Managing Budd-Chiari syndrome: a retrospective review of percutaneous hepatic vein angioplasty and surgical shunting

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Abstract

Background—The role of percutaneous hepatic vein angioplasty in the management of Budd-Chiari syndrome has not been well defined. Over a 10 year period at our unit, we have often used this technique in cases of short length hepatic vein stenosis or occlusion, reserving surgical mesocaval shunting for cases of diffuse hepatic vein occlusion or failed angioplasty.
Aims—To review the outcome of angioplasty and surgical shunting to define their respective roles.
Patients—All patients treated by angioplasty or surgical shunting for nonmalignant hepatic vein obstruction over a ten year period from 1987 to 1996.
Methods—A case note review of pretreatment features and clinical outcome.
Results—Angioplasty was attempted in 21 patients with patent hepatic vein branches and was succesful in 18; in three patients treatment was unsuccessful and these patients had surgical shunts. Fifteen patients were treated by surgical shunting only. Mortality according to definitive treatment was 3/18 following angioplasty and 8/18 following surgery; in most cases this reflected high risk status prior to treatment. Venous or shunt reocclusion rates were similar for both groups and were associated with subtherapeutic warfarin in half of these cases. Most surviving patients in both groups are asymptomatic although one surgical patient has chronic hepatic encephalopathy.
Conclusion—With appropriate case selection, many patients with Budd-Chiari syndrome caused by short length hepatic vein stenosis or occlusion may be managed successfully by angioplasty alone. Medium term outcome is good following this procedure provided that anticoagulation is maintained. Further follow up is required to assess for definitive benefits but we suggest that this should be included as a valid initial approach in the algorithm for management of Budd-Chiari syndrome.
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The management of symptomatic Budd-Chiari syndrome (BCS) continues to present considerable difficulties for the clinician, as a range of therapeutic options have been advocated. Most published series have advocated surgical portosystemic shunting as the most appropriate option for patients with stable liver function, with liver transplantation reserved for those presenting with fulminant liver failure or end stage chronic liver disease.[1–6] However, percutaneous hepatic vein angioplasty (PHVA) may also achieve symptomatic relief in patients with BCS caused by short length hepatic vein stenosis or occlusion. Since first described in the early 1980s,[7 8] we and others have described a favourable outcome for patients treated with this technique.[9–14] Successful PHVA restores physiological hepatic venous drainage and may thus be theoretically preferable to surgical shunting, with the attendant operative risks and diversion of blood from the liver resulting from the latter procedure. However, angioplasty is sometimes a challenging procedure for the radiologist and even when successful may be complicated by recurrent venous stenosis.

As many of the patients treated with PHVA at our centre may otherwise have been considered suitable candidates for surgical shunting, we have carried out a retrospective comparison of patients treated by each of these procedures in order to clarify their respective roles.

Patients and methods

PATIENTS
Between 1984 and 1996, 70 patients with clinically and radiologically validated BCS were referred to our centre for management. The primary treatment for these patients was surgical shunting (n=15), PHVA or other angioplasty (n=25), liver transplantation (n=10), transjugular intrahepatic portosystemic shunting (TIPSS) (n=4), or medical treatment (n=9) (fig 1). In the remaining cases, definitive treatment was not given either because of early death (n=5) or the patient refused treatment (n=2). Most of the patients undergoing liver transplantation were treated relatively early in our series; poor results from this procedure led us to choose surgical shunting or radiological angioplasty in preference to transplantation in patients without end stage liver disease. We have also recently adopted the TIPSS proce-

Figure 1   Definitive procedures for BCS at Queen Elizabeth Hospital Liver Unit, 1984–1996.

Table 1   Predisposing diseases leading to BCS (divided according to initial treatment)

Disease	PHVA (n=21)	Surgical shunting (n=15)
Myeloproliferative disease	4	6
Protein C deficiency	6	1
Protein S deficiency	1	0
Antithrombin III deficiency	0	1
Paroxysmal nocturnal haemoglobinuria	0	1
Oral contraceptive usage	1	0
Factor V Leiden heterozygote*	2	1
Unknown	9	6

*Each of these patients found to be heterozygous for the factor V Leiden mutation also had a concurrent predisposing cause for BCS.

