Emergency Care for Patients with Hemophilia

An instructional manual for Medical Professionals

Third Edition

Written by the Nursing Group of Hemophilia Region VI
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Determine if the patient has:
Hemophilia A or factor VIII (8) deficiency
Hemophilia B or factor IX (9) deficiency

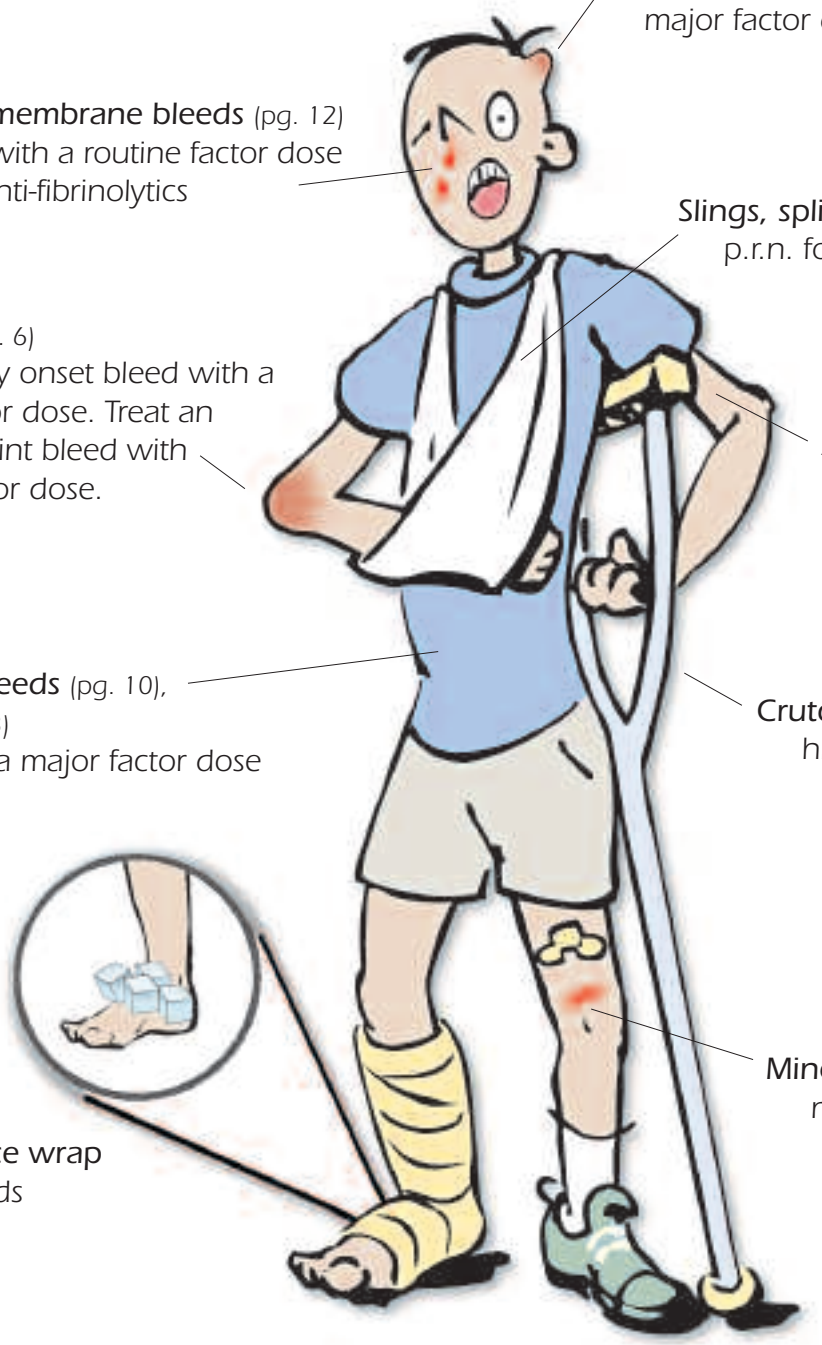
Head injury (pg. 4)
Always treat immediately with a major factor dose

Mucous membrane bleeds (pg. 12)
Treat with a routine factor dose and anti-fibrinolytics

Slings, splints, immobilizers
p.r.n. for joint bleeds

Joint bleeds (pg. 6)
Treat an early onset bleed with a routine factor dose. Treat an advanced joint bleed with a major factor dose.

