Recent advances in the management of immune mediated TTP

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Immune-mediated thrombotic thrombocytopenic purpura (iTTP) is a potentially life-threatening thrombotic microangiopathy caused by autoantibody-mediated severe ADAMTS13 deficiency. TTP should be suspected in patients with microangiopathic hemolytic anemia and thrombocytopenia without a definite cause. Early recognition of iTTP and prompt treatment with plasma exchange and corticosteroids are essential. Rituximab administration should be considered for refractory or relapsed iTTP. Rituximab can be used as a first-line adjuvant or preemptive therapy. Treatment with caplacizumab, a novel anti–von Willebrand factor nanobody, resulted in a faster time to platelet count response, a significant reduction in iTTP related deaths, and development of refractory iTTP. TTP survivors showed a higher rate of chronic morbidities, including cardiovascular disease and neurocognitive impairment, which can lead to poor quality of life and higher mortality rate. Meticulous long-term follow-up of TTP survivors is crucial.