LIVER DISEASE

Favourable medium term outcome following hepatic vein recanalisation and/or transjugular intrahepatic portosystemic shunt for Budd Chiari syndrome

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Background: We report our experience with management of patients with Budd Chiari syndrome over the past two decades. In 1996 we described a novel approach involving recanalisation of hepatic veins by combined percutaneous and transvenous approaches. This was incorporated into an algorithm published in 1999 in which our preferred treatment for all cases of Budd Chiari syndrome with short segment occlusion or stenosis of the hepatic veins involves recanalisation of the hepatic veins by transvenous or combined percutaneous-transvenous approaches. In symptomatic Budd Chiari syndrome where recanalisation is not possible, we perform transjugular intrahepatic portosystemic shunts (TIPS) because TIPS decompresses the portal circulation directly in an adjustable way. In this series of patients with Budd Chiari syndrome treated with radiological interventions alone, we assess their medium term outcome using two independent objective prognostic indices.

Methods: We retrospectively studied 61 patients with non-malignant Budd Chiari syndrome treated by radiological intervention alone in our centre.

Results: Actuarial survival for the entire cohort at one year and five years was 94% and 87%, respectively. Survival of our patients with mild disease (according to the Murad classification) was 100% at one year and at five years, with intermediate disease severity 94% at one year and 86% at five years, and with severe disease 85% at one year and 77% at five years.

Conclusion: Management of Budd Chiari syndrome by interventional radiology resulted in excellent medium term survival for patients in all categories of disease severity.

The wide variation in the management of Budd Chiari syndrome between centres could partly be explained by the lack of controlled clinical trials for this disease.¹ The prognostic index scores derived from and validated in patients with Budd Chiari syndrome permit objective assessment of disease severity and can be used to analyse efficacy of treatment.^{2–3} Of patients with Budd Chiari syndrome treated by medical treatment alone or by surgical portosystemic shunts, those with more severe disease, as reflected by higher prognostic index scores at presentation, had a significantly poorer survival compared with those with less severe disease.^{2–3}

We have previously reported our early results of hepatic vein recanalisation in Budd Chiari syndrome via interventional radiological methods, including balloon dilatation and hepatic vein stenting.^{4–5} We published an algorithm of the management of Budd Chiari syndrome emphasising the role of hepatic vein recanalisation and transjugular intrahepatic portosystemic shunt (TIPS).⁶ We hereby report the medium term outcome in patients with Budd Chiari syndrome who were treated by interventional radiology alone, applying recently published prognostic indices to objectively quantify disease severity at presentation.

METHODS

Study population and details of therapeutic intervention

We conducted a retrospective study of hospital records of patients with Budd Chiari syndrome managed in the Liver Unit in our hospital from 1984 to March 2004. In patients suspected of having Budd Chiari syndrome by clinical features or liver biopsy, hepatic venous outflow obstruction

was confirmed by imaging tests (Doppler ultrasound, computed tomography or magnetic resonance imaging scan, or hepatic venogram). Of 115 patients with Budd Chiari syndrome seen during the study period, three with malignancy obstructing hepatic venous outflow were excluded.

We further excluded 44 patients treated by modalities other than interventional radiology alone from the analysis. These 44 patients were treated by mesocaval shunt surgery (nine), mesoatrial shunt surgery (two), liver transplantation (11), mesocaval shunt surgery followed by percutaneous angioplasty of stenosed/thrombosed shunt or TIPS or hepatic vein recanalisation (seven), mesocaval shunt surgery after failed hepatic vein recanalisation (one), liver transplantation followed by TIPS (one), and medical treatment only (13), which consisted of anticoagulation, diuretics, and paracentesis as well as treatment of complications and the underlying cause. Case records of seven patients could not be traced. Thus our study population comprised 61 patients treated by interventional radiology alone.