Avoid intramuscular injections due to the possibility of causing a muscle bleed



Abdominal bleeds (pg. 10), **Trauma** (pg. 23)
Treat with a major factor dose

Crutches
hip, knee, ankle bleeds

Minor cuts / bruises
no treatment

Ice pack / Ace wrap
joint bleeds

TREATMENT FOR MINOR BLEEDS Minor bleeds include: Nose (epistaxis), Mouth (including gums) Joints (hemarthroses), Abrasions and superficial lacerations

Hemophilia A or Factor VIII (8) Severe to moderate deficiency:	Recombinant Factor VIII concentrate Dosage 20 - 30 units per kilogram
Hemophilia A or Factor VIII (8) Mild deficiency:	DDAVP responsive: DDAVP dosage: 0.3 micrograms per kilogram **(Maximim dose: 20 micrograms)** DDAVP non-responsive: Recombinant Factor VIII concentrate. Dosage 20 - 30 units per kilogram
Hemophilia B or Factor IX (9) deficiency Severe/Moderate/Mild:	Recombinant Factor IX concentrate Dosage 35 - 50 units per kilogram
For mucosal bleeds in all of the above add:	Tranexamic Acid (Cyklokapron) "Topical (mouthwash) or systemic" as per bleeding site (Contraindicated if hematuria)

TREATMENT FOR MAJOR BLEEDS/LIFE-THREATENING BLEEDS Head (intracranial) and Neck, Chest, Abdomen, Pelvis, Spine, Iliopsoas Muscle and Hip, Extremity muscle compartments, fractures or dislocations, any deep lacerations

Hemophilia A or Factor VIII (8) Severe/Moderate/Mild deficiency	Recombinant Factor VIII concentrate Dosage 40 - 50 units per kilogram
Hemophlia B or Factor IX (9) Severe/Moderate/Mild deficiency	Recombinant Factor IX concentrate Dosage 100 - 120 units per kilogram

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All major bleeding episodes should be considered potentially critical.
The goal is to raise the factor level to 80 - 100%.

Note: For patients with **Factor 8 and 9 inhibitors**, it is imperative that you contact the patient's hemophilia physician before any infusions. Contact information and numbers are located above.

Table of Contents

Hemophilia Treatment Centers

Purpose

This manual contributes to hemophilia care by enhancing the emergency department staff's understanding of hemophilia and its treatment. The goals of this manual are to:

- promote understanding of the complexities of hemophilia treatment
- provide a reference for the emergency center staff
- promote a consultative dialogue with the emergency department, hemophilia treatment center, and patient/family

Use

This manual provides a standardized format for evaluation and treatment of hemophilia emergencies. The content is segmented by systems and complications of hemophilia. Turn to an area of interest. The illustration on the left page provides information points for quick review. The text on the right page gives further detail of bleeding presentations, their possible complications and treatment.

It is suggested that the patient's hemophilia treatment center or hematologist be consulted for anything other than routine bleeding episodes.

To The Attending Medical Staff:

This manual is a guide for medical personnel who may be less familiar with hemophilia treatment. Its content consists of ***guidelines, recommendations and suggestions only***. The attending physician has the final responsibility for appropriate diagnosis and treatment.

Hemophilia basics

Definition

Hemophilia is a genetic disorder characterized by a deficiency or absence of one of the clotting proteins in plasma. The result is delayed clotting. Deficiencies of factor VIII (8) [Hemophilia A or Classic Hemophilia] and factor IX (9) [Hemophilia B or Christmas Disease] are the most common and referred to as hemophilia. Hemophilia mostly affects males due to the X-linked inheritance pattern.

Effects of hemophilia

Hemophilia prevents the formation of a firm, fibrin clot and results in a soft, unstable clot. Persons with hemophilia do not bleed faster than others; rather the bleeding is continuous. Significant blood loss can occur if treatment is delayed.

Incidence

The incidence worldwide is estimated to occur in 1:7,500 live male births; all races and ethnic groups are affected. Factor VIII (8) deficiency is four times more common than factor IX (9) deficiency but the clinical presentations and inheritance patterns are the same.

Severity

The amount of bleeding expected in an individual with hemophilia depends upon the severity of the deficiency. Normal plasma levels of factor VIII (8) and IX (9) range from 50-150%.

Those with less than 1% factor VIII (8) and IX (9) are considered to have **severe hemophilia**. Frequent bleeding episodes are common, particularly into joints. Bleeding can occur for no known reason or from trauma.

Persons with factor levels of 1-5% are considered to have **moderate hemophilia**. These persons may experience bleeding after minor trauma. After repeated bleeding into the same joint, persons with moderate hemophilia may experience bleeding in that joint with minor trauma.

Persons with more than 5% factor activity are considered to have **mild hemophilia** and bleed only after significant trauma or with surgery. Some carrier girls and women (called symptomatic carriers) can have lower than normal plasma levels of factor VIII (8) or IX (9) and thus may exhibit symptoms of mild hemophilia.

Serious bleeding sites

The six major sites of serious bleeding which threaten life, limb, or function are:

- **intracranial**
- **spinal cord**
- **throat**
- **intra-abdominal**
- **limb compartments**
- **ocular**

All of the above require immediate assessment and intervention, and are characterized by:

- **bleeding into an enclosed space**
- **compression of vital tissues**
- **potential loss of life, limb, or function**

Treatment

Treatment for bleeding involves replacing the deficient factor as the **first** course of action. This requires intravenous infusion of commercial factor concentrates. Specific doses, additional drugs and medical interventions depend upon the site and severity of bleeding. Once factor replacement therapy has been infused, diagnostic procedures and examinations can begin.

