ST. MICHAEL'S
UNITY HEALTH TORONTO

The Bott-Harrington Nursing Fellowship in Bleeding Disorders

LEARNING TOOL KIT

Developed by St. Michael's Hospital

PREFACE

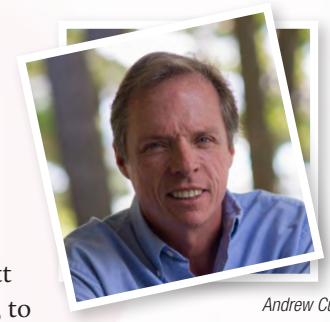
About The Bott-Harrington Nursing Fellowship in Bleeding Disorders

The Bott-Harrington Nursing Fellowship in Bleeding Disorders was created at St. Michael's Hospital with the goal of expanding the knowledge of bleeding disorder care beyond its Hemophilia Treatment Centre (HTC) to nurses working in other areas most likely to encounter patients with bleeding disorders, such as the emergency department, orthopedics and surgery.

Established in 2010, the six-month fellowship provides nurses with opportunities in the HTC under the mentorship of the Hemophilia Nurse Coordinator partnered with self-study learning modules. In return, the fellows are asked to share the knowledge and experience gained with colleagues in their clinical areas.

The fellowship was developed thanks to the incredible generosity and inspirational vision of Andrew and Hillary Cumming, supporters of St. Michael's.

Andrew's story is stirring. Born with severe hemophilia, Andrew has survived cancer, contracted Hepatitis C and HIV through contaminated blood products, and received a liver transplant. Through it all, he earned a PhD in physics, became a physics professor, and then launched a successful career in finance.



Andrew Cumming

Of his health battles, Andrew says he is one of the "lucky ones" thanks to the care he received at St. Michael's. He and people like the late Frank Bott and Denise Orieux, who lost two of their three sons, Gregory and Martin, to contaminated blood in the 1980s, are now proactive leaders in the bleeding disorders community, especially in promoting education, research and funding in hemophilia treatment.

For Andrew and wife Hillary, one of their great accomplishments is creating The Bott-Harrington Nursing Fellowship in Bleeding Disorders. The title invokes the Bott family name in honour of lives lost as well as those living today with bleeding disorders.

Ann Harrington

The name also pays tribute to the incredible nurses who care for so many dealing with hemophilia and its complications, as epitomized by Ann Harrington, the first nurse coordinator of the St. Michael's Hemophilia Treatment Centre, where both Andrew and the Botts' sons received treatment.

"Ann represents the best you can get in a hemophilia nurse. Her above-and-beyond knowledge, competence, dedication, accessibility and supportive care were a pillar of strength and hope to me and so many others," says Andrew.

"A bleeding disorder complicates almost every other illness a patient may have. It's my hope that this fellowship, and the knowledge-sharing it produces, creates condition and care awareness among nurses beyond hemophilia treatment clinics, especially for the new generation of bleeding disorder patients who are buoyed by excellent treatment outcomes."

The fellowship, created by Georgina Floros, Nurse Coordinator of the St. Michael's Hemophilia Program, has been modified to create the Bott-Harrington Nursing Fellowship in Bleeding Disorders Learning Tool Kit for nurses and other health care professionals caring for patients with bleeding disorders.

ACKNOWLEDGEMENTS

The fellowship team would like to thank the following people without whom this learning tool kit would not have been possible:

Financial support and original inspiration for this initiative:
Andrew and Hillary Cumming

Additional financial support:
David and Fiona Berry, The Budd Sugarman Foundation, David Young and the many friends who have generously supported this initiative

Dedicated to:
Ann Harrington, the first nurse coordinator of the St. Michael's Hospital Hemophilia Treatment Centre

The Bott family, who lost two wonderful young men to the contaminated blood crisis

Created by:
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St. Michael's Bott-Harrington Nursing Fellows who participated in the program and provided the insightful testimonials throughout this tool kit:
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The St. Michael's Hospital Foundation
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Jasmine Soliven, Interim Clinical Leader Manager, Specialty Clinics, CIBC Breast Center, Palliative Care, Occupational Health

With special thanks to the Canadian Hemophilia Society and the World Federation of Hemophilia for granting permission to include their information and resources within this tool kit.

MORE INFORMATION

If you require further information on this tool kit, how to introduce it into your organization, or have feedback, please contact the Hemophilia Treatment Centre office at St. Michael's Hospital, **416-864-5129**

EXCELLENCE IN NURSING CARE

Quality learning opportunities benefit both health care professionals and patients. The Bott-Harrington Nursing Fellowship in Bleeding Disorders exemplifies the six dimensions of quality here at St. Michael's: safety, outcomes, access, patient experience, equity and efficiency (SOAPEE). St. Michael's Hemophilia Treatment Centre, established in 1983, is a lifeline to thousands of patients with a bleeding disorder. Our ongoing quest to generate and share knowledge is a cornerstone of our nursing practice and patient care excellence. We are very pleased to be able to share this specialized knowledge with other nurses and health care professionals.

Dr. Robert Howard
President and Chief Executive Officer, St. Michael's

Improving the quality of patient care is the most important outcome of any learning experience at St. Michael's. The Bott-Harrington Nursing Fellowship in Bleeding Disorders is an excellent example of that. This incredible program afforded us the opportunity to educate our nurses in departments beyond the hospital's Hemophilia Treatment Centre. Further, this tool kit is supporting the broader health care community to do the same. Bleeding disorders entail highly specialized, comprehensive care; developing a knowledge base of the condition among the general nursing population will raise awareness of clinical symptoms in patients with hemophilia and further improve patient care outcomes.

Ella Ferris
Former Executive Vice-President, Programs, Chief Nursing Executive,
Chief Health Disciplines Executive, St. Michael's

Nurses make the difference in providing exceptional care. Andrew Cummings can attest to that. He helped establish The Bott-Harrington Nursing Fellowship in Bleeding Disorders in part to give back to St. Michael's and, in particular, to honour Ann Harrington, the first nurse coordinator of our Hemophilia Treatment Centre where Andy received care. This tool kit pays that generosity forward by giving nurses in other hospitals the chance to learn more about bleeding disorders and how to incorporate that knowledge into daily practice. Now that's inspired giving!

L. Alayne Metrick
President, St. Michael's Foundation

Comprehensive multi-disciplinary care delivered through dedicated Hemophilia Treatment Centres is well established as the optimal model for managing hemophilia and rare inherited bleeding disorders, allowing our patients to live full and productive lives. Nurses with specialized expertise and skills are important members of the comprehensive care team. Given the complexity and the rarity of these diseases, most health care providers are not well equipped to provide care for these patients outside of the specialty clinic setting. This learning tool kit, based on the The Bott-Harrington Nursing Fellowship's study program, is a creative way to help disseminate and share some of this expert knowledge with other nursing professionals.

Dr. Jerry Teitel
Medical Director, Specialized Complex Care,
Division of Hematology and Oncology

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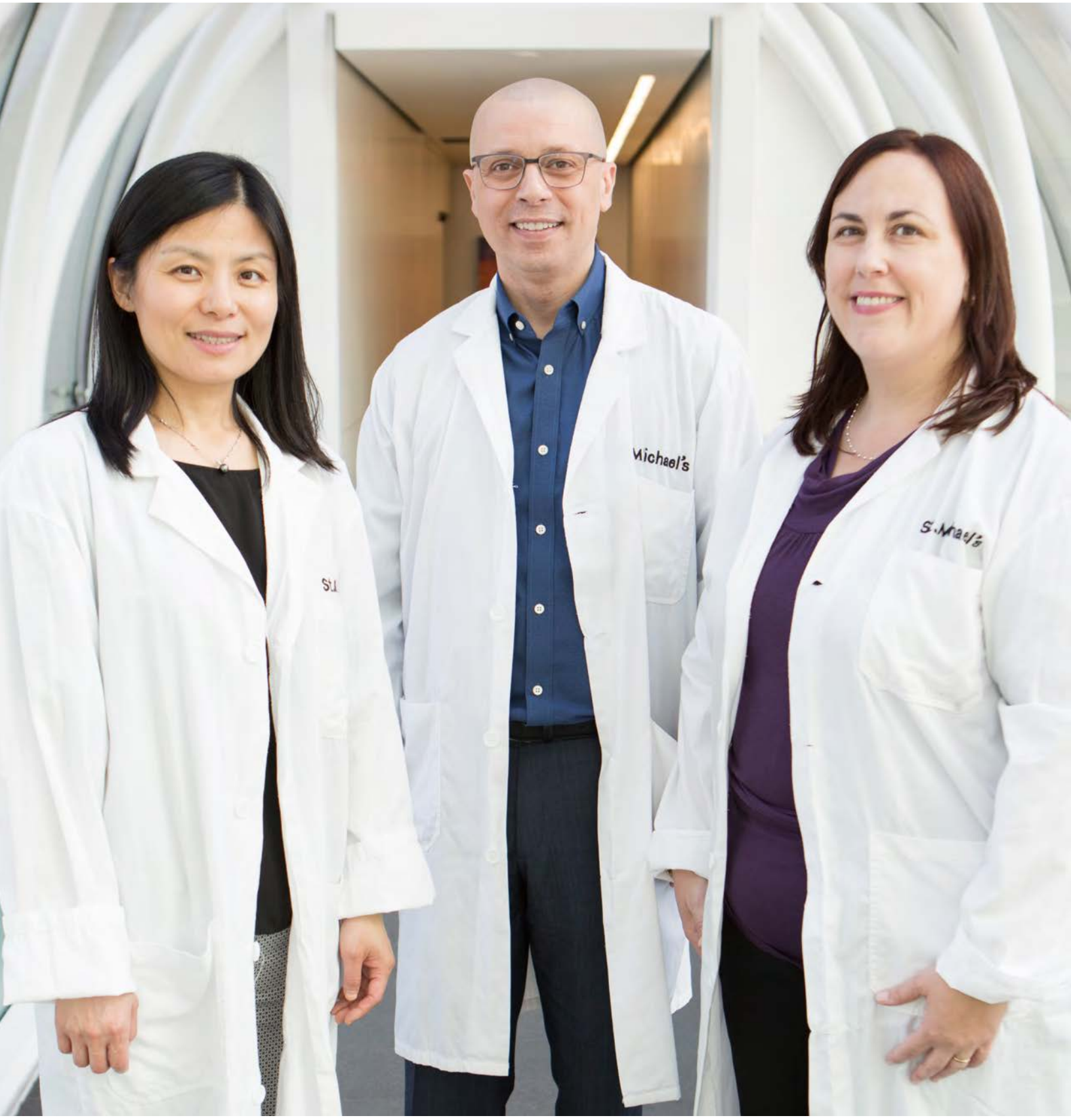
> *I undertook the fellowship because patients with bleeding disorders would present in the Emergency Department. I wanted to learn more so we had the knowledge to better expedite their care and improve the patient experience.*



USING THIS LEARNING TOOL KIT

Content for this tool kit has been adapted from the Bott-Harrington Nursing Fellowship in Bleeding Disorders at St. Michael's Hospital with the purpose of sharing its learning resources with health care organizations looking to enhance the knowledge of bleeding disorders among nursing staff.

