acquired or congenital, is not completely clear, but membranous obstruction is extremely rare in children[50,51]. The high incidence of primary diseases in these patients supports the concept of a thrombotic lesion preceding the membranous obstruction[52]. This view is strengthened by the careful study of Kage and colleagues[53], which favoured a thrombotic nature for the membrane after an analysis of 17 autopsy cases.

Pathology

The pathological picture is different in early and late Budd–Chiari syndrome. In the acute situation the liver may be extremely enlarged, macroscopically congested and (red-purple) discoloured with blunt edges. Histological examination shows normal periportal areas in the early stages, but the perivenular liver tissue is darkened by blood congestion caused by the venous thrombosis. This leads to atrophy and death of the centrolobular hepatocytes (*Fig. 1*). Following the collapse of stromal tissue, the central veins obliterate. The sinusoids become devoid of erythrocytes which are typically concentrated in the space of Disse[54]. Without adequate treatment the acute, and possibly reversible, situation becomes chronic and irreversible (*Fig. 2*). Fibrin thrombi organize as fibrous cores. Thin collagen strands replace the liver cords and the collapsed stroma, and regeneration occurs as the first sign of fibrosis and cirrhosis[55]. As the caudate lobe drains directly into the IVC by small and often unaffected veins, this segment may compensate for the loss of functioning liver tissue, sometimes by impressive enlargement; it may take over the function of the other affected liver segments.

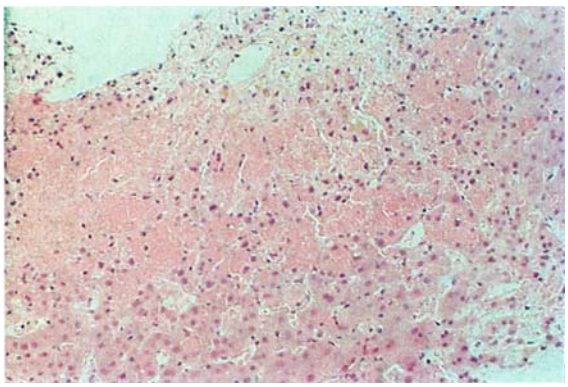

Fig. 1 Thrombosis of central veins with pericentral blood congestion and atrophy of hepatocytes

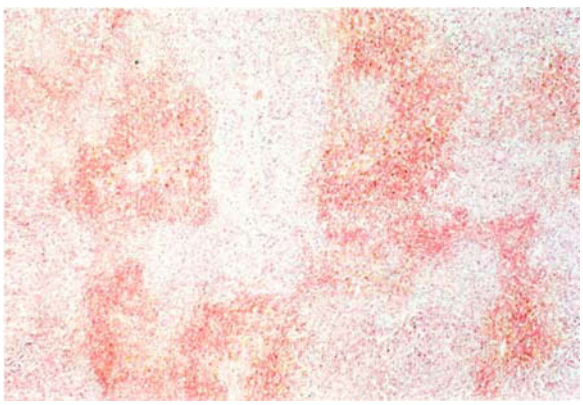

Fig. 2 Collapse of pericentral stromal tissue with bleeding and extensive death of hepatocytes.

Clinical manifestations

The onset of symptoms is often insidious with vague upper abdominal discomfort and progressive ascites caused by portal hypertension[47,56]. Mild jaundice may be present with minimal liver dysfunction at first presentation. At this early stage Budd–Chiari syndrome may mimic constrictive pericarditis[57]. Physical findings that suggest vena caval occlusion include leg oedema, episodes of pulmonary embolism and, sometimes, impressive varicose veins of legs, abdomen and lumbar region[58,59]. Some patients with IVC occlusion, however, have no symptoms at all[60]. With diuretic therapy, the clinical picture may appear reassuringly improved, but such treatment is only symptomatic and increasing portal hypertension leads to oesophageal variceal haemorrhage and progressive liver failure[61]. In a small proportion of cases, Budd–Chiari syndrome first presents as fulminant hepatic failure; within 8 weeks of onset encephalopathy, coagulopathy and massive hepatic necrosis may occur[62,63]. Bismuth and Sherlock[64] have proposed a classification for these different clinical manifestations of the syndrome according to the degree of concomitant liver failure: fulminant, acute, subacute and chronic.

