

Summary & Conclusion

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Budd Chiari syndrome is a heterogeneous group of disorders characterized by hepatic venous outflow obstruction at the level of the hepatic venules, large HVs, IVC, or the right atrium ⁽⁶⁾. But generally implies thrombosis of HVs & /or the intrahepatic or suprahepatic IVC. Up to 50% of cases of BCS are due to chronic myeloproliferative disorders like polycythemia vera or coagulopathies like factor V (Leiden) gene mutation. ⁽¹¹²⁾

The BCS may be primary, due to a membrane in IVC, or secondary to neoplasms, oral contraceptive use, pregnancy, hypercoagulable states, myeloproliferative disorders & paroxysmal nocturnal hemoglobinuria. In practice, no identifiable cause is found in up to 70% of patients. ⁽⁶²⁾

The clinical presentation is highly variable but may be categorized as acute & perhaps FHF, as subacute without evidence of cirrhosis or as chronic with evidence of portal hypertension & cirrhosis. ⁽¹¹³⁾

Doppler US of the liver is the initial investigation when BCS is suspected. CECT scanning, however, is better to detect parenchymal inhomogeneity & to delineate the venous anatomy of the liver in preparation for TIPS. MRI shows HVT well but is more expensive than CT scanning & less readily available. ⁽⁶¹⁾

Treatment depends on the underlying cause, the anatomic location, the extent of the thrombotic process & the severity of liver disease. Treatment options can be divided into medical treatment including anticoagulation & thrombolysis, radiological procedures such as angioplasty, TIPS & surgical procedures including porto-systemic shunting & OLT. ⁽¹¹⁴⁾

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Anticoagulation alone is unlikely to lead to sufficient recanalization of occluded vessels, or development of adequate collateral circulation. However, satisfactory long-term survival with only medical therapy has been reported.⁽¹¹⁵⁾ This may give rise to redefine the role of OLT which may now be preserved for patients failing TIPS.⁽⁸²⁾

TIPS was created using standard techniques by insertion of Palmaz stents, Wallstents or covered Viatorr stents. When a HV remnant was not present, PV was punctured directly from the IVC.⁽¹¹⁶⁾

After TIPS procedure patients underwent anticoagulation according to the guidelines. Patients underwent control angiography 3 months after TIPS & in addition if shunt dysfunction was suspected. An abdominal US was performed every 6 months . TIPS dysfunction was defined as an increase in portosystemic gradient above 10 to 12 mmHg & clinical signs of portal hypertension.⁽³⁸⁾

Therapy for BCS usually requires TIPS or OLT. The treatment modality is dependent on duration of illness, extent of thrombosis & degree of liver dysfunction. TIPS is successful as initial therapy as it promotes clinical improvement in the long run even though shunt revisions are generally needed. Recently, the necessity of subsequent OLT was remarkably low & thus TIPS could be regarded as definitive treatment option in BCS. OLT in BCS is associated with an increased risk of thrombembolic complications & early graft failure in spite of consequent anticoagulation therapy.⁽¹¹⁷⁾

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