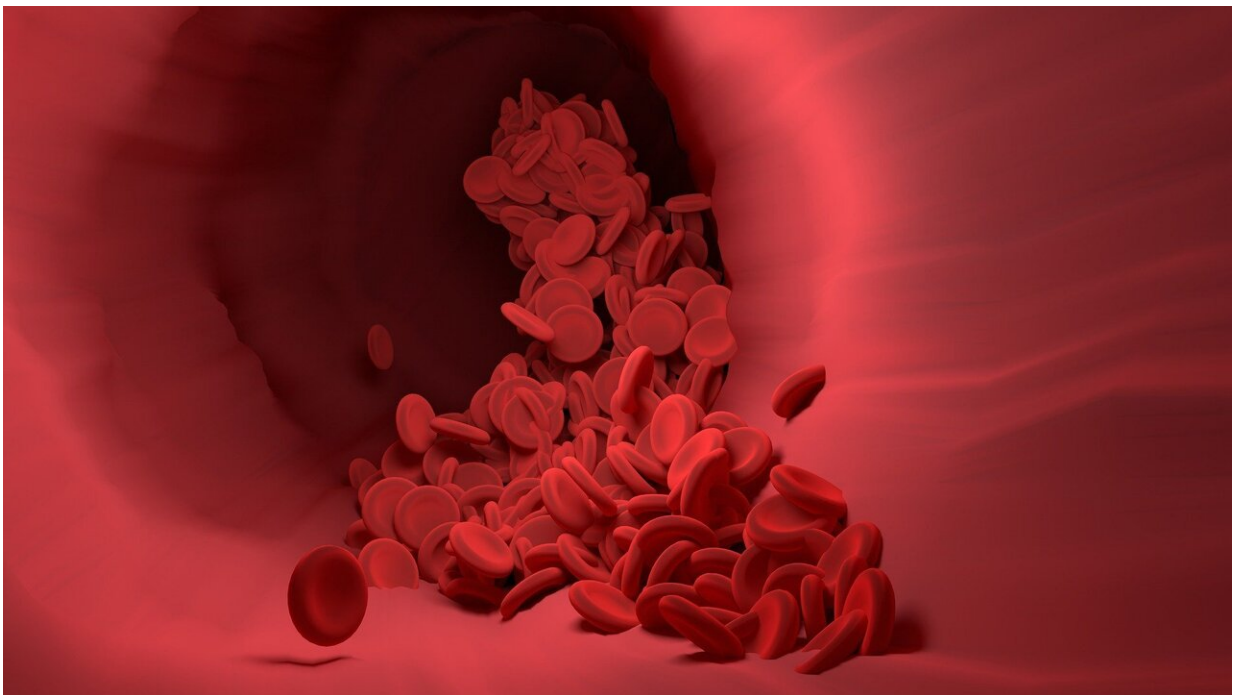


# Organizations collaborate to develop international von Willebrand Disease guidelines

January 12 2021

---



Credit: CC0 Public Domain

The American Society of Hematology (ASH), the International Society on Thrombosis and Haemostasis (ISTH), National Hemophilia Foundation (NHF), and World Federation of Hemophilia (WFH) have developed joint clinical practice guidelines on the diagnosis and

management of von Willebrand Disease (VWD), the world's most common inherited bleeding disorder. The guidelines were published today in *Blood Advances*.

VWD affects approximately 1% of the world's population, and it is the most common bleeding disorder. Although VWD occurs among men and women equally, women are more likely to notice the symptoms because of heavy or abnormal bleeding during their menstrual periods and after childbirth. This inherited condition results in the decreased production, absence, or abnormal function of the clotting protein von Willebrand factor.

VWD can cause unusual bleeding from small wounds or minor procedures, frequent nosebleeds, bruising, bleeding in joints, and heavy menstrual periods and post-partum bleeding in women. Symptoms may vary from patient to patient or in a single patient over the course of his or her life. Primary care providers, pediatricians, obstetricians, and gynecologists who observe unusual bleeding often refer their patients to a hematologist for further testing and management. Many individuals with mild symptoms do not receive a diagnosis right away and live for many years with untreated bleeding or do not realize they have VWD until they experience a severe bleed that could have been prevented.

