COMMENTARY

blood advances

TO THE EDITOR:

Commentary on the 2023 ASH guidelines for thrombophilia testing in venous thromboembolism

Manu Juneja[1] and Jeff Szer[1,2]

[1]Department of Clinical Haematology, Peter MacCallum Cancer Centre and Royal Melbourne Hospital, Melbourne, VIC, Australia; and [2]The University of Melbourne, Melbourne, VIC, Australia

We read with interest the latest American Society of Hematology (ASH) guidelines addressing thrombophilia testing in patients with venous thromboembolism.[1]

Unfortunately, we believe the following 2 significant issues were not addressed: (1) the importance of paroxysmal nocturnal hemoglobinuria (PNH) screening in patients with atypical sites of thromboses (eg, splanchnic or cerebral venous sinus thrombosis) in which there may be evidence of hemolysis or cytopenias and (2) the importance of warfarin as thromboprophylaxis in antiphospholipid syndrome as opposed to direct acting oral anticoagulants (DOACs).

In the recommendations, PNH is not addressed, except for the caveat that PNH thrombosis can occur at atypical sites.[1] Although PNH being the most vicious acquired thrombophilia, remains relatively rare; up to 10% of patients present with thrombosis, and it remains the leading cause of death and a significant cause of morbidity.[2,3] In our practice, thrombophilia testing, regardless of indication, includes a PNH screen, in part to avoid a potential fatal thrombotic event. This opinion is shared by other representative bodies.[4]

Similarly, the updated ASH guidelines neglect to describe the paucity of evidence in favor of DOAC thromboprophylaxis in antiphospholipid syndrome. As recent as last year, randomized controlled data favored warfarin over apixaban in thromboprophylaxis.[5] This built upon the previous data suggesting higher risk of recurrent thromboses in patients treated with rivaroxaban in lieu of vitamin K antagonists.[6] Moreover, many groups only recommend the use of DOACs in exceptional cases.[7]

We appreciate the immense challenges of providing uniform recommendations, particularly when the evidence base is low, but we believe that the above issues warrant addressing in future guidelines.

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Correspondence: Manu Juneja, Department of Clinical Haematology, Peter MacCallum Cancer Centre and Royal Melbourne Hospital, 300 Grattan St, Melbourne 3050, VIC, Australia; email: manu.juneja@mh.org.au.

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Data are available upon request from the corresponding author, Manu Juneja (manu.juneja@mh.org.au).

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