Not only does the information serve as a comprehensive primer on hemophilia and other bleeding disorders, but it also provides valuable care knowledge that can be incorporated into daily practice.

PRIMARY TARGET AUDIENCES ARE:

- Nurses who may encounter a patient with a bleeding disorder
- Nurses new to bleeding disorders care working in a Hemophilia Treatment Centre
- Other health care professionals looking to explore the unique treatment of bleeding disorders

THE TOOL KIT'S OBJECTIVES:

- Educate nurses and health care professionals about bleeding disorders
- Bridge the gap that currently exists with respect to knowledge and comprehension of bleeding disorders
- Provide the learner with insights into the lived experience of patients with bleeding disorders
- Enhance nursing comfort caring for patients with bleeding disorders which in turn improves the patient experience

> *For RNs who don't see bleeding disorder patients on a regular basis, it's the fear of the unknown when it comes to understanding and treating them. Learning about the condition helps build knowledge and confidence in providing the required care.*

Designed as a handy digital self-study guide, the learning content for each of its 10 modules is just a click away with online links to a vast library of bleeding disorder literature, videos and websites.

IMPORTANT NOTE

During the development of this tool kit, we made every effort to confirm that all references, resources and links included in its contents would be available online at the time of publication. However, it is beyond the publisher's control if that information availability changes in the future. This tool kit should be used only as reference material and nothing herein shall be construed as providing medical or expert advice. The education of all nurses and health care professionals is a shared responsibility between individual professionals and their respective health care organizations.

THE MODULES ARE BUILT AROUND FOUR CORE AREAS OF LEARNING:

(1) Understanding the role of comprehensive care in the treatment of bleeding disorders
(2) Learning the biology, pathophysiology and clinical manifestations of bleeding disorders
(3) Understanding the lived experience of people with bleeding disorders
(4) Taking newly acquired knowledge and applying it to clinical practice to improve care

While sequenced so each module builds on the previous ones, it is not necessary to approach them in that order. Each is structured with:

OBJECTIVES	Learning goals for the module
MODULE CONTENT	Identified resources to foster targeted learning
ADDITIONAL RESOURCES	Optional reading to expand understanding
KEY LEARNING	Outline of essential knowledge takeaways
TEST YOUR KNOWLEDGE	A multiple-choice quiz
REFLECTION	An opportunity to improve critical thinking, promot greater self-awareness and enhance personal and professional growth

Helpful facts, eye-opening statistics, interesting trivia and learning tips, along with comments and insights gleaned from the St. Michael's fellows who have completed the program, are highlighted throughout the tool kit.

Last, but not least, all those who have had a hand in envisioning, supporting, developing and contributing to this tool kit, ask that it become a true knowledge-sharing conduit for continuing to elevate the understanding and care of bleeding disorders throughout the nursing community.

SYMBOLOGY KEY

Did You Know 

Quick Fact 

By The Numbers 

Education In Action 

Learning Tip 

> *The learning modules are very comprehensive – read them!*
>
> *Then look on your unit for opportunities to put the information in action.*

ABBREVIATIONS KEY

Because bleeding disorders is a specialty care area, and for the information of our readers, here's a quick primer on the key medical and organizational abbreviations included in this tool kit's materials:

AHCDC – Association of Hemophilia Clinical Directors of Canada
CANHC – Canadian Association of Nurses in Hemophilia Care
CBDR – Canadian Bleeding Disorders Registry
CDC – Centres for Disease Control and Prevention
CHARMS – Canadian Hemophilia Assessment and Resource Management System
CHR – Canadian Hemophilia Registry
CHS – Canadian Hemophilia Society
FEIBA – Factor VIII inhibitor bypassing activity
GI/GU -- Gastrointestinal/Genitourinary
HCV – Hepatitis C Virus
HIP – Home Infusion Program
HIV – Human Immunodeficiency Virus
HTC – Hemophilia Treatment Centre
ITI – Immune Tolerance Induction
MSK – Musculoskeletal system
PTT – Partial thromboplastin time
VWD/VWF – von Willebrand disease /von Willebrand factor
WFH – World Federation of Hemophilia

MODULE 1

Comprehensive Care in Canada

This module examines the care history of bleeding disorders and how comprehensive care is currently provided in Canada.

The word hemophilia is rooted in two Greek words: haima, meaning blood, and phila, meaning affection or tendency toward.

Objectives

- Identify and describe the role of key team members of a comprehensive care program.
- Understand the integral role of the nurse coordinator within the comprehensive care team.
- Expand your understanding of how hemophilia care is provided in Canada.
- Increase your understanding of the challenges faced by people living with a bleeding disorder and apply this knowledge to develop strong therapeutic nurse-patient relationships.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

One of the roles of a nurse coordinator at a Hemophilia Treatment Centre is to teach people about bleeding disorders. This includes providing resources and education to others including family members, teachers, babysitters, workplace staff and local nurses.

> *I had no idea the amount of collaboration and range of healthcare professionals that make up the comprehensive care team in St. Michael's Hemophilia Treatment Centre.*

MODULE CONTENT

Please review the following:

Read:

All About Hemophilia: A Guide for Families
Chapter 1: An Introduction to Hemophilia ***(2010)***

All About Hemophilia: A Guide for Families
Chapter 3: Comprehensive Care for Hemophilia ***(2010)***

Canadian Association of Nurses in Hemophilia Care (CANHC) job description (2006)

In 1952, hemophilia was recognized to have two distinct disorders:

Hemophilia A
(factor VIII protein deficiency)

and Hemophilia B
(factor IX protein deficiency)

ADDITIONAL RESOURCES

Read:

Canadian Comprehensive Care Standards for Hemophilia and Other Inherited Bleeding Disorder (2007)

Science: Case Closed: Famous Royals Suffered From Hemophilia (2009)

Mortality among males with hemophilia: relations with source of medical care (2000)

Optimal haemophilia care versus the reality (2005)

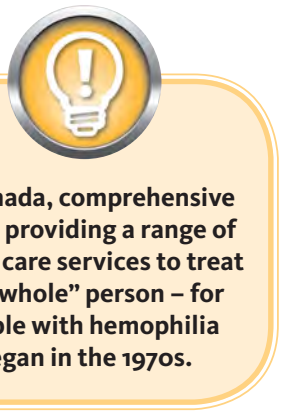
Contact:

Liaise with the Hemophilia Provincial Coordinator in Ontario

416.864.6060 EXT. 46981

- There are 26 Hemophilia Treatment Centres across Canada.
- In Toronto, there are two centres: St. Michael's Hospital and SickKids.

KEY LEARNING



In Canada, comprehensive care – providing a range of health care services to treat the "whole" person – for people with hemophilia began in the 1970s.

- Medical care for patients with bleeding disorders is specialized and should be provided by health care workers who have expert knowledge. This care is best met through Hemophilia Treatment Centres (HTC) rather than individual medical providers.
- An HTC core team should include: a medical director, nurse coordinator, physiotherapist, social worker, and an administrative assistant/data manager.
- Patients develop unique knowledge and experience of their bleeding disorder and thus they should be included as a valuable member of the care team.
- The development of a multi-disciplinary and comprehensive care model greatly improves the delivery of care and the accountability for factor concentrate use.
- Discovery of cryoprecipitate in the 1960s revolutionized care for patients with hemophilia and subsequent development of specific factor concentrates has further optimized care management.

Take each module's reflection assignment to work with you. It's amazing how applicable the knowledge becomes when faced with a hands-on situation.

> *A key learning was that patients with bleeding disorders, especially hemophilia, are extremely well informed about their condition. As a health care provider, you need to be too, not only to better understand the impact of a bleed on their health, life and job, but to also gain their confidence and respect.*

Canada

- 1 in 100 Canadians (300,000 people) carry an inherited bleeding disorder gene
- About 30,000 Canadians have symptoms severe enough to seek medical attention; yet many go undiagnosed
- Hemophilia A and B affect about 3,000 Canadians

Source: Canadian Hemophilia Society

Worldwide

- Over 287,000 people identified with a bleeding disorder
- Over 172,000 are diagnosed with Hemophilia A or B

Source: World Federation of Hemophilia, Annual Global Survey Data 2014

REFLECTION

After completing this module, reflect upon your newly acquired knowledge of comprehensive care and the role of the nurse coordinator.

Describe how this understanding of care delivery for bleeding disorders might influence your ongoing nursing practice.

MODULE 2

Coagulation and Genetics of Bleeding Disorders

The purpose of this module is to acquire an understanding of how blood clots, the cause of hemophilia and von Willebrand disease (VWD), and the emerging understanding of rare bleeding disorders followed by a Hemophilia Treatment Centre.

Objectives

- Identify the process of primary and secondary hemostasis.
- Compare the classifications of mild, moderate and severe hemophilia.
- Review the 3 main types of von Willebrand disease.
- Describe the inheritance pattern for hemophilia and von Willebrand disease.
- Review laboratory testing associated with diagnosing and managing various bleeding disorders.
- Review module content and demonstrate knowledge acquisition by completing the module quiz.

People with a bleeding disorder do not bleed any more profusely or faster than other people. However, they do bleed longer because their blood lacks a protein needed for normal clotting.

- Although hemophilia is an inherited bleeding disorder, 1 in 3 cases have no known family history of hemophilia.