dure in preference to surgical shunting; however follow up for these patients has been short. Our present review is therefore restricted only to those patients undergoing percutaneous radiological angioplasty or surgical shunting, for non-malignant hepatic vein obstruction. The 25 patients treated with radiological angioplasty included two treated with metal stent insertions for palliation of hepatic vein occlusion from solid malignancies, and two with isolated inferior vena caval obstruction; these patients were excluded from our analysis, leaving 21 patients in the PHVA and 15 in the surgical shunting groups. For the purposes of this review we recorded clinical, radiological, and

laboratory features prior to treatment and on follow up as of December 1997. Statistical analysis was used to compare clinical and laboratory variables before and after treatment, using the Mann-Whitney U test and Fisher's exact test as appropriate.

PRETREATMENT CLINICAL AND INVESTIGATIVE FEATURES

The spectrum of underlying diseases leading to BCS was similar for each group and is summarised in table 1; comparison of other features is summarised in table 2. All patients had symptomatic BCS at presentation. The frequency of clinical symptoms and laboratory abnormalities was similar in both groups, although patients in the surgical shunting group tended to have a higher incidence of hepatic encephalopathy (NS), higher serum bilirubin levels (p<0.01), and more severe ischaemic changes on liver histology as judged by a semiquantitative scoring system (p<0.05). The majority of patients (17/21 in the PHVA group and 14/15 in the surgical group) were of Western European origin. Radiological imaging (Doppler sonography and contrast venography) showed short length hepatic vein stenosis (SLHVS) in all 21 patients undergoing attempted PHVA. Doppler ultrasound reports were available for 17 patients and suggested patent intrahepatic vein branches in 16 (with more than one patent hepatic vein branch visualised in 10). In two patients, transjugular venography subsequently showed further patent hepatic vein branches. All patients underwent attempted transjugular hepatic venography; in 12 of these patent intrahepatic vein branches were shown while in the remaining nine patients hepatic veins were only confirmed by transhepatic venography.

In patients referred directly for surgical shunts, 4/15 had patent hepatic vein branches visible on Doppler sonography. In one case this was confirmed by transjugular venography (but angioplasty was not attempted) and in the remaining three cases hepatic veins were not accessible by the transjugular route. All of these cases were relatively early in our series and none of the patients underwent attempted transhepatic venography. The remaining 11 patients in the shunt group had diffuse hepatic venous occlusion shown by Doppler sonography (four of these also underwent hepatic venography, with no veins shown). Three of 21 patients in the PHVA group and 2/15 in the shunt group had associated inferior vena caval (IVC) stenosis with a pressure gradient of at least 10 mm Hg across the stenosis; in several remaining patients a degree of IVC stenosis was present but without a significant pressure gradient. None of the patients in either group had associated portal vein occlusion.

PERCUTANEOUS HEPATIC VEIN ANGIOPLASTY

The techniques used for this procedure have been described extensively elsewhere.[13] [14] Briefly, the procedure was generally undertaken where Doppler ultrasound scanning or contrast venography identified a short length stenosis or occlusion of the hepatic vein with a

Table 2   Clinical and investigative features at presentation

	PHVA (n=21)	Surgical shunting (n=15)	Significance (p value)
Age[1]	31 (16–65)	36 (20–57)	NS[2]
Duration of history, months[1]	3 (0–84)	3 (0–204)	NS[2]
Clinical features[4]			
Ascites	18	15	NS[3]
Variceal haemorrhage	4	3	NS[3]
Hepatic encephalopathy[5]	5	8	NS[3]
Muscle wasting[6]	7	6	NS[3]
Radiological features[4]			
Short length hepatic vein stenosis	21	4	
IVC occlusion	3	2	
Laboratory findings[1, 7]			
Serum bilirubin (µM/l)	31 (6–179)	60 (20–255)	p<0.01[2]
Serum AST (U/l)	48 (12–2410)	69 (23–504)	NS[2]
Serum albumin (g/l)	34 (20–45)	32 (26–39)	NS[2]
Serum creatinine (µM/l)[8]	87 (47–247)	104 (61–522)	
Prothrombin INR	1.4 (1.0–2.9)	1.4 (1.2–3.2)	NS[2]
Histological features[1, 9]			
Centrilobular necrosis score	1 (1–3)	2 (0–3)	p<0.05[2]
Fibrosis score	2 (0–3)	1 (0–3)	NS[2]