Selection of radiological intervention

Selection of the radiological intervention to be performed has been described elsewhere.⁷ Briefly, the intervention was chosen after assessment by ultrasound to see if there was a large patent segment(s) of hepatic vein(s) with potential for flow restoration by balloon dilatation or stenting. Our first choice was restoration of some hepatic venous outflow by these methods (recanalisation). In cases where there was a good segment(s) of patent hepatic vein(s), we attempted

Abbreviations: TIPS, transjugular intrahepatic portosystemic shunt; MELD, model for end stage liver disease

Figure 1 Hepatic vein recanalisation by combined transhepatic/transjugular procedure. Long patent segment of hepatic vein seen on ultrasound, amenable to recanalisation (A). Dilated obstructed hepatic venous segment seen on percutaneous transhepatic venogram with filling of intrahepatic collateral veins (B). Percutaneous transhepatic route used to pass a thin catheter and guidewire across hepatic vein block (C) and to pass a catheter into the inferior vena cava (IVC). Note transjugular catheter in the IVC. The transhepatic wire is snared in the IVC and brought up out through the jugular vein sheath. Hepatic venogram after transjugular balloon dilatation of the hepatic venous stenosis still shows relative obstruction (D). Normal flow of contrast from hepatic vein into the IVC after transjugular stenting of hepatic vein stenosis (E). I, catheter in IVC; P, percutaneous transhepatic catheter; T, transjugular transhepatic guidewire.

transjugular catheterisation with a view to balloon dilatation or stent insertion. If access could not be gained from the cava, then ultrasound guided transhepatic needle puncture of the vein was performed to allow traversal of the blocked vein segment out to the inferior vena cava. In such a case the wire was then snared and pulled out through the jugular vein to enable balloon dilatation or stent insertion without making the puncture track in the liver any larger (fig 1). Stent insertion was normally required if there was insufficient response to balloon dilatation alone or rapid re-stenosis/occlusion or recurrent re-stenosis after dilatation. In selected cases thrombolytic therapy was used as an adjunct to these procedures.⁸ If there were no suitable hepatic veins for recanalisation found by ultrasound or hepatic venography or if the procedure failed, then we offered TIPS to symptomatic patients to decompress the liver. Our previously described algorithm⁶ for treatment of patients with Budd Chiari syndrome was followed in all new patients since 1999.

All radiological interventions were classified as minor if transjugular and/or transfemoral balloon dilatation of the hepatic vein and/or inferior vena cava was performed. A major intervention was defined as combined transhepatic/transjugular balloon dilatation of the hepatic vein and/or stenting of the hepatic vein or TIPS procedure.

Follow up after radiological intervention

Venography and pressure studies were performed at six months following the radiological intervention (or earlier if hepatic vein or TIPS occlusion was suspected) to assess the patency of hepatic venous outflow or TIPS. Subsequent follow up of hepatic venous outflow or TIPS was done by six

monthly Doppler ultrasound with annual venography for TIPS cases. All patients were followed up regularly, both to monitor hepatic venous outflow or TIPS patency and to detect overt manifestation or evolution of the underlying thrombophilic condition.

Table 1 Baseline characteristics of patients with Budd Chiari syndrome treated by interventional radiology alone

	Hepatic vein recanalisation (n = 31)	TIPS (n = 26)	Both procedures (n = 4)
Sex (M/F)	14/17	7/19	1/3
Age (y)*	36 (16–60)	44 (21–67)	30 (23–42)
Follow up (months)*	53 (0–181)	35 (0–93)	80 (40–120)
Child's score			
A	8	0	1
B	19	10	1
C	4	16	2
MELD score*	10 (2–23)	17 (7–34)	16 (8–28)
PI (Zeitoun ²)†			
≤ 5.4	19	5	1
> 5.4	12	21	3
PI (Murad ³)†			
Class I	12	3	1
Class II	17	14	1
Class III	2	9	2

*Median (range).

†Prognostic index.

MELD, model for end stage liver disease (Mayo model).