> *The fellowship's modules and opportunity to work in the hospital's Hemophilia Treatment Centre were eye openers. They taught me to look for clues when a small bleed turns into a certified event.*

MODULE CONTENT

Please review the following. An * denotes previously referenced sources:

Read:

All About Hemophilia: A Guide for Families
Chapter 1: An Introduction to Hemophilia ***(2010)***

All About Hemophilia: A Guide for Families
Chapter 2: How a Child Gets Hemophilia ***(2010)***

All About von Willebrand Disease ...
for people with von Willebrand disease and their families ***(2011)***

Guideline for the diagnosis of von Willebrand disease (2012)

Laboratory Testing Reference List - London Health Sciences Centre

Watch:

The Coagulation Cascade:
A computer-generated movie on hemophilia

Hemophilia is also known as the "Royal Disease." England's Queen Victoria was a carrier of hemophilia. Daughters Alice and Beatrice were carriers, too, passing on the genetic disorder through marriage into the German, Spanish and Russian Royal Families. Her son Leopold had hemophilia, dying at 31 from a massive bleed after falling.

This is an opportunity to delve deeper into a topic that you only learned the basics about in nursing school.

ADDITIONAL RESOURCES

Read:

Current understanding of hemostasis (2011)

What every nurse should know about hemophilia (2013)

Disorders of Platelet Function (2002)

von Willebrand Disease: advances in pathogenetic understanding, diagnosis, and therapy (2013)

Factor I Deficiency (2004)

Factor II Deficiency (2006)

Factor V Deficiency (2006)

Factor VII Deficiency (2014)

Factor X Deficiency (2006)

Factor XI Deficiency (2007)

Factor XII Deficiency (2004)

Factor XIII Deficiency (2014)

Bernard-Soulier Syndrome (2008)

Glanzmann Thrombasthenia (2014)

Daughters whose fathers have hemophilia are obligate carriers of the genetic mutation that causes it. Daughters whose mothers are carriers of hemophilia are known as potential carriers and have a 50% chance of inheriting the gene mutation.

Watch:

Platelet Adhesion and Aggregation (2010)

Visit:

DNA Learning Centre: Hemophilia

> *There are more people out there with undiagnosed bleeding disorders than recognized. Learning more about the condition helps you take that into account when doing an assessment.*

KEY LEARNING

Christmas Disease, the original name for factor IX deficiency, was named after Stephen Christmas, a Torontonian. He was the first person in the world to be diagnosed with this distinct form of hemophilia.

- Primary hemostasis involves platelets and von Willebrand factor. Secondary hemostasis involves coagulation factors. Both primary and secondary hemostasis are required to create a stable clot.
- A defect in primary or secondary hemostasis will result in prolonged and excessive bleeding.
- Hemophilia is an X-linked genetic disorder, affecting predominately males.
- VWD is an autosomal dominant (recessive in type 3 and 2N) genetic disorder affecting men and women equally, but more women show symptoms.
- In mild hemophilia there is 6% to 30% of normal clotting factor activity; in moderate hemophilia there is 1% to 5% of normal clotting factor activity; in severe hemophilia there is <1% of normal clotting factor activity.
- There are three main categories of von Willebrand disease:
    - Type 1: Symptoms are usually very mild with lower than normal levels of von Willebrand factor
    - Type 2: Symptoms are moderate due to the von Willebrand factor (VWF) protein not working properly, causing lower than normal VWF activity
    - Type 3: Symptoms are more severe due to very little or no von Willebrand factor and low factor VIII levels

> *Even though there's been no formal diagnosis, you can see how family histories are very important in ascertaining the possibility of hemophilia or other bleeding disorder.*

- Von Willebrand disease is the most common type of bleeding disorder affecting both men and women, with 1% of the world's population having low VWF levels but most don't satisfy diagnostic criteria for VWD.

TEST YOUR KNOWLEDGE

The answers appear on page 52 of this tool kit.

1. **Hemophilia is:**
   a. Characterized by a low or absent factor VIII (8) or factor IX (9) level
   b. Contagious
   c. Inherited
   d. Characterized by rapid blood loss
   e. a and c

2. **Hemophilia A is also referred to as:**
   a. Classical hemophilia
   b. Factor VIII deficiency
   c. Christmas Disease
   d. a and b
   e. All of the above

3. **The severity of hemophilia changes from one generation to another:**
   a. True
   b. False

4. **Hemophilia is carried on the "X" chromosome:**
   a. True
   b. False

5. **Boys born to men with hemophilia will pass the condition on to future generations:**
   a. True
   b. False

6. **Von Willebrand disease**
   (choose all correct answers):
   a. Can affect both men and women
   b. Has 3 main classifications
   c. Has an X-linked inheritance pattern
   d. Is the most common bleeding disorder

7. **VWD levels rise in the blood in response to:**
   a. Hormone changes
   (eg. Pregnancy, menstrual cycle, oral contraception, etc)
   b. Stress
   (eg. Surgery, infection, emotions, etc)
   c. Exercise
   d. All of the above

Von Willebrand disease is named after Finnish physician Erik von Willebrand who, in 1925, first noted the condition to be different from hemophilia, which mainly affected males.

This learning has enabled me to troubleshoot some bleeding situations right away.

I've also become an initial go-to resource on our floor in addition to the support from the Hemophilia Treatment Centre team.

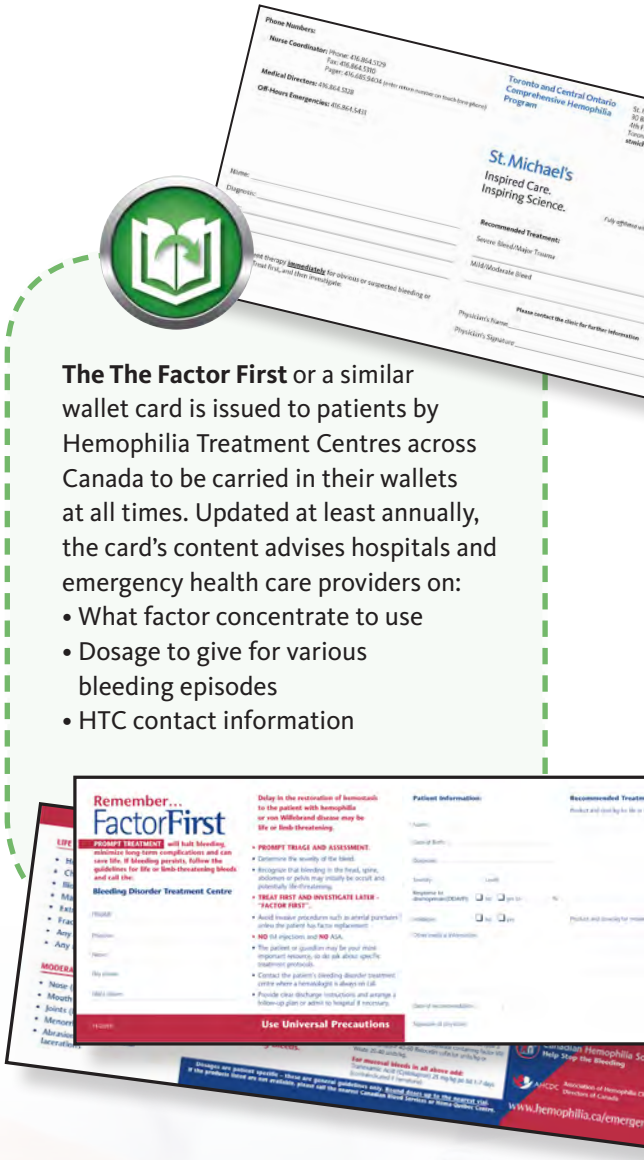
MODULE 3

Clinical Manifestations and Bleed Assessment

The goal of this section is to gain a greater understanding of how various bleeding disorders manifest clinically and apply this knowledge to bleeding assessments.

Objectives

- Review the signs and symptoms of the following bleeds:
  - Intracranial hemorrhage
  - Hemarthrosis (joint bleed)
  - Muscle
  - Soft tissue
  - Mucocutaneous
  - Gastrointestinal/Genitourinary
- Develop an understanding of how to triage and assess a bleed safely in a variety of clinical settings:
  - Telephone triage
  - Clinical setting assessment
- Understand the consequences of hemarthrosis and the importance of bleed prevention.
- Review module content and demonstrate knowledge acquisition by completing the module quiz.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.



The The Factor First or a similar wallet card is issued to patients by Hemophilia Treatment Centres across Canada to be carried in their wallets at all times. Updated at least annually, the card's content advises hospitals and emergency health care providers on:

- What factor concentrate to use
- Dosage to give for various bleeding episodes
- HTC contact information

> *Learning more about bleeding disorders has allowed me to introduce that knowledge into our Emergency Room assessments.*

MODULE CONTENT

Please review the following:

Read:

All About Hemophilia: A Guide for Families
Chapter 4: Management of Bleeds ***(2010)***

Telephone Guidelines (2013)
Journal of Haemophilia Practice:
Decision-support tool for telephone triage – the Canadian Experience

Identifying Common Joint & Muscle Bleeds (2014)

Watch:

Video: Joint Bleeds and Complications (2009)

ADDITIONAL RESOURCES

Read:

WFH Treatment Guidelines 3rd Edition

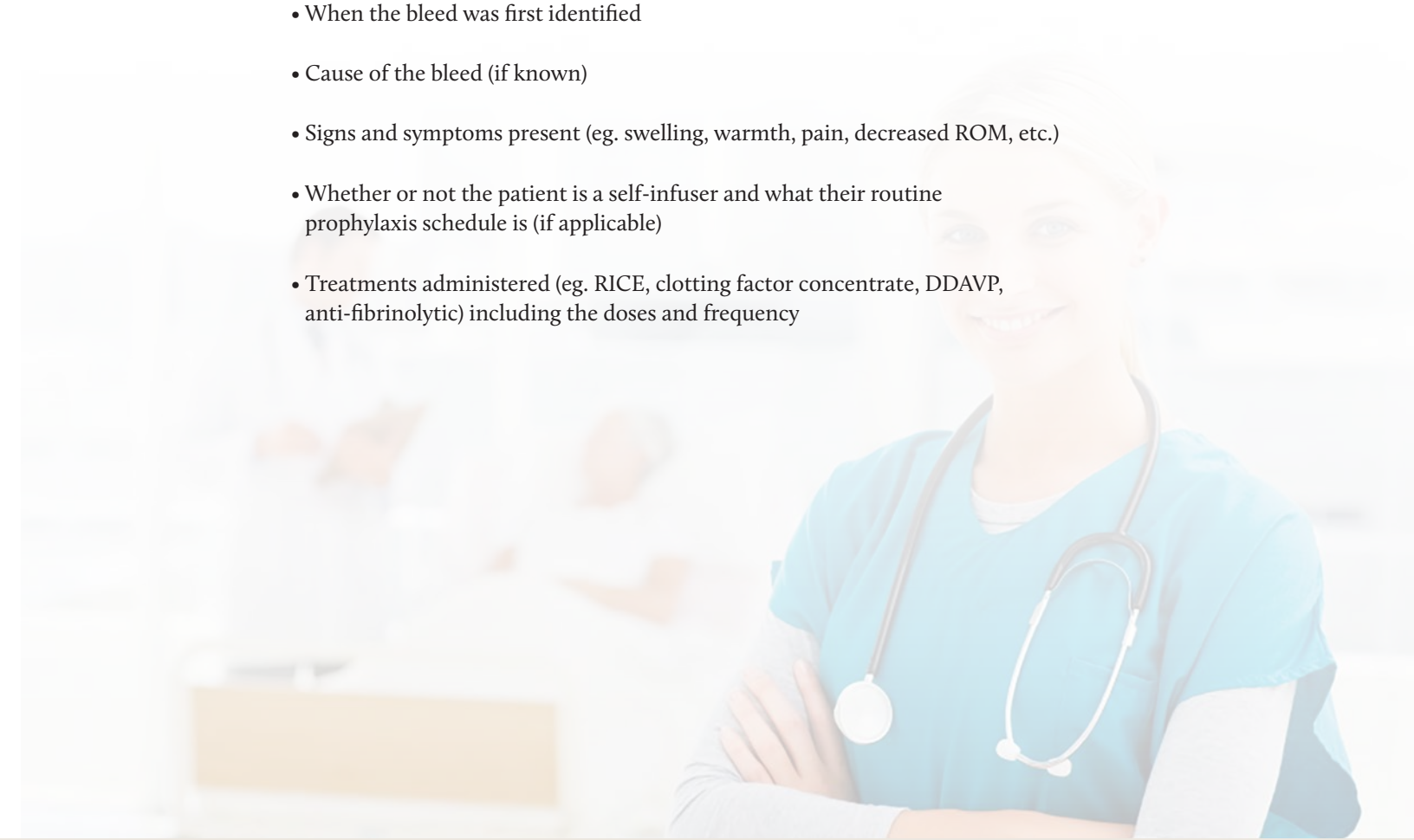
Watch:

Joint Bleeding in Haemophilia (2009)

> *My hematology department's doctors also work in the Hemophilia Treatment Centre.*
>
> *Not only did the program help me better understand bleeding disorders, but it was amazing*
>
> *to see how the centre's comprehensive care team works together.*

KEY LEARNING

- Bleeding into the head, neck, chest and abdomen are potentially life threatening and must be treated promptly.
- Muscle bleeds, such as iliopsoas, forearm and calf, are particularly concerning sites due to the threat of compartment syndrome.
- Early signs of joint bleeding include a "bubbling and tingling" feeling in the joint. Later signs include warmth, swelling, pain and decreased range of motion.
- An effective nursing assessment of an acute bleed is vital to providing appropriate treatment. Assessments and history taking should include questions such as:
    - Type and severity of bleeding disorder
    - When the bleed was first identified
    - Cause of the bleed (if known)
    - Signs and symptoms present (eg. swelling, warmth, pain, decreased ROM, etc.)
    - Whether or not the patient is a self-infuser and what their routine prophylaxis schedule is (if applicable)
    - Treatments administered (eg. RICE, clotting factor concentrate, DDAVP, anti-fibrinolytic) including the doses and frequency



REFLECTION | Reflect on the challenges of providing a clinical assessment over the telephone and how the manifestations of a bleed can impact a patient's day-to-day life.

TEST YOUR KNOWLEDGE

The answers appear on page 52 of this tool kit.

1. **Signs of a soft tissue bleed may include:**
   a. Bruising
   b. Pain
   c. Warmth
   d. Swelling
   e. All of the above

2. **Signs of a muscle bleed may include:**
   a. Pain
   b. Warmth and swelling
   c. Restricted movement
   d. All of the above

3. **Signs of a joint bleed include:**
   a. Pain
   b. Loss of movement
   c. Swelling and warmth
   d. Difficulty weight bearing
   e. All of the above

4. **An iliopsoas bleed** (choose all that apply):
   a. Is a large muscle bleed
   b. Is a limb threatening bleed
   c. Is located in the calf
   d. Requires immediate and ongoing treatment with clotting factor concentrates

5. **Signs and symptoms of a bleed into the brain include:**
   a. Drowsiness and lethargy
   b. Vomiting
   c. Irritability
   d. Headache
   e. All of the above

6. **Possible life-threatening bleed sites include:**
   a. Head
   b. Neck
   c. Chest
   d. Abdomen
   e. All of the above

To fully explore this tool kit's learning, the larger experience would be to arrange for placement, volunteer or drop-in time at a local HTC in order to shadow the clinicians, meet the patients and bring the knowledge to life.

I will continue to do one day a month in the Hemophilia Treatment Centre for the hands-on practice that will keep my learning fresh.

MODULE 4

Coagulation Therapy and Management of Bleeding

The content of this module will help you develop an understanding of the guiding principles of bleed management.

Objectives

- Identify treatment options based on diagnosis, severity of disease and bleed characteristics, available to patients with bleeding disorders.
- Develop a plan of care in consultation with the physician based on your assessment.
- Develop an understanding of the mechanism of action and use of Desmopressin acetate, anti-fibrinolytics and clotting factor concentrates.
- Understand the process of ordering/prescribing, acquiring, storing and distributing products used for the management of a bleeding disorder.
- Demonstrate competence in the preparation and administration of coagulation therapy.
- Consider, if appropriate and available, the role of the various comprehensive care team members in the management of bleed episodes.
- Review module content and demonstrate knowledge acquisition by completing the module quiz.

Aspirin and other non-steroidal anti-inflammatories such as Advil and Ibuprofen can affect clotting. Consult the HTC before prescribing or administering these medications.

> *Other nurses now come to us (the fellows) with questions about patients they're afraid might bleed, or to help with the different factor products and how to push them. It's nice being a resource.*

MODULE CONTENT

Please review the following. An * denotes previously referenced sources:

Read:

All About Hemophilia: A Guide for Families
Chapter 4: Management of Bleeds ***(2010)***

All About Hemophilia: A Guide for Families
Chapter 5: Clotting Factor Therapy ***(2010)***

Clotting Factor Concentrates

Desmopressin: A Guide for Patients and their Caregivers (2015)

Cyklokapron: A Guide for Patients and their Caregivers

Guidelines for Emergency Management of Hemophilia and von Willebrand Disease - Factor First

To Do:

Review institution-specific transfusion medicine and pharmacy manuals

Contact:

Liaise with personnel at local transfusion medicine department and pharmacy

The World Federation of Hemophilia (WFH) was founded in 1963 by Montreal businessman Frank Schnabel, who was born with severe hemophilia A. Dedicated to improving, educating and sustaining care for people with inherited bleeding disorders globally, WFH, a network of patient organizations in 127 countries, was officially recognized by the World Health Organization (WHO) in 1969. Its head office remains in Montreal.

ADDITIONAL RESOURCES

Read:

***Guidelines for the Management of Hemophilia* (2012)**

***Emergency Care Issues in Hemophilia* (2007)**

All About Hemophilia: A Guide for Families
Chapter 6: The Role of Prophylaxis ***(2010)***

All About Hemophilia: A Guide for Families
Chapter 7: Home Infusion ***(2010)***

***Desmopressin (DDAVP) in Treatment of Bleeding Disorders* (2012)**

***Fibrinolytic Inhibitors in the Management of Bleeding Disorders* (November 2012, revised edition)**

In addition to clotting factor concentrate treatments when necessary, bleeding can be slowed, or swelling and pain reduced, with First Aid steps called R.I.C.E.:

Rest
Ice (once hemostatic agents administered)
Compression
Elevation

KEY LEARNING

- Early intervention at the first sign of bleeding can minimize or eliminate the complications associated with most bleeds.
- In patients with more severe forms of hemophilia, home self-infusion is the ideal treatment management strategy.
- The patient, nurse, hematologist and physiotherapist all play an important role in managing acute bleeding episodes.
- Communication between patient and an HTC is a crucial part of caring for this unique population.
- There are a variety of factor concentrates available in Canada both recombinant and plasma derived.
- All factor concentrates come from the Canadian Blood Services /HemaQuebec via a hospital blood transfusion department and are supplied from the manufacturer as a freeze-dried powder with diluent for reconstitution prior to administration.

> *One weekend, we had a patient whose factor levels were seriously fluctuating. The on-call HTC physician noted how valuable it was that, since I had the (fellowship) knowledge and training, I could push the factor she ordered right away. It really helped our patient get the care they needed, when they needed it.*

TEST YOUR KNOWLEDGE

The answers appear on page 52 of this tool kit.

1. **The amount of factor administered depends on** (circle all that apply):
   a. The weight of the person
   b. The reason for treatment (type of bleed, routine prophylaxis schedule and dosing, surgery)
   c. The baseline factor level of the patient
   d. Knowledge of previous response to therapy

2. **Treatment of a muscle bleed may include:**
   a. Clotting factor concentrate/ Desmopressin acetate (DDAVP)
   b. R.I.C.E.
   c. Crutches
   d. All of the above

3. **As a general rule, for every 1 unit/kg of factor VIII infused, factor VIII levels in the blood increase by:**
   a. 5%
   b. 1%
   c. 2%
   d. 10%

4. **As a general rule, for every 1 unit/kg of factor IX infused, factor IX levels in the blood increase by:**
   a. 5%
   b. 1%
   c. 2%
   d. 10%

5. **When a patient presents with hematuria, recommendations may include** (choose all that apply):
   a. Increasing fluid intake
   b. Administration of anti-fibrinolytics (eg. Cyklokapron)
   c. Clotting factor concentrate/Desmopressin acetate (DDAVP)
   d. Medical imaging to assess cause of hematuria

6. **When a patient presents with a nosebleed, recommendations may include** (choose all that apply):
   a. Blow your nose before applying pressure
   b. Bend your head slightly forward
   c. Pinch just below the bridge of your nose
   d. Give Cyklokapron
   e. Give clotting factor concentrate/ Desmopressin acetate (DDAVP)
   f. Refer to an ear, nose and throat specialist

7. **Other medications that are used to control bleeding are** (circle all that apply):
   a. Desmopressin acetate (DDAVP)
   b. Anti-fibrinolytics
   c. Topical thrombin
   d. Fibrin glue

