von Willebrand Disease

Hannah Smith
hs276009@yahoo.com

Follow this and additional works at: http://digitalcommons.otterbein.edu/stu_msn
Part of the Nursing Commons

Recommended Citation
Smith, Hannah, "von Willebrand Disease" (2017). Master of Science in Nursing (MSN) Student Scholarship. 244.
http://digitalcommons.otterbein.edu/stu_msn/244
Inheritance patterns: The subtypes are VWD 1, VWD2A, VWD2B, VWD2M, VWD2N and VWD 3. VWD 1 is the mildest and VWD 3 is the most severe. VWD 1, VWD2B, VWD2M are an autosomal dominant inheritance pattern. VWD2N and VWD 3 are a recessive inheritance pattern and VWD2A can be both (Federici, 2016, p. 42). Type 3 VWD contains null alleles which results in the complete nonappearance on von Willebrand factor. Type 1 VWD has incomplete penetrance and involves defects in intracellular mRNA splicing, secretion and final part of the von Willebrand factor. Type 3 is fully penetrant and results in restriction of the von Willebrand factor protein (Leebeek & Eikenboom, 2016, p. 2073).

Normal von Willebrand function: von Willebrand factor (VWF) is synthesized in the endothelial cells and is either secreted or stored in the endothelial cells where they are kept until they are stimulated. VWF contains amino acids that play a role in the structure or function of the molecule. VWF is made specifically in the endoplasmic reticulum of the cell where it becomes a large high molecular weight protein that binds collagen and contains the platelet receptors GPIb and GPII/IIa (Schneppenheim, 2011, p. 53-54).

Subtype pathophysiology: The subtypes are classified as such due to their mechanism of action. VWD 1, the mildest form, has less than the normal amount of von Willebrand factor. VWD2A has decreased platelet adhesion. VWD2A loses its platelet adhesion because it loses its multimer shape and is no longer able to bind to the receptor (Brooks et al., 2016, p. 360). VWD2B has increased affinity for the platelet GPIb alpha. VWD2B decreases binding for factor VIII. Factor VIII is a protein that plays a role in the clotting cascade. Finally, VWD 3, is the most severe, is complete loss of any VWF (Federici, 2016, table 1).

Implications for nursing care

When a patient presents with the signs and symptoms of von Willebrand disease the nurse practitioner should begin with a detailed history, especially a familial health history, and a physical examination. Preliminary laboratory tests include a platelet count, thyroid level, prothrombin time, activated partial thromboplastin time and a hemoglobin and hematocrit level (Brooks, Brooks, & Aluva, 2016, p. 102). If these results come back as abnormal, if there is a familial history of bleeding disorders or any of the severe bleeding signs are present (see above) the patient should be screened for a bleeding disorder. VWD screening tests include VWF antigen (VWF:Ag), VWF ristocetin cofactor activity (VWF:RCo), factor VIII activity (FVIII:C) and VWF:Ag/VWF:RCo (Roberts & Flood, 2015, p. 12).

Once a definitive diagnosis for VWD has been made the nurse practitioner should refer the patient to a hematologist. From there, the patient will be given the appropriate treatment depending on the subtype VWD the patient has. After the patient has received initial appropriate treatment and is stable the nurse practitioner can care for the patient to perform routine laboratory work and prescribe medication (Brooks et al., 2016, p. 104).

Type 1 VWD (and sometimes type 2) is commonly treated with desmopressin acetate (DDAVP). The use of this drug increases factor VIII and of VWF. The drug causes temporary vasoconstriction which elicits the endothelial cells to release clotting factors (Kaufman, 2014, p. 91). In type 3 and in the majority of type 2 patients factor replacement is necessary (Leebeek & Eikenboom, 2016, p. 2075-2076).

Transgenic and aminocaproic acid are fibrinolysis inhibitors which can be given in a mouthwash form prophylactically to patients undergoing dental procedures (Leebeek & Eikenboom, 2016, p. 2077).

Antibiotics may be given to individuals with type 2B VWD due to the mutated GP-IB/IX platelet receptor. The mutated receptor has a dysfunctional affinity for platelets making the VWF “tied up” with the platelets rather than being able to aid in the clot formation of a real vascular injury (Ware, 2013, p. 5004).

Oral contraceptives can be given to women with symptoms of heavy menstrual cycles but special care must be taken to not mask the symptoms before getting a definitive von Willebrand disease diagnosis (Brooks et al., 2016, p. 104).

The nurse practitioner should be aware that the patient will have a higher risk for infection, medical complications, venous thromboembolism and blood transfusions following trauma or surgical procedures (Cancienne et al., 2015, p. 228).

During surgery the patient must be educated about pre-surgical preparation, new medications, how to care for oneself and given information about support groups, community resources and genetic counseling (Brooks et al., 2016, p. 102).

Conclusion

Von Willebrand factor’s function is to bind to collagen sites during vascular injury, play a role in platelet adhesion and aggregation, and is a carrier protein for factor VIII. Malfunctioning symptoms of pathological bleeding occur. (Leebeek & Eikenboom, 2016).

VWD2A is neither autosomal dominant nor recessive. (“How von Willebrand is inherited,” 2014). The types and subtypes are determined by inheritance patterns and underlying pathophysiology. The mildest (type 1) is caused by insufficient VWF and the most severe (type 3) is complete absence of VWF (Federici, 2016).

Diagnoses is made from a wide range of laboratory results that each type (and subtype) follow (Roberts & Flood, 2015, p. 12).

Treatment also varies based on the type of VWD diagnosed. DDAVP, anti-platelets and intravenous infusions are used based on all options. Despite treatment patients are at higher risk for infection and other complications following surgery or childbirth (Leebeek & Eikenboom, 2016).

References