THE CHILDREN'S MEDICAL CENTER OF DAYTON

Pediatric Clips NURSING

Von Willebrand disease

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Pediatric Nursing Clips by Pediatric Advanced Practice Nurses at Dayton Children's provides quick reviews of common pediatric conditions.

The Children's Medical Center of Dayton is the region's pediatric referral center for a 20-county area. As the only facility in the region with a full-time commitment to pediatrics, Dayton Children's offers a wide range of services in general pediatrics as well as in 35 subspecialty areas for infants, children and teens. We welcome your inquiries about services available – call 937-641-3666 or e-mail marketing @childrensdayton.org.

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CASE STUDY

Kelsey, a 14-year-old female, was referred to the hematology department with a two-year history of menorrhagia, easy bruising and occasional episodes of epistaxis. Her father had epistaxis as a child and her paternal grandmother had a hysterectomy at an early age due to menorrhagia. Menorrhagia is menstruation at regular cycle intervals but with excessive flow and duration. Clinically, it is defined as total blood loss exceeding 80 mL per cycle or menses lasting longer than seven days. She had no history of surgery and denied gum bleeding. Her mother stated, "All the females in Kelsey's dad's family have bad periods."

DIAGNOSTIC WORKUP

Initial work-up:

CBC with platelet count: Normal
PT: 11.8 seconds (11.8-16.4)
APTT: 36 seconds (25.7-37.8)
Ristocetin Cofactor (VWF:RCO): 31.6% (37.5-117.1)
Von Willebrand Antigen (VWF:Ag): 37.4% (48.1-146)
Blood Type: O+
Multimers: Normal
Factor VIII: 62% (50-150%)
Platelet Function Screen: EPI 98 (<167)

Repeat testing:

VWF:RCO: 28.4%
VWF:Ag: 35.1%

DIAGNOSIS

Von Willebrand disease, type 1

CASE DISCUSSION

Von Willebrand disease (VWD) is a hereditary bleeding disorder caused by the decreased production or the decreased function of the von Willebrand Factor (VWF). VWF is a protein in the blood stream that is necessary for normal blood clotting. Persons with VWD produce less VWF or a molecule that does not function normally. Dr. Erik von Willebrand, a physician from Helsinki, Finland, first described the disorder in 1925. He studied a large family whose numerous family members had minor to major bleeding episodes. Both males and females exhibited bleeding symptoms, one of the daughters bleeding to death during her fourth menstrual cycle. She most likely had VWD, type 3, which is a severe form of the disorder.

VWD is the most common bleeding disorder and has worldwide distribution. It is estimated that one to three percent of the population are affected. Males and females of all races can be affected and diagnosed at any age. Symptoms are generally mild and include:

- Epistaxis
- Gum or mouth bleeding
- Easy bruising
- Heavy and/or prolonged menses
- Mucus membrane bleeding
- Prolonged bleeding at time of surgery
- Post-partum bleeding
- Rare joint or muscle bleeding (except in severe VWD)

The four types of VWD include:

- Type 1 - Autosomal dominant, reduced levels of VWF, VWF that is present functions normally, mild and most common type, quantitative disorder
- Type 2 - Autosomally inherited, several different subtypes, may have enough VWF, but the protein present is missing a portion of the molecule and functions abnormally, qualitative disorder
- Type 3 - Autosomal recessive, total or near total absence of VWF, marked reduced levels of Factor VIII, severe disorder, rare incidence 1:1,000,000
- Acquired - New onset of hemorrhagic symptoms, usually develops in association with unrelated underlying disorders (eg, autoimmune diseases, cancer, congenital heart disease)

Persons with VWD do not bleed faster than those without. However, they do not clot as effectively and therefore can bleed longer. When the VWF is not present or does not function properly, platelets do not stick readily to the damaged blood vessel. Normally, when a blood vessel is injured, the wall of the vessel narrows or constricts to reduce the blood flow. Blood platelets then aggregate to the injured site and stick together to form a platelet plug. A series of blood proteins weave together to form a fibrin or mature blood clot, then bleeding stops and healing can occur. Those deficient in VWF make a weak platelet plug that does not stick together and can break off prematurely. When this happens the cycle must be repeated and bleeding then continues for a longer period of time. VWF has been referred to as the glue that holds the blood clot together.

MANAGEMENT

Generally bleeding symptoms from VWD, type 1, are mild and do not require treatment. Medications may only be required to prevent bleeding associated with surgery, dental extractions

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or for trauma. Several medications can be used to treat VWD. Desmopressin acetate is a synthetic hormone that following administration releases VWF stored in the lining of blood vessels. Two forms of Desmopressin acetate include: DDAVP, administered intravenously, and Stimate, a highly concentrated nasal spray. If DDAVP is ineffective, a VWF containing Factor VIII concentrate is recommended. Alphanate and Humate-P are the only FDA approved concentrates indicated for the treatment of VWD.

Evaluation of the response to DDAVP or Stimate is important. Most patients with type 1 will have a recommended Stimate trial to determine the effectiveness. Kelsey came to the hematology clinic for the Stimate trial. A von Willebrand activity level (VWF:RCO) was drawn and resulted at 35% (37.5-117.1). One spray of Stimate was administered intranasally. A second VWF:RCO was drawn one hour after the Stimate was given and Kelsey had a three-fold increase in VWF:RCO levels. These results indicate that Stimate is an appropriate medication for Kelsey to treat or prevent bleeding episodes.

CONCLUSION

Kelsey was referred to a pediatric gynecologist familiar with VWD. Menorrhagia is often treated with oral estrogens such as birth control pills. Kelsey and her mother decided to try Stimate first and it was effective. Kelsey treats with Stimate on the second and fifth day of her menstrual cycle, the bleeding has decreased and her period generally lasts six days.

Patients with VWD should avoid the use of aspirin and limit nonsteroidal anti-medications NSAIDS such as ibuprofen, Aleve and Motrin. Patients should inform their health care providers of their diagnosis. Special precautions may be needed before any invasive or surgical procedures. Patients are advised to contact the hemophilia nurses prior to these procedures so proper management and treatment can be coordinated.

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FEATURED NURSE SPECIALIST

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THE WEST CENTRAL OHIO HEMOPHILIA CENTER AT DAYTON CHILDREN'S

The West Central Ohio Hemophilia Treatment Center (HTC) is located inside the hematology/oncology department at The Children's Medical Center of Dayton. HTC provides comprehensive coordinated care for over 550 patients with bleeding disorders. The multidisciplinary staff consists of a pediatric hematologist, nurse coordinator, resource nurse and social worker. Also available are a physical therapist, nutritionist, genetic counselor and other pediatric and adult specialists. Hemophilia follow-up care is also available at Dayton Children's Specialty Care Center – Warren County in Middletown. For more information, call 937-641-5877 or 937-641-3207.

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