A Collaborative Approach to Clinical Practice Guidelines

ASH ISTH NHF WFH 2021 Clinical Practice Guidelines on the Diagnosis and Management of von Willebrand Disease

American Society of Hematology | ISTH International Society on Thrombosis and Haemostasis | National Hemophilia Foundation for all bleeding disorders | WFH World Federation of Hemophilia, Fédération Mondiale de l'Hémophilie, Federación Mundial de Hemofilia

CLINICAL GUIDELINES

blood advances

ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease

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Background: von Willebrand disease (VWD) is a common inherited bleeding disorder. Significant variability exists in management options offered to patients.

Objective: These evidence-based guidelines from the American Society of Hematology (ASH), the International Society on Thrombosis and Haemostasis (ISTH), the National Hemophilia Foundation (NHF), and the World Federation of Hemophilia (WFH) are intended to support patients, clinicians, and health care professionals in their decisions about management of VWD.

Methods: ASH, ISTH, NHF, and WFH formed a multidisciplinary guideline panel. Three patient representatives were included. The panel was balanced to minimize potential bias from conflicts of interest. The University of Kansas Outcomes and Implementation Research Unit and the McMaster Grading of Recommendations Assessment, Development and Evaluation (GRADE) Centre supported the guideline development process, including performing and updating systematic evidence reviews (through November 2019). The panel prioritized clinical questions and outcomes according to their importance to clinicians and patients. The panel used the GRADE approach, including GRADE Evidence-to-Decision frameworks, to assess evidence and make recommendations, which were subject to public comment.

Results: The panel agreed on 12 recommendations and outlined future research priorities.

Conclusions: These guidelines make key recommendations regarding prophylaxis for frequent recurrent bleeding, desmopressin trials to determine therapy, use of antiplatelet agents and anticoagulant therapy, target VWF and factor VIII activity levels for major surgery, strategies to reduce bleeding during minor surgery or invasive procedures, management options for heavy menstrual bleeding, management of VWD in the context of neuraxial anesthesia during labor and delivery, and management in the postpartum setting.

Summary of recommendations

These guidelines are based on updated and original systematic reviews of evidence conducted under the direction of the Outcomes and Implementation Research Unit at the University of Kansas Medical Center (KUMC). The panel followed best practices for guideline development recommended by the Institute of Medicine and the Guidelines International Network (G-I-N).[1-3] The panel used the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach to assess the certainty in the evidence and formulate recommendations.[4-10]

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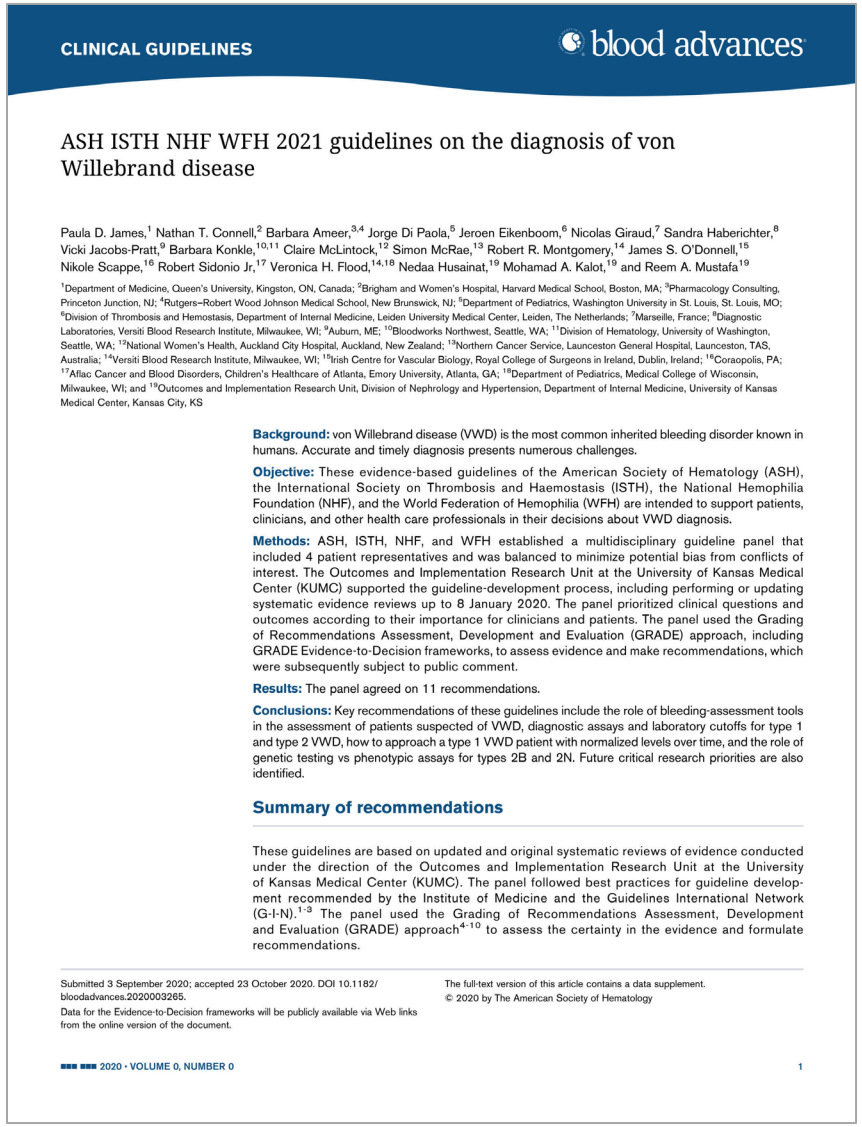
*N.T.C. and V.H.F. contributed equally to this study as first authors.

