Hemophilia A: Pathophysiology and Treatment Strategies

Daniel Dinneen  
*Otterbein University*, daniel.dinneen@otterbein.edu

Follow this and additional works at: https://digitalcommons.otterbein.edu/stu_msn

Part of the Hemic and Lymphatic Diseases Commons, Medical Pathology Commons, and the Nursing Commons

**Recommended Citation**

Dinneen, Daniel, "Hemophilia A: Pathophysiology and Treatment Strategies" (2014). *Nursing Student Class Projects (Formerly MSN)*. 25.  
https://digitalcommons.otterbein.edu/stu_msn/25

This Project is brought to you for free and open access by the Student Research & Creative Work at Digital Commons @ Otterbein. It has been accepted for inclusion in Nursing Student Class Projects (Formerly MSN) by an authorized administrator of Digital Commons @ Otterbein. For more information, please contact jwu@otterbein.edu.
Introduction

Hemophilia is a sex-linked recessive coagulation disorder that varies widely in severity and age of diagnosis depending on the family history. The complications of hemophilia can be life-threatening and pathogenic in nature, so early identification and appropriate management are critical to minimize the risk of developing adverse outcomes. Hemophilia is a genetic disorder that primarily affects the male population and is due to the absence or deficiency of certain clotting factors. Hemophilia A is caused by a deficiency of factor VIII, while Hemophilia B results from a deficiency of factor IX. Both disorders are inherited in an autosomal recessive pattern, and the severity of the condition can vary greatly. Hemophilia is more common in the male population due to the sex-linked nature of its inheritance, with a prevalence of 1 in 5,000-10,000 male births in the United States. The management of hemophilia is focused on preventing or treating bleeding episodes, and the goal is to maintain adequate factor levels to prevent bleeding and maintain hemostasis.

Pathophysiology

Hemophilia A Pathophysiology

Common early manifestations are 
- Tranexamic acid (2011)
- 
- Research indicates that current treatment is not sufficient to 
- lifespan as hemostasis is a constant process. The treatment of hemophilia is necessary to provide adequate hemostasis and prevent bleeding episodes. Nurses treat a variety of hemophilia patients throughout their lifespan, and nurses play a significant role in the management of these patients.


diagnosed during childhood and is a lifelong condition. Pathology is usually associated with the portion of the gene affected; a functional factor VIII level 1% or less than 5% can be associated with severe or joint bleeding, respectively. Several bleeding episodes can occur as these proteins provide significant structural clot stability (Hall & Guyton, 2012). Factor VIII and IX are proteins manufactured in the liver that are necessary for the clotting cascade and coagulation (Stoelting, 2012). In the absence of factor VIII, the intrinsic pathway of the clotting cascade is impaired, leading to impaired coagulation and increased risk of bleeding.

- The intrinsic coagulation pathway is triggered by the activation of factor X and is essential in activating factor V, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Impaired activation of factor VIII results in impaired coagulation and increased risk of bleeding episodes.

- The completion of the intrinsic coagulation pathway of hemostasis; without intrinsic coagulation, the development of thrombin and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with hemophilia A treatment through early adulthood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- As is the case with many disease processes, severity of Hemophilia A can be varied and directly correlated to the severity of factor VIII deficiency (Srivastava, A., Brewer, A. K., Mauser-Bunschoten, E. P., et al., 2013). Table 3 illustrates the relationship of bleeding and factor level. As stated, factor VIII is critically important to the completion of the intrinsic coagulation pathway of hemostasis; without intrinsic activation, the sole means of adequate coagulation is dependent upon activation of the extrinsic pathway, namely factor X (Hall & Guyton, 2011). These factors indicate that the primary goal of hemophilia treatment is early childhood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- Patients that develop factor VIII inhibitors are classified as either high inhibitors or antibodies (Kasper, 2004). The primary goal of hemophilia treatment is early childhood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- Hemophilia A is a genetic disorder that primarily affects the male population and is due to the absence or deficiency of certain clotting factors. Hemophilia A is caused by a deficiency of factor VIII, while Hemophilia B results from a deficiency of factor IX. Both disorders are inherited in an autosomal recessive pattern, and the severity of the condition can vary greatly. Hemophilia is more common in the male population due to the sex-linked nature of its inheritance, with a prevalence of 1 in 5,000-10,000 male births in the United States. The management of hemophilia is focused on preventing or treating bleeding episodes, and the goal is to maintain adequate factor levels to prevent bleeding and maintain hemostasis.