[1]Median values (range given in brackets).
[2]Mann-Whitney U test; [3]Fisher's exact test. p values ≥0.05 are denoted as not significant (NS).
[4]Absolute numbers.
[5]Clinical grade 1 or higher.
[6]Defined where clinically obvious.
[7]Normal laboratory values: bilirubin, 1–22 µM/l; AST, 5–43 U/l; albumin, 34–51 g/l; creatinine, 60–100 µM/l.
[8]Two patients in shunt group treated with haemodialysis prior to surgery.
[9]Centrilobular necrosis and fibrosis scores: 0=none; 1=mild; 2=moderate; 3=severe.
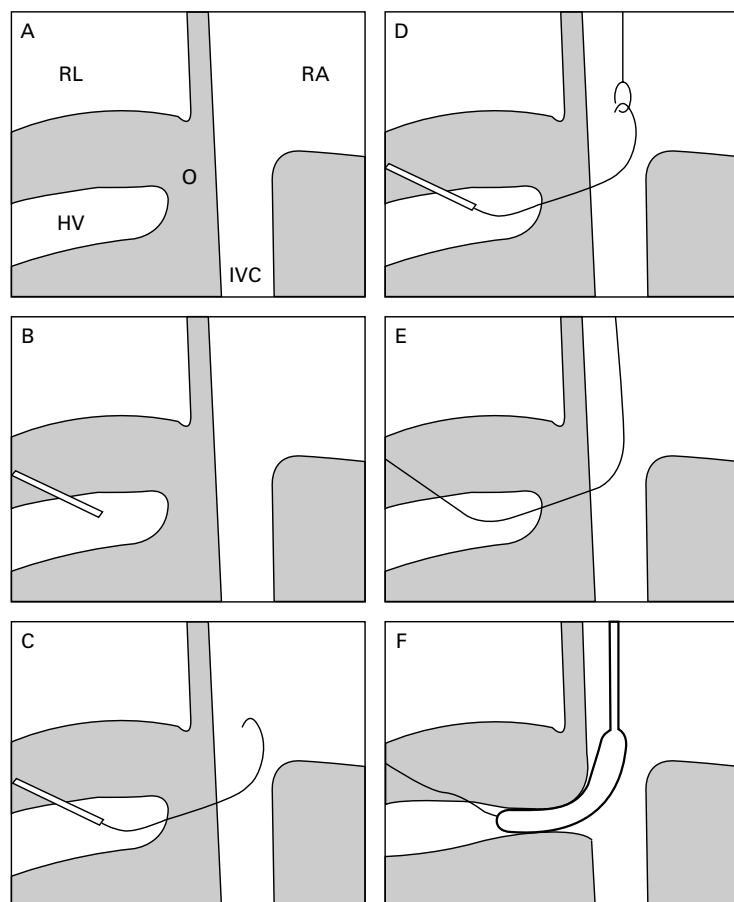AST, aspartate aminotransferase.

Figure 2   Schematic illustration of combined transjugular/transhepatic balloon angioplasty procedure. (A) Hepatic vein occlusion. O = occlusion, HV = hepatic vein, IVC = inferior vena cava, RA = right atrium, RL = right lung. (B) 5F or 6F sheath introduced into HV under ultrasound guidance. (C) Guidewire introduced into IVC by manipulation or by puncture. (D) Transjugular snare wire used to capture guidewire from above. (E) Guidewire pulled proximally out of jugular vein. (F) Balloon catheter introduced from above and angioplasty carried out.

patent venous lumen proximally; this lesion has been defined as a short length hepatic vein stenosis.[15] The initial therapeutic approach was transjugular in all cases; in selected cases where this approach failed further attempts were made via a transhepatic route with insertion of a percutaneous transhepatic guidewire and snaring of the guidewire with a wire loop inserted from the transjugular route. Transjugular balloon angioplasty to a maximum of 14 mm diameter was then done. In selected cases repeated procedures were undertaken to confirm or improve hepatic venous outflow, and in selected cases where residual thrombus was visualised the transvenous catheter was left in place for regional thrombolysis. If there was early reocclusion or severe residual stenosis, a self expanding metal stent was inserted to maintain venous patency. Figures 2, 3, and 4 illustrate these techniques. Angioplasty was considered to have been successful if restoration of or clear improvement in hepatic venous outflow was shown by contrast angiography and if the patients' symptoms stabilised or improved. In the first three cases of PHVA, patients did not receive anticoagulant treatment following the procedure. Thereafter we revised our management to include anticoagulation, with heparin followed by warfarin with

dosage adjustment to maintain the prothrombin INR between 2.5 and 4.0. Further ultrasound with Doppler flow imaging was carried out at regular (six monthly) intervals, and we have adopted a policy of contrast venography at annual intervals for most patients with repeat angioplasty if any recurrent stenosis has occurred. If PHVA was technically impossible or failed to relieve symptoms, the patient was referred for surgical portosytemic shunting.