Investigations

Ultrasonography is today the first and most important tool in the diagnosis of Budd–Chiari syndrome, with a sensitivity of over 85 per cent[65,66]. The absence of major hepatic vein images and the classical enlargement of the caudate lobe may be demonstrated by this technique[67,68]. With duplex scanning the portal and hepatic venous flow, as well as flow in the IVC, can be measured[69–72] and these correlate well with venography[73]. Magnetic resonance imaging is capable of providing static and functional information about the hepatic veins and the IVC, but its additional value remains to be assessed[74–77]. Parenchymal abnormalities are best demonstrated by computed tomography, as shown in a comparative study by Miller and colleagues[78]. Necrotic masses or nodular regenerative hyperplasia may resemble metastases, which should be excluded by fine-needle aspiration cytology[79–81]. Radiocolloid scintigraphy of the liver, typically showing an increase of isotope centrally in the organ and hypertrophy of the caudate lobe, is today of only historical interest[56,82].

Angiographic catheterization of the IVC and hepatic veins may show occlusion or a narrowing of the vena cava due to compression by the caudate lobe[83]. If the hepatic veins can be visualized, a typical intrahepatic 'spider's

web' may be seen[56,83]. Intraluminal IVC pressure measurment and, if possible, wedge pressure measurement of the hepatic veins are mandatory. This haemodynamic information, especially a pressure gradient across the retrohepatic IVC from the bifurcation distally up to the right atrium, should be obtained and considered when surgical therapy is planned.

Of prime importance in the investigations is a liver biopsy to assess hepatocyte damage and the extent of fibrosis and cirrhosis.

Management

Untreated Budd–Chiari syndrome causes death in months or years[84,85]; spontaneous resolution is rarely reported. Incidental case reports are available describing successful thrombolysis in the acute situation. Urokinase, streptokinase and recombinant tissue plasminogen activator are advocated as thrombolytic agents[86–88]. It is difficult to understand why there should by any benefit from treatment with anticoagulants alone other than prevention of thrombus extension[89], but this, in combination with diuretics, is all too often the first and sometimes the only step in the treatment of the syndrome. In general, surgery is almost always mandatory, its nature depending on the location of the outflow obstruction. Direct local treatment may be appropriate for a membrane or web in the IVC. A variety of shunt procedures are also available to which the transjugular intrahepatic portasystemic shunt (TIPS) has recently been added. Finally, liver transplantation may be the appropriate treatment for a few patients.

Direct treatment

As a web or membrane in the IVC as a cause of Budd–Chiari syndrome is more common in the East, the majority of series and case reports regarding direct treatment of this lesion are of Asian, Japanese and also South African origin. Percutaneous balloon angioplasty has been successful in selected cases, with a long-term follow-up of 6 years or more, sometimes after multiple procedures[90–92]. Lifelong anticoagulation is advised in these patients. To prevent restenosis and occlusion, prophylactic use of intravascular metal stents, such as the Wallstent (Schneider, Bulach, Switzerland) and the Gianturco stent (William Cook, Bjærverskov, Denmark) may be advisable[93–95]. Stenting and laser treatment of locally narrowed ostia of the hepatic veins has also been described[96,97]; an alternative treatment is transcardiac membranotomy or membrane excision[98,99]. In more extensive obstruction involving stenosis of the IVC, cavoplasty with autologous pericardial patching under hypothermia and cardiopulmonary bypass may be carried out[100–103]. Obstruction of the IVC and hepatic veins has been corrected by resection of the part of the liver containing the terminal portions of the major hepatic veins and the diseased part of the IVC, followed by direct anastomosis to the right atrium. This procedure, first described by Senning[104], allows hepatic venous blood to enter the right heart directly[105,106].

Shunts

The key concept of any shunt procedure is the conversion of one of the large splanchnic veins into an outflow tract

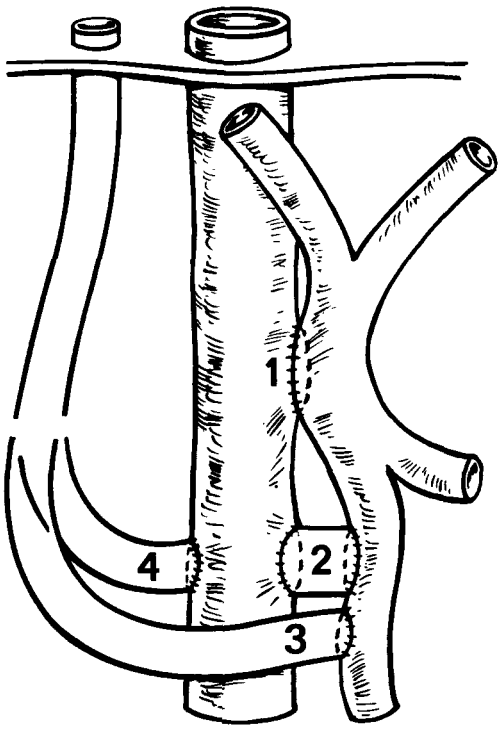

Fig. 3 Different forms of shunt: 1, portacaval shunt; 2, mesocaval shunt; 3, mesoatrial shunt; 4, cavoatrial shunt