Table 2 Actuarial survival in patients with Budd Chiari syndrome treated by interventional radiology alone

Baseline disease severity*	n	1 y survival (95% CI)	5 y survival (95% CI)
PI $\leq$ 5.4	25	100%†	89% (86–100%)
PI $>$ 5.4	36	95% (79–99%)	82% (69–95%)

Actuarial survival in the two subgroups of patients with a prognostic index (PI) $\leq$ 5.4 and $>$ 5.4 was not significantly different (log rank test, $p=0.082$).

*Disease severity stratified using PI described by Zeitoun and colleagues.² In a study of 120 patients with Budd Chiari syndrome, 88 of whom had surgical portosystemic shunts, they reported actuarial one and five year survival of 100% and 94% (SEM 5) in patients with PI $\leq$ 5.4 and 77% (SEM 8) and 62% (SEM 10) in patients with PI $>$ 5.4, respectively.

†100% survival, so confidence intervals not available.

Gradation of disease severity at presentation and statistical analysis

Severity of disease at initial presentation to our centre was graded by two different prognostic indices.^{2,3} Actuarial survival at one and five years was analysed in patients in different categories of baseline disease severity using the Kaplan-Meier method.

RESULTS

Description of study population

Sixty one patients were treated by interventional radiology alone, of which 32 were referred to our unit after 1999 when the treatment algorithm came into place.⁶ They were treated with hepatic vein recanalisation (31), TIPS (26), or both procedures (four). (In two patients who presented in 1997 and 1999, TIPS procedure failed and they were treated by mesocaval shunt, one of whom subsequently underwent TIPS in 2002 when he had occlusion of the mesocaval shunt—these 2 patients are not included in the study.) The 61 patients (22 males; median age 36 years (range 16–67)) were followed up for a median of 52 months (range 0–181). The time interval between the diagnosis of Budd Chiari syndrome to radiological intervention was $<$ 1 month (43 patients), 1–2 months (four patients), 2–3 months (three patients), and $>$ 3 months (11 patients). In 15 patients minor radiological procedures were performed as the initial definitive intervention (transjugular balloon dilatation of the hepatic vein in 12 patients, transjugular and/or transfemoral balloon dilatation of the inferior vena cava in three patients). Forty six patients required major interventions for initial definitive

Table 3 Actuarial survival in patients with Budd Chiari syndrome treated by interventional radiology alone

Baseline disease severity*	n	1 y survival (95% CI)	5 y survival (95% CI)
Class I	16	100%†	100%†
Class II	32	94% (86–100%)	86% (73–99%)
Class III	13	85% (65–100%)	77% (53–100%)

Overall, the difference in survival between all three classes was not significant (log rank test, $p=0.2$). Difference in survival between classes I and III approached significance (log rank test, $p=0.062$).

*Disease severity stratified using prognostic index described by Murad and colleagues.³ In a study of 237 patients with Budd Chiari syndrome, 117 of whom had surgical portosystemic shunts, they reported actuarial survival at one year of 93% (95% CI 86–100%) in class I, 85% (95% CI 78–92%) in class II, and 60% (95% CI 47–73%) in class III disease severity, and at five years 89% (95% CI 79–99%) in class I, 74% (95% CI 65–83%) in class II, and 42% (95% CI 28–56%) in class III disease severity, respectively.

†100% survival, so confidence intervals not available.

treatment (combined transhepatic/transjugular balloon dilatation of the hepatic vein in nine patients, stenting of the hepatic vein in eight patients, TIPS in 29 patients). While polytetrafluoroethylene covered TIPS stents were inserted at the initial intervention in six patients and at subsequent reintervention after initial bare TIPS stent placement in three patients, all other TIPS procedures were performed using bare stents. Additionally, thrombolysis using recombinant tissue plasminogen activator was performed as an adjunctive procedure in six patients.

Baseline disease severity in the study cohort

As shown in table 1, patients who underwent hepatic vein recanalisation had lesser degrees of liver failure at baseline (assessed by Child's score, model for end stage liver disease (MELD) score, and prognostic indices) compared with those who underwent TIPS.