SURGICAL SHUNTING

Patients were generally selected for shunting if they had no identifiable hepatic veins amenable to angioplasty, or if thrombolysis and/or angioplasty had failed to relieve symptoms, provided that the portal and mesenteric veins were considered to be patent. Mesocaval shunts were used preferentially, with mesoatrial shunts reserved for patients with suprahepatic IVC occlusion. Initially patients were shunted using woven Dacron grafts (n=6); we later used polytetrafluoroethylene grafts (n=5) and have more recently used autologous jugular vein grafts (n=7). All shunt procedures except one were performed by the same surgeon (JACB). All patients were anticoagulated postoperatively with heparin followed by warfarin, with dosage adjusted to maintain the prothrombin INR between 2.5 and 4.0. Patients with extensive portal vein thrombosis or end stage liver disease were not considered for shunting.

Results

PROCEDURES AND OUTCOME DURING FIRST ADMISSION

PHVA

In this group, 10 patients were treated by transjugular angioplasty alone (with more than one therapeutic transjugular procedure in five patients) and 11 were treated by combined transjugular and transhepatic procedures (with more than one therapeutic transhepatic procedure in four, including one patient with associated caval stenosis who underwent transjugular, transfemoral, and transhepatic procedures). Six of 21 patients had thrombolytic agents infused at the site of occlusion following transvenous catheterisation, and 4/21 had hepatic vein stents inserted. Intrahepatic luminal venous pressure measurements both before and after angioplasty were available for seven patients; median pressure before treatment was 34 mm Hg (range 25–39); after treatment median pressure was 19 mm Hg (range 12–30), with a median reduction in pressure of 15 mm Hg (range 7–24). Intraluminal pressures were not generally measured in patients undergoing transhepatic procedures. Early mortality was 2/18 (fig 5); both deaths occurred early in our series (in 1988 and 1990 respectively). The first death was in a patient with combined hepatic vein (HV) and IVC occlusion with hepatic encephalopathy and hepatorenal syndrome; a hepatic vein-right atrial channel was formed by the transjugular route but the patient died within 24 hours from circulatory failure; permission for postmortem examination was refused. The second case was a patient who had undergone transjugular
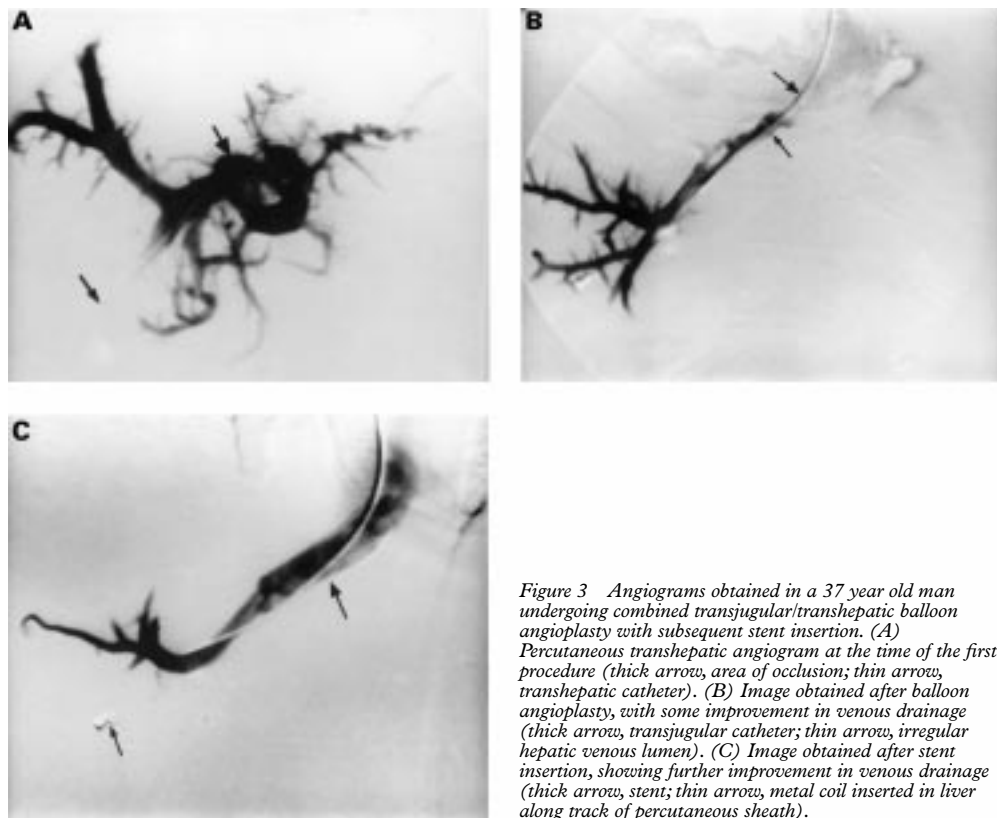
Figure 3 Angiograms obtained in a 37 year old man undergoing combined transjugular/transhepatic balloon angioplasty with subsequent stent insertion. (A) Percutaneous transhepatic angiogram at the time of the first procedure (thick arrow, area of occlusion; thin arrow, transhepatic catheter). (B) Image obtained after balloon angioplasty, with some improvement in venous drainage (thick arrow, transjugular catheter; thin arrow, irregular hepatic venous lumen). (C) Image obtained after stent insertion, showing further improvement in venous drainage (thick arrow, stent; thin arrow, metal coil inserted in liver along track of percutaneous sheath).

