The Budd-Chiari syndrome

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R É S U M É

Prérequis : Le syndrome de Budd-Chiari est une affection rare, souvent fatale en l'absence de traitement optimal. Il est caractérisé par une obstruction des voies de drainage veineux hépatique.
But : Préciser la physiopathologie, les étiologies, les moyens diagnostiques et les modalités thérapeutiques de ce syndrome.
Méthodes : Revue de la littérature
Résultats : Le syndrome de Budd-Chiari est une entité complexe avec un large spectre d'étiologies et de présentations. Les anomalies hématologiques, en particulier le syndrome myéloprolifératif, sont la cause la plus fréquente du syndrome de Budd-Chiari. La présentation clinique dépend de l'étendue et la rapidité de l'obstruction veineuse hépatique. L'échodoppler, la tomodensitométrie et l'imagerie par résonnance magnétique des veines hépatiques et de la veine cave inférieure sont des moyens non invasifs permettant le diagnostic de la maladie. Les anticoagulants sont utilisés en première ligne avec les diurétiques et le traitement prophylactique de l'hypertension portale. Pour les sténoses courtes, une angioplastie peut être proposée. En cas d'échec, le recours au TIPS est indiqué et en dernier lieu, une transplantation hépatique. Cette stratégie thérapeutique a permis une survie à 5 ans de 70% environ. Le pronostic à moyen terme dépend de la sévérité de la maladie hépatique.
Conclusion : Le diagnostic du syndrome de Budd-Chiari doit être considéré chez tout patient ayant une maladie aigue ou chronique du foie. La prise en charge thérapeutique devrait suivre une stratégie par étapes.

M o t s - c l é s
Syndrome Budd-Chiari, Ascite, Thromboses, Shunt Transjugulaire Intrahépatique Portosystémique

S U M M A R Y

Background: The Budd-Chiari syndrome is a rare disease, often fatal if not treated optimally. It is characterized by a blocked hepatic venous outflow tract.
Aim: This review attempted to present pathophysiology, aetiologies, diagnosis and therapeutic modalities of the Budd-Chiari syndrome.
Methods: Review of literature.
Results: Budd-Chiari syndrome is a complex disease with a wide spectrum of aetiologies and presentations. Hematologic abnormalities, particularly myeloproliferative disorders, are the most common causes of the Budd-Chiari syndrome. The clinical presentation is governed by the extent and rapidity of the hepatic vein occlusion. Doppler-ultrasound, computed tomography or magnetic resonance imaging of hepatic veins and inferior vena cava are usually successful in demonstrating non-invasively the obstacle or its consequences. A therapeutic strategy has been proposed where anticoagulation, correction of risk factors, diuretics and prophylaxis for portal hypertension are used first; then angioplasty for short-length venous stenosis; then Transjugular Intrahepatic Portosystemic Shunt (TIPS); and ultimately liver transplantation. Treatment progression is dictated by the response to previous therapy. This strategy has achieved 5-year survival rates approaching 70%. Medium-term prognosis depends on the severity of liver disease.
Conclusion: The diagnosis of the Budd-Chiari syndrome must be considered in any patients with acute or chronic liver disease. Management of this syndrome should follow a step by step strategy.

K e y - w o r d s
Budd-Chiari syndrome, Ascites, Thrombosis, Transjugular Intrahepatic Portosystemic Shunt (TIPS)

The Budd-Chiari syndrome is an uncommon disease, often fatal if not treated optimally. Firstly reported in 1845 by George Budd, who described the classic triad of abdominal pain, hepatomegaly and ascites. In 1899, Hans Chiari documented the histopathological features of the syndrome [1, 2]. Budd-Chiari syndrome is defined, according to the European Group for the Study of Hepatic Vascular Diseases, as hepatic venous outflow obstruction at any level from the small hepatic veins to the junction of the inferior vena cava and the right atrium, regardless of the cause of the obstruction. Cardiac etiologies of hepatic congestion or veno-occlusive disease (sinusoidal obstruction syndrome) are not included in this definition [3]. Budd-Chiari syndrome is a complex disease with a wide spectrum of aetiologies and presentations. When the blockage is caused by invasion or compression by a tumor, Budd-Chiari syndrome is considered secondary. Otherwise, it is related to thrombosis and considered primary. This article reviews the current literature with respect to presentation, management and prognosis of the disease.

