Hemophilia of North Carolina

Learning About Bleeding Disorders

Bleeding disorder is the general term used to describe a range of rare, hereditary conditions, which can lead to poor blood clotting and uncontrolled bleeding.

What is Hemophilia?
Hemophilia is the most well-known of the bleeding disorders. It occurs when the clotting factor (usually factor VIII or factor IX) is missing or doesn't clot properly. There are three different severities of hemophilia: mild, moderate, and severe.

Bleeding caused by minor cuts and scrapes is usually not as big a problem for a person with hemophilia. The more serious threat is internal bleeding, which can develop spontaneously or as a result of routine medical and dental procedures, trauma and excessive physical activities. Bleeding into major joints and muscles is very common. Repeated episodes can lead to long-term disabilities such as crippling arthritis or, if left untreated, even death.

There is no known cure for hemophilia. With proper treatment, individuals can lead healthy, normal lives.

Who Gets Hemophilia?
The Centers for Disease Control (CDC) estimates that roughly 20,000 individuals in the United States have hemophilia. In about a third of the cases, there is no known family history of hemophilia. There are an estimated 1,000 families affected by hemophilia living in North Carolina. In addition, von Willebrand disease affects 1-2% of the U.S. population, or 80,000 people in North Carolina alone.

How is Hemophilia Treated?
The standard treatment for hemophilia is infusions of plasma (created from human blood) or recombinant products (created in a lab) to replace the missing clotting factor proteins in the blood. Most individuals are on some kind of home infusion regimen; others go to a hemophilia treatment center or a hospital for treatment. On average, the clotting factor products used to treat hemophilia cost between $60,000-$300,000 annually. This can be over a million dollars a year if there are complications like inhibitors to the medicine.

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What is von Willebrand Disease?
Von Willebrand Disease (VWD) is the most common bleeding disorder, affecting about one in every 100 persons. The condition occurs when the von Willebrand factor, a protein that works in unison with factor VIII, is missing or defective. Symptoms often include frequent nosebleeds, easy bruising and excessive bleeding, particularly following surgery. Like hemophilia, there are three different severities: mild, moderate, and severe.

Who Gets VWD?
VWD is thought to affect between 1-2% of the U.S. population. VWD affects women and men equally. In women, VWD may also cause heavy or prolonged menstrual cycles and excessive bleeding after childbirth. VWD often goes undiagnosed or incorrectly attributed to gynecologic conditions and, as a result, many women end up having unnecessary hysterectomies.

How is VWD treated?
The main treatment options for patients with VWD are desmopressin (DDAVP), recombinant von Willebrand factor (rVWF), and von Willebrand factor/factor VIII (vWF/FVIII) concentrates.

Rare Bleeding Disorders
In addition to hemophilia and VWD, there are other bleeding disorders affecting a number of North Carolinians. Examples include platelet deficiencies and deficiencies in factors I, II, V, VII, X, XI, XII or XIII. Most people are unfamiliar with these conditions because they are so rarely diagnosed. The treatment of choice in the United States for most of these conditions is cryoprecipitate and fresh frozen plasma. For individuals with factor VII, there is a recombinant product, which can be used to treat bleeding.