Data for the Evidence-to-Decision frameworks will be publicly available via Web links from the online version of the document.

The full-text version of this article contains a data supplement.



CLINICAL GUIDELINES

blood advances

ASH ISTH NHF WFH 2021 guidelines on the diagnosis of von Willebrand disease

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Background: von Willebrand disease (VWD) is the most common inherited bleeding disorder known in humans. Accurate and timely diagnosis presents numerous challenges.

Objective: These evidence-based guidelines of the American Society of Hematology (ASH), the International Society on Thrombosis and Haemostasis (ISTH), the National Hemophilia Foundation (NHF), and the World Federation of Hemophilia (WFH) are intended to support patients, clinicians, and other health care professionals in their decisions about VWD diagnosis.

Methods: ASH, ISTH, NHF, and WFH established a multidisciplinary guideline panel that included 4 patient representatives and was balanced to minimize potential bias from conflicts of interest. The Outcomes and Implementation Research Unit at the University of Kansas Medical Center (KUMC) supported the guideline-development process, including performing or updating systematic evidence reviews up to 8 January 2020. The panel prioritized clinical questions and outcomes according to their importance for clinicians and patients. The panel used the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach, including GRADE Evidence-to-Decision frameworks, to assess evidence and make recommendations, which were subsequently subject to public comment.

Results: The panel agreed on 11 recommendations.

Conclusions: Key recommendations of these guidelines include the role of bleeding-assessment tools in the assessment of patients suspected of VWD, diagnostic assays and laboratory cutoffs for type 1 and type 2 VWD, how to approach a type 1 VWD patient with normalized levels over time, and the role of genetic testing vs phenotypic assays for types 2B and 2N. Future critical research priorities are also identified.

Summary of recommendations

These guidelines are based on updated and original systematic reviews of evidence conducted under the direction of the Outcomes and Implementation Research Unit at the University of Kansas Medical Center (KUMC). The panel followed best practices for guideline development recommended by the Institute of Medicine and the Guidelines International Network (G-I-N).[1-3] The panel used the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach[4-10] to assess the certainty in the evidence and formulate recommendations.

Submitted 3 September 2020; accepted 23 October 2020. DOI 10.1182/bloodadvances.2020003265.

Data for the Evidence-to-Decision frameworks will be publicly available via Web links from the online version of the document.

The full-text version of this article contains a data supplement.

Connell NT, Flood VH, Brignardello-Petersen R, et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand disease. *Blood Adv.* 2021;5(1):301-325.

James PD, Connell NT, Ameer B, et al. ASH ISTH NHF WFH 2021 guidelines on the diagnosis of von Willebrand disease. *Blood Adv.* 2021;5(1):280-300.

Meet The Speakers

Nikole Scappe

Manager of Education,

National Hemophilia Foundation

Ellen Riker

Consultant,

National Hemophilia Foundation

American Society of Hematology

Kailee Boedeker, MPH

Manager of Clinical Quality Improvement, American Society of Hematology

A Collaborative Approach to Clinical Practice Guidelines: The Patient Lens

Nikole Scappe
Manager of Education
National Hemophilia Foundation

NATIONAL HEMOPHILIA FOUNDATION
for all bleeding disorders

Disclosures

N/A

OUR MISSION

The National Hemophilia Foundation (NHF) is dedicated to finding cures for inheritable blood disorders and to addressing and preventing the complications of these disorders through research, education, and advocacy, enabling people and families to thrive.

NUESTRA MISIÓN

La Fundación Nacional de Hemofilia (NHF) se dedica a encontrar curas para los trastornos sanguíneos hereditarios y a abordar y prevenir las complicaciones de estos trastornos a través de la investigación, la educación y la abogacía permitiendo que las personas y familias prosperen.