MODULE 5

Home Infusion Program (HIP)

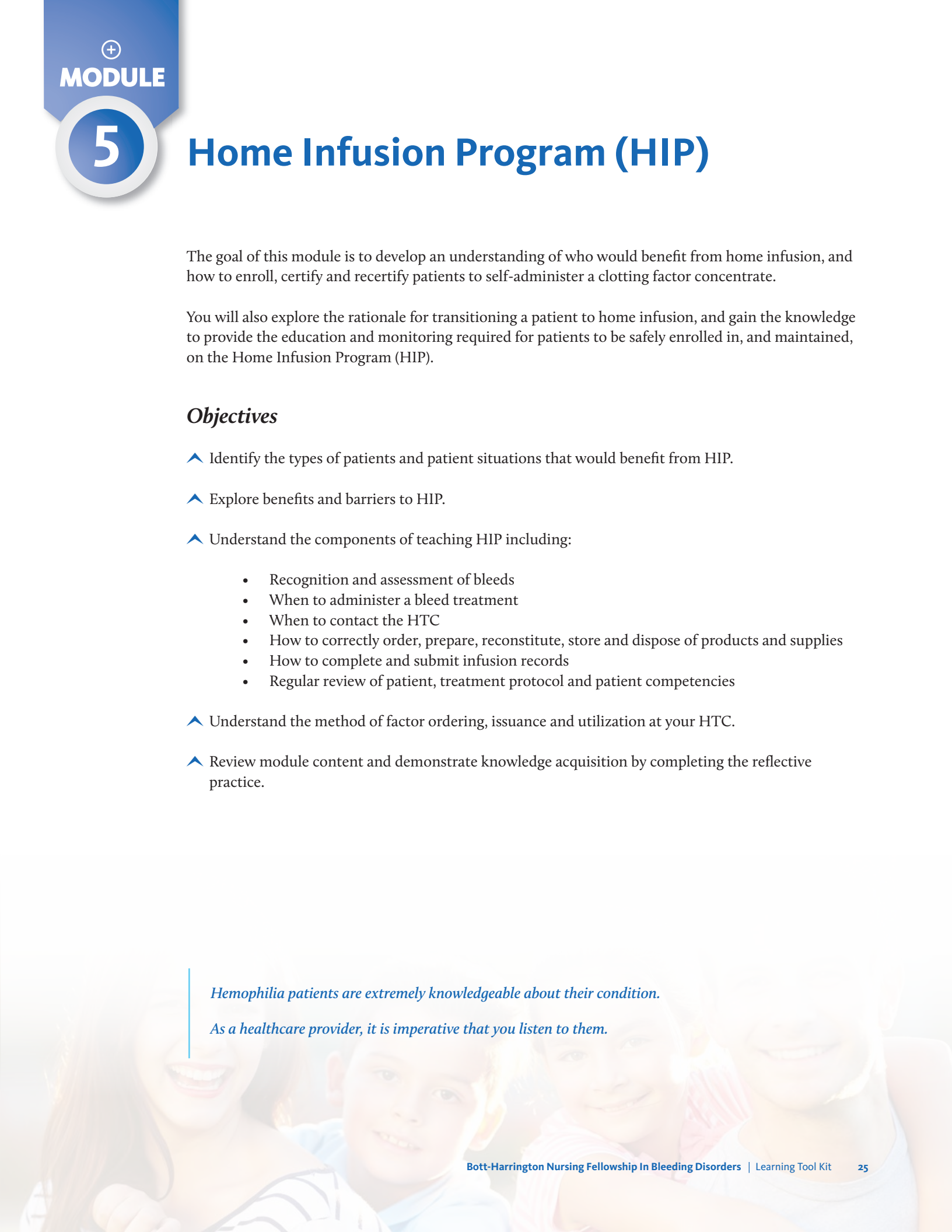
The goal of this module is to develop an understanding of who would benefit from home infusion, and how to enroll, certify and recertify patients to self-administer a clotting factor concentrate.

You will also explore the rationale for transitioning a patient to home infusion, and gain the knowledge to provide the education and monitoring required for patients to be safely enrolled in, and maintained, on the Home Infusion Program (HIP).

Objectives

- Identify the types of patients and patient situations that would benefit from HIP.
- Explore benefits and barriers to HIP.
- Understand the components of teaching HIP including:
    - Recognition and assessment of bleeds
    - When to administer a bleed treatment
    - When to contact the HTC
    - How to correctly order, prepare, reconstitute, store and dispose of products and supplies
    - How to complete and submit infusion records
    - Regular review of patient, treatment protocol and patient competencies
- Understand the method of factor ordering, issuance and utilization at your HTC.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

> *Hemophilia patients are extremely knowledgeable about their condition.*
>
> *As a healthcare provider, it is imperative that you listen to them.*



MODULE CONTENT

Please review the following. An * denotes previously referenced sources:

Read:

All About Hemophilia: A Guide for Families
Chapter 7: Home Infusion ***(2010)***

Home Treatment Guide for People with Bleeding Disorders (2009)

In 1969, Dr. Hanna Strawczynski launched the first Canadian Home Infusion Program (HIP) for hemophilia at the Montreal Children's Hospital. Home infusion of factor concentrates by a patient or caregiver delivers many advantages including better results with prompt treatment for bleeds, and improved lifestyle, especially for patients who live many kilometers away from a hemophilia clinic or hospital.

ADDITIONAL RESOURCES

Read:

***What is Prophylaxis (2014)**

Looking After the Veins (2003)

Home Infusion Manuals
(London Health Sciences Centre)

All About Hemophilia: A Guide for Families
Chapter 6: The Role of Prophylaxis ***(2010)***

KEY LEARNING

- HIP allows for prompt treatment of bleeds and easier adherence to prophylaxis regimens.
- HIP allows for fewer interruptions to school and work life for patients and families and allows for increased independence and control.
- HIP certification is a process that may take families differing amounts of time to complete. Ongoing support and monitoring is necessary to maintain competency and optimal outcomes.
- Venous access is critical for life-long care of patients with a bleeding disorder. Maintaining good vein health will permit some patients to use the same site throughout their life.
- A contract between patient/family and the HTC, at point of certification, can ensure the responsibilities of HIP are clearly understood.
- Good communication between patients/families and the HTC is an essential component of home infusion.

There are several strategies in place to provide care for patients who live far distances from a Hemophilia Treatment Centre (HTC):

- **Home infusion if possible**
- **HTC-coordinated care with local practitioners**
- **Tele-connection between HTC nurse coordinators, patients and families**
- **Conduct HTC outreach clinics**
- **Funding to support patient travel for annual HTC assessments**

> *Bleeding disorder information and the skills that accompany the actual care are very usable in daily practice.*

KEY LEARNING (con't)

❍ Expectations for patients enrolled in an HIP include:

- Ensuring an adequate supply of factor concentrate to meet treatment requirements
- Providing a record of infusion details (diary/infusion log)
- Attending routine assessment appointments
- Review self-infusion skills annually
- Maintain regular communication with the HTC about bleeds and relevant health concerns (upcoming surgeries/procedures)

❍ Potential risks of HIP include:

- Bleeds may not be assessed adequately therefore incorrect treatment may be given
- Poor response to treatment may not be identified
- Improper storage of clotting factor concentrates
- Improper disposal of sharps and supplies
- Infection or damage to veins or venous access device may go unrecognized

Patients with bleeding disorders who require frequent infusions of clotting factor concentrate and who have poor venous access will often require an alternative route of venous access. Though a Port-A-Cath® is the most common alternative, a PICC or arteriovenous (AV) fistula are other possible solutions.

REFLECTION

After completing this module, reflect upon your newly acquired knowledge and experience by describing how this might influence your ongoing nursing practice. To help you get started, reflection suggestions might include:

- The unique responsibility of caring for patients who are enrolled on HIP
- The effect of HIP on quality of life

MODULE 6

Musculoskeletal (MSK) Complications of Hemophilia and Other Bleeding Disorders

This module will help you understand the effects of bleeding in the musculoskeletal (MSK) system and the associated impact on the overall health of patients living with a bleeding disorder.

Objectives

- Discover the complexities of MSK assessments to improve your own assessment skills.
- Identify the risks associated with muscle bleeds and how to reduce the related complications.
- Delineate the effect of bleeding into a joint, focusing on both long- and short-term complications/damage.
- Review the broad variety of treatments used to manage an acute bleed, minimize long-term complications and alleviate the related pain of an MSK bleed.
- Describe orthopedic interventions used to manage hemophilic joint disease.
- Describe how physiotherapy can be utilized for the prevention and recovery of MSK bleeds.
- If you have access to a HTC:
    - Observe detailed MSK assessments with a skilled clinician (physiotherapist, rheumatologist, hematologist)
    - Demonstrate competency in conducting MSK bleed assessments and reporting your findings to the hemophilia care team in order to collaboratively develop a treatment plan
- Review module content and demonstrate knowledge acquisition by completing the module quiz.

> *Orthopedics has a special connection with hemophilia because many patients with bleeding disorders can develop joint problems due to joint bleeds.*

MODULE CONTENT

Please review the following. An * denotes previously referenced sources:

Read:

All About Hemophilia: A Guide for Families
Chapter 8: Complications of Hemophilia ***(2010)***

****Identifying Common Joint and Muscle Bleeds (2014)***

Assessment and management of pain in haemophilia patients (2011)

April 17th is World Hemophilia Day, created in 1989 to raise awareness for hemophilia and other inherited bleeding disorders. The annual date chosen honours Frank Schnabel, founder of the World Federation of Hemophilia (WFH) – it's his birthday

•

The number of red blood cells, white blood cells and platelets is the exact same in people with and without hemophilia. The only difference is the person with hemophilia lacks factor VIII or IX protein in their blood.

> *This tool kit is an excellent learning resource about bleeding disorders.*
>
> *It should be required reading, especially for emergency room nurses.*

ADDITIONAL RESOURCES

Read:

Musculoskeletal Complications of Hemophilia (2010)

Musculoskeletal Complications of Hemophilia: The Joint (1997)

Musculoskeletal health of subjects with hemophilia A treated with tailored prophylaxis:
Canadian Hemophilia Primary Prophylaxis Study (CHPS) ***(2012)***

Rehabilitation of Muscle Dysfunction in Hemophilia (2012)

All About Hemophilia: A Guide for Families
Chapter 12: Physical Activity, Exercise and Sports ***(2010)***

Exercises for People with Hemophilia (2006)

In joint bleeding, the components of red blood cells can accumulate in the joint space. One of these components is hemosiderin, an iron-containing complex that can cause damage if it accumulates in tissues.

