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About the Rotterdam Score as a predictor of Budd-Chiari Syndrome prognosis

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In 1845, Dr. George Budd commented, for the first time, on 3 patients with hepatic vein obstruction in his Liver Diseases seminar. At that time, sepsis was assumed to be the cause of thrombosis in two of the patients. Fifty-three years later, Dr. Hans Chiari published a case series and, while practicing as a pathologist in Prague, observed 3 patients with hepatic vein thrombosis; he studied these cases along with a literature review that included 7 other patients. Despite the rarity of this condition, he concentrated on "a disease that could soon lead to death," which he named obliterative phlebitis. All 3 livers were severely clogged and necrotic, with portomesenteric venous thrombosis and large-volume ascites. Histology revealed a minimal adventitious reaction with no significant perivascular involvement. Thrombosis was considered a complication of an endophlebitis process caused by syphilis, and this theory remained unconfirmed in the years that followed. However, this description is a cornerstone in the discovery of what is now known as Budd-Chiari syndrome (1).

I have read with great interest the study recently published by Muñoz-Maya et al. in the Revista Colombiana de Gastroenterología, which describes the etiology, treatment, and outcomes in a retrospective cohort of 35 patients diagnosed with Budd-Chiari syndrome. Due to the rarity of this condition, there are no prospective data in the literature and the largest studies to date are mainly comprised of data collected from retrospective cohorts. As the study authors themselves state, it is one of the largest series of patients diagnosed with Budd-Chiari syndrome published in Colombia. In their publication, disease severity was classified using the MELD Score (Model for End-Stage Liver Disease) and the Rotterdam Score to predict therapeutic success and assess response earlier, so that definitive invasive measures are not delayed. and prognosis is improved (2).

Although Budd-Chiari syndrome is a rare disorder, it has a variable prognosis and can result in early mortality (death before 3 months) in up to 20% of cases. Furthermore, the performance of severity indexes such as the Child-Pugh classification, MELD, Rotterdam score, and Clichy criteria for establishing early mortality is still uncertain. The Rotterdam score is calculated using the following equation:

1.27 x encephalopathy + 1.04 x ascites + 0.72 x prothrombin time international normalized ratio (INR) + 0.004 x bilirubin μmol/L

Ascites and hepatic encephalopathy are classified as present [1] or absent [0], and prothrombin time as greater [1] or less [0] than an INR of 2.3. The total score ranged from 0.02 to 4.03. In this way, three kinds of patients are observed

- Class I (good prognosis): with a total score between 0 and 1.1;
- Class II (intermediate prognosis): with scores between 1.1 and 1.5;
- Class III (poor prognosis): with a score greater than 1.5.

The best index to establish 3-month mortality in patients with Budd-Chiari syndrome is the Rotterdam score, with an area under the curve (AUC) of 0.84 (95% confidence interval [CI]: 0.68-0.98, p=0.005). Therefore, it has been concluded that the Rotterdam score is the best index to predict mortality at 3 months in the context of the Budd-Chiari syndrome and, for this reason, it should be used to determine the urgency of treatment (3).

Concerning the prognosis of patients with Budd-Chiari syndrome, the clinical practice guidelines of the European Association for the Study of the Liver (EASL) more recently established that there have been several attempts to determine parameters or combinations of parameters that can predict disease prognosis in these patients. Although these prognostic indexes are valid for the assessment of transplant-free survival and invasive therapy-free survival, their predictive accuracy is suboptimal for use in individual patients in daily clinical practice. The development of hepatocellular carcinoma, or even the progression of the underlying hematological disease, can modify the prognosis of Budd-Chiari syndrome (4).

At this point, a rather momentous question arises: Is it better to use anticoagulation, transjugular intrahepatic portosystemic shunt (TIPS), or liver transplantation for the treatment of Budd-Chiari syndrome? At present, there is no reason to choose one over the other, and all are worthy of consideration for therapy. In fact, the centers that serve these patients should have extensive experience in all these techniques to ensure the best treatment, thus these patients should be referred to the centers with more experience. The suggested approach to patients with Budd-Chiari syndrome is a step therapy that progresses from less invasive (anticoagulation) to more invasive therapies (liver transplantation) (5).

A timely diagnosis is required due to the high mortality rate associated with this condition in the absence of treatment. Early clinical manifestations should lead to ruling out other diagnoses, since resolution of the syndrome may be achieved in some cases when treating concomitant diseases. It is important to bear in mind that treatment depends on the characteristics of each patient and step therapy is suggested from the least to the most invasive procedure, in order to provide better prognostic outcomes to patients, who, despite the treatment they receive, must be strictly monitored (6). One could reasonably conclude that the sickest patients require urgent liver transplantation, but the possibility that emergency shunt or TIPS placement may also be effective cannot be excluded depending on local experience.

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