Family

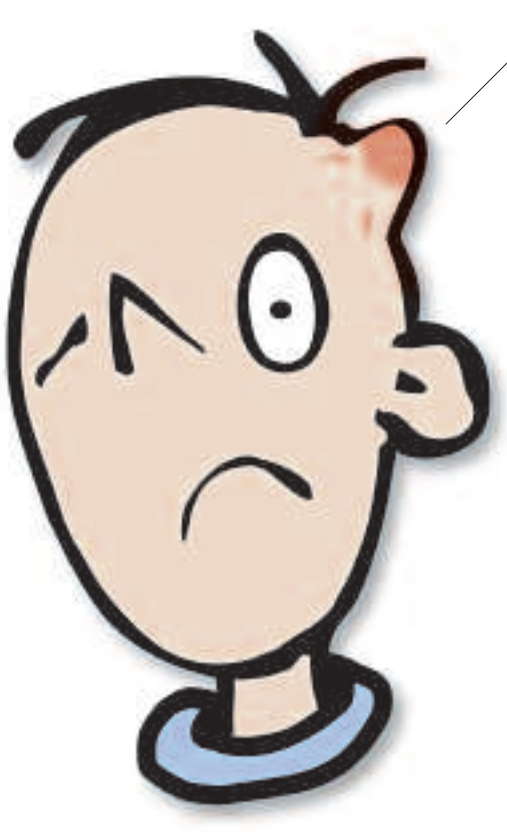
Parents and persons with hemophilia are knowledgeable about the management of the disorder and their input should be sought and heeded. Most hemophilia families are medically sophisticated and should not be dismissed as novices.

Interview the family about whether factor concentrate has been administered prior to arriving at the ED; if so, determine when and at what dose. Additional factor may be required, depending upon the time lag and severity of the bleed. Establish who the treating hematologist or hemophilia treatment center is and contact them for other than routine bleeding.

Head injury

Treat all head injuries with or without swelling.

First, infuse a major factor dose.*
Then perform diagnostic studies, such as CT scan, neuro exam.



Discharge Instructions

Call the hemophilia treatment center or the patient's hematologist for follow-up factor doses.*

Report any signs or symptoms to the hemophilia treatment center or the patient's hematologist.

Head injury instructions for a two week period (instead of the usual instructions for 24 - 48 hour period).

*Dose chart inside front cover

Intracranial hemorrhage (ICH) is the leading cause of death from bleeding in all age groups. Without early recognition and treatment, death or severe neurologic impairment can occur. ICH may be spontaneous, without history of injury. Early neurologic symptoms may not be evident due to the slow, oozing nature of hemophilia bleeding.

Treatment

All significant head trauma, with or without hematoma, must be treated promptly with the major dose of factor replacement* before any diagnostic tests.

Diagnostic imaging

Obtain an emergency CT scan to rule out ICH after the major factor dose* has been given. Notify the patient's hematologist or hemophilia treatment center of the ED admission and the diagnostic findings.

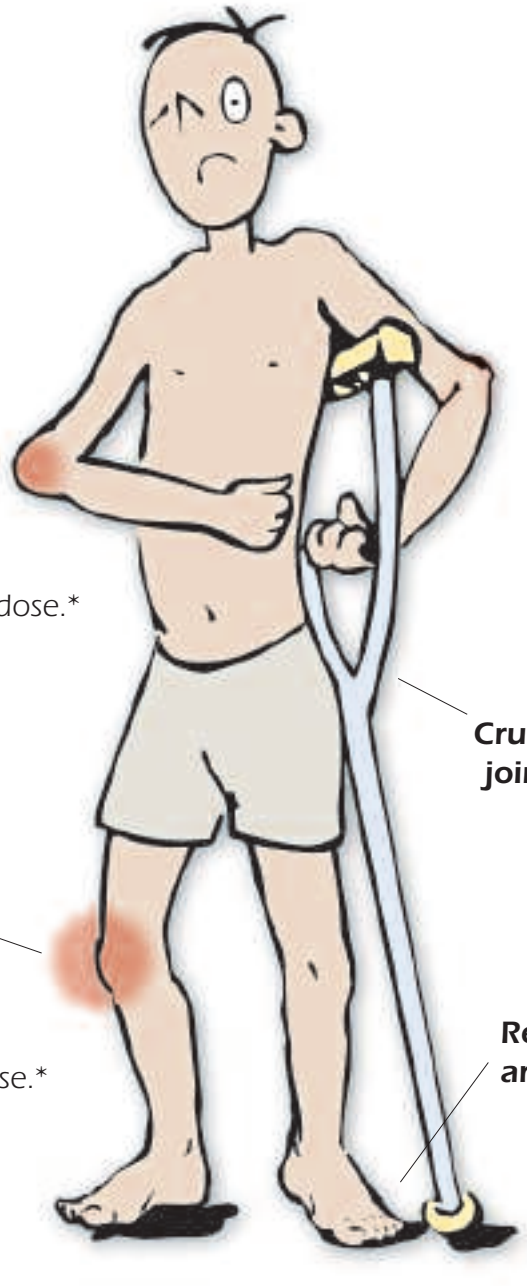
Possible admission

The patient should be admitted to the hospital for observation if he suffered a severe blow to the head or if he exhibits any neurologic symptoms such as headache with increased severity, irritability, vomiting, seizures, vision problems, focal neurologic deficits, stiff neck, or changes in level of consciousness. **Patients with a past history of ICH are at increased risk of repeated head bleeds.**

