

## BUDD-CHIARI: ENDOVASCULAR MANAGEMENT

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### BUDD –CHIARI SYNDROME

Budd-Chiari syndrome (BCS) is characterized by obstruction of the venous outflow of the liver. The course of the disease can be rapidly fatal, or in the chronic form, be confused with other causes of cirrhosis and portal hypertension. Most cases of BCS in Western countries are related to thrombosis of the major hepatic veins (HV) secondary to hypercoagulable disorders. Idiopathic obstruction of the hepatic portion of the IVC is the most common form of BCS in developing countries. Membranous obstruction of the IVC is now considered to be a sequela of previous thrombosis. IVC obstruction can also be related to post-transplant states and tumors.

Budd initially described in 1845, three cases of hepatic vein thrombosis due to infectious phlebitis. Chiari described in 1899 three additional cases of hepatic vein occlusion due to phlebitis, Inferior Vena Cava (IVC) involvement was present in one of the cases. Since Pleasants's literature review in 1911 of 296 cases, the Budd-Chiari syndrome (BCS) has included both hepatic vein and IVC obstruction/stenosis.

Okuda in 2001 emphasized that the classic BCS also called hepatic vein thrombosis (HVT) and the membranous obstruction of the IVC (MOVC), also called primary thrombosis of the IVC, are epidemiologically, pathologically and clinically different and should be treated as two different clinical entities. HVT and MOVC have a different onset, clinical manifestations and natural history. Whereas HVT is a severe disease with an acute onset, MOVC presents as a mild disease at the onset, which can eventually turn into a fibrous occlusion of the IVC. Okuda proposed that the term Hepatic Vein Outflow Block (HVOB) should replace the term Budd-Chiari Syndrome and be divided into: a) Obliterative Hepatocavopathy, replacing the terms, primary IVC thrombosis/membranous obstruction of the IVC (MOVC); and b) Primary hepatic vein thrombosis.

Okuda also stated that Hepatic Vein Outflow Block can be classified into primary and secondary. Primary: a) Hepatic vein thrombosis/obstruction: w/s IVC involvement; and b) IVC thrombosis/obstruction: mainly IVC obstruction with hepatic venous ostia involvement. Secondary HVOB is most commonly due to compression by a hypertrophied caudate lobe or by tumor involvement.

The obliterative hepatocavopathy type of HVOB selectively affects the intrahepatic segment of the IVC at the diaphragmatic level, with involvement of the hepatic vein orifices. Membrane like obstruction commonly found angiographically, has been found to consist of organized old thrombus arranged in layers of different ages. The clinical onset is mild and causes liver damage by congestion. A distinct pattern of subcutaneous venous collaterals typically develops together with retroperitoneal collaterals through the ascending lumbar and the iliolumbar veins into the hemiazygos and azygos veins. This type of the disease is more common in Asia and Africa.

Hepatic vein thrombosis has an acute onset with severe liver damage often requiring urgent portocaval decompression and might even require liver transplantation depending on the

extent of venous involvement. The collateral venous pathways are similar to those seen in portal hypertension. This type of the disease is more common in Western countries. HVT is most commonly associated with a hypercoagulable state. Hypercoagulable states are uncommon in obliterative hepatocavopathy, with infections and parasites having been more commonly the suspected causes in Asia and Africa. Extrinsic compression by tumors or a hypertrofied caudate lobe are also common causes of obliterative hepatocavopathy. The pathophysiology of Hepatic Vein Outflow block is related to a sudden rise of the sinusoidal pressure following the hepatic vein outflow blockage, with centrilobular congestion and sinusoidal dilatation. Prolonged congestion leads to loss of hepatocytes, fibrous scarring and cirrhosis. The diagnosis of Hepatic Vein Outflow Block is established by the clinical history and physical examination. The best imaging methods are: Ultrasound, Computerized Tomography and Magnetic Resonance Imaging. Once the diagnosis is established, inferior vena cavography is needed to guide endovascular techniques. The prognosis of patients with Hepatic Vein Outflow Block is best expressed by the study of H. Okuda who followed 157 patients with hepatic vein outflow obstruction for 15 years and found a 20% mortality, higher for men (25%) than for women (15%). The Cause of death on his study was: liver failure 12/33; variceal bleed 7/33; HCC 4/33. The incidence of HCC varies between countries with the highest incidence in South Africa, 47.5% in obliterative hepatocavopathy cases. HCC is rare in HVT patients except in Behcet's disease

Prompt diagnosis with definition of the specific anatomic and hemodynamic factors with hepatic and IVC venography and liver biopsy is crucial to define the appropriate treatment in order to obtain decompression of the congested liver and to relieve portal hypertension before cirrhosis develops. Surgical portosystemic shunts are effective in relieving the portal hypertension but have a high rate of complications. Liver transplant is the preferred procedure for advanced cirrhosis and poor hepatic function, and for fulminant liver failure. Percutaneous transluminal angioplasty (PTA), stent placement and transjugular intrahepatic shunts (TIPS), are minimally invasive procedures with encouraging initial results. The purpose of this presentation is to discuss the different endovascular techniques that could be used in the treatment of HVOB. A clear understanding of the anatomy and hemodynamic factors is essential to choose the adequate endovascular technique for each patient.

