von Willebrand Disease in Pregnancy

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Imagine this scenario: A young woman presents to the labor and birth unit with no prenatal care. She is a 16-year-old gravida 1 para 0-0-0-0 with obesity who is unsure of her dates. Her fundus measures 34 cm, and fetal movement is palpated. She is complaining of bright red vaginal bleeding that started while she was sleeping, and she reports no pain. She saturated the bed, reporting a stain of blood about a foot across. Her father, who had previously been unaware of her pregnancy, is with her and is very concerned, because her mother had died in childbirth from bleeding after birth.

When questioned about her past medical history, the young woman reports always having very heavy periods, using 10 to 12 tampons per day on her first 2 days. She sometimes has bleeding when she brushes her teeth too hard. On physical examination, she has multiple bruises that are of unknown origin. She states that she has always bruised very easily.

Abstract von Willebrand disease is the most prevalent inherited bleeding disorder, affecting up to 1.3% of the population. It is caused by a defect or deficiency of the von Willebrand factor. Women with the condition may not be aware of their condition at the time of childbirth, but they are at high risk of postpartum hemorrhage even days after birth. In this article we briefly review the condition and specific considerations for the antepartum, intrapartum, and postpartum phases. It is important for nurses who care for women during childbirth to have a keen understanding of this condition.
http://dx.doi.org/10.1016/j.nwh.2016.08.002

Keywords bleeding disorder | postpartum hemorrhage | pregnancy | von Willebrand disease

von Willebrand Disease

von Willebrand disease is the most prevalent inherited bleeding disorder, affecting up to 1.3% of the population (American College of Obstetricians and Gynecologists [ACOG], 2013). It is caused by a defect or deficiency of the von Willebrand factor, which is located on chromosome 12 (Pacheco et al., 2010). The von Willebrand factor functions to support platelet aggregation and formation of clotting at an injury site and is a carrier protein for Factor VIII (James et al., 2009).

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Symptoms

A person with von Willebrand disease presents with bleeding symptoms including excessive bruising or bleeding after injury or procedure, epistaxis, or gingival bleeding (ACOG, 2013). Menorrhagia is the most common symptom in women, often at the onset of menses (James, 2006). Women with von Willebrand disease are also at increased risk to develop hemorrhagic ovarian cysts (James, 2006).

Disease Classifications

There are three main classifications of von Willebrand disease: type 1, type 2 (which has four variations), and type 3.

Type 1

Type 1 is the most common type and accounts for about 80% of cases; it is caused by deficiency of von Willebrand factor (Pacheco et al., 2010). This type is autosomal dominant, and symptoms vary from mild to moderate (Pacheco et al., 2010).

Type 2

Type 2 accounts for 20% of cases and is caused by structural or functional defects in von Willebrand factor (Pacheco et al., 2010).

Type 2A. Type 2A is characterized by normal or slightly reduced von Willebrand factor and dysfunctional platelet aggregation (Pacheco et al., 2010). This type can be inherited as autosomal dominant or recessive (Pacheco et al., 2010).

Type 2B. Type 2B is characterized by a defect that increases platelet binding, causing mild thrombocytopenia that may worsen with pregnancy (Kujovich, 2005; Pacheco et al., 2010). This type is autosomal dominant (Pacheco et al., 2010). Many women with type 2B are misdiagnosed with idiopathic thrombocytopenic purpura and are treated ineffectively (Kujovich, 2005).

Type 2M. Type 2M is caused by decreased platelet adhesion due to low affinity between platelets and von Willebrand factor (Pacheco et al., 2010). Type 2M can be inherited as autosomal dominant or recessive (Pacheco et al., 2010).

Type 2N. Type 2N is caused by a defect that inactivates the Factor VIII binding site on von Willebrand factor, resulting in low Factor VIII levels but normal von Willebrand factor levels (Kujovich, 2005). This type is autosomal recessive and often presents similarly to hemophilia A (Pacheco et al., 2010).

Type 3

Type 3 is the least common but most severe form of von Willebrand disease; it accounts for 5% to 10% of cases (Kujovich, 2005). In type 3, von Willebrand factor levels are undetectable and Factor VIII levels are extremely low (Pacheco et al., 2010). Type 3 is autosomal recessive (Pacheco et al., 2010).

Treatment

Treatment options for women with von Willebrand disease presenting with menorrhagia include combined oral contraceptive pills or progestin-only contraceptives, including pills, implant, or intrauterine device (ACOG, 2013). After childbearing is complete, a hysterectomy or endometrial ablation may be appropriate to control heavy uterine bleeding (James, 2006).

Prophylactic treatment during pregnancy is recommended when Factor VIII levels are less than 25% of the normal range, which varies by age, and von Willebrand factor is less than 50 IU/dL (Kujovich, 2005). Treatment options for von Willebrand disease during pregnancy include desmopressin, transfusional therapy,

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