



## Comprehensive Care - Does source of care impact patient outcomes?

The hemophilia comprehensive care model was first established in the mid-1970s to improve care to individuals with hemophilia; the comprehensive care model aggregated expertise for hemophilia and provided multi-disciplinary services to address the many facets and broad based ramifications of these disorders including the medical, musculoskeletal, psychological, educational, vocational and financial needs of each patient. Individuals that receive care at a hemophilia comprehensive treatment center not only have access to the highest level of medical expertise for these rare disorders, but also to a team of dedicated, well trained professionals that understand the specific obstacles individuals with bleeding disorders face.

Currently in the United States, there are approximately 140 comprehensive hemophilia treatment centers (HTCs) that comprise the federal network. **The Indiana Hemophilia and Thrombosis Center (IHTC) is the only federally recognized comprehensive treatment center in Indiana.**

Each treatment center has a core team of professionals including a hematologist, nurse coordinator, physical therapist and social worker who spend the **majority** of their time caring for patients with bleeding disorders; in addition, treatment centers may include a variety of other professionals such as dental hygiene, dietetics, genetic counseling, research staff etc. The IHTC exceeds national staffing standards and offers an extensive spectrum of care through dedicated trained professionals. The IHTC team includes genetic counseling, dental hygiene, nutrition, career counseling, pharmacy services, physical therapy, orthopedic services, HIV and hepatitis testing and counseling, research protocols, and nursing services including phone consultation, travel coordination, infusion training, prophylaxis and immune tolerance programs, surgical coordination, and community and medical provider education. Programs and services at the IHTC are wide ranging — from school in-services and travel letters, to disease management for cost control and assurance of quality care.

In 1995, a landmark three year study involving hemophilia treatment centers in six states was initiated by the CDC. Patients

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## Bleeding Disorder IQ: Common Misperceptions

### Only men have hemophilia.

- Females may have factor XI deficiency or hemophilia C.
- Female carriers of factor VIII and IX deficiency (hemophilia A and B) may have levels that place them in the mild range of deficiency.
- Rarely, females may have moderate to severe hemophilia due to Lyonization imbalance, Turner syndrome or the co-inheritance of a carrier gene from mother and abnormal gene from a father with hemophilia.

### Patients with hemophilia have a family history of the disorder.

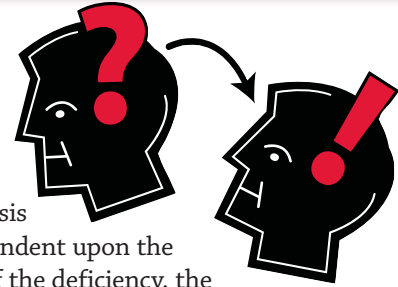
- Some families are not aware of the presence of hemophilia in their family due to rare bleeding episodes with mild or moderate deficiency or due to inaccurate carrier evaluation or communication.
- The gene causing hemophilia may be passed down for several generations in females before it presents clinically in an affected male.
- Spontaneous genetic mutations account for approximately 30% of cases.

### Individuals with hemophilia are diagnosed at birth; therefore you will not see undiagnosed patients after the first few years of life.

- Many patients with hemophilia are not diagnosed at birth. Only 50% of males with severe hemophilia will bleed in association with circumcision.
- Individuals with mild or moderate hemophilia often present later in life, and commonly not until they experience injury or undergo an invasive procedure or surgery.
- Infants who do not have a family history of hemophilia are not tested at birth and may not be diagnosed until after the neonatal period; the length of time

before

diagnosis is dependent upon the level of the deficiency, the hemostatic stress the individual has experienced, and the level of suspicion of the care provider.



### Hemophilia is a disorder that primarily affects Caucasians.

Hemophilia occurs equally in all races and ethnic groups; therefore incidence rates among Caucasians, African Americans and Hispanic males in the US are similar.

