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# Letter to the editor about: “Eculizumab First” in the Management of Posttransplant Thrombotic Microangiopathy



**To the Editor:** We read with interest the article by Maritati *et al.*<sup>1</sup> describing their experience with eculizumab in the treatment of very early posttransplant thrombotic microangiopathy. The reported incidence of almost 10% is remarkably high and prompted us to look more closely. Diagnosing thrombotic microangiopathy at such an early stage (median posttransplantation time of 3 days) is inherently challenging due to the multiple insults the kidney transplant may have experienced. For example, fibrin thrombi in the glomerular capillaries have been documented in cases of donor disseminated intravascular coagulation.<sup>2</sup> Various factors, including the use of antilymphocyte serum and surgical site hematoma, may lead to thrombocytopenia and hemolysis, and schistocytes may even be found positive under these conditions.<sup>3</sup> A more detailed characterization, particularly with regard to the schistocyte threshold and the inclusion of pathological details in line with the criteria recently established by the Banff working group would have been beneficial.<sup>4</sup> The authors highlight the identification of the MCPggaac haplotype in 16 of 29 patients (55%). This haplotype has been documented as an aggravating risk factor for atypical hemolytic uremic syndrome in patients with established pathogenic mutations but its presence, when isolated and heterozygous, is not deemed pathogenic, a condition observed in 39% of the European population. In this regard, we question the authors' mention of 2 references that would support the role of the MCPggaac haplotype as a predisposing factor *per se* in some conditions. Indeed, Le Clech *et al.*<sup>5</sup> found that only the homozygous MCPggaac haplotype was associated with secondary hemolytic uremic syndrome. Furthermore, Sánchez-Moreno *et al.*<sup>6</sup> reported a case of atypical hemolytic uremic syndrome in a pediatric patient with a pathogenic factor B mutation and the MCPggaac haplotype who did not relapse after transplantation, even in the absence of eculizumab, leading the authors to speculate that the absence of the MCPggaac haplotype in the donor may have protected

the endothelial cells. Finally, in the absence of a control group, we challenge the authors' claim that their results suggest an association between the use of eculizumab in their ill-defined thrombotic microangiopathy and the normalization of hemolysis along with an improvement in graft function. Such results are usual in the immediate posttransplant period, particularly after an initial period of delayed graft function.

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