NATIONAL HEMOPHILIA FOUNDATION
for all bleeding disorders

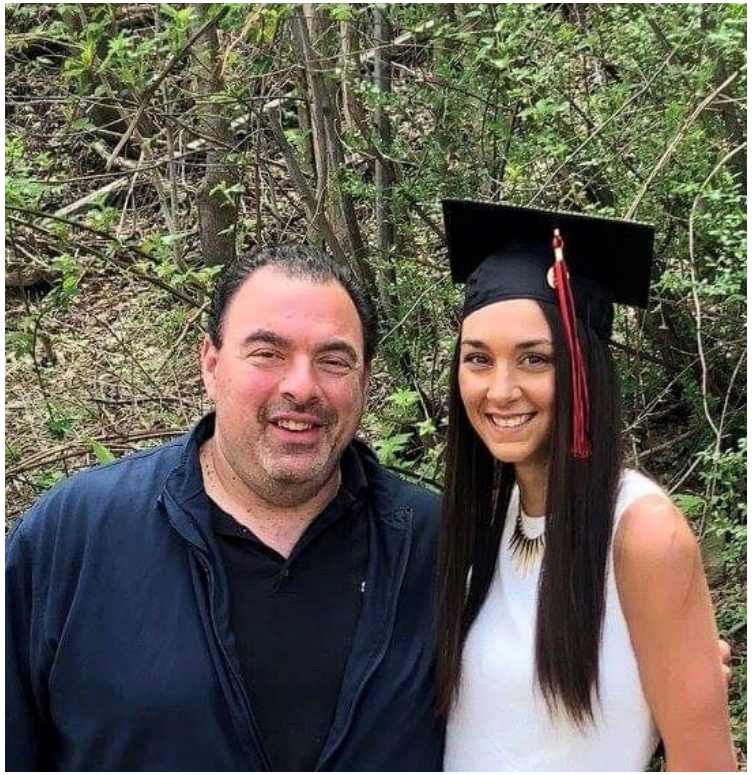
Agenda

1. Background of von Willebrand disease (VWD)
2. My Story
3. Team approach to guidelines and training
4. Working for NHF and education around the guidelines

Goal: By the end of my presentation, I hope that you will see the direct impact that patient's involvement can have on the creation of guidelines and other similar documents and policies that affect the very lives of those patients

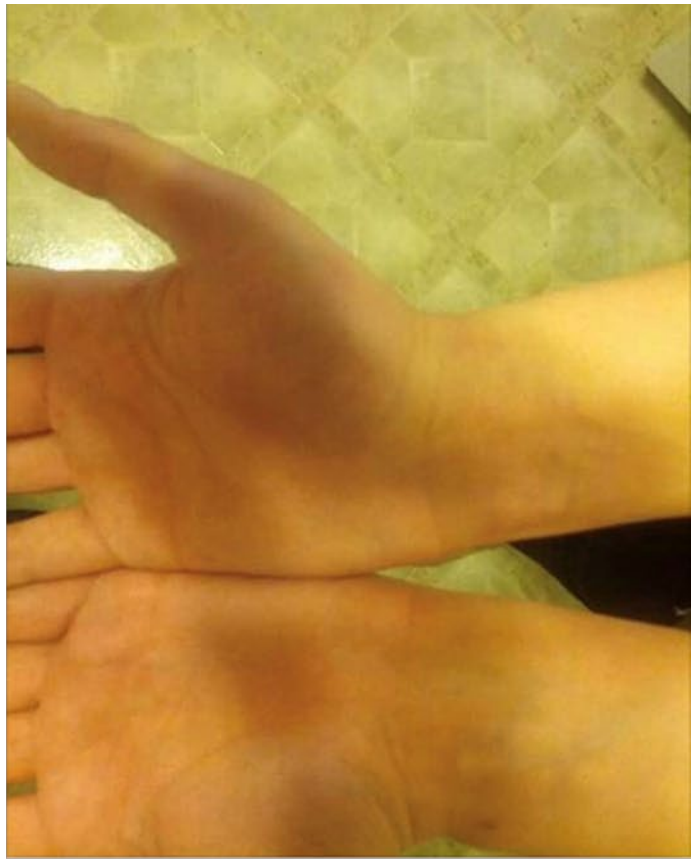
Von Willebrand Disease (VWD)

