Hemophilia and Hospitalization
A Self Learning Program

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This is a self-learning program designed to give the health care provider an overview of hemophilia and the care of patients with this disorder.

DIRECTIONS

Read this module, and answer the post-test questions. Review your answers; if you have any incorrect answers please refer back to the text.

Hemophilia A and Hemophilia B

Hemophilia A, factor VIII deficiency, and hemophilia B, factor IX deficiency, are hereditary bleeding disorders. There are many proteins in the blood necessary for it to clot properly. Factor VIII and factor IX are two of these proteins. When a patient has less than the lower limit of normal of one of these proteins, they are given a diagnosis of hemophilia.

The genes involved in the production of factor VIII and factor IX are carried on the X chromosome. They are frequently referred to as sex-linked disorders. Because this is an x-linked inherited disorder, it affects males and women are carriers of the disorder. Approximately 20% of carrier women actually have low factor levels themselves and can have bleeding with significant trauma or surgical procedures just like a hemophiliac.

In the majority of hemophilia patients there is a family history of the disorder; however a new mutation can occur for the first time in the birth of an infant. About one out of three persons with hemophilia have no family history of the disorder.

Hemophilia A and B can occur in different severities depending on the amount of clotting protein circulating in the blood, mild >5-50%, moderate >1-5% and severe 0-1%. The severity of the disorder remains the same in families. Patients with severe hemophilia have the most frequent bleeding and those with mild hemophilia typically bleed following trauma or an invasive procedure. All patients with hemophilia regardless of their severity will require specialized treatment while hospitalized.

Bleeding episodes in patients with hemophilia can occur anywhere there is an injury to a blood vessel wall, however most bleeding episodes occur in soft tissue, muscles, and joints. Most patients are able to identify a cause of their bleed, but this is not always true, especially in patients with severe hemophilia, as they can have what is referred to as a "spontaneous hemorrhage". In very young children the cause of the bleed may not be known due to non verbal communication.

Treatment of hemophilia bleeding episodes is most often done by replacing the missing clotting factor either factor VIII or factor IX, in adequate amounts to stop bleeding. The clotting concentrates are administered intravenously. The amount or dose of factor is determined by a hematologist or primary care physician caring for the patient. Patients with mild hemophilia A may use clotting concentrates to treat bleeding episodes like their more severe counterparts, or they often are able to use a synthetic hormone known as DDAVP to treat bleeding. This medication is administered intravenously, and also comes in an intra nasal high concentration form. DDAVP works by causing the release of stores of factor VIII from endothelial cells into the blood stream. This raises the factor VIII level in the blood stream. The patient should have a documented response to this medication prior to its use. DDAVP should never be given to a child under the age of 2. **There is no equivalent product for factor IX deficient patients.**

What Brings a Child with Hemophilia to the Hospital?

UNCONTROLLED OR SERIOUS BLEEDING

Patients with all types and severity of hemophilia can present to the hospital emergency room with a bleeding episode. Most parents/patients with severe or moderate hemophilia have been trained in home infusion by the age of 5 and are able to administer the clotting concentrate at home when a bleeding episode starts. Patients with severe hemophilia generally present when a bleed is causing significant pain or is not responding to the treatment. Children who are not yet on home infusion will come in for factor replacement when they are having a bleeding episode. Most patients/families with mild hemophilia are not able to administer the clotting factor or intravenous DDAVP at home so they will present for evaluation and treatment. Due to infrequent bleeding in mild hemophilia these patients often do not recognize a bleed until it has gone on for some time, so they often have a significant hemorrhage in progress when they present.

Intracranial hemorrhage is a serious and life threatening complication of hemophilia. When a child with hemophilia presents to an emergency room with signs and symptoms of this type of bleed or any other life threatening hemorrhage, **factor concentrate should be administered immediately.** A full evaluation including lab work, CT scans, MRI, and x-rays can then be performed. This could mean the difference between life or death to these patients.

Patients with serious bleeding problems may be admitted to the hospital for follow up treatment or they may be discharged from the emergency department. Patients with life or limb threatening hemorrhage will always be admitted for observation and follow up treatment.

SURGERY

All surgeries in persons with hemophilia require factor replacement or treatment. If surgical procedures are not done with adequate factor replacement, significant hemorrhage can take place during and after the procedure and wound healing will be impaired.

In children with hemophilia the most common surgical procedure is placement of a venous access device, usually a port-a-cath. Dental procedures done under anesthesia are also common.

Older adolescents with hemophilia may have significant arthritic changes in joints as a result of repetitive bleeding into these joints. Severe arthritis often warrants orthopaedic procedures. Very minor procedures can be done on an outpatient basis if the patient is able to remain locally or lives close by the hospital in case there are problems.

