Laboratory Diagnosis of von Willebrand Disease (VWD): The 2008 NHLBI Guidelines

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This presentation reviews selected information from the 2008 NHLBI evidence-based guidelines for diagnosis and management of von Willebrand disease (VWD), focusing on recommendations for clinical and laboratory assessment and diagnosis. The full guidelines document, as well as a synopsis ("Pocket Guide") and a patient education brochure, are availalble via the NHLBI web site (www.nhlbi.nih.gov/guidelines/vwd), and an edited version of the full guidelines document has been published (Haemophilia 2008 [March]14[2]:171-232).

Results and Conclusions: From among the main and ancillary diagnostic and management recommendations (>54), several are briefly reviewed in this presentation and a few are summarized as follows. 1) Initial diagnostic evaluation relies on assessment of the hemostatic and medical history using specific queries about key points of the history that are the most useful for assessing the probability of a bleeding disorder, including VWD. 2) Recommended initial testing for VWD includes 3 tests: plasma von Willebrand factor antigen (VWF:Ag), ristocetin cofactor (VWF:RCo) activity, and factor VIII coagulant activity (FVIII); with reflexive or subsequent VWF multimer analysis and optional specialized VWF assays for evaluation of abnormal results, preferably performed in the absence of conditions associated with elevation of VWF, and with careful attention to collection and processing of the blood sample. 3) Calibrators for VWF:Ag, VWF:RCo and FVIII assays should be referenced to the World Health Organization (WHO) plasma standard. 4) Referencing VWF testing results to the population reference range, rather than to ABO-stratified reference ranges, may be clinically useful. 5) The recommended “cut-off” level of VWF:RCo and/or VWF:Ag for definite diagnosis of VWD is <30 IU/dL (%), reflecting multiple considerations; however individuals with 30-50 IU/dL VWF may have VWD or risk of bleeding with invasive procedures and may merit treatment to elevate VWF. 6) Diagnosis of VWD requires correlation of clinical assessment with supportive laboratory testing results.