Watch:

****The Coagulation Cascade:***
A computer-generated movie on hemophilia

****Video: Joint Bleeds and Complications (2011)***

KEY LEARNING

- Prophylaxis is standard of care for severely affected patients with bleeding disorders.
- Maintaining a healthy weight and staying active helps to build muscle and reduce the impact on joints.
- Hemophilic arthropathy is the most common complication of hemarthroses.
- Pain and mobility issues associated with hemophilic arthropathy will impact a patient's quality of life.
- A basic assessment of a bleeding joint should include an assessment of the patient's pain as well as a MSK assessment comparing affected to unaffected side for: range of motion (ROM), swelling, temperature, and neurovascular changes.
- Target joints unresponsive to secondary prophylaxis may benefit from joint aspiration, steroid injection and radiosynovectomy.
- Orthopedic surgery, such as arthrodesis and arthroplasty, are often needed to alleviate the pain of chronic hemophilic arthropathy.

> *Over the years of working in orthopedics, we're slowly starting to see a decrease in joint problems caused by bleeding disorders. The advent of home therapy starting in childhood has had a positive impact on adulthood joint degeneration since patients can self-administer to catch a bleed right away.*

TEST YOUR KNOWLEDGE

The answers appear on page 52 of this tool kit.

1. **Some medications that are used to treat the pain associated with acute bleeds and chronic joint disease are** (circle all that apply):
   a. Opioids
   b. COX-2 inhibitors (Celebrex, Mobicox)
   c. Acetaminophen
   d. Acetylsalicylic acid (ASA)

2. **The function of the synovium is:**
   a. Lubricate the joint
   b. Innervate the stabilizing muscle
   c. Important source of nutrients for the cartilage
   d. a and c

3. **The symptoms of arthritis are:**
   a. The joint may be painful when used
   b. The muscles around the joint may be weak and atrophied from lack of use
   c. The joint becomes stiff, especially in the morning or after long periods of sitting
   d. All of the above

4. **A patient with hemophilic arthropathy may have difficulty differentiating between arthritic pain and swelling, and bleed pain and swelling:**
   a. True
   b. False

5. **Synovitis and arthritis can be prevented by:** (choose all that apply)
   a. Prophylaxis (giving factor to prevent bleeding)
   b. Recognizing and treating bleeds immediately
   c. Applying R.I.C.E. to promote healing
   d. Treating with Aspirin (ASA)

MODULE 7

Psychosocial Aspects of Care

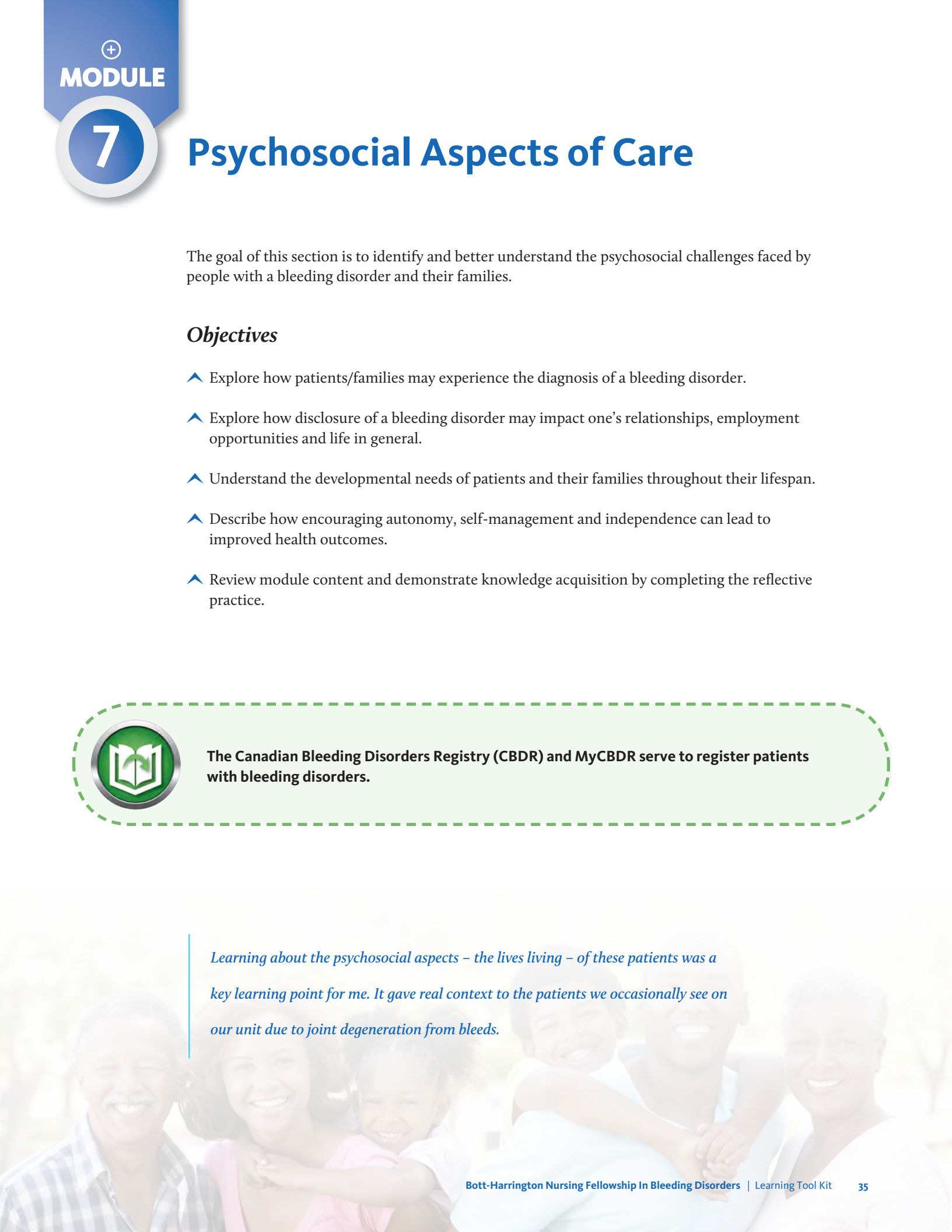
The goal of this section is to identify and better understand the psychosocial challenges faced by people with a bleeding disorder and their families.

Objectives

- Explore how patients/families may experience the diagnosis of a bleeding disorder.
- Explore how disclosure of a bleeding disorder may impact one's relationships, employment opportunities and life in general.
- Understand the developmental needs of patients and their families throughout their lifespan.
- Describe how encouraging autonomy, self-management and independence can lead to improved health outcomes.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

The Canadian Bleeding Disorders Registry (CBDR) and MyCBDR serve to register patients with bleeding disorders.

> *Learning about the psychosocial aspects – the lives living – of these patients was a key learning point for me. It gave real context to the patients we occasionally see on our unit due to joint degeneration from bleeds.*



MODULE CONTENT

Please review the following:

Read:

All About Hemophilia: A Guide for Families
Chapter 10: Growing with Hemophilia ***(2010)***

All About Hemophilia: A Guide for Families
Chapter 13: Transitions Towards Independence ***(2010)***

Contact:

Meet with an HTC's program social worker to gain an understanding of the issues facing this unique population, and strategies that can be used to support the patients in their experiences.

Visit:

Teens Taking Charge: Managing Hemophilia *(2014)*

Healthy living tips for people with bleeding disorders include:

- **Stay in close communication with your Hemophilia Treatment Centre**
- **Routine checkup at an HTC**
- **Maintain a regular prophylaxis regimen – treat bleeds early and adequately**
- **Exercise and maintain a healthy weight to protect joints**
- **Get vaccinated – Hepatitis A and B are preventable**

> *Knowledge is the key to increasing care-provider confidence.*

ADDITIONAL RESOURCES

Read:

All About Carriers (2007)

Bon Voyage! Travelling with a bleeding disorder (2012)

A pilot study on the effects of the transition of paediatric to adult health care in patients with haemophilia and in their parents: patient and parent worries, parental illness-related distress and health-related Quality of Life (2008)

Towards comprehensive care in transition for young people with haemophilia (2010)

Genetic Counselling for Hemophilia (2002)

Psychosocial Care for People with Hemophilia (2007)

Take Control: Transitioning to Adult Care – "What should I know?" (2015)

Watch:

Camp Wanakita Part 1 (2009)
Camp Wanakita Part 2 (2009)

Celebrities and high-profile people affected by hemophilia include movie star Richard Burton, actor-comedian Alexandrea Borstein (the voice of Lois Griffin on Family Guy) **and professional cyclist Alex Dowsett.**

KEY LEARNING

- The diagnosis of a bleeding disorder can trigger feelings of loss of one's potential and fears of the future. Care must be taken to address these feelings at the time of diagnosis and throughout a patient's lifespan.
- Living with a life-long health condition while managing work and family responsibilities can be very stressful.
- Disclosing the diagnosis of a bleeding disorder can be an emotionally stressful experience. The HTC can play a crucial role in supporting patients and caregivers regarding whether to disclose, when and how to disclose, and human rights around work and disclosure.
- Body image concerns arising from joint damage and side effects of medications for HCV and HIV can impact patients' feelings of intimacy. Some may physically find sexual intercourse difficult due to joint immobility.

The National Inherited Bleeding Disorder Genotyping Lab was established in 2000 at Queen's University, Kingston, ON. The lab provides genetic testing and database documentation for genetic analysis of inherited bleeding disorders.

Elevate your learning by not just reading the tool kit modules for facts, but also by really reflecting on how hemophilia and other bleeding disorders affect the whole person and the life they're living.

> *As a nurse, there's the potential of encountering patients with bleeding disorders in many departments – educate yourself.*

- Programs supported by the Hemophilia Society, such as camp and Families in Touch events, can provide opportunities for education, promote normalization and foster independence.
- Transition is a process that should begin early so a gradual shift in responsibility occurs. Transitioning responsibilities from parents to the child can be difficult for all involved.
- Transitioning from pediatric to adult care can be an additional stressor on young adults as well as parents. The HTC must be involved to ensure a smooth transfer of care.