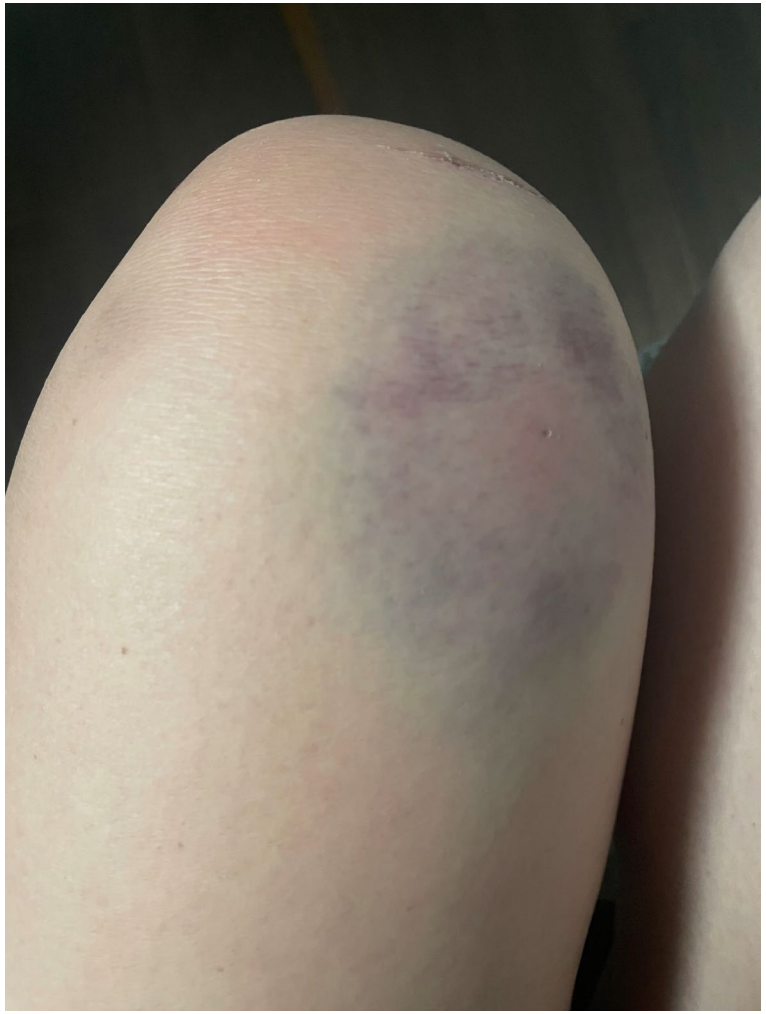
- **Most common inherited bleeding disorder:**
  - VWD is the most common bleeding disorder, affecting up to 1% of the US population – or approximately 1 in every 100 people.
- **Can cause unusual bleeding**
  - From small wounds or “minor” procedures
  - Frequent nosebleeds and bruising into joints
- **Inherited equally by men and women**
- **Often impacts women disproportionally through**
  - Heavy or abnormal periods
  - Post-partum bleeding

