

PLASMIC SCORE APPLICABILITY FOR THE DIAGNOSIS OF THROMBOTIC MICROANGIOPATHY ASSOCIATED WITH ACQUIRED ADAMTS13 DEFICIENCY IN EMERGENCY SETTINGS AT THE HUB LABORATORY OF AUSL ROMAGNA: STATE OF THE ART AND FUTURE PROSPECTS.

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Background and aims: Thrombotic thrombocytopenic purpura (TTP) is a potentially fatal microangiopathic haemolytic anaemia (MAHA) that requires early diagnosis. It is classically characterised by haemolytic anaemia, thrombocytopenia, and renal and neurological dysfunction. The PLASMIC score, which is based on seven laboratory and clinical criteria, enables patients at high risk of TTP to be identified rapidly. A decreased or absent level of activity of the von Willebrand factor-cleaving protease ADAMTS13 (activity below 10%) is diagnostic of TTP. Timely evaluation of the ADAMTS13 level is crucial for guiding optimal therapy and urgent intervention. The Romagna Health Service provides diagnostic assistance to an area covering 6,500 km² with a population of 1,200,000. In the field of haemorrhagic and thrombotic diseases, AUSL Romagna provides a 24-hour second-level coagulation on-call service, including factor and inhibitor dosages and ADAMTS13 activity testing. This study aims to evaluate the applicability of the PLASMIC score for patients with suspected TTP in emergency settings at AUSL Romagna's Hub Laboratory.

Methods: This retrospective study examined 36 patients who were either admitted to hospital or attended the emergency department with suspected thrombotic microangiopathy between 2023 and 2024. Data on ADAMTS13 activity (as measured by the HemosIL AcuStar chemiluminescent immunoassay) and medical and laboratory information were obtained from the patients' medical records. The patients were analysed in two groups according to their ADAMTS13 activity (below or above 10%) and were then stratified according to their PLASMIC score. The correlation between the PLASMIC

score and ADAMTS13 activity was then evaluated to determine sensitivity, specificity, and diagnostic accuracy in the high-risk group (score 6–7), using the area under the ROC curve (AUC).

Results: Of the 36 patients, 5 had ADAMTS13 levels below 10%, while 31 had levels above 10%. Of the latter group, 2 had HUS, 4 had sepsis, 1 had pancytopenia due to vitamin B12 and folate deficiency, 4 had autoimmune diseases, 7 had solid cancers, 4 had haematological diseases (myelofibrosis, myelodysplastic syndromes and lymphoma), 2 had MAHA induced by infection, 1 had drug-induced haemolysis and 6 cases showed no potential risk factors. In the high-risk group (scores 6–7), five out of seven patients (71.4%) had ADAMTS13 levels below 10%, meaning they were classified as having TTP. In the intermediate-risk group (score 5) and the low-risk group (score 0–4), no patients had an ADAMTS13 level below 10%. The PLASMIC score had a sensitivity and specificity of 100% and 93%, respectively, for screening for TTP. The PLASMIC score was found to be a highly effective tool for distinguishing patients with severe ADAMTS13 deficiency from those without, with an area under the curve (AUC) of 0.965. The positive predictive value (PPV) and negative predictive value (NPV) were 71% and 93%, respectively.

Conclusions: The high-risk PLASMIC score has a strong ability to discriminate between patients with and without severe ADAMTS13 deficiency. This enables clinicians to promptly initiate appropriate diagnostic investigations. Integrating artificial intelligence (AI) into healthcare could transform patient care and outcomes in future. AI-driven predictive analytics could enhance the accuracy, efficiency and cost-effectiveness of disease diagnosis and clinical laboratory testing.

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