Instructions

If the patient is discharged home, instruct the family to monitor the patient for signs and symptoms of neurologic deterioration and report any abnormalities to the hematologist. Consult the hematologist for follow-up factor replacement doses if the patient is discharged home from the emergency department.

Joint bleeding



Advanced joint bleed
- heat
- pain
- swelling
- decreased range of motion

Treatment: a major factor dose.*

Crutches for weight bearing joints and crutch instructions.

Early onset joint bleed
- tingling
- pain
- limited range of motion

Treatment: a routine factor dose.*

Remember, toes and fingers are joints too.

Discharge Instructions

For the next 24 hours:

- RICE (rest, ice, compression [ace wraps], elevation)
- sling or splinting if support is needed (i.e. Aircast® for ankles)

Follow-up with the hemophilia treatment center or with the patient's hematologist.

**Dose chart inside front cover*

The hallmark of hemophilia is joint and muscle bleeding. Spontaneous joint and muscle bleeding can occur without a definite history of trauma. The patient may not be able to identify a specific event that resulted in bleeding.

While persons with hemophilia may bleed into any joint space, the joints which most frequently bleed are the ankles, knees, and elbows. Other possible bleeding joint sites include the shoulders and hips. As repeated bleeding occurs, the synovial tissue thickens and develops even more friable blood vessels. A vicious cycle of bleeding and rebleeding may set in and the affected joint is referred to as a "target joint." Eventually, repeated bleeding into joints leads to a form of chronic arthritis with destruction of cartilage and the eventual destruction of bone resulting in decreased joint mobility and function.

Signs and symptoms

Outward signs of joint bleeding include restriction of movement, swelling, heat, and erythema on and around the joint. The patient may report symptoms of a bubbling or tingling sensation with no physical signs. Later symptoms include a feeling of fullness within the joint and moderate to severe pain as the bleed worsens.

Treatment

Some patients may present for treatment with no other outward signs of bleeding than decreased range of motion and a complaint of pain or tingling. This is indicative of an **early onset joint bleed** and is the optimal time to treat. The patient should be infused as quickly as possible with a routine dose of factor* in order to minimize pain and joint destruction. Extreme pain, swelling, heat, and immobility are signs and symptoms of an **advanced joint bleed** which occurs only after blood has filled the joint space. Symptoms suggestive of an advanced joint bleed require a major factor dose.*

Infuse before any diagnostic procedures such as x-ray. If a joint bleed is treated early before obvious outward signs occur, then the need for expensive follow-up infusions may be lessened or avoided altogether. **Before dislocated joints are reduced, infuse with a major factor dose**.*

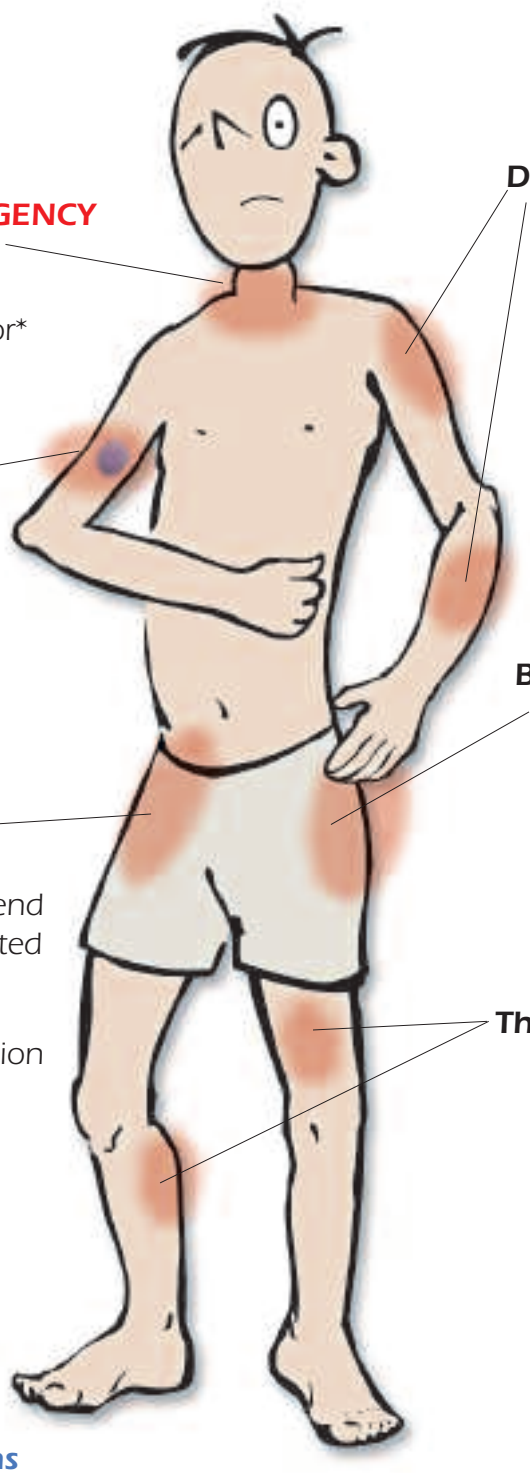
Joint aspiration: Caution!