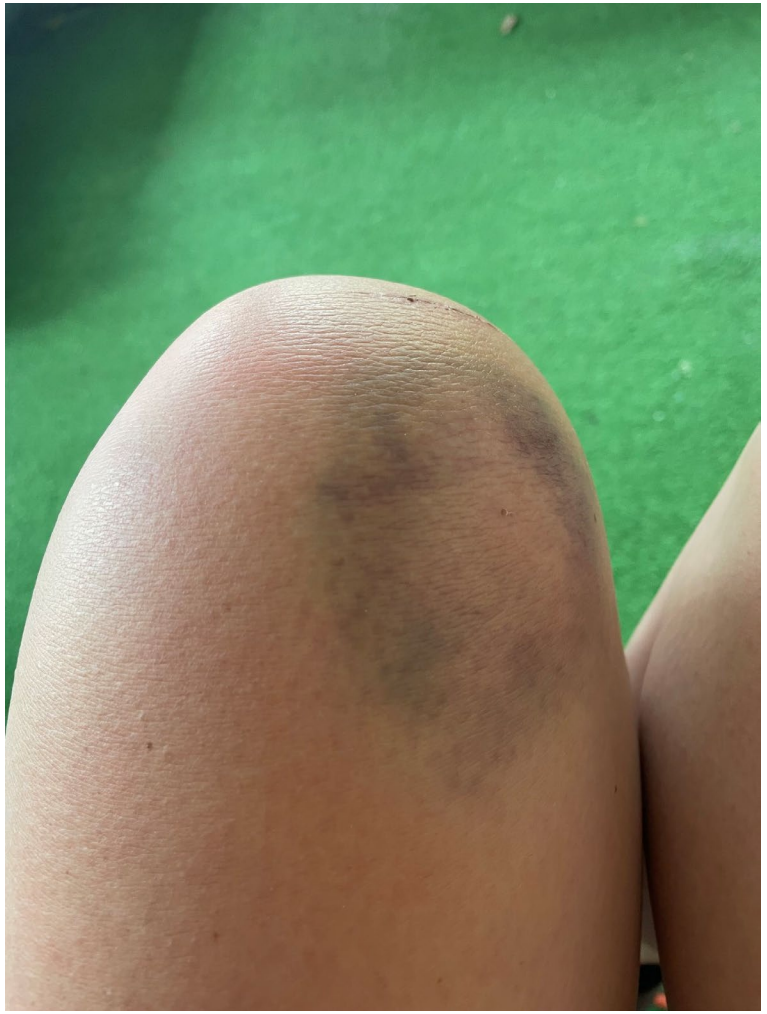


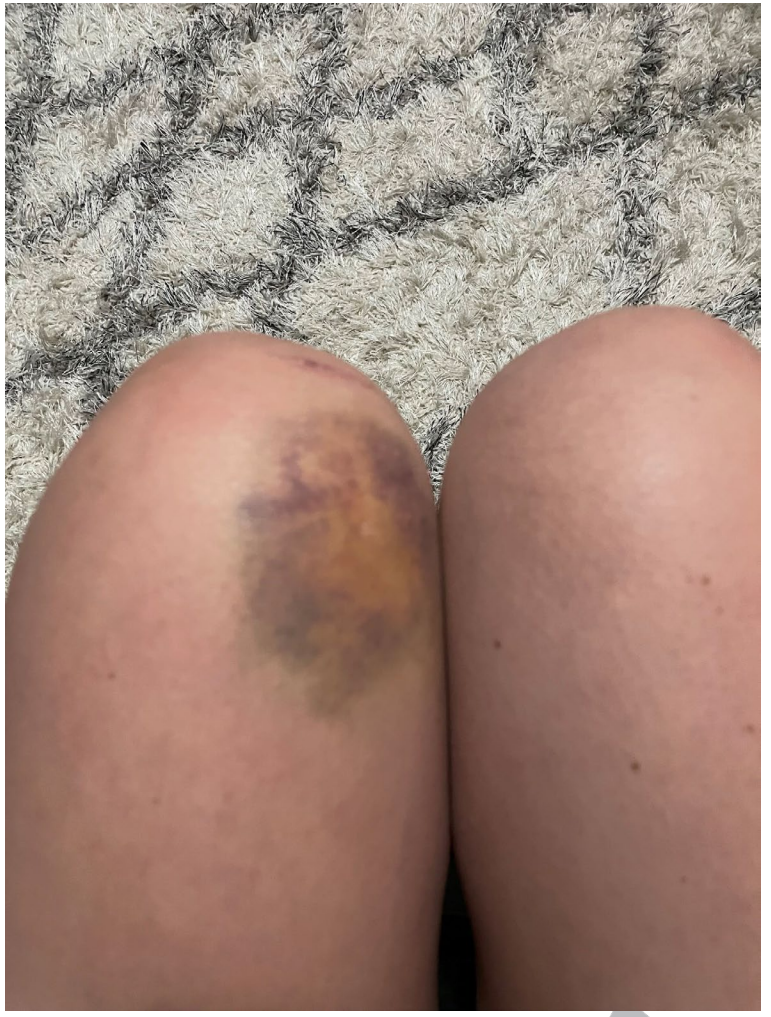

I SERVED WITH PRIDE


MOON
25
Wilson





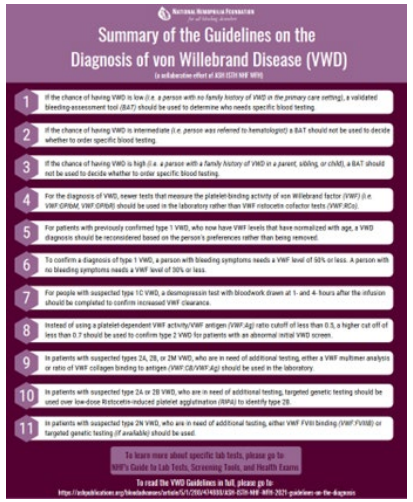


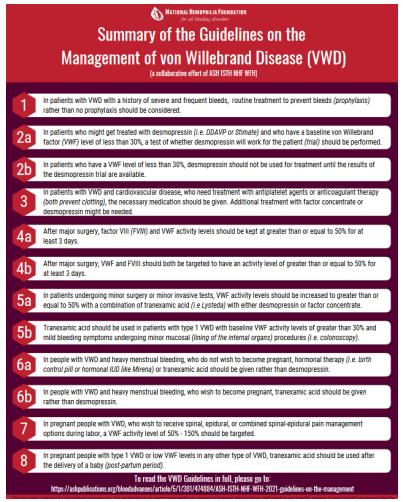


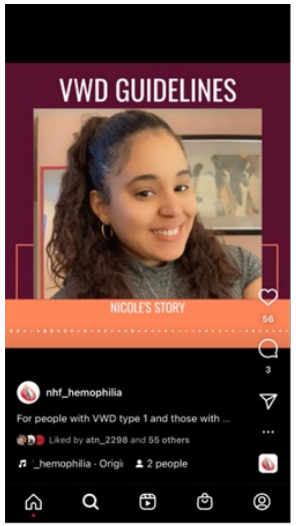
Number of patients on panels:

Diagnosis: 4

Management: 3



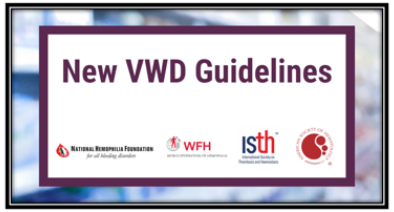




VWD Guidelines: What They Mean For You

Nikole Scappe
Manager of Education

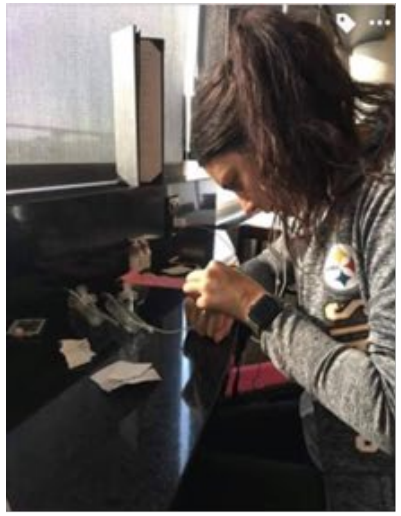
NATIONAL HEMOPHILIA FOUNDATION
for all bleeding disorders



Wednesday Webinar Series

4-part Wednesday Webinar Series

- Wednesday Webinar: New Guidelines for VWD: Patient Perspective | National Hemophilia Foundation
- VWD Guidelines: Diagnoses | National Hemophilia Foundation
- The New VWD Guidelines: Management | National Hemophilia Foundation
- VWD Guidelines and Shared Decision-making | National Hemophilia Foundation




I SERVED WITH PRIDE


National Hemophilia Foundation
for all bleeding disorders

A Collaborative Approach to Clinical Practice Guidelines

Presentation by

Ellen Riker

Principal, Artemis Policy Group

Disclosures

- Consultant to American Society of Hematology
- Consultant to National Hemophilia Foundation

2015

"A well-qualified and authoritative organization, or a consortium of such organizations, should develop a new or updated evidence-based clinical practice guideline on VWD."

Report of November 2014 National Hemophilia Foundation Strategic Summit on VWD

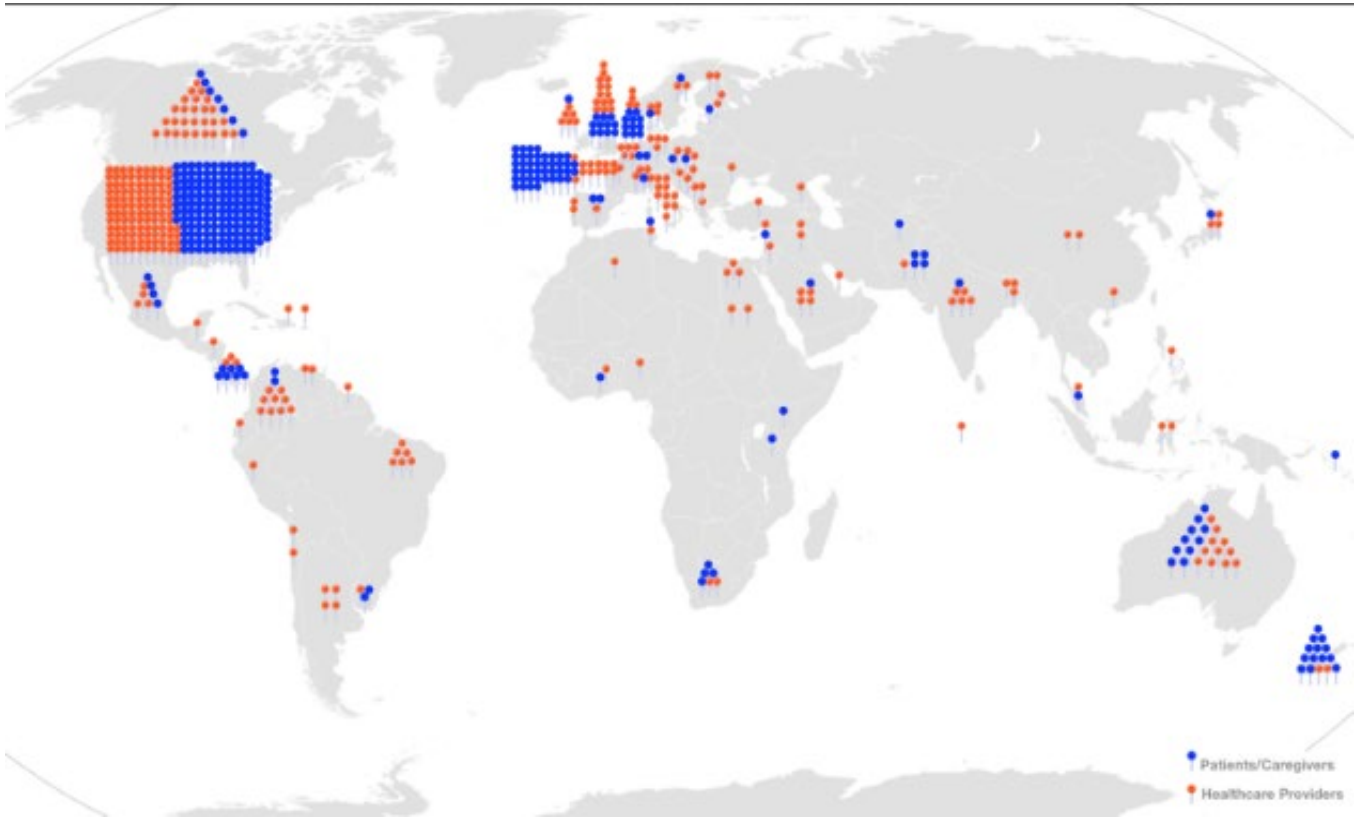
NATIONAL HEMOPHILIA FOUNDATION
for all bleeding disorders

