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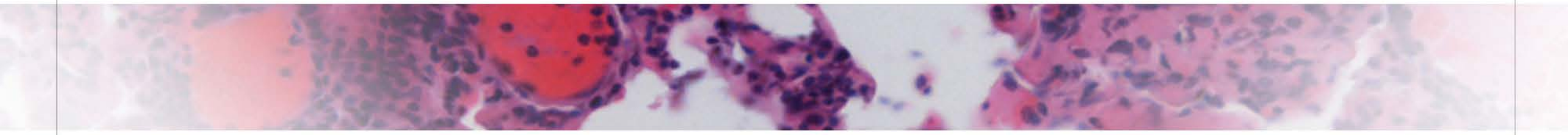

ICD-10-CM Diagnosis Topic: von Willebrand disease

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von Willebrand disease (VWD)

- Most common inherited bleeding disorder, in which the blood does not clot properly.
- Estimated that between 1 in 1,000 and 1 in 10,000 people are affected by symptomatic bleeding.*
- Multiple subtypes of VWD that require individualized treatment based on specific diagnosis, with a range of symptoms and multiple therapies available to treat them.
- Symptoms can disproportionately affect women, who may experience menstrual and postpartum hemorrhage.

*Barbara A. Konkle and Steven W. Pipe. Diagnosis and management of von Willebrand disease: A community-wide effort to deliver evidence-based clinical practice guidelines. *Haemophilia*. 2021;00:1-3.

Impetus for Change

- Currently, only one ICD-10-CM code, D68.0, Von Willebrand's disease, exists, making it difficult to accurately document, diagnose and in turn, appropriately treat the different subtypes of VWD.
- In 2006, the International Society on Thrombosis and Haemostasis (ISTH) published classification for the subtypes of VWD.
- In January 2021, American Society of Hematology, ISTH, the National Hemophilia Foundation, and the World Federation of Hemophilia published guidelines on the diagnosis and management of VWD.

Request for Additional Codes

- **TYPE 1 VON WILLEBRAND DISEASE**: partial quantitative deficiency of von Willebrand factor (VWF)

TYPE 2 VON WILLEBRAND DISEASE: qualitative defects of von Willebrand factor (VWF):

- **TYPE 2A VON WILLEBRAND DISEASE**: qualitative defects of VWF includes variants with decreased platelet adhesion characterized by selective deficiency of high-molecular-weight VWF multimers.
- **TYPE 2B VON WILLEBRAND DISEASE**: qualitative defects of VWF that identifies “hyper-adhesive” VWF forms also usually associated with high-molecular-weight VWF loss (includes variants with increased affinity for platelet glycoprotein lb).
- **TYPE 2M VON WILLEBRAND DISEASE**: qualitative defects of VWF includes variants with markedly defective platelet adhesion with a normal size distribution of VWF multimers.
- **TYPE 2N VON WILLEBRAND DISEASE**: qualitative defects of VWF includes variants with markedly decreased affinity for factor VIII. (identifies defective VWF:FVIII binding).

Request for Additional Codes cont.

- **TYPE 3 VON WILLEBRAND DISEASE**: total quantitative deficiency of VWF (complete or near complete absence of VWF).

- **ACQUIRED VON WILLEBRAND SYNDROME**: The acquired von Willebrand syndrome (AVWS) is a deficiency in the amount or function of VWF that is due to acquired rather than inherited causes. AVWS may arise due to autoantibody formation such as that seen in immune dysregulation disorders or VWF may be directly adsorbed onto malignant cells as observed in patients with Wilms tumors or Waldenstrom macroglobulinemia. Treatment consists of supportive therapy with VWF concentrate, desmopressin, and/or antifibrinolytic therapy along with correction of the underlying cause (e.g. valve replacement therapy or immunosuppression).

- **PLATELET-TYPE VON WILLEBRAND DISEASE**: Platelet-type von Willebrand disease is due to a functional defect in the platelet receptor for von Willebrand factor. Often misdiagnosed as type 2B von Willebrand disease, treatment consists of platelet transfusions in addition to standard VWD therapies such as VWF concentrate or antifibrinolytic therapy.
    - ***Inclusion term within VWD, other code***

Support for Proposal

- Letters of Support Received from:
  - American College of Physicians
  - National Hemophilia Foundation
  - International Society on Thrombosis and Haemostasis
- Proposal reviewed by:
  - American Academy of Pediatrics
  - American College of Obstetricians and Gynecologists

QUESTIONS?