All other elective surgeries can be done and often are scheduled in children with hemophilia when needed. Hemophilia alone should not prevent a child from having an elective procedure. Adequate factor replacement is required.

The hematologist will manage the factor replacement for a surgical procedure in consultation with the surgeon. Members of the Hemophilia Treatment Center staff are available to assist the patient, his family and hospital staff with any questions or concerns.

RELATED ISSUES

A few older adolescents with hemophilia have been exposed to hepatitis C. This chronic illness has complications that can bring a hemophiliac to the hospital. Liver biopsies may be required for those patients who are hepatitis C positive; hospitalization may be needed with this procedure.

Patients with hemophilia are not exempt from medical problems that plague other people. Hemophiliacs should be cared for like all other patients.

Basic Care

ROUTINE CARE

Routine care for the hospitalized child with hemophilia is no different than that of any other hospitalized child. Do not assume that these children are accustomed to hospital settings. The goal of the hemophilia center is to keep children out of the hospital, so even though this child has a chronic illness this may be their first hospitalization.

PROCEDURES

Any invasive procedure in a child with hemophilia will require careful discussion to determine if factor replacement is necessary. The hematologist or primary care physician should be notified prior to the procedure by the physician ordering the procedure. Following is a list of procedures where factor replacement is necessary; this is not an all inclusive list but a starting point:

Suturing of any type
Arterial blood gases
Skin biopsy
Endoscopy
Gastric lavage
Spinal tap
Dental procedures/dental surgery

TIPS FOR THE CARE OF CHILDREN WITH HEMOPHILIA

1. When lab work (especially a factor level) is ordered on a hemophiliac and a time is specified, be sure that the blood is drawn at that time. These labs are necessary in determining optimal use of factor replacement. The half-life of factor VIII is between 8-12 hours and the half-life of factor IX is between 18-24 hours. This necessitates the timely running of these samples.

2. While many of these patients have never been hospitalized before, they or their parents are very familiar with their venous access. If a patient/parent suggests that a certain vein not be used, please honor that request. If the child has a port-a-cath that needs to be accessed it is advisable to allow the parent to access the port as they are most familiar with their child.

3. If the child has a central venous access device, lab may be drawn from the line after a 10 cc discard. If at all possible, factor levels should not be drawn from a line that factor concentrate has been or is being administered through.

4. **Do not use heat** on a child with hemophilia. This can cause vasodilatation and may exacerbate a bleeding episode.

5. In addition to no heat, all aspirin-containing medications should be avoided as well as IM injections. Some hematologist will allow IM injections if the patient has adequate factor coverage.

Factor and Factor Administration

TYPES OF FACTOR

There are three categories of factor replacement products; intermediate purity human derived factor, high purity human derived factor, and recombinant (genetically engineered) factor. Each of these types of products has a place in the care of persons with hemophilia. Most children should receive a recombinant factor product.

Intermediate Purity Human Derived Factor: This type of factor is derived from human blood, is virally inactivated to prevent the transmission of viruses, and contains more than one specific factor protein. The factor VIII products contain von Willebrand protein as well as the factor VIII and can be used for the treatment of von Willebrand disease. The factor IX products contain other vitamin K dependent clotting proteins, factor II, VII, and X in varying amounts and be used to treat patients who are deficient in these clotting proteins.

High Purity Human Derived Factor: This type of factor is derived from human blood, is virally inactivated to prevent the transmission of viruses, and contains either factor VIII alone or factor IX alone. They contain no other coagulation proteins and can not be used to treat other factor deficiencies.

Recombinant (genetically engineered) Factor: This type of factor is manufactured in a laboratory in a controlled environment. Some of these brands do come in contact with or contain human albumin, while others have no contact with human blood at all. The end product contains only factor VIII or factor IX, no other coagulation proteins and cannot be used for other factor deficiencies.

FACTOR ADMINISTRATION

If you have never administered factor concentrate please read the product insert that accompanies the product. It will walk you through how to mix and administer this medication.

Steps for administration of Factor Concentrate

1. The order would be written by a physician or physician designee. It should include the type of factor (human or recombinant). A specific brand may be requested by either the patient or the physician. The order should also include the number of units and the route of administration, (IV push or continuous infusion), and frequency. An order from a physician is required to change a brand or type of factor. An example of an order:

   Kogenate FS 850 units IV push (number of units are located on the box as well as the vial)

2. The order should be sent to the pharmacy. The nurse may want to make a follow up phone call to the pharmacy to assure that the dose will be sent to the floor prior to the time the next dose is due.