Collaboration Gets Underway…

- ASH and NHF spoke about co-sponsoring the VWD Guidelines and together decided to include the international organizations – International Society of Thrombosis and Hemostasis (ISTH) and the World Federation of Hemophilia (WFH).
- NHF's Medical and Scientific Advisory Council and Chapter leaders identified clinicians and people living with VWD to nominate to serve on the panel. Patient participation was critical to NHF
- As part of our MOU with ASH, NHF and the other organizations would allow staff to attend all panel meetings.
- Recognizing the limitations in the literature, NHF and WFH asked that we survey clinicians and people living with VWD internationally seeking their input on priorities related to the diagnosis and management of VWD.

Scoping Survey

71 countries	6 continents	601 participants
9,500 discrete comments	51% patients	49% healthcare providers
21% male	54% female	26% no gender identified
18% low / middle-income	82% high-income countries	



Kalot, MA, et al; An international survey to inform priorities for new guidelines on von Willebrand disease. *Haemophilia*. 2020; 26: 106– 116 https://onlinelibrary.wiley.com/doi/full/10.1111/hae.13881

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Results - New ICD-10 VWD Diagnosis Codes

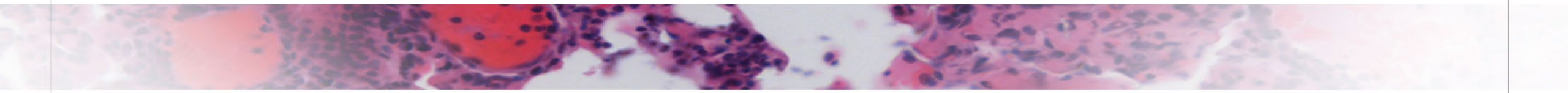
- Previously, one ICD-10 diagnosis code, D68.0, Von Willebrand disease, for VWD and all subtypes
- New clinical practice guidelines for diagnosis and treatment of VWD - recommendations varied by type of VWD
- Submitted to CDC ICD-10 Coordination and Maintenance Committee March 2021
- Proposal was accepted and codes were published in the Medicare Inpatient PPS proposed rule for FY 2023

New ICD-10 VWD Diagnosis Codes as of 10/1/22

- D68.00, Von Willebrand disease, unspecified
- D68.01, Von Willebrand disease, type 1
- D68.020, Von Willebrand disease, type 2A
- D68.021, Von Willebrand disease, type 2B
- D68.022, Von Willebrand disease, type 2M
- D68.023, Von Willebrand disease, type 2N
- D68.029, Von Willebrand disease, type 2, unspecified
- D68.03, Von Willebrand disease, type 3
- D68.04, Acquired von Willebrand disease
- D68.09, Other von Willebrand disease

American Society *of* Hematology

Helping hematologists conquer blood diseases worldwide



Disseminating and Implementing Guideline Recommendations

Kailee Boedeker, MPH

American Society of Hematology

Disclosures

- N/A

Strategic Approach

Raise Awareness

Dissemination

Implementation

Raising Awareness of the Guidelines

- Joint Press Release
- Social Media Campaigns
- E-newsletters
- Video Interviews with Panelists
- Presentation and Promotional Activities at Relevant Meetings

PRESS RELEASES

AMERICAN SOCIETY OF HEMATOLOGY / NEWSROOM / PRESS RELEASES / ORGANIZATIONS COLLABORATE TO DEVELOP INTERNATIONAL STATE-OF-THE-A

Organizations Collaborate to Develop International State-of-the-Art Guidelines on the Diagnosis and Management of von Willebrand Disease

