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Heterogeneity of diagnosis, treatment, and management for immune thrombotic thrombocytopenic purpura: Are we still peering through the looking glass

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LETTER TO THE EDITOR

Heterogeneity of diagnosis, treatment, and management for immune thrombotic thrombocytopenic purpura: Are we still peering through the looking glass?

Dear Editor

The accurate diagnosis, timely treatment, and thoughtful management of patients with immune thrombotic thrombocytopenic purpura (TTP) are critical. Despite advances made in ADAMTS13 testing, initiation of emergent therapeutic plasma exchange (TPE), and expanded options for immunosuppression, there is heterogeneity with regard to the care of TTP patients in North America.[1,2] This has created numerous challenges in conducting meaningful multicenter studies.

At the 2017 AABB Annual Meeting, a session was offered entitled "Don't Play 'Hot Potato' with TTP: A 360 Degree Approach to TTP Patient Care." In this interactive session with audience response systems in place, the approximately 200 participants in attendance were asked a series of questions related to TTP diagnosis, treatment, and management. The answers demonstrated a diversity of thoughts about perceived optimal strategies, as well as highlighted sources of variability that may influence decisions regarding care of patients with this potentially life-threatening condition (Figure 1).

With such wide-ranging responses, harmonization is needed. The American Society for Apheresis TTP Consensus Conference attempted to address this concern, as

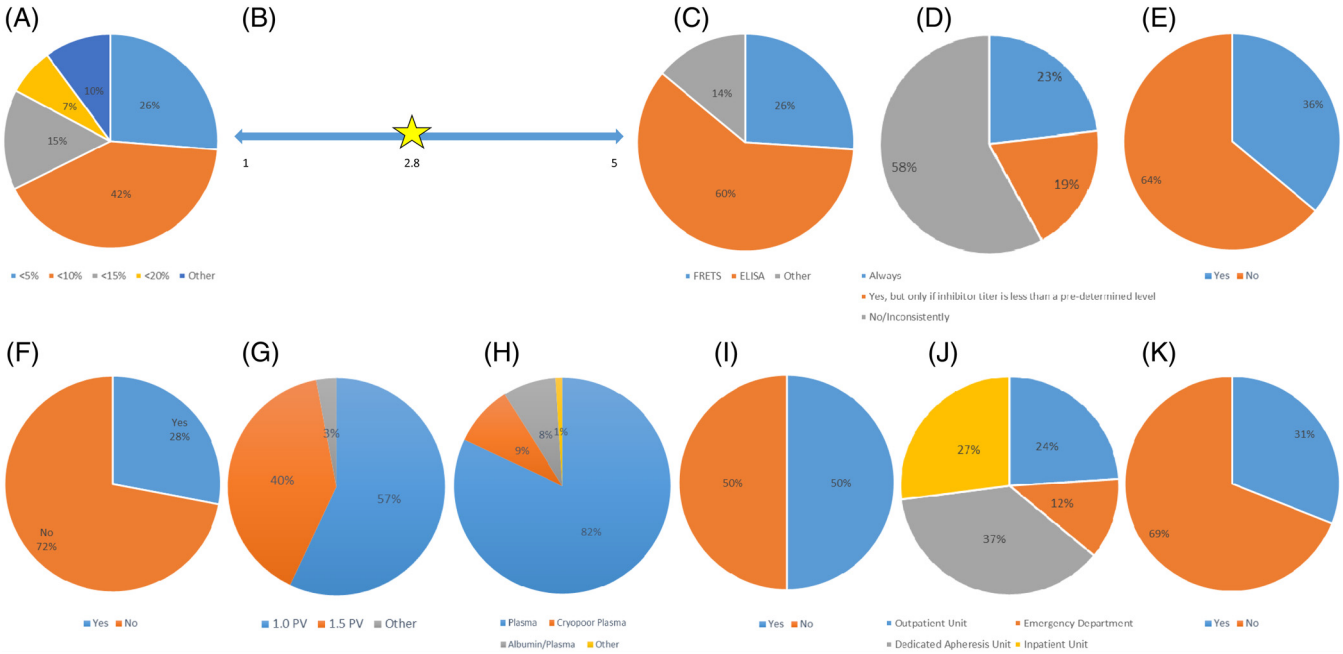

FIGURE 1 A, What cutoff does your institution use to define deficiency of ADAMTS13 activity in TTP? B, On a scale of 1 to 5 (one being least confident and five being most confident), how confident do you feel about your ADAMTS13 activity test cut-off accurately reflecting clinical TTP that responds to TPE? C, What type of ADAMTS13 activity testing platform do you use? D, Do you order an ADAMTS13 autoantibody test? E, Do you have an in-house ADAMTS13 assay? F, Do you have routine access to STAT (results in <4 hours) ADAMTS13 activity testing? G, How many plasma volumes do you routinely exchange when performing TPE for the majority of TTP patients? H, What replacement fluid do you typically use during TPE for TTP? I, Do you perform a TPE taper when treating TTP patients? J, For a patient with asymptomatic TTP exacerbation, in what setting should TPE be performed? K, Should we be treating TTP patients to a biochemical remission (i.e., normalization of ADAMTS13 activity) instead of a normal platelet count?

have other groups.[3-9] Most recently, the International Society for Thrombosis and Haemostasis has drafted preliminary guidelines for the diagnosis, treatment, and management of TTP that are now being finalized.[10] However, these guidelines also have recommendations based on low certainty or very low certainty evidence. Given the iterations of recommendations that have come previously based on similarly shaky findings, future steps must include targeted analyses of specific TTP patient outcomes that address critical gaps in our knowledge. Until compelling and meaningful outcome-based data drives guideline reformations, opinions about best practices will undoubtedly continue to vary and we will still be partially peering through the looking glass with regard to TTP diagnosis, treatment, and management.

CONFLICT OF INTEREST

J.S.R. and M.A.M. are consultants for Sanofi Genzyme; the other authors have no conflicts of interest to declare.

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