angioplasty of a short length hepatic vein stenosis but developed early recurrence of stenosis, and died of a variceal haemorrhage while further procedures were being considered. Postmortem examination confirmed hepatic vein occlusion with evidence of recent thrombus. There were no other complications relating to the procedures or anticoagulation.

Three patients were referred for surgery because of failed angioplasty. In the first patient transhepatic angioplasty was attempted but the hepatic vein occlusion could not be traversed. The second patient had transjugular angioplasty of one hepatic vein branch, and was being considered for a transhepatic procedure on a second patent hepatic vein branch;

however this branch became occluded with progression of symptoms and so the patient underwent emergency surgery. The third patient had transhepatic angioplasty with the catheter left in situ for local thrombolysis; however the catheter became blocked and plans for further procedures were abandoned.

The median length of hospitalisation for surviving patients was eight days (range 2–41).

Surgical shunting
In this group, mesocaval shunts were used for 16/18 cases (including the three patients who had failed angioplasty). Mesoatrial shunts were used in two cases where complete IVC occlusion was present. Postoperative mortality (less than 30 days) was 5/18, including both patients undergoing a mesoatrial shunt (fig 5). Four of five patients who died had hepatic encephalopathy and 2/5 were receiving renal support prior to surgery. Four of five of these cases had diffuse hepatic vein occlusion while
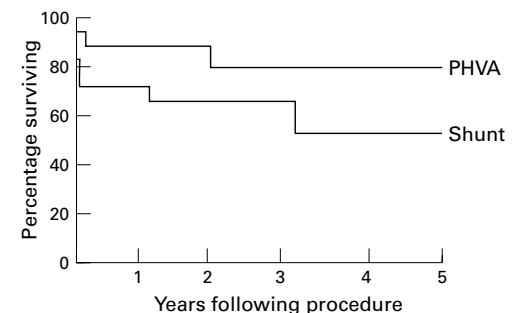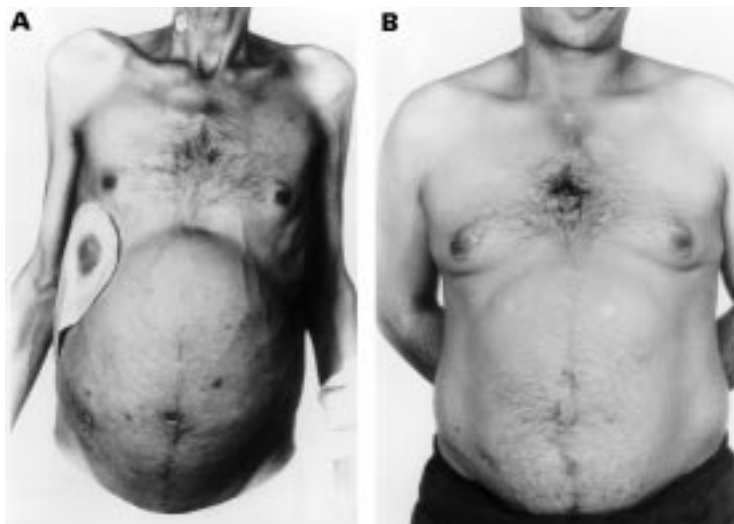