Joint bleeds in hemophilia are generally not treated by aspirating the joint. This procedure can make matters worse by creating another site from which the patient may bleed. If joint aspiration is deemed necessary, such as in suspected joint sepsis, then the joint should be aspirated by an orthopedic surgeon associated with a hemophilia treatment center. The patient's hematologist must be involved in order to arrange for factor coverage before and after the procedure.

Discharge and follow-up care

Upon discharge, the hemophilia patient with a joint bleed should be instructed to keep the affected joint at rest, elevate the affected limb, and apply ice packs. Additional support to the affected joint may be applied by wrapping with an ace bandage. For ankle bleeds, an Aircast® may be a useful splinting device. Crutches are useful to help individuals when they have lower extremity joint bleeds and need to be non-weight bearing. Follow-up should be made to the local hemophilia treatment center or to the patient's hematologist as soon as possible.

Muscle / soft tissue bleeding



Neck swelling: EMERGENCY
- potential airway compromise
- major dose of factor*

Deltoid / forearm bleed
- routine factor dose*
- major factor dose* if a compartment syndrome is suspected

Soft tissue bleeds and bruising
- no functional impairment
- tenderness, but no severe pain
- no factor needed

Buttock bleeds
- pain
- with/without swelling
- routine dose of factor*
- major dose of factor* if the leg on the affected side exhibits tingling or swelling

Iliopsoas bleeds
- flexed hip
- pain/inability to extend the leg on the affected side
- major factor dose* and hospital admission

Thigh/calf bleed
- pain
- with/without swelling
- impaired mobility
- routine factor dose*
- major factor dose* if a compartment syndrome is suspected

Discharge Instructions

- rest
- ice
- non-weight bearing
- follow-up with the hemophilia treatment center or the patient's hematologist

**Dose chart inside front cover*

Signs, symptoms, and sites

Muscle bleeding is common in persons with hemophilia. Any muscle group may be subject to bleeding. Muscles that exhibit warmth, pain, and swelling should be treated with a routine dose of factor.* Common bleeding sites include the upper arm, forearm, thigh, and calf muscles. Toddlers frequently have buttock and groin bleeds. Abdominal wall muscles and iliopsoas muscles are also common bleeding sites. These abdominal muscles generally do not have observable swelling, yet they may hold a large amount of blood. Patients who complain of low abdomen or groin pain, especially with signs of nerve compression, are probably experiencing an iliopsoas bleed. These patients should receive an emergency hematology consult and possible admission for observation and several major doses of factor coverage.* The hallmark sign of iliopsoas bleeding is spontaneous flexion of the leg on the affected side with an inability to extend the leg without pain.

Consequences of muscle bleeds

Muscle bleeds can result in serious consequences if not treated promptly. Extensive blood loss may occur in large muscle groups. Muscle bleeding places pressure on nerves and blood vessels and, if left untreated, these bleeds may result in permanent disabilities such as foot drop and Volkmann's contracture. It is important that the patient's hematologist be consulted before any surgical consults. Most muscle bleeds respond well to medical management and do not require fasciotomy. Such an extreme measure will usually generate problems for the patient and require a tremendous amount of post-surgical factor infusions, more so than if the patient was treated medically.