CITATIONS

PUBLISHED ON:
JAN 12 2021

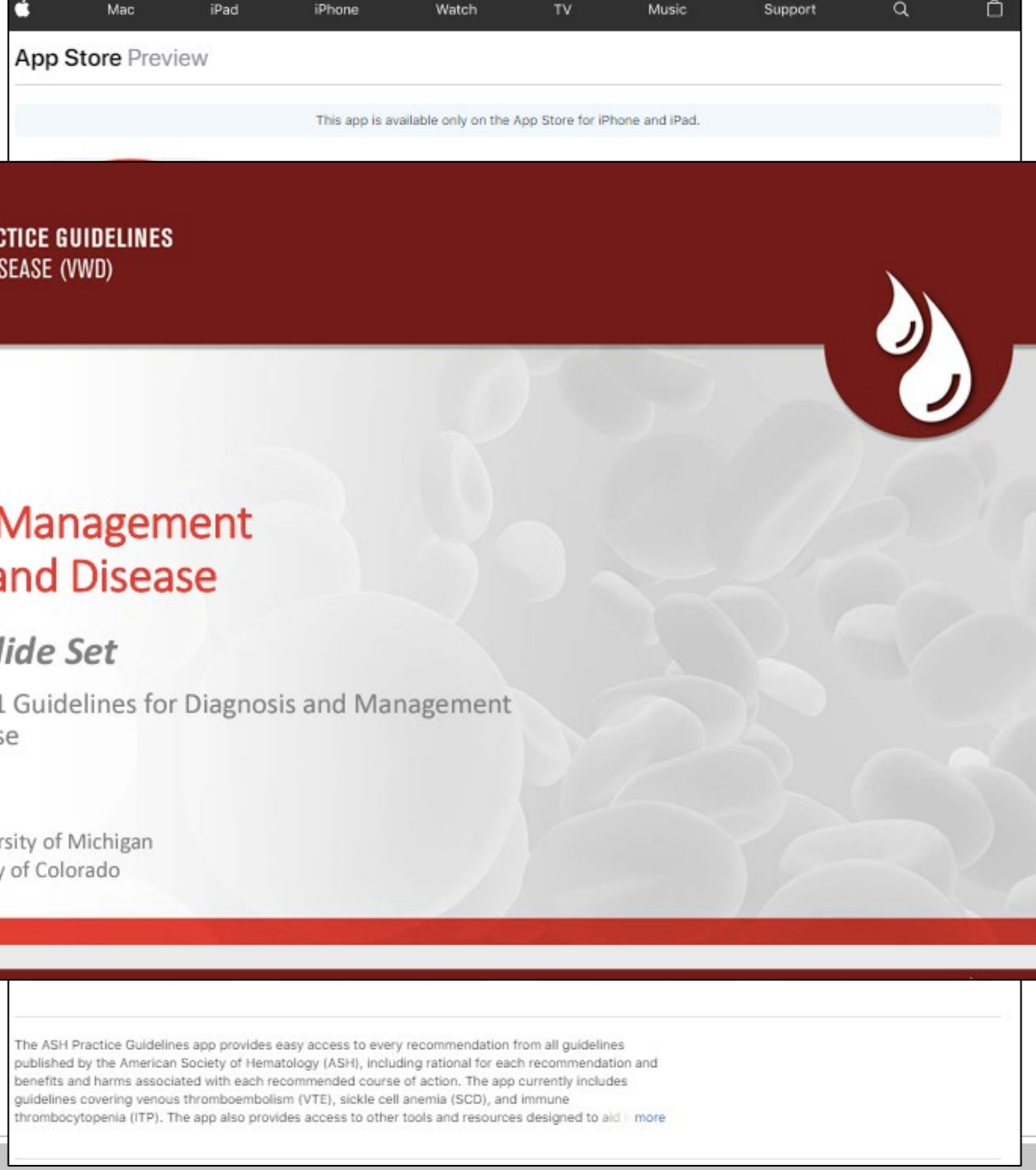
(WASHINGTON – January 12, 2021) The American Society of Hematology (ASH), the International Society on Thrombosis and Haemostasis (ISTH), National Hemophilia Foundation (NHF), and World Federation of Hemophilia (WFH) have developed joint clinical practice guidelines on the diagnosis and management of 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.

Dissemination Activities

- Infographics and Snapshots
- Translations
- Patient Summaries
- Mobile App
- Educational Teaching Slides
- Guideline International Library Repository
- Podcasts
- Community Workshops



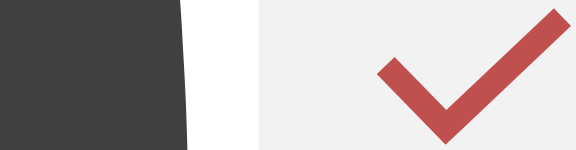
Dissemination Activities

- Virtual Summits
- Commentaries
- CDC Blood Disorders
- Pocket Guides
- Annual Meeting Sessions
- Webinar Series

Implementation

- ICD Codes
- Patient Decision Aids
- Guideline Implementation Champions
- Examples of possible future activities
  - Quality Measures
  - QI Toolkits
  - Clinical Decision Support

Next Steps



Impact Evaluation

Monitoring and Updating

Thank You!

Questions?