METHODS

A systematic search of MEDLINE, PubMed and the Cochrane library was performed for articles relevant to the Budd-Chiari syndrome, published between 1980 and 2012. The following search terms were used to search all databases: Budd-Chiari syndrome, portal veins, hepatic venous outflow obstruction, thrombosis, ascites, Transjugular Intrahepatic Portosystemic Shunt (TIPS). The following types of studies were excluded: interviews, case-reports, letters, comments and editorials. The table 1 shows the American recommendation adapted from the American College of Cardiology and the American Heart Association Practice Guidelines.

DISCUSSION

Pathogenesis

Obstruction is usually caused by a thrombus, but may result from extrinsic compression (tumour, abscess, cysts), membranous webs within the inferior vena cava (IVC) [4], or postoperative complications following liver transplantation [5, 6]. Obstruction of the hepatic venous outflow tract results in increased hepatic sinusoidal pressure and portal hypertension. The portal venous stasis and congestion cause hypoxemic damage in adjacent hepatocytes. Afterwards centrilobular fibrosis, nodular regenerative hyperplasia, and ultimately cirrhosis occur. Main alterations in hepatic morphology include atrophy of peripheral regions and hypertrophy of the caudate lobe and central portions of the liver [7, 8]. However, if the hepatic sinusoidal pressure is reduced by the creation of a portosystemic shunt or by the development of a portal venous collateral system, liver function improves [9].

Etiology

Factors that confer a predisposition to the development of the Budd-Chiari syndrome, including hypercoagulable states, both hereditary and acquired, and a variety of other causes, can be identified in about 75 percent of patients. The presence of multiple causes in the same patient is increasingly recognized [10]. In one third of the patients the cause is not visible. Causes of Budd-Chiari syndrome include [10, 11] :

Table 1: Grading System for Recommendations (adapted from the American College of Cardiology and the American Heart Association Practice Guidelines) [13]

Classification	Description
Class I	Conditions for which there is evidence and/or general agreement that a given diagnostic evaluation, procedure or treatment is beneficial, useful, and effective.
Class II	Conditions for which there is conflicting evidence and/or a divergence of opinion about the usefulness/efficacy of a diagnostic evaluation, procedure or treatment.
Class IIa	Weight of evidence/opinion is in favor of usefulness/Weight of evidence/opinion is in favor of usefulness/efficacy.
Class IIb	Usefulness/efficacy is less well established by evidence/opinion.
Class III	Conditions for which there is evidence and/or general agreement that a diagnostic evaluation/procedure/treatment is not useful/effective and in some cases may be harmful.
Level of evidence	**Description**
Level A	Data derived from multiple randomized clinical trials or meta-analyses.
Level B	Data derived from a single randomized trial, or nonrandomized studies.
Level C	Only consensus opinion of experts, case studies, or standard-of-care.

Hematological disorders:
Hematologic abnormalities, particularly myeloproliferative disorders, are the most common causes of the Budd-Chiari syndrome. Their diagnosis can be made by showing the V617F mutation in Janus tyrosine kinase-2 (JAK2). V617F JAK2 has been found in about 80% and 50% of patients with polycythemia Vera and essential thrombocythemia or idiopathic myelofibrosis, respectively [12]. Bone marrow biopsy should be performed in all Budd-Chiari syndrome patients with negative test for the mutation [11]. We do not rule out a diagnosis of myeloproliferative disease solely on the basis of normal or low peripheral blood cell counts (Class I, Level B) [13].
Other hematological causes of the syndrome include paroxysmal nocturnal hemoglobinuria, characterized by complement-induced haemolytic anaemia, and inherited deficiencies of protein C, protein S, and anti thrombin III [14]. However, in patients with inherited deficiencies of these proteins, the levels are below 10 to 20 percent of normal values. When coagulation factor levels are below the normal range, do not regard decreased levels of protein C, protein S or anti thrombin as a primary, possibly inherited, deficiency in the absence of a positive family history or screening (Class I, Level C) [13].
The factor V Leiden mutation the prothrombin-gene mutation, and the methylene tetrahydrofolate reductase mutation have also been noted in patients with the Budd-Chiari syndrome [10]. Antiphospholipid syndrome which is another cause of the Budd-Chiari syndrome because of an increased tendency to form abnormal blood clots in blood vessels is a disorder of coagulation, which causes blood clots in both arteries and veins [15].
Other factors :
Pregnancy is a danger factor for thrombosis of hepatic veins and especially after the third trimester of delivery.
Contraceptive pill: the relative risk of hepatic-vein thrombosis among women who use oral contraceptives is 2.37 [16].
Chronic inflammatory diseases such as sarcoidosis , systemic lupus erythematosus, inflammatory diseases of intestine, Sjogrens syndrome and Behcets disease cause non particular superficial phlebitis that reduces the prostaglandin levels which results in thrombosis of hepatic veins [17].
Chronic infections such as hydatoid cyst, aspergillosis, syphilis, tuberculosis and amoeboid cyst can also cause mechanical thrombotic or non thrombotic stenosis of the inferior vena cava [18].
Malignant diseases such as hepatocellular carcinoma, atrial myxoma, liomyosarcoma and Wilms' tumor, kidneys and adrenal glands cancer can also cause thrombosis or stenosis of hepatic veins [19, 20].