### Individuals with bleeding disorders bleed faster and may bleed to death from minor injuries.

- People who are affected with bleeding disorders do not bleed faster, but bleed longer due to an inability to form a stable fibrin clot.
- Minor bleeding is usually not life-threatening even to patients with hemophilia. Oftentimes, vessel vasoconstriction and platelet aggregation are enough to stop minor bleeding. Some notable exceptions include oral bleeding where estimation of the amount of blood loss may be difficult due to swallowing blood.

### Playing sports is not recommended for people with bleeding disorders.

- Regular activity is an important measure in decreasing the number of bleeding episodes. Regular exercise and development of good muscle tone assist individuals with the stabilization of joints and prevention of injury.
- Physical activity is very important in weight management; patients with bleeding disorders who are overweight have decreased joint range of motion

**Disorder IQ continued on back**

## JOIN US APRIL 30, 2008 CLOTTERS' CLUB

*It's not all in your genes:  
acquired hemophilia*

Westin Hotel • Indianapolis • 5:00 p.m.

*A nationally renowned guest lecturer, Dr. David Green, and physicians from the IHTC will present both scientific and clinically relevant information.*

**For more information, contact Sherry Potts, Program Coordinator, at 317-871-0000.**



*The Clotters' Club is an educational program designed for physicians and associated healthcare providers providing clinically current topics in hemostasis and thrombosis.*

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and an increased risk of morbidity from their bleeding disorder.

- Many sports once thought to be forbidden for patients with bleeding disorders are now encouraged. For a rating of sports activities please see *Playing it Safe: Bleeding Disorders, Sports and Exercise* (National Hemophilia Foundation). Sports outside of those rated as category #3 are now considered safe and appropriate for patients with bleeding disorders as long as appropriate counseling, protective measures and replacement therapy are included in the planning for participation.

### Comprehensive care at a federally recognized hemophilia treatment center is no more effective than care from other provider sources.

Treatment through a federally recognized HTC has been proven to confer an important decrease in morbidity and mortality for patients with bleeding disorders. The CDC documented a 40% decrease in mortality when patients were associated with their federally recognized hemophilia treatment center, compared to treatment provided through all other sources, including non-federally recognized medical facilities and centers. Treatment at a federally recognized center is also associated with decreased hospitalizations and complications, and increased independence and productivity.

### The aPTT is always prolonged in someone with hemophilia.

- The aPTT may not be prolonged in mild factor IX or VIII deficiency. In addition, the sensitivity of each laboratory in detecting moderate deficiencies may vary; therefore, the aPTT should not be used as a screening test in patients with a suspicious clinical history.
- The aPTT is relatively insensitive in patients with Von Willebrand disease; it is often normal in this disorder and again should not be used as a screening test.
- Rare bleeding disorders exist in which the PT and aPTT are expected to be normal; consultation with the IHTC is available to discuss diagnostic issues and problems.

### Intracranial hemorrhage in patients with hemophilia occurs after a head injury and is associated with a visible hematoma on the scalp or head.

Intracranial hemorrhage may occur without loss of consciousness and without development of a visible bruise or hematoma. A report of injury may not always be present. Symptoms of some

intracranial hemorrhages may evolve over time and take several days to emerge; therefore a report of temporally related head injury may not be present. Intracranial hemorrhage remains an important cause of morbidity and mortality in patients with bleeding disorders and therefore, a high degree of suspicion and aggressive approach to evaluation is important in detection of these bleeding episodes.

### Bleeding disorders are rare.

Von Willebrand Disease affects 1 to 2% of the population; VWD affects all racial groups and both sexes equally.

### Women are not affected with bleeding disorders.

- In addition to being a hemophilia carrier, women may be affected with other bleeding disorders, the most common of which is Von Willebrand Disease. VWD affects men and women equally. It is the most common bleeding disorder in the general population affecting 1-2 per 100 population. It is an under-diagnosed entity; when not identified, affected patients are at increased risk of suffering morbidity.
- Diagnosing a woman with VWD may be difficult; symptoms may be subtle, and laboratory analysis may be misleading. Referral to an HTC is often helpful for diagnosis and allows appropriate education and determination of the treatment modality specific to the sub-type of Von Willebrand Disease.
- In addition, many rare bleeding disorders exist, affecting females equally compared to males.