Pathophysiology cont.

Signs and Symptoms

- Common early manifestations are 
- Tranexamic acid (2011)
- 
- Research indicates that current treatment is not sufficient to 
- lifespan as hemostasis is a constant process. The treatment of hemophilia is necessary to provide adequate hemostasis and prevent bleeding episodes. Nurses treat a variety of hemophilia patients throughout their lifespan, and nurses play a significant role in the management of these patients.

- The intrinsic coagulation pathway is triggered by the activation of factor X and is essential in activating factor V, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Impaired activation of factor VIII results in impaired coagulation and increased risk of bleeding.

- The completion of the intrinsic coagulation pathway of hemostasis; without intrinsic coagulation, the development of thrombin and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with hemophilia A treatment through early adulthood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- As is the case with many disease processes, severity of Hemophilia A can be varied and directly correlated to the severity of factor VIII deficiency (Srivastava, A., Brewer, A. K., Mauser-Bunschoten, E. P., et al., 2013). Table 3 illustrates the relationship of bleeding and factor level. As stated, factor VIII is critically important to the completion of the intrinsic coagulation pathway of hemostasis; without intrinsic activation, the sole means of adequate coagulation is dependent upon activation of the extrinsic pathway, namely factor X (Hall & Guyton, 2011). These factors indicate that the primary goal of hemophilia treatment is early childhood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- Patients that develop factor VIII inhibitors are classified as either high inhibitors or antibodies (Kasper, 2004). The primary goal of hemophilia treatment is early childhood to maintain/preserve musculoskeletal function and prevent or minimize the risk of joint damage.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- Hemophilia A is a genetic disorder that primarily affects the male population and is due to the absence or deficiency of certain clotting factors. Hemophilia A is caused by a deficiency of factor VIII, while Hemophilia B results from a deficiency of factor IX. Both disorders are inherited in an autosomal recessive pattern, and the severity of the condition can vary greatly. Hemophilia is more common in the male population due to the sex-linked nature of its inheritance, with a prevalence of 1 in 5,000-10,000 male births in the United States. The management of hemophilia is focused on preventing or treating bleeding episodes, and the goal is to maintain adequate factor levels to prevent bleeding and maintain hemostasis.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- Hemophilia A is a genetic disorder that primarily affects the male population and is due to the absence or deficiency of certain clotting factors. Hemophilia A is caused by a deficiency of factor VIII, while Hemophilia B results from a deficiency of factor IX. Both disorders are inherited in an autosomal recessive pattern, and the severity of the condition can vary greatly. Hemophilia is more common in the male population due to the sex-linked nature of its inheritance, with a prevalence of 1 in 5,000-10,000 male births in the United States. The management of hemophilia is focused on preventing or treating bleeding episodes, and the goal is to maintain adequate factor levels to prevent bleeding and maintain hemostasis.

- The primary goal of treatment for patients with hemophilia A is to correct factor VIII deficiency, and one major role of the intrinsic pathway of the clotting cascade is to activate factor X, which in turn activates factor VII, leading to the synthesis of thrombin and fibrin. Inhibitors of coagulation and eventually fibrin, stable clot formation fails (Björnsen & Holme, 2012). The majority of patients present with severe bleeding episodes, with some patients requiring frequent bleeding episodes and frequent hospitalizations.

- Hemophilia A is a genetic disorder that primarily affects the male population and is due to the absence or deficiency of certain clotting factors. Hemophilia A is caused by a deficiency of factor VIII, while Hemophilia B results from a deficiency of factor IX. Both disorders are inherited in an autosomal recessive pattern, and the severity of the condition can vary greatly. Hemophilia is more common in the male population due to the sex-linked nature of its inheritance, with a prevalence of 1 in 5,000-10,000 male births in the United States. The management of hemophilia is focused on preventing or treating bleeding episodes, and the goal is to maintain adequate factor levels to prevent bleeding and maintain hemostasis.