Figure 5 Actuarial survival curves following PHVA or surgical shunting for BCS.

Table 3 Current clinical status in survivors (divided according to definitive treatment)

-	PHVA (n=15)	Surgical shunting (n=10)	Significance (p value)
Duration of follow up, months[2]	36 (12–103)	67 (18–115)	NS[1]
Return to employment or education	9/9	8/9	
Receiving treatment for			
myeloproliferative disease	1	3	
Clinical features[3]			
Ascites	1	2	
Oedema	1	2	
Diuretic treatment	2	3	
Hepatic encephalopathy	0	1	
Laboratory findings[2]			
Serum bilirubin (µM/l)	13 (4–39)	28 (14–42)	p<0.01
Serum AST (U/l)	24 (15–44)	34 (25–56)	p<0.05
Serum albumin (g/l)	45 (37–49)	41 (34–44)	p<0.01

[1]Mann-Whitney U test; p values ≥0.05 are denoted as not significant (NS).
[2]Median values (range given in brackets).
[3]Absolute numbers.
AST, asparate aminotransferase.

the fifth had a short segment stenosis but had not had angioplasty attempted. In all cases death was due to development of multiorgan failure, occurring a median of two days (range 2–16) postoperatively. The median length of hospitalisation for surviving patients was 15 days (range 8–28).

LATE MORBIDITY AND MORTALITY
PHVA
Complete or near complete occlusion of previously recanalised hepatic veins occurred in 6/16 surviving patients, leading to recurrence of symptoms in 4/6 cases. The median delay to reocclusion was 2 years 11 months (range 1 year 8 months to 5 years 3 months). Recurrent occlusion was associated with subtherapeutic or absence of warfarin treatment in 3/6 cases. Further angioplasty was successful in 5/6 cases; one of these was complicated by haemobilia which was succesfully treated by embolisation of an intrahepatic arterial pseudoaneursym. In the remaining case reocclusion was asymptomatic and no further attempts at recanalisation were made. Late death occurred in 1/16 patients (fig 5). This was a patient with combined hepatic vein and IVC stenosis who had two stents inserted at the time of her initial procedure; the upper stent later migrated towards the atrium leading to recurrent tachycardia. Combined cardiac and abdominal surgery to remove this stent was undertaken 18 months later, but the patient died from multiorgan failure after prolonged hospitalisation. There were no other episodes of serious morbidity in this group although most patients are undergoing annual transjugular venograms to check for hepatic venous patency, with further dilatation if any restenosis has occurred.

Surgical shunting
Shunt occlusion occurred in 6/13 surviving patients, leading to recurrent symptoms in all cases. The median delay to reocclusion was 2 years 7 months (range 2 months to 7 years 10 months), involving four Dacron and two jugular grafts. Shunt occlusion was associated with subtherapeutic warfarin dosage in 3/6 cases (all Dacron shunts). One of these cases was a patient who was unable to obtain warfarin treatment after returning to his native country

in the Middle East; this patient died. The remaining five cases of shunt occlusion were managed by percutaneous transfemoral dilatation (two patients, with stenting of the stenosis in one case), attempted portacaval shunt, insertion of TIPSS at another centre, and thrombolytic treatment (one patient each). The attempted portacaval shunt and the thrombolytic treatment were both unsuccessful and these patients died (fig 5). There were no other deaths in this group.