Treatment and follow-up care

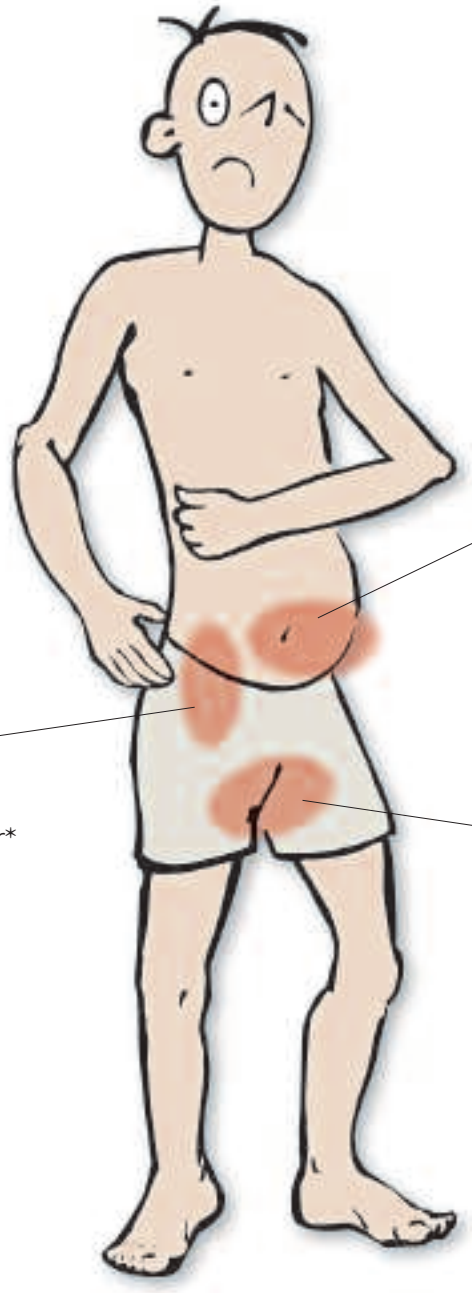
Most muscle bleeds are treated with a routine dose of factor.* Large abdominal muscle groups and iliopsoas bleeds should be treated with a major dose of factor.* Rest and ice packs are also helpful. The patient should be referred back to his hemophilia treatment center or to his hematologist for follow-up as soon as possible. **If any suspicion of compartment syndrome and nerve compression exists, then the patient should have an emergency hematology consult and should be admitted to the hospital.**

Soft tissue and superficial bleeds

Soft tissue bleeds usually do not require aggressive treatment. Superficial hematomas and bruises may appear anywhere on the body and, if they do not threaten function and mobility, they do not need to be treated.

Gastrointestinal / urinary tract bleeding

Nausea and vomiting may indicate intracranial hemorrhage as well as gastrointestinal problems.



Abdominal pain

Treat immediately with a major dose of factor* for:

- flank pain
- melena
- vomiting blood

Iliopsoas bleeding

- flexed hip
- pain on extension
- major dose of factor*

Hematuria

- bed rest for 24 hours
- force fluids
- consult the hemophilia treatment center or the patient's hematologist

Discharge Instructions

- force fluids for hematuria
- rest
- no weight lifting
- report any symptoms
- follow-up with the hemophilia treatment center or the patient's hematologist

 **Dose chart inside front cover*

Initial presentation

Acute abdominal pain in a patient with hemophilia may have many origins, such as GI tract hematomas (both spontaneous or trauma induced), pseudotumors, iliopsoas or retroperitoneal bleeding. Bleeding may also occur with hemorrhoids or the passage of kidney stones. Notify the hemophilia treatment center or the patient's hematologist.

Patients who present to the emergency department with abdominal or flank pain, melena or hematemesis should be triaged for immediate examination and given factor replacement therapy at the major dosage.* After factor therapy, then diagnostic x-rays, scans and endoscopy procedures can be carried out.

Abdominal trauma and benign events such as forceful coughing or vomiting can precipitate an abdominal bleed. Blood loss can be significant before outward signs and symptoms appear. Infants can have bleeds with gastroenteritis, intussusception or Meckel's Diverticulum.

A history of lifting heavy objects, weight lifting, falling on bicycle handlebars or stretching the groin can precipitate abdominal wall, iliopsoas (see pages 8 and 9), or retroperitoneal bleeding.

Symptoms

Symptoms of abdominal muscle bleeding (rectus, pectorals, latissimus, obliques) are a palpable mass, rigidity, and pain. Concurrent bleeding in the abdominal cavity may be present and go unnoticed for days with a steadily dropping hemoglobin. Rupture of the liver, spleen, or pancreas should be considered when the hemoglobin falls dramatically following trauma.

For nausea and vomiting without an obvious cause, consider that these may be symptoms of intracranial bleeding. Inquire about head injury, mental status changes, and other neurologic signs and symptoms, and consider CT scan of the head.

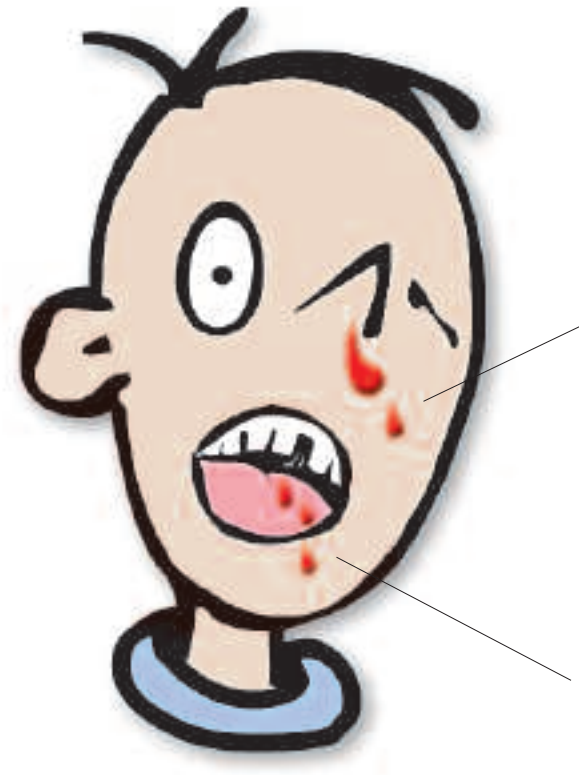
Genitourinary bleeding

Hematuria is often frightening to the patient but not a serious event. Instruct the patient to remain at bed rest and force fluids the next 24 hours. Protracted hematuria may require a routine dose of factor coverage.* **Anti-fibrinolytics are contraindicated with hematuria.** Contact the hematologist.