Clinical findings

Patients with Budd-Chiari syndrome present with varying degrees of symptomatology and acuity. The clinical presentation of Budd-Chiari syndrome is governed by the extent and rapidity of the hepatic vein occlusion and on whether a venous collateral circulation has developed to decompress the hepatic sinusoids [15]. Four main clinical variations have been described: fulminant, acute, subacute and chronic liver disease. Patients with the fulminant form of the syndrome present with hepatic encephalopathy within eight weeks after the development of jaundice. This presentation is unusual. Patients with the acute form have symptoms of short duration, intractable ascites, and hepatic necrosis without the formation of venous collaterals. The sub acute syndrome, which is the most common, has a more insidious onset; ascites and hepatic necrosis may be minimal, because the hepatic sinusoids have been decompressed by a portal and hepatic venous collateral circulation. When the Budd-Chiari syndrome is acute, thrombosis of all the major hepatic veins is usual, whereas in the sub acute form it is present in only a third of patients. The chronic form is manifested as complications of cirrhosis [15]. Abdominal pain, hepatomegaly, and ascites are present in almost all patients with the Budd-Chiari syndrome [21, 22]. However, asymptomatic patients with hepatic-vein thrombosis have also been described in 20 percent of cases, in whom the liver sinusoids were decompressed by large intra hepatic and portosystemic collaterals [23].

DIAGNOSIS

Laboratory examinations
Serum aspartate and alanine aminotransferase levels may be more than five times the upper limit of the normal range in the fulminant and acute forms of the Budd-Chiari syndrome, whereas increases are smaller in the subacute form. Serum alkaline phosphatase and bilirubin levels also increase to a varying extent, along with a decrease in serum albumin. The examination of ascetic fluid shows a total protein level more than 2.5 g per deciliter and white blood cells are usually less than 500/μL. Hematological studies are needed to evaluate for hypercoagulability [15].
Para clinical examinations
Ultrasound combined with Doppler imaging of the liver has a diagnostic sensitivity and specificity of 85 percent or more and is the first line investigation when the Budd-Chiari syndrome is suspected [24]. Doppler-sonography done by an experienced examiner, aware of the diagnostic suspicion, is considered as a very effective and reliable diagnostic means (Class I, Level C) [13]. The frequent non-specific sonomorphological signs are splenomegaly (78%), unhomogeneous liver parenchyma (76%), caudate lobe hypertrophy (67%), ascites (56%) and extrahepatic collaterals (44%) [15]. Hepatic veins devoid of flow signal, collateral hepatic venous circulation, a spider-web appearance usually located in the vicinity of the hepatic vein ostia and stagnant, reversed or turbulent flow can all be indicative of Budd-Chiari syndrome [25, 26]. When it is technically difficult to obtain an adequate sonographic evaluation or when the diagnostic features cannot be demonstrated, computed tomography (CT) or, preferably, Magnetic Resonance Imaging (MRI) should be performed as a second line of investigation (Class I, Level C) [13, 25, 27]. CT scans allow for the evaluation of ascites, hepatic vein patency, vena cava patency

and caudate lobe hypertrophy. MRI is better for visualizing the whole length of the inferior vena cava and may permit differentiation of the acute form of the Budd-Chiari syndrome from the subacute and chronic forms [28]. The third line of investigation should be retrograde cannulation of the hepatic veins for venography and liver biopsy [29]. Venography is a specific examination useful in the assessment of the extent of outflow obstruction and also allows for pressure measurements. X-ray venography can be considered as a diagnostic procedure in patients where the diagnosis remains uncertain (Class I, Level B) [13]. Whereas the liver biopsy can be considered only when an obstructed hepatic venous outflow tract has not been demonstrated with noninvasive imaging (Class I, Level C). Histopathologic clues of venous outflow obstruction are centrizonal congestion, hemorrhage and cell necrosis, sinusoidal dilatation with or without central vein obliteration and congestive pattern of cirrhosis [30].

