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Safety and Efficacy of Caplacizumab vs the Standard of Care in Immune Thrombotic Thrombocytopenic Purpura

This infographic is intended for researchers and healthcare professionals and reflects the contents of the following article:

Izquierdo, C. P., Mingot-Castellano, M. E., Fuentes, A. E. K., Peinado, J. G., Cid, J., Jimenez, M. M. ... & Del Rio-Garma, J. (2022). Real-world Effectiveness of Caplacizumab vs the Standard of Care in Immune Thrombotic Thrombocytopenic Purpura. *Blood Advances*, *6*(24), 6219–6227. https://doi.org/10.1182/bloodadvances.2022008028

Immune thrombotic thrombocytopenic purpura (iTTP) is a rare disease characterized by microangiopathic haemolytic anaemia



Severe thrombocytopenia (<30 × 10⁹/L)



Variable degree of ischaemic organ damage

Mechanism underlying iTTP

Acquired by the deficiency of a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13 (ADAMTS13)



Production of autoantibodies against ADAMTS13

iTTP is associated with high

morbidity and mortality



Lowered ADAMTS13 enzyme activity



Reduced interaction of ADAMTS13 with von Willebrand factor (vWF)



Appropriately treated, the mortality rate falls to 5%–20%

Death from iTTP occurs predominantly during initial the days

If untreated, the mortality

rate rises to 90%

90%

Early diagnosis of iTTP and appropriate treatment are crucial in reducing mortality risk



Reduced vWF

clearance

Caplacizumab is approved for adults with an acute episode of iTTP in conjunction with plasma exchange (PEX) and immunosuppression

Accumulation of

platelet/vWF multimers

and increased clotting

PEX Immunosuppressive therapy (corticosteroids with or without rituximab) Caplacizumab (emerging treatment) Caplacizumab is a novel humanized nanobody targeting vWF It blocks platelet-vWF aggregation and thrombi formation It is fast-acting, showing therapeutic benefits within hours of administration



Text excerpts reprinted from *Blood Advances*. Vol. 6, Izquierdo CP, Mingot-Castellano ME, Kerguelen Fuentes AE, *et al*. Real-world Effectiveness of Caplacizumab vs the Standard of Care in Immune Thrombotic Thrombocytopenic Purpura, 6219–6227. Copyright 2022, with permission from The American Society of Hematology



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