CURRENT CLINICAL STATUS IN SURVIVORS
Duration of follow up for survivors has been longer for the group undergoing surgical shunts (n=10; median 67 months, range 18–115) compared with the PHVA group (n=15; median 36 months, range 12–103, difference not significant). Table 3 summarises clinical and laboratory features. There are no significant differences in the incidence of residual symptoms, which is low in each group, and none of the patients are clinically jaundiced although serum levels of bilirubin remain higher (p<0.01) in survivors of surgical shunting. In the surgical shunting group, one patient is being treated for chronic hepatic encephalopathy and 2/10 have developed isolated hepatic nodules noticed at routine follow up ultrasound scanning. Both patients have undergone targeted liver biopsy to determine the nature of the nodules and histology showed normal or regenerative liver in each case. The majority or all of the patients in each group who were in employment or full time education prior to their treatment have been able to return to these activities.

Discussion
Our review suggests that excellent medium to long term recovery from BCS may be expected in the majority of patients in whom hepatic venous drainage is restored by PHVA. The lower mortality in this series in comparison with surgical portosystemic shunting is probably largely reflective of the more severe risk factors at presentation in patients who were managed by shunting alone. A lower mortality in patients with BCS caused by SLHVS compared with those with diffuse hepatic vein occlusion has been recognised previously.[12 15] Nevertheless, some patients in our series who were managed successfully with PHVA had presented with advanced liver disease characterised by variceal bleeding and hepatic encephalopathy, which suggests that the role for PHVA in BCS is not limited merely to those with mild disease.

From this retrospective comparison we are unable to draw firm conclusions regarding the relative benefits of PHVA and surgical shunting in patients wtih SLHVS. Long term morbidity may occur from both procedures and in patients undergoing PHVA we require annual check venograpy to ensure patency of the hepatic vein. However, we believe that restoration of physiological venous drainage from the liver is preferable to the diversion of blood from the liver that is consequent on portosystemic shunting, and may be associated with a lower

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          ┌──────────────────────────┐
          │  Symptomatic Budd-Chiari  │
          │   syndrome suspected      │
          └──────────────────────────┘
                     │
      Doppler US or other non-invasive imaging
          (e.g. contrast CT or MR)
            ╱                    ╲
  Abnormal venous outflow    Normal venous outflow
            │                    │
  Hepatic venography with     Consider other diagnosis
  intraluminal pressure
  measurements
        ╱          ╲
Short length hepatic   Diffuse venous occlusion
vein stenosis or
occlusion                    ╱           ╲
    │              Mesenteric angiography¹
 Angioplasty        +/– anticoagulation
   ╱     ╲              ╱          ╲
Successful  Failed  PV + MV patent  PV + MV occluded
             │                          │
      Transhepatic approach     Consider any porto-
          ╱        ╲            systemic shunt possible²
    Successful    Failed—Portosystemic shunt²
        │                (TIPSS or surgical)
  Anticoagulate + arrange
  further elective procedures
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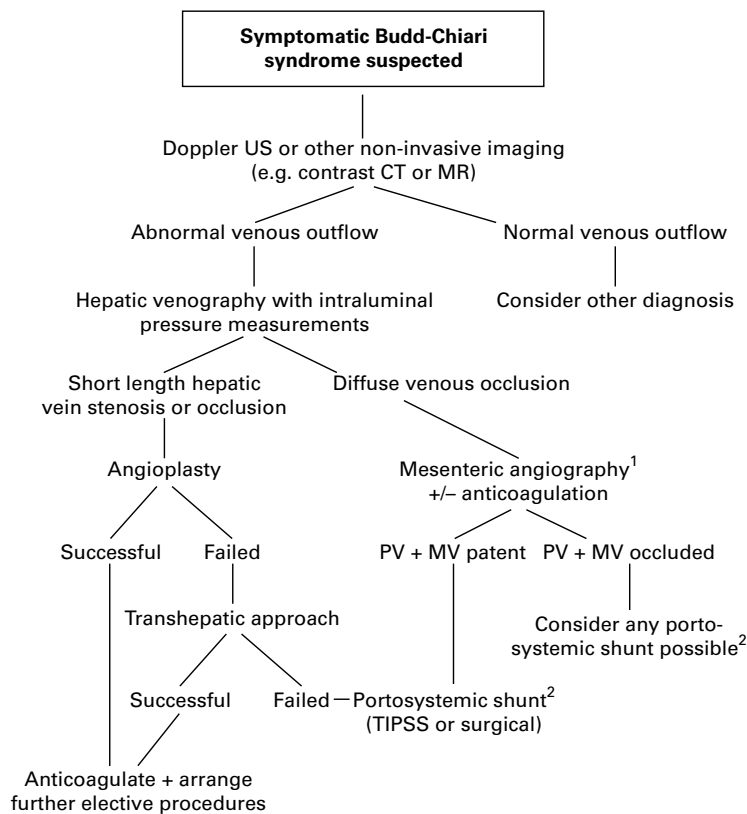
Figure 6   Suggested algorithm for management of BCS. ¹Contrast CT or MR may obviate need for direct angiography. ²Liver transplantation may be appropriate for fulminant or end stage disease.