At the very least, use this information to better understand the reality of *living* with the condition as opposed to just *learning* about the condition

There are several hemophilia-related organizations that patients, family members and health care providers can turn to for more information:

- **Canadian Hemophilia Society**
- **World Federation of Hemophilia**
- **National Hemophilia Foundation**
- **Association of Hemophilia Clinical Directors of Canada**
- **Canadian Blood Services**
- **HémaQuébec**

REFLECTION

After completing this module, apply your newly acquired knowledge and experience by describing how this might influence your daily nursing practice. Areas to reflect upon might include:

- How the hereditary nature of hemophilia may influence a patient's experience of the disease
- How decisions made in childhood and young adulthood can impact the patient's experience of aging with a life-long condition
- How bleeding risk and complications may impact sexual intimacy and relationships

MODULE 8

The Role of Inhibitors

This module is designed to help you demonstrate an understanding of the unique challenges associated with providing care to people with an inhibitor.

Objectives

- Understand the pathophysiology of congenital and acquired inhibitors.
- Identify risk factors of inhibitor development.
- Understanding the importance of screening to allow for early detection of inhibitors.
- Identify the signs of inhibitor development and understand the importance of patients and families having the ability to identify the signs of inhibitor development
- Gain an understanding of the complexities of caring for patients with inhibitors:
    - Immune Tolerance Induction (ITI)
    - Treatment and management of bleeding episode
    - Prophylaxis
    - Preparation for invasive procedures
- Understanding the importance of frequent assessment by the HTC team, including routine laboratory testing, to optimize patient outcomes.
- Review module content and demonstrate knowledge acquisition by completing the module quiz.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

> *Just after finishing the fellowship, an elderly patient was admitted to our department with a right-flank bleed due to a fall. Diagnosis was acquired hemophilia A. In conjunction with our HTC professionals, we were able to help support and teach her, and her caregiver, about the condition.*

- Incidence of acquired hemophilia is 2 per million population
- 20% to 30% of people with severe hemophilia A (factor VIII deficiency) will develop some level of inhibitory antibody to their treatment
- 5% to 10% of people with mild or moderate hemophilia A will develop inhibitors
- Less than 5% of people with severe hemophilia B (factor IX deficiency) will develop high-level inhibitors to treatment

MODULE CONTENT

Please review the following. An * denotes previously referenced sources:

Read:

All About Hemophilia: A Guide for Families
Chapter 8: Complications of Hemophilia ***(2010)***

All About Inhibitors (1999)

ADDITIONAL RESOURCES

Read:

Diagnosis and treatment of factor VIII and IX inhibitors in congenital haemophilia (2012)

A Guide to the Management of Patients with Inhibitors to Factor VIII and Factor IX (2010)

What Are Inhibitors? (2009)

Diagnosis and Management of Inhibitors to Factor VIII and Factor IX (2004)

Inhibitors in Hemophilia: A Primer (2008)

Acquired Hemophilia (2005)

KEY LEARNING

- Inhibitors are antibodies that target clotting factor protein:
    - In congenital hemophilia, this is an alloantibody that is directed at the infused clotting factor concentrate
    - In acquired hemophilia, this is an autoantibody that is directed at the patient's endogenous factor
- Inhibitors are the most serious complication of hemophilia.
- Steps should be taken to minimize the risks of antibody formation to clotting factor concentrates whenever possible.
- Quality of life is impacted due to:
    - Increased burden of care
    - Unpredictability of response to therapy
    - Increased incidence of hemophilic arthropathy
- Care of patients with inhibitors is targeted at eradication of the antibody through immune tolerance induction (ITI) and management of bleeding with bypassing agents.

While hemophilia is most often thought of as a genetic disorder, there is a type of hemophilia that is not inherited called acquired hemophilia. Acquired hemophilia results in the formation autoantibodies that inactivate factor VIII. It can be associated with various autoimmune disorders, cancers, pregnancy or have no known cause.

Launched in 2015, the Canadian Bleeding Disorders Registry (CBDR) is a national clinical patient database managed by McMaster University, Hamilton, ON, on behalf of the Association of Hemophilia Clinic Directors of Canada (AHCDC). The system is currently being phased into Hemophilia Treatment Centres across the country to assist in the treatment and research of bleeding disorders. Software tool MyCBDR was also launched to enable patients to record condition updates and view their information.

> *Life-long learning is part of our nursing mandate, so professional development opportunities like this tool kit are good.*

REFLECTION

After completing this module, think about your newly acquired knowledge and experience and describe how you might apply it in your ongoing nursing practice. This may include reflections upon:

- How an inhibitor impacts a patient's quality of life and care
- How this is relevant to your clinical setting

TEST YOUR KNOWLEDGE

The answers appear on page 52 of this tool kit.

1. **Inhibitors are antibodies that are directed at specific coagulation factors in the body:**
   a. True
   b. False

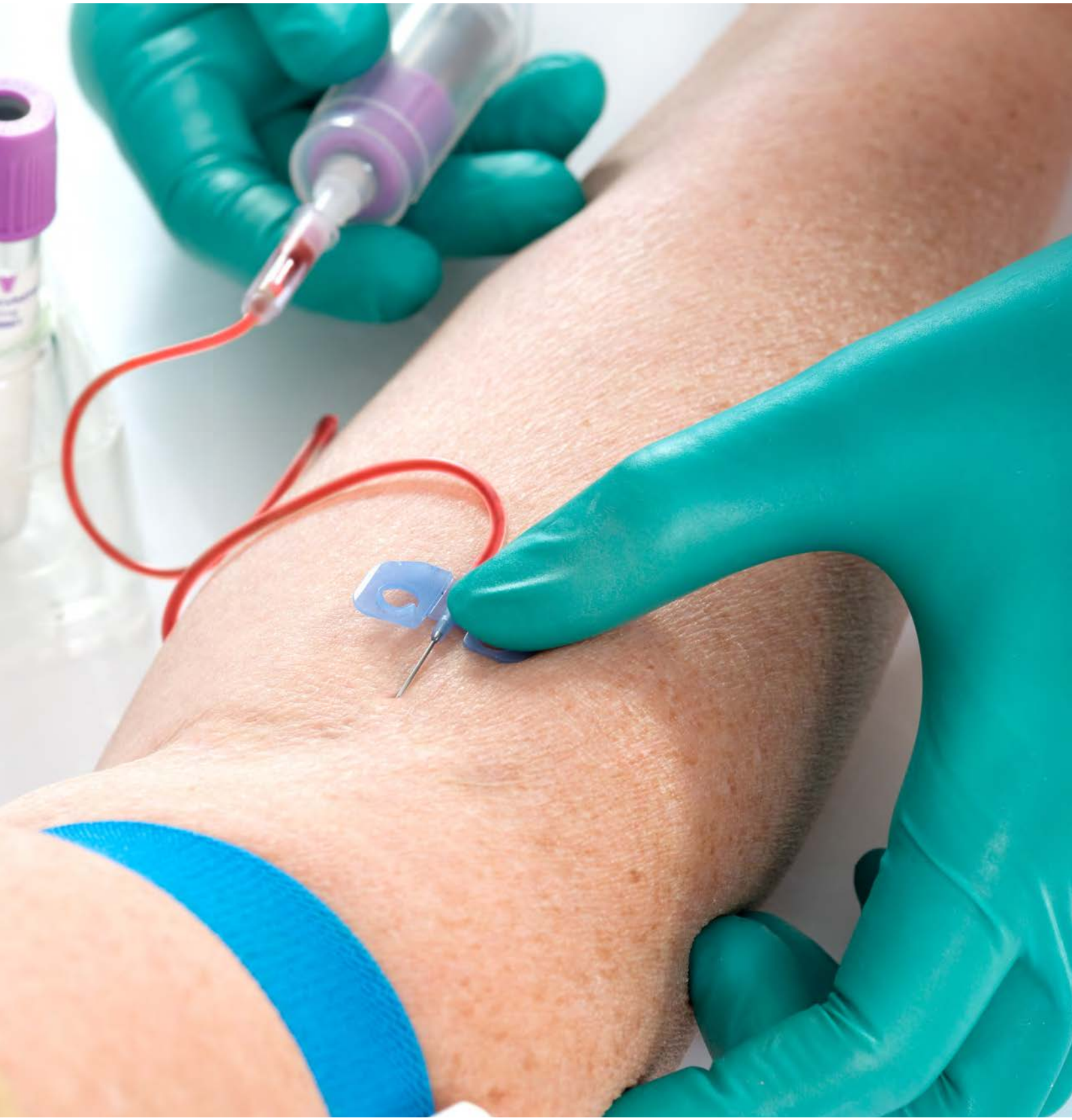
2. **Inhibitors are more common in hemophilia A than hemophilia B:**
   a. True
   b. False

3. **Immune Tolerance Induction (ITI)** (circle all that apply)**:**
   a. Involves giving high doses of factor at frequent intervals
   b. Overwhelms the immune system
   c. May take over three years to eradicate the inhibitor
   d. Requires excellent venous access and a strong commitment by the patient/family

4. **The unit of measurement for an inhibitor is Bethesda Units (BU):**
   a. True
   b. False

5. **FEIBA** (factor eight inhibitor bypassing agent) **and Niastase** (recombinant factor VIIa) **are both coagulation factor concentrates that are used to treat and prevent bleeding in patients with inhibitors:**
   a. True
   b. False

6. **First-line treatment to eradicate acquired hemophilia involves immunosuppression with cyclophosphamide and prednisone:**
   a. True
   b. False



MODULE 9

Preparing for Procedures

This module examines the care history of bleeding disorders and how comprehensive care is currently provided in Canada.

The Canadian Association of Nurses in Hemophilia Care (CANHC) was formalized in 1997.