of the congested liver and congested splanchnic area. Under these conditions the liver should be able to resume its normal function, with a reduction in portal hypertension preventing further complications. Different forms of shunting have been described in the treatment of Budd–Chiari syndrome, the choice depending on the patency of the IVC and the pressure gradient between the portal vein and vena cava. In Budd–Chiari syndrome confined to the hepatic veins, intraoperative angiography may show compression of the IVC by the caudate lobe, but no occlusion. In these circumstances the caval pressure will be in the typical range for patients with ascites, varying between 4 and 12 mmHg. In patients with caval obstruction caused by thrombosis or web formation, the IVC pressure rises easily to 20–23 mmHg. Portal or superior mesenteric vein pressures in Budd–Chiari syndrome[107] are between 20 and 25 mmHg. If intraoperative manometric studies demonstrate pressure in the retrohepatic IVC equal to or greater than those in the portal vein, portacaval or mesocaval shunting will be ineffective and result in thrombosis, as a minimum pressure gradient of 10 mmHg is required to guarantee long-term shunt patency. In these circumstances a mesoatrial shunt is indicated (*Fig. 3*)[108,109].

Portacaval shunt

If a shunt is indicated, the portacaval side-to-side construction is the procedure of choice only if the IVC pressure is substantially lower than the portal pressure. This shunt is performed preferably without interposition of a graft. Direct side-to-side anastomosis between the portal vein and the intrahepatic IVC is technically demanding, but it carries the highest patency rate of all

shunts[108,110–112]. Some authors regard a large caudate lobe as a contraindication or advise a partial resection, but Orloff and colleagues in their large series of more than 1400 cases have described a technique in which the direct side-to-side portacaval shunt remains free from compression by the caudate lobe; an internal jugular vein interposition graft is needed only in exceptional cases[113,133]. The hypertrophied caudate lobe may compress the IVC but, except in severe cases (more than 75 per cent diameter compression), this narrowing seems to be of no haemodynamic relevance. Moreover, after a successful shunt procedure the caudate lobe may return to its previous volume.

Mesocaval shunt

The mesocaval interposition H shunt using homologous vena cava was first reported by Lord *et al.*[114] and Read *et al.*[115]. In 1972 Drapanas and colleagues[116,117] advocated use of a wide calibre (18–22-mm) Dacron (DuPont, Wilmington, Delaware, USA) prosthesis. The H shunt is simple and avoids hilar dissection; the portal vein and the infrahepatic suprarenal IVC are not compromised, allowing possible future liver transplantation[64,118]. However, the price for this simplicity is a high rate of thrombotic complications, varying between 24 and 53 per cent[119,120]. Cameron and colleagues[121] introduced the mesocaval C shunt on the wider retropancreatic part of the superior mesenteric vein. This variation prevented thrombotic complications in a series of 30 patients. Today, the widest part, which is the preferred site for the anastomosis, of the superior mesenteric vein can easily be assessed by intraoperative ultrasonography. The introduction of externally reinforced polytetrafluorethylene (PTFE) vascular grafts prompted the use of this material in mesocaval shunting, but long-term results are lacking. To overcome the thrombotic complications Bismuth and Sherlock[64] prefer to use the internal jugular vein, with good results. Sixteen patients were treated with a venous mesocaval shunt, two of which thrombosed after technical complications relating to the shunt; both were successfully treated by thrombectomy.

The survival rate after shunt procedures varies widely and is directly related to the pressure difference between the portal vein and IVC, and hence to the flow in the shunt[85,86,122]. Once the patient has successfully recovered from surgery the prognosis and prospects of long-term survival are dictated by the underlying disease[123,124].

