

## Predictors of survival in thrombotic thrombocytopenic purpura

Scully *et al.* reported that rituximab as an adjunct to plasma exchange and steroids for initial therapy of acquired thrombotic thrombocytopenic purpura (TTP) significantly reduces the risk of relapse compared with historical controls.<sup>1</sup> In recent years increasing attention has been focused on improving our ability to identify patients at high risk for mortality or relapse. The recent results from the French TMA Reference Center (*Benhamou et al., Haematologica 2012*),<sup>2</sup> where a predictive model for death has been proposed are interesting but one major consideration gives pause. Only patients with severe ADAMTS13 deficiency were included in the study. Although severe ADAMTS13 deficiency has high specificity for the diagnosis of TTP, its sensitivity is low, and as yet, undetermined. The Oklahoma TTP-HUS registry reported that only 13% of patients have severe ADAMTS13 deficiency and even in the category of idiopathic TTP this number was only 33%. Patients with severe ADAMTS13 deficiency were more likely to need more plasma exchanges to achieve remission, though they were also more likely to suffer relapses. Interestingly, this was a linear trend across all levels of ADAMTS13 activity and not merely a characteristic of patients with severe deficiency.<sup>3</sup> By choosing this particular subset of patients for analysis, we run the risk of developing a model that is not applicable to the majority of patients with TTP, even idiopathic TTP. Also, several patients who are at risk for late mortality (>30 days) may elude identification by the proposed score. It is necessary to validate this score in a less selective group of patients before its utility can be established.

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