risk of liver failure and/or hepatic encephalopathy. One survivor of surgical shunting in our series has chronic hepatic encephalopathy, a recognised occasional sequel of portocaval shunting for BCS which has not been reported following PHVA.[16] Furthermore, restoration of hepatic venous drainage may theoretically reduce the extent of architectural disturbance that normally follows BCS treated conservatively or by portosytemic shunting, as nodular regenerative hyperplasia is presumed to be a consequence of disordered vasculature such as that occurring in BCS.[17] Persisting fibrosis and nodule formation have been reported in a high proportion of long term survivors of BCS treated by conventional means, with development of hepatocellular carcinoma in some cases.[18] [19] The duration of follow up for patients treated with PHVA in our series has been relatively short, but at present we have not encountered any cases of nodule formation in these patients on routine follow up ultrasonography; this is now the subject of a prospective evaluation by computed tomography (CT) or magnetic resonance (MR) imaging.

Our experience of reocclusion of hepatic veins following angioplasty where warfarin was not given (in early cases) or was subtherapeutic has led us to believe that strict anticoagulation (with prothrombin INR greater than 2.5) is mandatory to help prevent reocclusion, as is the case for surgical shunts.[20] Excellent long term results following angioplasty for caval webs have been reported without the use of warfarin in oriental series,[21] [22] but our experi-

ence and that of others suggests that, following angioplasty, hepatic vein webs are more likely to restenose compared with caval webs.[22]

We suggest that in patients with symptomatic BCS in whom patent intrahepatic vein branches can be identified by Doppler studies, strong consideration should be given towards therapeutic radiological procedures to restore hepatic venous outflow and we propose an algorithm for management of BCS incorporating this therapeutic approach (fig 6). If BCS is suspected clinically, ultrasound and Doppler studies should be done and this is likely to confirm or refute the diagnosis in the majority of cases.[23] If ultrasound confirms BCS or if there is uncertainty then contrast hepatic venography should be undertaken and angioplasty attempted if SLHVS is confirmed. This may require repeated, including transhepatic, procedures and/or stent insertion as described earlier in order to obtain sustained improvement in hepatic venous outflow. If this approach is successful then clinical recovery should follow although full recovery is occasionally slow (as illustrated in fig 4). A portosystemic shunt should be considered where angioplasty is unsuccessful or clearly fails to resolve symptoms, or if diffuse hepatic vein occlusion is present, provided that the portal and mesenteric veins are patent. The choice of portocaval, mesocaval, or mesoatrial shunt may depend on the preference of the surgeon and the presence or absence of a caval stenosis; similar outcomes have been reported for each of these procedures,[1] [2] [4] [6] although the use of the mesenteric vein rather than the portal vein for shunting will be less likely to compromise any future liver transplant in the rare circumstance where this may be required. As an alternative to a surgical shunt, a TIPSS may be considered; there have been few reports of this procedure for BCS but the experience of ourselves and others is that this may relieve symptoms effectively in selected cases.[24] If portal vein thrombosis is present at the outset, therapeutic options are limited; in this situation mesenteric angiography and early anticoagulation may help while consideration is given to any possible portosystemic shunting procedure or liver transplantation.[25] Following shunt surgery or angioplasty long term anticoagulation should be given (after screening for any possible thrombophilic disorder) with close radiological follow up and further interventional procedures as described earlier. Patients with advanced liver failure and diffuse hepatic vein occlusion should be considered for liver transplantation.

In summary, we have found that patients with symptomatic BCS and moderate to severe liver failure may be managed successfully by PHVA with appropriate case selection. Short and medium term morbidity and mortality for this procedure compare favourably with surgical shunting, although further follow up is required to assess for putative long term benefits.

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