Adjunctive therapy

Immediately after hepatic vein recanalisation or TIPS, all patients were anticoagulated with heparin which was changed to warfarin in a few days (aiming for a target international normalised ratio of 2.5–3.5). Patients with myeloproliferative diseases were also treated with hydroxyurea or venesection. In 15 patients with essential thrombocythemia or recurrent shunt thrombosis despite adequate anticoagulation, aspirin 150 mg daily was also added.

Underlying prothrombotic state

An underlying prothrombotic state was identified in 31 of 61 patients. These included 17 patients with myeloproliferative disorders—polycythemia rubra vera ($n=7$), essential thrombocythemia ($n=6$), myelofibrosis ($n=1$), and unclassified myeloproliferative disorders ($n=3$). One patient with homozygous and four with heterozygous mutations of factor V Leiden gene were detected out of 21 patients tested. The other thrombophilic states included paroxysmal nocturnal haemoglobinuria ($n=4$), systemic lupus erythematosus ($n=1$), antiphospholipid syndrome ($n=1$), methylene tetrahydrofolate reductase gene mutation ($n=1$), and prothrombin gene mutation ($n=1$). Ten of the 39 women were on oral contraceptives, prior to the diagnosis of Budd Chiari syndrome. Other associated diseases included ulcerative colitis ($n=1$), coeliac disease ($n=1$), myelodysplastic syndrome ($n=1$), neurosarcoidosis ($n=1$), and history of multiple cancers in the past ($n=1$).

Survival after radiological intervention

Actuarial survival in the entire study cohort at one and five years was 94% (95% confidence interval (CI) 88–100%) and 87% (95% CI 78–96%), respectively. As shown in tables 2 and 3, excellent medium term outcome was achieved after interventional radiology for patients in all categories of baseline disease severity. Although a significant difference in survival was not noted between patients with varying grades of baseline disease severity, there was a trend towards a significant difference in survival between patients with mild disease (class I) and those with severe disease (class III), as shown in table 3. Of the nine patients who died, the cause of death was attributable to radiological intervention in two patients (patient Nos 3 and 9 in table 4).

Complications and follow up after radiological intervention

Complications of radiological intervention were bleeding hepatic artery pseudoaneurysm after transjugular liver biopsy performed subsequent to hepatic vein recanalisation and thrombolysis in one—managed by embolisation—and contrast induced renal failure in two (one died, she also had

Table 4 Cause of death in 61 patients treated by interventional radiology alone

	Age (y)/ sex	Treatment	Cause of death	Comorbid diseases	Time to death after treatment
1	36/F	Hepatic vein recanalisation	Decided against further active treatment in view of poor general condition	Neurosarcoid	13 months
2	48/M	TIPS	Massive cerebral infarct, severe haemolysis, renal failure	PNH	16 months
3	56/F	TIPS	Culture negative neutrocytic ascites, septicaemia, acute renal failure, worse after contrast	PRV	3 days
4	59/M	TIPS	Subdural haematoma	PNH	1 month
5	36/F	Hepatic vein recanalisation	Not known	Coeliac disease	120 months
6	27/M	Hepatic vein recanalisation	Not known	None	41 months
7	23/F	Hepatic vein recanalisation	Not known	None	139 months
8	52/F	TIPS	Subdural haematoma	None	6 months
9	67/M	TIPS	Worsening hepatic encephalopathy	None	12 days

PNH, paroxysmal nocturnal haemoglobinuria; PRV, polycythemia rubra vera; TIPS, transjugular intrahepatic portosystemic shunt.

culture negative neutrocytic ascites and septicaemia). Five patients developed grade I hepatic encephalopathy after TIPS (they were not in encephalopathy prior to TIPS) which responded to treatment with lactulose.

