von Willebrand Disease (VWD): Disease Overview

General Information

Von Willebrand Disease is a genetic bleeding disorder caused either by a defect in or lack of von Willebrand clotting factor. Von Willebrand factor is the clotting factor that interacts with platelets immediately after an injury to form the initial plug which prevents blood from flowing freely at the site of injury. People who have von Willebrand Disease are unable to effectively form this initial plug.

Von Willebrand Disease can be inherited from either parent and affects both males and females equally. A parent with VWD has a 50% chance of passing the gene on to each of his or her children. VWD is estimated to occur in 1 to 2 percent of the population.

Four Subtypes of VWD

• Type I – This is the most common type of VWD, in which levels of von Willibrand clotting factor are reduced. There may also be a reduction in the normal levels of factor VIII. This is also typically the mildest form of VWD.
• Type II – In this type of VWD, there is an abnormality in the von Willibrand clotting factor. There are further sub classifications of this type based on the abnormality.
  - Type IIa – the level of von Willebrand factor is reduced, as is the ability of platelets to clump together.
  - Type IIb – although the von Willebrand factor is defective, the ability of the platelets to clump together is actually increased.
• Type III – Patients with this type have no von Willebrand clotting factor and often have factor VIII levels less than 10%. This is severe von Willebrand Disease.
• Pseudo (Platelet Type) – This type resembles Type IIb, only the defects are in the platelets rather than the von Willebrand factor.

Symptoms

- Increased or excessive bruising
- Recurrent nose bleeds
- Heavy menstrual bleeding
- Heavier than normal bleeding after any type of surgery

Diagnosis and Treatment

Diagnosis of VWD can be difficult, as the tests used to detect levels and function of the von Willebrand factor protein can be misleading and often must be repeated multiple times. It is recommended that if a person is suspected of having VWD, that they be referred to a hematologist who specializes in the diagnosis and treatment of bleeding disorders for evaluation.

Treatment of VWD is dependent upon the severity and type of bleeding.

Stimate®, or desmopressin acetate (DDAVP), is usually the treatment of choice for mild VWD. This drug, a nasal spray, works by boosting the patient's own levels of von Willebrand factor and factor VIII. Thus, it can be given to stop bleeding or to prevent bleeding for mild procedures such as a tooth extraction.

Excessive bleeding may require the use of factor replacement products containing both von Willibrand factor and factor VIII. These products directly replace missing or defective clotting factors. Products that have FDA approval for the treatment of VWD include: Humate P, Alphanate, and Wilate. Koate DVI is another product containing both factors and may be used, but it has not gained FDA approval specifically for the treatment of VWD.

** For further information or questions, please feel free to contact Family Factor at 251-633-8090 or Toll Free at 1-877-611-0004.*

REFERENCES

1. National Hemophilia Foundation, "von Willebrand disease". National Hemophilia Foundation. 06/02/2011 http://www.hemophilia.org/NHFWeb/MainPgs/MainNHF.aspx?menuid=182&contentid=47&rptname=bleeding.