### Women who give birth without complication or abnormal bleeding can not have Von Willebrand Disease.

Both hormone levels during pregnancy and oral contraceptives increase the production of Von Willebrand factor 2 to 3 fold, thereby reducing the likelihood of abnormal bleeding. Some sub-types of VWD therefore have less bleeding symptoms during pregnancy, especially in later trimesters; however, affected individuals may experience increased bleeding after delivery when levels fall towards baseline. Delayed bleeding may occur in some individuals. Breastfeeding may help to maintain a higher baseline level of VWF; therefore, it is encouraged not only for the health of the infant, but for decreasing maternal risk of post-partum hemorrhage as well.

**For more information on bleeding disorders and treatment, please visit the IHTC website at [www.ihtc.org](http://www.ihtc.org).**

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with bleeding disorders were identified in each state through the treatment centers, state board of health and other sources; approximately 3,000 patients were followed over the course of the study to determine the effects of source of care, outcome and hospitalization rate. **An important finding that emerged from this study was that those patients who received care through an HTC had lower mortality rates compared to those who sought care from outside the federal treatment center network; in fact, mortality was 60% higher among those who received care outside of a federal center, which translated into a 40% reduction in the risk of death observed among persons who utilized an HTC [1].** This finding had additional significance given that more patients with severe disease and co-morbid conditions including inhibitors, hepatitis and HIV were cared for within the federal HTC network; the population that would be expected to have an increased risk of mortality in fact had less. This compelling study underscores the importance of referral of hemophilia patients to your federal hemophilia comprehensive treatment center in Indiana; the demonstrated effectiveness in management through a comprehensive center for this chronic disease emphasizes the importance for patients to be connected to their HTC.

In addition to lower mortality rates, patients receiving care from a federally recognized comprehensive center had fewer bleeding related hospitalizations and complications, and missed fewer days from work and school, translating into increased productivity. A higher degree of independence and an ability to experience a higher quality of life can be achieved through comprehensive care.

In the fall of 2004, the IHTC instituted a disease management program (DMP) for patients with bleeding disorders covered by Indiana's high-risk insurance health plan. The goal of the program

was to reduce the overall cost of care and improve health outcomes through a variety of mechanisms, including increased medical oversight and intervention [2]. The DMP brings together expertise, experience, and advanced technology in an outcome-oriented, patient-focused approach. The program is unique due to its focus on high-risk, high-cost patients and its strategy of proactive, preventive interventions to optimize patient outcomes while decreasing the need for high-cost ER visits, hospitalization, and surgery. In its 4th year of operation, the program continues to identify new opportunities for progress in the management of bleeding disorders; each element of the program seeks to attain the most accurate, effective and efficient method to diagnose and manage bleeding disorders.

Patients treated at an HTC have superior access to clinical studies, innovative treatments and the most current education and resources. The federal network of HTCs are the mainstay for ongoing education for patients and families affected by bleeding disorders. HTCs effectively coordinate home therapy and preventive services and work closely with hemophilia consumer organizations. Referring your patient to the IHTC impacts the quality of care they receive and their outcome.

Further information about hemophilia care and hemophilia treatment centers is available through the National Hemophilia Foundation (800-424-2634) or from Hemophilia of Indiana (317-396-0065). Visit the IHTC website for a more detailed look at the center, including the IHTC pharmacy, specialty clinics, detailed programs and services, and a media hub offering relevant newsletters, physician and staff publications.

*References: 1 Soucie JM, et al. Blood. 2000; 96(2): 437-442. 2 Tencer T, et al. Haemophilia 2007; 13: 480-488.*