During the follow up period, 40 of the 61 patients underwent repeated radiological interventions (median minor procedures 1 (range 0–13); median major procedures 0 (range 0–4)) when necessary to maintain adequate patency of hepatic venous outflow or TIPS. We analysed the need for reinterventions over the different time periods of the study. From 1984 to 1988, of two patients treated, both had 0 major and two minor reinterventions each. From 1989 to 1993, the five patients treated underwent 0–3 major (median 0) and 1–13 minor (median 5) reinterventions. From 1994 to 1998, the 22 patients treated underwent 0–1 major (median 0) and 0–9 minor (median 1) reinterventions. From 1999 to March 2004, the 32 patients treated underwent 0–3 major (median 0) and 0–5 minor (median: 0) reinterventions.

Patients who were in hepatic encephalopathy at presentation

Of the 61 patients, one was in grade I hepatic encephalopathy and another was in grade IV hepatic encephalopathy at presentation; two other patients were sedated and on mechanical ventilation at transfer to our hospital. Three of the four patients who were in hepatic encephalopathy at presentation had a correctable precipitating factor for hepatic encephalopathy (oesophageal variceal bleed in two, bacteraemia in one) which was treated (endoscopic therapy for oesophageal varices, antibiotics) and underwent hepatic vein recanalisation (in one patient) or TIPS (in two patients). All three patients were alive at follow up, ranging from 12 to 116 months. In contrast, one patient (patient No 9 in table 4) who was in grade I hepatic encephalopathy at presentation did not have any precipitating factor for encephalopathy other than hepatic venous outflow obstruction. He developed rapid deterioration of sensorium and progressive liver failure after TIPS and died 12 days after the procedure.

Patients with underlying myeloproliferative disorder

Of the 17 patients with underlying myeloproliferative disease, one died—the cause of death was attributable to radiological intervention and not to the myeloproliferative disorder per se (table 4). Median follow up in the 17 patients with underlying myeloproliferative disorder was 90 months (range 0–108).

DISCUSSION

This is the first study of the efficacy of interventional radiology in the management of Budd Chiari syndrome which has applied prognostic indices to stratify patients into different grades of disease severity at presentation. We report excellent medium term survival in all categories of disease severity. A trend towards statistically significant difference in survival was noted between patients with mild and severe disease (table 3) and this needs to be analysed further in studies including larger number of patients.

The prognostic index for Budd Chiari syndrome derived by Zeitoun *et al* was based on a multicentre study, conducted from 1970 to 1992, of 120 patients with Budd Chiari syndrome (88 treated by surgical portosystemic shunts and 38 had only medical treatment).² The prognostic index derived by Murad *et al* was based on a multicentre study of 237 patients, conducted from 1984 to 2001, in which 117 patients had surgical portosystemic shunts and 39 had only medical treatment.³ The present study was conducted in a single centre from 1984 to March 2004 and analysed patients who were treated by interventional radiology alone. Thus differences in study design do not permit comparison of the efficacy of different treatment modalities. Also, in view of the retrospective analysis of our study it is not possible to say whether patients died before they reached our unit. However, none of the patients in whom the diagnosis of Budd Chiari syndrome was made and who were transferred to our centre died while waiting for radiological intervention.

The treatment offered to patients with Budd Chiari syndrome at our centre over the past 21 years has evolved from liver transplantation or surgical portosystemic shunts to percutaneous hepatic vein recanalisation or TIPS (fig 2). Benefits from restoring physiological hepatic venous outflow and its less invasive nature makes hepatic vein recanalisation superior to the other treatment modalities. In patients with diffuse occlusion of hepatic veins precluding recanalisation or when recanalisation is not technically feasible, TIPS is the next preferred treatment modality.⁶

Our learning curve for these procedures parallels the evolution of interventional radiology techniques over the years. Initially we were only able to provide balloon dilatation of short length venous stenoses or occlusions. With the introduction of metal stents, it became possible to stent hepatic veins which did not respond to dilatation or which rapidly restenosed. We were cautious about the use of stents at first but recently stented patients need less follow up venography and/or redilatation compared with those treated