Objectives

- Understand the bleeding risk in relation to patient diagnosis and planned procedure and how this impacts decisions around the plan of care.
- Appreciate the risk associated with inhibitor development around the time of interventions/surgeries and the need for inhibitor screening pre-operatively.
- If you have access to a HTC:
    - Develop skill and expertise at creating individualized treatment plans based on established guidelines, clinical knowledge and previous experience in collaboration with the HTC medical director
    - Understand the communication required with multiple stakeholders in preparing for procedures:
        - ~ Patient
        - ~ Anesthetist, surgeon, dentist, interventional radiologist, Obstetrics/Gynecology
        - ~ Coagulation Lab
        - ~ Transfusion medicine services (blood bank)
        - ~ Pharmacy
        - ~ Nursing unit caring for patient before and after procedure
        - ~ Post-discharge nursing support service (rehabilitation services, community nursing agencies, industry sponsored patient support programs)
    - Gain an understanding of the importance of having surgery done at the HTC, appreciating the need to have a competent coagulation laboratory available to process blood samples for factor assays in a timely manner
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

> *My fellowship days in the HTC coincided with its pre-natal care sessions. It was incredible to witness the enormous support and specialized care that was preparing these young women for potentially having a son with hemophilia or a daughter who carries the gene.*

MODULE CONTENT

Please review the following:

Read:

Nursing Guidelines for the Treatment of Hemophilia & Other Inherited Bleeding Disorders (London Health Sciences Centre) ***(2014)***

Guidance on the dental management of patients with haemophilia and congenital bleeding disorders (2013)

ADDITIONAL RESOURCES

Read:

Comprehensive care of the patient with haemophilia and inhibitors undergoing surgery: practical aspects (2013)

Dental Care for People with Bleeding Disorders: What you Need to Know (2012)

Joint Replacement Surgery in Hemophilia (2010)

What every nurse should know about hemophilia (2013)

> *In orthopedic surgery, we sometimes must give anti-coagulants to patients told to stay away from blood thinners. During my fellowship time, I worked with the HTC team to improve communication and clarity around these orders; this is a prime example of how the fellowship bridged my front-line career.*

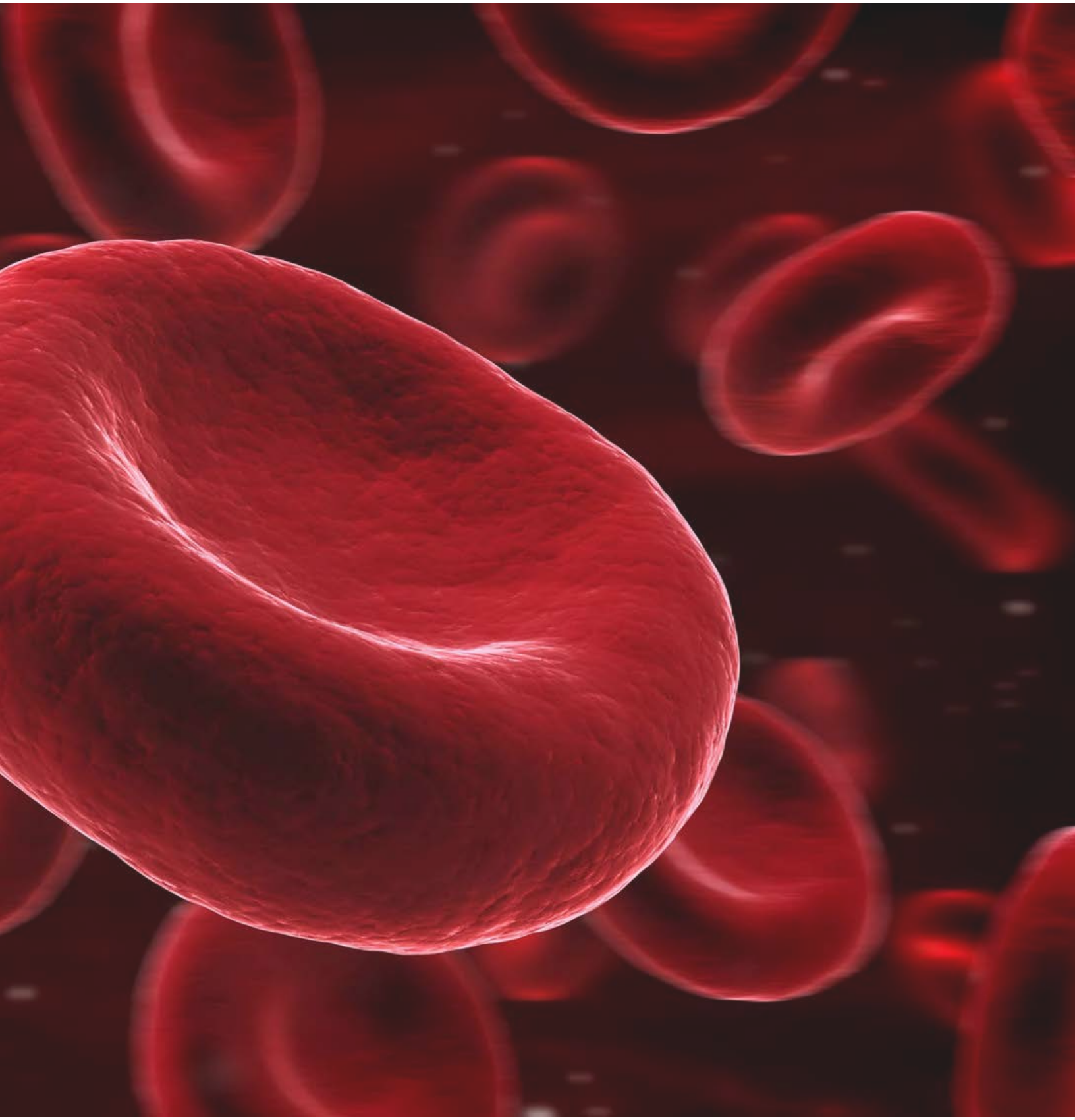
KEY LEARNING

- Patients are educated to inform the hemophilia treatment centre (HTC) prior to undergoing ANY invasive procedure.
- Adequate correction of the hemostatic deficiencies prevents intra- and post-procedure bleeding and complications.
- Communication between the HTC and the responsible MD (surgeon, dentist, etc) is crucial to ensuring appropriate management of the patient with a bleeding disorder.
- Many patients with milder bleeding disorders often fail to disclose their disorder and due to the milder nature of their deficiency are not identified. This may lead to patients undergoing invasive procedures without informing the HTC or receiving treatment.
- Routine pre-op screening with a partial thromboplastin time (PTT) may falsely reassure a surgeon/anesthetist that it is safe to proceed putting a patient with mild factor deficiency at risk of bleeding due to a lack of hemostatic coverage.
- Recent inhibitor monitoring is essential for appropriate assessment of risk around the time of procedures.
- Many health care professionals outside of the HTC are unfamiliar with hemophilia and other bleeding disorders and knowledge may be lacking with respect to the importance of vigilant monitoring and intervention at the time of procedures.
- Invasive procedures are ideally performed at the HTC and must occur at the HTC for inhibitor patients.

> *I am now able to expedite care for bleeding disorder patients in our area, not only by championing their care and getting and administering factor product, but also teaching other nurses how to provide the required treatment.*

REFLECTION

How might your newly acquired knowledge and experience from this module influence your daily nursing practice? Are there ways you can apply the learning? Keep in mind that your confidence and ability to create and implement individual treatment plans will increase as your knowledge and experience of bleeding disorders develops.



MODULE 10

The Impact of HIV and Hepatitis C on Patients with Bleeding Disorders: A Historical Perspective

The purpose of this section is to help you gain a deeper understanding of the impact that HIV and Hepatitis C, transmitted by blood products in the 1980s and 1990s, has on patients with hemophilia, their families and the bleeding disorders community.

Objectives

- Develop an understanding of the lived experience of hemophilia patients with HIV and/or Hepatitis C (HCV) by exploring their collective and individual experiences and challenges through both the module content and interviews with select patients.
- Explore the psychosocial impact and stigma on patients who acquired life-threatening infections through contaminated blood products.
- Review module content and demonstrate knowledge acquisition by completing the reflective practice.

> *The modules in this (fellowship) program provide excellent learning, especially if you work with HIV patients with hemophilia or Hepatitis C, or encounter other autoimmune diseases.*

MODULE CONTENT

Please review the following:

Read:

The Tragic History of AIDS in the Hemophilia Population 1982-1984

Watch:

The Inside Story Pt. 1: Red Cross blood scandal victim speaks out

The Inside Story Pt. 2: Red Cross blood scandal victim speaks out

The Casey Awards 2013 - John Plater

CBC Video Archives 1983 - 2007

(1983)	(1996)
(1984)	(1997)
(1985)	(1997)
(1994)	(1998)
(1994)	(2002)
(1995)	(2007)

Contact:

Meet with patients living with HIV and/or HCV

ADDITIONAL RESOURCES

Read:

Canadian Hemophilia Society – report cards on the Canadian blood System

Visit:

CATIE: Canada's source for HIV and Hep C Information

Canadian Hemophilia Society – HCV/HIV

KEY LEARNING

- Some patients with hemophilia who were infected with HIV and/or Hepatitis C through their prescribed treatments blame the health care team for their infection. As a result, they may feel a sense of betrayal and mistrust in the health care system.

- In the 1980s, the development of what seemed to be a mild and unknown form of hepatitis (later identified as hepatitis C) was considered an acceptable risk.

- In 1982, the Centre for Disease Control (CDC) began special investigations that resulted in the discovery that AIDS was not just a sexually transmitted disease particular to the homosexual population – but perhaps a blood-borne pathogen. This investigation was launched because hemophilia patients, having only one known risk factor, blood transfusions, began to present with symptoms resembling the early signs of AIDS (e.g. pneumocystis pneumonia (PCP)).

In 1993, Justice Horace Krever was appointed to lead a Royal Commission investigating the contaminated blood products of the early 1980s. Krever's 50 recommendations, contained in a 1,200-page report, were the result of:

- The four-year inquiry
- 274 days of hearings
- 474 witnesses appearing before the Commission
- 389 individuals and organizations delivering telephone or written statements
- 50,000 pages of testimony
- 175,000 documents totaling about one million pages collected

REFLECTION

Reflect upon how this module's learning experience and knowledge about HIV and Hepatitis C infections in the hemophilia population might alter your daily nursing practice. Things to think about might include:

- How your personal bias or fear may present a challenge when caring for this patient population
- How to reduce stigma and misunderstandings about HIV and Hepatitis C among your colleagues
- The impact of receiving contaminated blood and how it might affect the relationship between a patient and the health care team

TEST YOUR KNOWLEDGE ANSWERS

Module 2

1. e
2. d
3. b
4. a
5. b
6. a, b, d
7. d

Module 3

1. e
2. d
3. e
4. a, b, d
5. e
6. e

Module 4

1. a, b, c, d
2. d
3. c
4. b
5. a, c, d
6. b, c, d, e, f
7. a, b, c, d

Module 6

1. a, b, c
2. d
3. d
4. a
5. a, b, c

Module 8

1. a
2. a
3. a, b, c, d
4. a
5. a
6. a

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