## DISCUSSION

Most patients have been traditionally managed with surgery. Unfortunately, there is a high incidence of early postoperative complications including thrombosis of surgical shunt in up to 50% of patients, and an operative mortality ranging from 19%-31%. Orthotopic liver transplantation is the preferred procedure for advanced cirrhosis and poor hepatic function and for fulminant liver failure with a long survival rate from 45%-87% at 5 years.

Endovascular treatment of HVOB has been reported with high success rates. Depending on the site of the obstructions, different interventional techniques can be performed. For membranous obstruction, segmental stenosis and occlusion of the IVC, PTA and stent placement are recommended as a first line of treatment in HVOB patients with IVC obstruction. Thrombolytic therapy has been successful in lysing fresh thrombi in the hepatic veins but this presentation is uncommon in the clinical practice.

The results of the endovascular treatment of short hepatic veins occlusion with stents are encouraging. Most post-thrombotic occlusions of the HV appear as short, focal lesions at the level of the origin while the distal vein is usually patent. PTA has been used on these cases,

but recurrence is common. Metallic stents have shown better results. Even with obstruction of the three hepatic veins, endovascular treatment of at least one hepatic vein appears to be adequate for decompression of the liver. Unlike metallic stents placed during TIPS, the stents placed into the hepatic veins rarely provoke intimal hyperplasia.

The endovascular recanalization of the obstruction of the origin of the hepatic veins is a challenging task. Under fluoroscopy it is difficult to establish the relationship of the cava and the right atrium, when a stump of the hepatic vein is not seen, recanalization from the antegrade jugular approach can be impossible. Ultrasonographic guidance to puncture the vein transhepatically allows the recanalization of the vein in most cases. Once a guide wire has crossed the obstruction, a combined technique can be used to dilate and stent the vein from a transjugular approach. Although the risk of hemorrhage using the transhepatic approach is higher, the use of ultrasonic guidance and subsequent embolization of the liver tract make the procedure very safe in most patients.

TIPS in patients with HVOB have been used as a bridge for hepatic transplantation and also as a primary treatment, with survival rates comparable to the surgical literature. TIPS creation in HVOB patients is technically difficult due to the obstruction of the hepatic veins and the inferior and anterior displacement of the portal vein secondary to caudate lobe enlargement. Direct IVC to portal shunts are necessary in many patients, alternatively if the stump of the RHV is found, the transhepatic puncture can be started from that point. Although marked improvement in the hepatocellular function and symptomatology is noted after TIPS, the long-term permeability of TIPS is poor, especially in patients with hypercoagulable states. In recent series the primary permeability reported was 43% to 50%. Multiple revisions are usually required to improve patency.

Combined obstruction of HV and IVC:

The traditional management of the combined obstruction of the IVC and the HV has been either liver transplantation or a mesoatrial shunt. Using modern endovascular techniques it is possible to simultaneously reconstruct the obstructions of the IVC and the HV. Usually the IVC is recanalized and stented first, followed by an attempt to recanalize the origin of the HV using a transjugular or transhepatic approach. If the HV is completely obliterated, a direct TIPS between the stented IVC and the portal vein could be performed.

Thrombolytic therapy:

Although acute occlusion is uncommon in the clinical practice, thrombolytic therapy is indicated for the treatment of fresh thrombi in the IVC and/or HV.

IVC: Primary IVC obstruction can present as a stenosis, a membrane, or a long segment occlusion, it can also involve the HV origin. PTA and stent placement are recommended as a first line of treatment in patients with IVC obstruction. Secondary IVC compression by an enlarged caudate lobe is a common finding in patients with HVOB; the compression may regress after HV decompression and it may not require direct treatment in the early stages of HVOB.

IVC obstruction or stenosis with gradients greater than 20 mm Hg, may interfere with the success of surgical portosystemic shunts, IVC stent placement combined with surgical shunting may avoid the high morbidity and mortality associated with mesoatrial shunts. In

some cases of IVC obstruction, it may be impossible to cross the lesion from either a femoral or a jugular approach. Sharp needle recanalization under careful multiplanar guidance is possible in most cases.

Metallic stents have improved the long-term results of endovascular treatment of IVC obstruction. The Gianturco stent (Cook, Bloomington, IN) is the most commonly used stent; its wide interstices may minimize compromise of the origin of the HV. Metallic stents in the IVC may interfere with a future liver transplant, especially when they protrude into the right atrium, or too distally into the intrahepatic IVC.

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