Figure 2 Trends in the treatment of Budd Chiari syndrome at our centre. From 1999, we have followed an algorithmic approach to the treatment of Budd Chiari syndrome.⁶ Since 1999, one patient needed more than one modality of treatment (after a failed attempt at a transjugular intrahepatic portosystemic shunt (TIPS) procedure he had mesocaval shunt surgery; following shunt thrombosis three years later he underwent a successful TIPS) and four patients had medical management alone (asymptomatic diffuse hepatic vein occlusion in one patient, symptomatic diffuse hepatic and portal vein occlusion in two patients, and long segment of discontinuity between the inferior vena cava and right atrium in one patient which precluded interventional radiological treatment).

by balloon dilatation alone. Therefore, we now advocate stent placement immediately after a transhepatic hepatic vein recanalisation or when balloon dilatation is not immediately effective in a stenosis. Also, surgical shunts are now reserved for TIPS failures. TIPS can be more technically challenging in Budd Chiari syndrome but we have only had two complete technical failures (1997 and 1999) and one of these had successful TIPS after late occlusion of a surgical mesocaval shunt. Budd Chiari patients have required more TIPS reinterventions than other patients, which might be because of the increased thrombotic tendency or longer shunts required. Polytetrafluoroethylene covered TIPS stents are superior to bare stents in Budd Chiari syndrome, with improved TIPS patency, lower TIPS dysfunction rate, lower number of reinterventions, and fewer prosthesis requirements.⁹

Short length stenoses of hepatic veins amenable to recanalisation remain under recognised and percutaneous hepatic vein recanalisation is currently an under utilised treatment modality in the management of Budd Chiari syndrome. In a study from France, short length stenoses of hepatic veins were identified in 25 of 86 patients (29%) with hepatic venous outflow obstruction.¹⁰ In 1999, we published an algorithm for managing patients with Budd Chiari syndrome with hepatic vein recanalisation as the preferred first line modality of treatment.⁶ From 1999 to March 2004, applying this management algorithm at our centre, 14 of 34 patients (41%) with Budd Chiari syndrome were detected to have discrete proximal stenoses of hepatic veins and underwent percutaneous recanalisation of the same. As shown in table 1, patients with hepatic vein stenosis amenable to percutaneous recanalisation had lesser degrees of liver failure (assessed by Child's score, MELD score, and prognostic indices) compared with patients with diffuse hepatic vein occlusion.

Chronic hepatic venous outflow obstruction can be well compensated and may be asymptomatic. The current management algorithm for Budd Chiari syndrome at our centre is shown in fig 3. We advocate hepatic vein recanalisation, even if the patient is asymptomatic, and medical treatment as the sole treatment modality is reserved

for the asymptomatic patient in whom recanalisation is not technically feasible due to diffuse occlusion of hepatic veins. Restoration of physiologically directed sinusoidal blood flow not only encourages recovery of liver function but is also likely to remove the stimuli for macronodular regeneration which characterises chronic Budd Chiari syndrome and may predispose to hepatocellular carcinoma.¹¹ Also, restoration of physiology should improve symptoms, quality of life, and decrease the need for diuretics and the risk of complications. These factors will be addressed in future studies.

In our own experience and in other series reported in the literature, surgical portosystemic shunting for Budd Chiari syndrome is associated with significant risk of early death from liver failure.¹²⁻¹³ We postulate that this is in part caused by inadequate decompression of the liver due to shunt malfunction by factors such as torsion of the interposition graft (jugular vein bridging superior mesenteric vein to inferior vena cava) or to high pressure in the inferior vena cava due to Budd Chiari induced compression by the caudate lobe. The results of the current study suggest that radiological recanalisation is a safe alternative. By draining the portal circulation directly into the supracaudate portion of the inferior vena cava, not only does TIPS abrogate these risks entirely but it has the added advantage that the pressure gradient between the portal vein and inferior vena cava can be accurately measured and further adjusted if necessary at the end of the procedure. Although TIPS has been advocated as a bridge to liver transplantation,¹ our experience is to the contrary, with good outcome after TIPS as definitive long term treatment in Budd Chiari syndrome.