"While VWD is a common bleeding disorder, it is also complex, presenting challenges in the timely diagnosis and appropriate management of bleeding for patients," said 2021 ASH President Martin S. Tallman, MD, of Memorial Sloan Kettering Cancer Center. "Because diagnosis is not straightforward and symptoms range in severity, there is a need for trustworthy guidelines to help improve the quality of care for patients. There are no better partners for ASH in this effort than ISTH, NHF, and WFH, whose collective expertise in VWD from the scientific and patient perspectives around the world enhanced this effort."

Notably, the guidelines make key statements on thresholds of laboratory testing to classify VWD to be more inclusive of patients who experience bleeding, but whose blood work does not meet currently accepted thresholds for diagnosis. In addition, the guidelines for the first time recommend that VWD patients who suffer from frequent, severe bleeding that decreases quality of life should receive routine VWD prophylaxis—an injectable concentrate of clotting protein—several times a week. Together, the guidelines on diagnosis and management include a combined 19 recommendations.

The guidelines were developed by two expert panels made up of 32 individuals, including U.S.-based and international hematologists, individuals living with VWD, and scientists with expertise in evidence synthesis and appraisal and guideline development methodology. Clinical questions were developed by the panels and prioritized, and an international survey was completed to identify the most important clinical questions. A systematic review of available evidence was conducted by the University of Kansas Medical Center and the panel referred to this evidence to make recommendations. This process shed light on the lack of strong evidence on which to base recommendations, and therefore the report calls for more research.

"These guidelines are an excellent example of collaboration across multiple societies ensuring that the guidelines developed represent the most up to date and relevant advice about diagnosis and treatment of this common, but often misunderstood bleeding disorder," said ISTH Past President Claire McLintock, MD. "Working as part of the incredible team that developed the guidelines in diagnosis of VWD was hugely rewarding and inspiring." People with VWD made up approximately one quarter of each guideline panel's composition. As full voting members they contributed their expertise to every aspect of the rigorous guideline development process, from the prioritization of clinical questions, through the evaluation of the available evidence, to the consideration of

factors such as stakeholder values and preferences, equity, and feasibility. Their involvement, actively supported by the panel chairs and collaborating organizations, was critical to ensuring the development of recommendations with the greatest pertinence and in which the community will have great confidence. Collaboration with WFH and NHF was essential to engaging the broader VWD patient community.

With their extensive established functional networks with healthcare professionals in centers for comprehensive care of bleeding disorders and patient organizations throughout the US and around the world, the four collaborating organizations, ASH, ISTH, NHF, and WFH will all play important roles in the creation and dissemination of resources based on the guidelines, and their implementation in individual care and broader advocacy efforts.

"These guidelines are an extremely important step in our quest to address the difficulties individuals go through to obtain an accurate, timely diagnosis, and appropriate treatment. The challenge ahead will be for us to educate both those living with VWD and [healthcare professionals](#) on the guideline recommendations," said Leonard Valentino, MD, President and CEO of NHF. "We were excited to work with ASH, ISTH, and WFH to create these and introduce them to the community." "In 2017 we gathered members of the global VWD community for the first WFH International Symposium on VWD. Delegates from countries around the world told us that the number one tool they needed in order to provide comprehensive care to people with VWD was clinical practice guidelines," explained WFH President Cesar Garrido. "The WFH is very proud to deliver these guidelines today, with our partners from ASH, ISTH, and NHF, to the global community. In addition to offering guidance for clinicians and individuals with VWD as they partner in personalized care decisions, they constitute the foundation for important advocacy initiatives to improve access to the recommended diagnostic techniques and treatment options in all countries."

The publication of the guideline will be accompanied by tools and educational resources to help patients, hematologists and other health care providers understand and implement the recommendations.

**More information:** Paula D. James et al. ASH ISTH NHF WFH 2021 guidelines on the diagnosis of von Willebrand disease. *Blood Adv* (2021) 5 (1): 280–300. [doi.org/10.1182/bloodadvances.2020003265](https://doi.org/10.1182/bloodadvances.2020003265)

Nathan T. Connell et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease. *Blood Adv* (2021) 5 (1): 301–325. [doi.org/10.1182/bloodadvances.2020003264](https://doi.org/10.1182/bloodadvances.2020003264)

Provided by American Society of Hematology

Citation: Organizations collaborate to develop international von Willebrand Disease guidelines (2021, January 12) retrieved 6 May 2024 from <https://medicalxpress.com/news/2021-01-collaborate-international-von-willebrand-disease.html>

This document is subject to copyright. Apart from any fair dealing for the purpose of private study or research, no part may be reproduced without the written permission. The content is provided for information purposes only.