Scrotal bleeding may occur after trauma, especially in toddlers. Infuse with a routine factor dose* and have the family contact the patient's hematologist for follow-up care.

Mucous membrane bleeding

A Dental or E.N.T. consult may be needed.



Nose bleeds may respond to other measures. Refer to "Controlling Epistaxis" table on page 14.

Mouth bleeds (gum, tooth, frenulum or tongue laceration) need factor* and anti-fibrinolytics. Refer to the "Anti-fibrinolytics" table on page 15.

Anti-fibrinolytics are not routinely available from all pharmacies. Pharmacies associated with the hemophilia treatment centers will be able to fill a prescription.

Discharge Instructions

Patients should follow-up with their hemophilia treatment center or hematologist the next day.

Instruct the patient on: Controlling Epistaxis, Anti-fibrinolytics, Diet Modification tables (pages 14 - 15) as needed.

Mucous membrane bleeding may require medical care in the emergency department. Patients should receive factor who:

- are experiencing profuse and/or prolonged bleeding
- have sustained a known injury to the mouth, tongue, or nose
- have severe swelling in the mouth or naso-pharynx area
- are experiencing respiratory distress
- have difficulty swallowing

The patient may not know the reason for the symptom or bleeding. It may have been caused by trauma, infection, or the bleed may be spontaneous. If airway blockage is suspected, the patient should immediately receive a major dose of factor.* After the factor level has been raised, further interventions including invasive procedures may be done.

Do not make a person with hemophilia wait for factor replacement. The longer he waits, the more bleeding takes place. If the bleed is in a closed space, the accumulation of blood will cause surrounding tissue damage, airway obstruction, and enhance pain.

Epistaxis

A person with epistaxis who is unable to control the bleed himself may need a routine dose of factor* and anti-fibrinolytic treatment (Amicar). Be sure the person knows how to control and stop the bleeding. (See table on page 14.)

Oral Cavity

Bleeding in the mouth can be hard to control. The patient will probably need factor. A single infusion of a routine dose of factor* may temporarily stop the bleeding, but clots break down normally on days 3-5 and bleeding may start again at that time. An anti-fibrinolytic may be indicated to maintain hemostasis. (See table on page 15.) Anti-fibrinolytics may be available through the patient's home health company for next day delivery. A modified diet should be started at the same time as factor therapy. (See table on page 15.)

Bleeding may occur with erupting or exfoliating teeth. It is more common with exfoliating teeth, especially a tooth that is very loose. A dental consult may be needed to extract the tooth since it will continue to lacerate the tooth socket as long as it is in place. A major or routine dose of factor* should be given prior to extraction. A frenulum or tongue laceration will require a major or routine dose of factor.*

Retropharyngeal

After the major dose of factor* is given, further observation, x-rays and admission may be required.

Controlling Epistaxis
Instruct the patient:

1. To gently blow his nose to remove mucous and unstable clots which will interfere with hemostasis.
2. Tilt his head forward so any blood will come out the nares and not down the back of the throat.
3. Apply firm pressure to the entire side of the nose that is bleeding for 15 minutes.
4. Release the pressure to see if bleeding has stopped, blow out any soft clots.
5. If the bleeding continues, reapply pressure for another five minutes.
6. Factor replacement* at a routine dose and/or anti-fibrinolytic agents (see next page) may be needed.
7. During active bleeding, or when the bleeding has stopped, spray or apply two drops of oxymetazoline (ex. NeoSynephrine®, Dristan®, or Afrin®) nasal spray/drops to the side that was bleeding. These can be used at home PRN for epistaxis.
8. Instruct the patient to use olive oil in the nares to keep the membranes soft and moist, and prevent the formation of hard crusts which might crack and restart bleeding.
9. An ENT consult may be required for possible cauterization of a vessel.

 **Dose chart inside front cover*

Anti-Fibrinolytics

Anti-fibrinolytics may also be indicated in nasal or oral bleeding. Amicar and Cyklokapron are both anti-fibrinolytic agents. Either may be prescribed for mucous membrane bleeding to promote clot adhesion in conjunction with factor replacement at a routine dose.* In some cases they may be prescribed without factor replacement.