Liver transplantation has been considered for patients with Budd Chiari syndrome presenting with hepatic encephalopathy. However, hepatic encephalopathy is an uncommon presentation of Budd Chiari syndrome and there are scant data on treatment outcomes in this situation.¹⁴ In our experience, hepatic vein recanalisation is safer than TIPS in patients with Budd Chiari syndrome presenting with hepatic encephalopathy. If hepatic vein recanalisation is not technically feasible, the potential benefit of recovering hepatocyte function by decompressing the engorged liver by TIPS has to be weighed against the risk of worsening encephalopathy. Of the four patients who presented in hepatic encephalopathy in the current series (one underwent hepatic vein recanalisation, three had TIPSS), three patients had a correctable precipitating factor for hepatic encephalopathy other than Budd Chiari syndrome, and reversal of the precipitating factor, in addition to hepatic venous decompression, probably contributed to the positive outcome in those three patients.

One advantage of interventional radiology in treating Budd Chiari syndrome is the reduced incidence of complications compared with procedures requiring laparotomy. A potential drawback is risk of contrast induced renal failure which can be minimised by less nephrotoxic contrast agents.¹⁵⁻¹⁶

Lifelong anticoagulation, which is advised in Budd Chiari syndrome, needs close monitoring. In the current series, two patients died of subdural haemorrhage during anticoagulation (one of whom had thrombocytopenia due to paroxysmal nocturnal haemoglobinuria) while another died of massive cerebral infarct, despite being on anticoagulation. Liver transplantation for Budd Chiari syndrome can theoretically cure the underlying thrombophilic state, thus obviating the need for lifelong anticoagulation, although this is not always the case as multiple aetiological factors may be present in a single patient.¹⁷

The role of surgical portosystemic shunts and liver transplantation in Budd Chiari syndrome is likely to diminish, in view of the excellent survival possible with hepatic vein recanalisation and/or TIPS. Simultaneous portal vein thrombosis in patients with Budd Chiari syndrome

Figure 3 Current algorithm for management of Budd Chiari syndrome. *Complete portal vein thrombosis precludes transjugular intrahepatic portosystemic shunt (TIPS) (and shunt surgery). Early re-thrombosis on follow up of any patient may be treated by local thrombolysis.

remains a challenging situation with limited treatment options, although occasionally, TIPS insertion may be feasible.^{18, 19}

In conclusion, we have shown that excellent medium term survival is achieved by hepatic vein recanalisation and/or TIPS for Budd Chiari syndrome in all categories of baseline disease severity.

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EDITOR'S QUIZ: GI SNAPSHOT

Answer

From question on page 763

A repeat Doppler duplex ultrasound was performed which revealed a filiform circumscribed stenosis at the site of the portal vein anastomosis, leading to blood flow acceleration up to 100 cm/s (fig 1A). A computed tomography scan confirmed the stenosis in the extrahepatic part of the portal vein.

Transjugular measurement of the hepatic venous pressure gradient (5 mm Hg; normal 4–8 mmHg) by wedged pressure measurement confirmed the suspected prehepatic stenosis. As surgical reanastomosis was considered high risk due to substantial cachexia, a portal angioplasty was planned. Massive ascites prevented percutaneous portal access. Therefore, the portal system was accessed via transjugular transhepatic puncture of the right portal vein. A 95% stenosis was detected by fluoroscopy with a pressure gradient of 30 mm Hg (38/8 mm Hg distal/proximal of the stenosis) (fig 1B). A gradual angioplasty up to 15 mm was performed resulting in complete resolution of the pressure gradient with only minimal residual narrowing (8 mm) (fig 1C, D, E). After intervention, ascites resolved and oesophageal varices disappeared. In the following months the patient gained 18 kg in weight. Unfortunately, 17 months after the acute Budd-Chiari syndrome breast cancer was detected.

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Figure 1 Doppler duplex ultrasound (A) and fluoroscopy (B) of the portal vein stenosis. Gradual angioplasty of stenosis up to 15 mm (C). Fluoroscopy (D) and Doppler duplex ultrasound (E) after dilation with minimal residual narrowing.