



COMMENTARY

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## Fibrin Formation Challenges in Hemophilia and Clot Stabilization

Helge Leung\*

Department of Hematology, Xuzhou Medical University, Shuyang, China

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### About the Study

Hemophilia is a rare genetic disorder that affects the blood's ability to clot properly. This condition is characterized by a deficiency or dysfunction of certain clotting factors, leading to prolonged bleeding and difficulty in forming stable blood clots. There are two main types of hemophilia. They are hemophilia A, which is caused by a deficiency or dysfunction of clotting factor VIII, and hemophilia B, which is due to a deficiency or dysfunction of clotting factor IX. The pathophysiology of hemophilia involves a complex interplay of genetic, biochemical, and physiological factors. Hemophilia is an X-linked recessive disorder, meaning that the faulty gene responsible for the condition is located on the X chromosome. Since females have two X chromosomes (XX), they can be carriers of the hemophilia gene without displaying symptoms. However, males, who have only one X chromosome (XY), are more likely to manifest the disorder if they inherit the faulty gene from their mothers. The genes encoding clotting factors VIII and IX are located on the X chromosome, and mutations in these genes lead to the production of dysfunctional or insufficient clotting factors. As a result, individuals with hemophilia lack the ability to form stable blood clots in response to bleeding, making those susceptible to prolonged bleeding episodes.

The clotting process involves a series of steps known as the clotting cascade. This cascade is a tightly regulated sequence of events that culminate in the formation of a stable blood clot. Hemophilia disrupts this cascade, primarily affecting the intrinsic pathway, where factors VIII and IX play crucial roles. Vasoconstriction and platelets attaching to the injured blood artery are the first steps in the healing process after a bleeding injury. Hemophilia affects these mechanisms without affecting the succeeding steps in the clotting cascade.

The intrinsic pathway is activated when blood comes into contact with damaged tissue. Factor XII initiates the cascade, leading to the activation of factors XI, IX, and VIII in a sequential manner. In hemophilia, the deficiency or dysfunction of factor VIII (hemophilia A) or factor IX (hemophilia B) interrupts this pathway, preventing the formation of the tenase and prothrombinase complexes. Tenase, formed by factors VIIIa, IXa, and phospholipids, activates factor X. Subsequently, the prothrombinase complex, consisting of factors Xa, Va, calcium ions, and phospholipids, converts prothrombin into thrombin. Thrombin is a key enzyme that converts fibrinogen into fibrin, forming the meshwork of the blood clot. Fibrin strands weave through the platelet plug, creating a stable clot that prevents further bleeding. In hemophilia, the absence or dysfunction of factors VIII or IX impairs fibrin formation, leading to fragile and unstable clots that are easily disrupted.

The disruption of the clotting cascade in hemophilia results in a range of clinical manifestations. Individuals with hemophilia may experience spontaneous bleeding or prolonged bleeding following minor injuries or surgeries. Common sites of bleeding include joints, muscles, and internal organs.

Bleeding into joints, known as hemarthrosis, is a feature of hemophilia. This recurrent joint bleeding can lead to chronic joint damage, inflammation, and pain. Target joints, often affected in severe cases, may experience repeated bleeding episodes. Hematomas in muscles and soft tissues are common in individuals with hemophilia. These hematomas can cause swelling, pain, and limited range of motion in affected areas. In severe cases, internal bleeding can occur in vital organs such as the brain or gastrointestinal tract, leading to life-threatening complications. Bleeding into the central nervous system can result in neurological deficits and long-term disabilities.

Contact: Helge Leung, Email: HelgeL222@yahoo.com

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Diagnosing hemophilia involves assessing the patient's clinical history, family history, and laboratory tests. The levels of clotting factors VIII and IX are measured to confirm the diagnosis and determine the severity of the condition. Genetic testing may be conducted to identify specific mutations in the clotting factor genes. Management of hemophilia focuses on preventing and treating bleeding episodes. Replacement therapy with clotting factor concentrates is the primary treatment. Patients with hemophilia A receive factor VIII concentrates, while those with hemophilia B are treated with factor IX concentrates. These clotting factors can be given as needed to treat active bleeding or as a preventative measure to stop bleeding.