Mesoatrial shunt

Bypass of the liver and IVC is indicated in patients in whom the IVC is (sub)totally occluded, especially in the case of coexisting obstruction of the hepatic veins for which angioplasty or membranotomy is neither indicated nor possible[125]. Caval compression by the caudate lobe alone is seldom an indication for mesoatrial shunting, unless the difference in venous pressure in the mesenteric vein and IVC, measured directly during operation, is so small that the patency of a portacaval or mesocaval shunt is jeopardized. The haemodynamic consequences of the stenosis, as has previously been noted, should be quantified by cavography and pressure measurements[126,127]. Since the first description by Cameron and Maddrey[125] in 1978, different varieties of mesoatrial shunts have been suggested[128]. The patency of these mesoatrial shunts varies between 75 and 100 per cent in some small series; the mean follow-up in all series is less than 2 years[129–131]. Others have had more discouraging results, with success rates of only 33–66 per cent[109]. Much attention has been paid to the technical details of the operation, and improved results have been claimed by using an external silicone rubber sleeve around a 16-mm externally reinforced PTFE prosthesis to prevent compression of the graft by the sternum[128]. Warren and colleagues[132] proposed a two-step procedure: first, decompression of the venous splanchnic bed by a mesoatrial shunt, resolving the swelling of the liver and the caudate lobe, followed by a second operation after several months with closure of the mesoatrial shunt and creation of a side-to-side portacaval shunt. Orloff *et al.*[133] reported unsatisfactory results in eight patients after mesoatrial shunting with 16-mm PTFE externally reinforced grafts. In five patients the graft occluded soon after operation, uniformly leading to death. After experimental work on cavoatrial shunting combined with side-to-side portacaval shunting, these authors' clinical results in five patients were improved, with no graft occlusion and no death. In this procedure the vascular graft receives venous blood from the obstructed splanchnic bed and from the obstructed IVC, leading to a high rate of blood flow through the graft, resulting in fewer thrombotic complications.

Transjugular intrahepatic portasystemic shunt

The elegant TIPS procedure was recently introduced for relief of portal hypertension; it has been performed in a few patients with Budd–Chiari syndrome[134–137]. The long-term results of TIPS insertion have not yet been evaluated and, especially in Budd–Chiari syndrome with (sub)total occlusion of the hepatic veins, cannulation is not always possible. Sometimes the shunt can be introduced through a large vein of the caudate lobe, but it is not clear whether positioning a TIPS through the only functioning part of the liver, i.e. the caudate lobe, does any good. A possible indication for a TIPS is acute bleeding, as it may allow a short-term stabilization of the patient who is awaiting definitive surgical treatment[135,137]. Thrombotic complications with a TIPS are common and only short-term follow-up studies are so far available. The indications for a TIPS are not completely clear and the procedure should be reserved for patients with acute fulminant Budd–Chiari syndrome to relieve variceal bleeding before liver transplantation.

Liver transplantation

The choice between shunt or liver transplantation for Budd–Chiari syndrome is affected by several factors, the most important of which is the estimated hepatic reserve. Salient features are the presence of encephalopathy, hypoalbuminaemia and hyperbilirubinaemia and, most significantly, the result of liver biopsy[138]. The proportion of patients with Budd–Chiari syndrome receiving a liver transplant varies widely in different series. Three groups of patients with the syndrome requiring liver transplantation in preference to a shunt may be identified: those with Budd–Chiari syndrome presenting as fulminant hepatic failure, those with end-stage chronic liver disease at first presentation and those who rapidly deteriorate after a shunt procedure[64,139,140]. Some studies of liver transplantation for Budd–Chiari syndrome contain a few

patients with fulminant hepatic failure[138,139], but the clinical picture may be very impressive without irreversible loss of hepatic function, so no liver transplant should be done before adequate evaluation by hepatic biopsy. In patients presenting with end-stage chronic liver failure, the decision in favour of transplantation is more straightforward[141]. Even here, however, a liver biopsy showing extensive fibrosis and cirrhosis is mandatory before the surgical option of a shunt is discarded[142–144]. The survival rate after liver transplantation for Budd–Chiari syndrome varies from 45 to 80 per cent after 5 years, but these figures are influenced by small numbers and better results in later series. All authors describe thrombotic complications in some patients, sooner or later, after transplantation, and so lifelong anticoagulant therapy is advised except in patients in whom a thrombogenic condition, such as antithrombin III deficiency, is corrected by the liver transplantation. Depending on the underlying aetiology, liver transplantation for Budd–Chiari syndrome may be considered as palliative, requiring long-term adjuvant therapy, or as curative, with correction of a metabolic defect[145–147].

Conclusion

Suspected Budd–Chiari syndrome should be confirmed by radiology, including venograms and pressure measurements in the IVC. Haematological or other underlying disorders should be evaluated. Based on the outcome of liver biopsy, three treatment pathways are possible. In exceptional cases liver biopsy may show neither necrosis nor fibrosis and this feature, combined with a mild clinical picture, may allow treatment by anticoagulation and administration of a diuretic. Re-evaluation should be thorough and frequent. In the majority of patients centrolobular necrosis is present without irreversible signs of fibrosis or cirrhosis. In these patients decompression by a shunt is the treatment of choice, the specific shunt depending on IVC haemodynamics. A small group of patients with severe fibrosis, or even cirrhosis, presenting with end-stage liver disease will benefit from liver transplantation, as do those with acute liver failure resulting from Budd–Chiari syndrome.

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