3. **Bolus Infusion:** The factor will arrive on the unit in a syringe. Often times the dose that is sent from the pharmacy may differ slightly from the physician order. This is due to the fact that units per vial vary from lot to lot and as long as the dose is +/ 10% of the ordered dose it is acceptable to give. These are the guidelines that are given to the pharmacy by the hematologist on staff. Vial sizes are not exact; factor should never be thrown away. If the factor is to be administered through a running IV, the maintenance fluid should be stopped, the line flushed with normal saline, the factor administered according to the rate specified in the product insert, the line should be flushed again with normal saline and then the maintenance IV restarted.

4. **Continuous Infusion:** Continuous infusion of factor VIII or factor IX is used following major surgery, intracranial hemorrhage or other significant trauma where a continuous level of factor needs to be maintained; avoiding the peaks and troughs that happen with bolus infusion. Continuous factor infusion must

always be on an infusion pump. The rate of infusion will be specified in the orders.

Tips for continuous infusion:

a. Always run a dedicated line
b. Factor should hang no longer than 12 hours, after 12 hours it should be discarded and a new infusion set up.
c. Brand name **Recombinate** is the only recombinant factor VIII that can be used in continuous infusion.
d. When you have one hour's worth of factor left in the buretrol, contact the pharmacy for another syringe. Continuous means continuous!

PROBLEM SOLVING

The number one thing to understand when taking care of a child with hemophilia and/or administering factor is **if you don't know, ask.** Hemophilia is not common. This may be the first time you have cared for one of these children or administered factor. There is a hemophilia nurse available to assist Monday through Friday 8-5 on pager (303) 266-4517. After hours and on the weekend the hematology fellow on call can be accessed through the hospital operator.

Discharge Planning

It is important to begin discharge planning as soon as the child is admitted to the hospital. If a hemophiliac is admitted to the hospital it is likely that he will need follow up care, including factor administration upon discharge. The unit discharge planner can work with the patient and his family to assess what will be needed. Factor concentrate is generally supplied by the homecare company that routinely services the patient (on an out-patient basis) and those orders can be sent to them. They will usually supply all of the infusion supplies including dressing change kits if a central line or venous access device is used. If the child and /or his family are not able to administer the clotting concentrate at home then a visiting nurse will be needed. The hemophilia team will set up any follow up appointments regarding the patient's hemophilia. Other follow up appointments and discharge prescriptions should be handled in the unit's usual manner.

POST TEST

TRUE OR FALSE

1. When a patient is admitted for a significant hemarthosis it is appropriate to apply heat to the joint.

   True False

2. Hemophiliacs can have surgery safely without factor replacement if it is a minor procedure.

   True False

3. Patients with mild hemophilia do not bleed very often and may not be well educated about their disorder.

   True False

4. A continuous infusion of factor VIII or factor IX should is good for 16 hours.

   True False

5. When factor is ordered for a patient it is alright to use what ever brand the pharmacy sends up.

   True False

MULTIPLE CHOICE

1. Persons with hemophilia A are missing factor:

   a. IX
   b. VII
   c. VIII
   d. all of the above

2. Orders for bolus infusions of factor should contain all of the following except:

   a. number of units
   b. type/brand of factor
   c. frequency
   d. time of next factor level

3. The most common type of surgery done on persons with hemophilia is:

   a. Port placement
   b. General Surgery
   c. Cardiac Surgery
   d. Orthopaedic Surgery

4. Which is not a reason a hemophiliac would be admitted to the hospital:

   a. Uncontrolled bleeding episode
   b. Comprehensive hemophilia evaluation
   c. Medical emergency
   d. Elective surgery

5. Prior to drawing a factor level from venous access device how much blood should be discarded?

   a. 2 cc
   b. 10cc
   c. 5cc
   d. None of the above

POST TEST ANSWERS

TRUE OR FALSE

1. False
2. False
3. True
4. False
5. False

MULTIPLE CHOICE

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

REFERENCES

Coyne, Michael. *Avoiding Indecision and Hesitation with Hemophilia-Related Emergencies.* Emergency Medicine Reports. November 2, 1992. pp 165-176.

Furie, Bruce, Steven A Limentani and Cathy G. Rosenfield. *A Practical Guide to the Evaluation and Treatment of Hemophilia.* Blood. Volume 84, Number 1. 1994:pp3-9.

Goodnight, Scott and William Hathaway. Hemostasis and Thrombosis, A Clinical Guide. McGraw –Hill Publishing. New York.

Miller, Connie. *Inheritance of Hemophilia.* Publication of the national hemophilia Foundation. 1998.

RESOURCES

Local Hemophilia Treatment Center:	Mountain States Regional Hemophilia and Thrombosis Center 303-724-0724
National Hemophilia Foundation- HANDI	1-800-42-HANDI

WEB SITES

www.hemophilia.org
www.wfh.org
www.hemophiliagalaxy.org
www.hemophilia.net
www.hemophiliavillage.com
www.mmhc.com
www.web-depot.com/hemophilia