Management

Initially and whenever possible the underlying risk factors for venous thrombosis must be corrected without delay (Class I, Level C). Recently published guidelines suggest medical therapy (anticoagulation, treatment of underlying disease, symptomatic therapy of portal hypertension complications) as the first-line treatment, angioplasty/stenting the second-line (in patients with short-length stenosis not responding to medical therapy), Transjugular Intrahepatic Portosystemic Shunt (TIPS) the next step (in patients not responding to medical therapy and in case of stenosis unsuitable for angioplasty/stenting) and liver transplantation as the last chance when TIPS is not effective [31].

Medical therapy

The first step of therapeutic strategy is based on immediate initiation of anticoagulation with low molecular-weight heparin, targeting anti-Xa activity to 0.5-0.8 IU/mL. A rapid shift to vitamin K antagonists when clinically appropriate, targeting an international normalized ratio (INR) 2 to 3, is indicated (Class I, Level B) [13, 32]. Maintaining permanent anticoagulation therapy is possible, unless a major contra-indication is present or a complication of anticoagulation therapy occurs (Class I, Level C).

Screening and management of gastroesophageal varices, which constitute the main source of major bleeding in Budd-Chiari syndrome patients treated with anticoagulation, must be done as recommended for other types of liver disease until more data are available (Class I, Level C) [13, 33].

Beta-adrenergic blockers or endoscopic therapy for patients with large varices should be carried out. Patients with ascites should be treated with diuretics. Discontinuing anticoagulants should be considered before paracentesis because of the increased risk of bleeding [33].

Percutaneous recanalization

A check for a venous obstruction amenable to percutaneous angioplasty/stenting in all symptomatic patients is recommended and a treatment is indicated accordingly (Class I, Level C) [13]. In the case of short-length stenosis of major hepatic vein or inferior vena cava, angioplasty and/or stenting is a therapeutic approach suitable for Budd-Chiari syndrome [34, 35]. The efficacy and the inocuity of this procedure have been confirmed [32, 34]. In asymptomatic patients, the benefit-risk ratio of this therapeutic option is still debated [11]. Anticoagulation and angioplasty appear to succeed in controlling Budd-Chiari syndrome in only 20 to 30 percent of patients [32].

Transjugular Intrahepatic Portosystemic Shunt (TIPS)

In patients without ongoing improvement on anticoagulation therapy (with or without angioplasty), TIPS insertion can be considered (Class I, Level C) [13, 36]. A common complication of TIPS is hepatoencephalopathy in 25% and also the constriction of anastomoses which appears in 34% to 75% of patients. However, the influence of TIPS on patient survival and factors that predict the outcome of TIPS in Budd-Chiari syndrome patients remain unknown. Long-term outcome for patients with severe syndrome treated with TIPS is excellent even in high-risk patients, suggesting that TIPS may improve survival [15].

Liver transplantation

In patients with failure of TIPS treatment, or in whom TIPS insertion is judged unfeasible or is unsuccessful, liver transplantation is the remaining option (class I, Level C) [13]. A group of patients with poor prognosis might benefit early liver transplantation [37, 38]. The impact of liver transplantation is difficult to assess. Two studies of the outcome of transplanted patients have shown 5-year survival rates reaching 80% [39, 40]. Liver transplantation can be considered in patients with fulminant hepatic failure (Class I, Level C) [13].

This therapeutic strategy has allowed for achieving 5-year survival rates in 90 percent [36]. This improvement in survival has been obtained with complete resolution of clinical signs and improvement of liver function tests which results in excellent quality of life [35].

PROGNOSIS

The natural development of the disease does not have good results. Symptomatic forms have a poor spontaneous course as it has been estimated that 90% of untreated patients will die within 3 years [11]. Death can be related to liver failure function, bleeding of varicose veins or refractory ascites. Asymptomatic forms of Budd-Chiari syndrome carry a good prognosis [23]. Diagnosis in young age, low Child-Pugh score, absence of ascites or easily controlled ascites, low levels of creatinine, sodium, albumin and bilirubin, seem to be related with better prognosis [15]. Patients with long-standing should be monitored, to control BCS for late development of hepatocellular carcinoma and transformation of underlying myeloproliferative disease (Class I, Level C).

The 5-year survival is calculated at 38-87% following systematic by-pass of portal vein and the 5-year survival percentage after the transplantation of liver is 70% [41, 42].

CONCLUSION

Budd-Chiari syndrome is a complex disease with a wide spectrum of aetiologies and presentations. This disease should be treated following a step by step strategy. Anticoagulation and medical therapy should be the first line treatment. Revascularization or TIPS are indicated in case of no response to medical therapy.

Evidence base medicine

None of the above recommendations are based on a high level of evidence due to the difficulty in making appropriate clinical studies on a sufficient number of patients with this rare disease. Most of the above recommendations are based on expert consensus [13].

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