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TIPS for Budd-Chiari Syndrome: Long-Term Results and Prognostics Factors in 124 Patients

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Background & Aims: Budd-Chiari syndrome (BCS) is a rare and life-threatening disorder secondary to hepatic venous outflow obstruction. Small series of BCS patients indicate that transjugular intrahepatic portosystemic shunt (TIPS) may be useful. However, the influence of TIPS on patient survival and factors that predict the outcome of TIPS in BCS patients remain unknown. *Methods:* One hundred twenty-four consecutive BCS patients treated with TIPS in 6 European centers between July 1993 and March 2006 were followed until death, orthotopic liver transplantation (OLT), or last clinical evaluation. *Results:* Prior to treatment with TIPS, BCS patients had a high Model of End Stage Liver Disease and high Rotterdam BCS prognostic index (98% of patients at intermediate or high risk) indicating severity of liver dysfunction. However, 1- and 5-year OLT-free survival were 88% and 78%, respectively. In the high-risk patients, 5-year OLT-free survival was much better than that estimated by the Rotterdam BCS index

OLT-free survival was much better than that estimated by the Rotterdam BCS index (71% vs 42%, respectively). In the whole population, bilirubin, age, and international normalized ratio for prothrombin time independently predicted 1-year OLT-free survival. A prognostic score with a good discriminative capacity (area under the curve, 0.86) was developed from these variables. Seven out of 8 patients with a score >7 died or underwent transplantation vs 5 out of 114 patients with a score <7 . **Conclusions:** Long-term outcome for patients with severe BCS treated with TIPS is excellent even in high-risk patients, suggesting that TIPS may improve survival. Furthermore, we identified a small subgroup of BCS patients with poor prognosis despite TIPS who might benefit from early OLT.



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Abbreviations used in this paper

BCS, Budd-Chiari syndrome; HE, hepatic encephalopathy; OLT, orthotopic liver transplantation; TIPS, transjugular intrahepatic portosystemic shunt

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