Amicar - aminocaproic acid
Recommended dosage:
child: oral dose 50-100 mg/kg (not to exceed 4 Gms) every 6 hours for 3 - 10 days
adult: oral dose 3-4 Gms every 6 hours for 3 -10 days

Cyklokapron - tranexamic acid (may not be available in the USA)
Recommended dosage:
child and adult: oral dose 25 mg/kg every 8 hours for 3 - 8 days

These medications must be given around the clock to keep blood levels constant.

These medications may be available through the family's home health company, the hemophilia treatment center, or the family may have a supply at home. They are difficult to obtain from most pharmacies.

Follow-up through the hemophilia treatment center or patient's hematologist.

Topical agents such as Topical Thrombin and Gelfoam may also be used to help control mucous membrane bleeding.

Antibiotics and pain medications may also be indicated.

Diet Modifications

Directions for the patient:

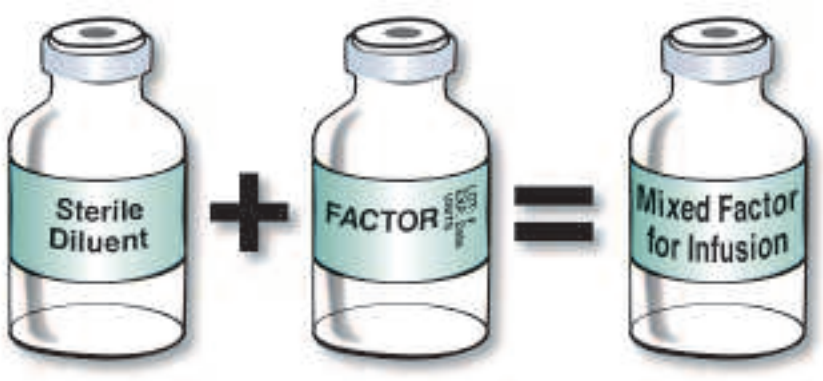
1. Diet should be restricted to soft, cool, or lukewarm foods until the area is fully healed. Suggested foods: Jello, noncarbonated drinks, sherbert, lukewarm soups (not cream soups), baby foods, blenderized or pureed foods, spaghetti.

2. Avoid milk products and foods made with milk. Milk products may contribute to clot breakdown and may also cause nausea and vomiting if the patient has swallowed blood.

3. Avoid using a straw. Negative pressure from the sucking action can dislodge the clot and aggravate the bleeding site.

4. Avoid hard foods like chips, popcorn, tacos, etc.

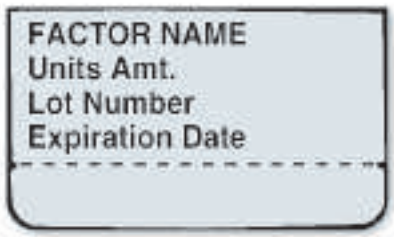
Factor Reconstitution



Examples: Antihemophilic factor (AHF) or factor VIII (8)*
plasma derived or recombinant

Coagulation factor IX (9)*
plasma derived or recombinant

Factor box top



Mixing instructions and the rate of administration are found on the drug insert. *Document the lot number(s), expiration date(s), factor concentrate trade name and total number of units infused.*

Patients may request the box top for their records.

Some patients are instructed to bring unmixed factor concentrate with them to the ED to minimize treatment delay and cost. Occasionally, patients will bring prepared factor concentrate after unsuccessful home venipuncture attempts. Please assist with venipuncture and allow the patient or family to infuse the prepared factor concentrate if less than 3 hours have elapsed since reconstitution.

 **Dose chart inside front cover*

Dosage

Each bottle of factor concentrate is labeled with the activity expressed as International Units (IU, example: 287 IU). The dosage to be administered is based on the patient's body weight in kilograms (kg)*.

IMPORTANT!

These are examples. Please refer to the inside front cover for specific dose ranges set by your local hemophilia treatment center.

Factor VIII (8) is calculated using the formula:
1 IU/kg = 2% rise in Factor VIII (8) activity
50 IU/kg = 100% correction (example)

Factor IX (9) is calculated using the formula:
1 IU/kg = 1% rise in Factor IX (9) activity
80 IU/kg = 80% correction (example)

The **ENTIRE** contents of all the vials reconstituted for an infusion should be used, even if it exceeds the calculated dosage. A larger dose will only prolong the period of normal coagulation. Due to its expense, **factor concentrate should never be discarded**!

The half-life of factor VIII (8) is 8-12 hours; the half-life of factor IX (9) is 18-24 hours.

Factor types

Recombinant

Refers to genetically engineered Factor VIII (8), IX (9) or VII (7) concentrates which are not derived from plasma.

Plasma derived product

Factor VIII (8) or IX (9) concentrates of high specific activity and purity of the specific clotting factor achieved through the use of an affinity column matrix. Some contain von Willebrand factor and are used to treat von Willebrand Disease.

Activated prothrombin complex concentrates - plasma product

Prothrombin complex concentrates purposely "activated" so they contain some Factor IX (9), Factor X (10), etc. in the active form for use with Factor VIII (8) inhibitor patients only. The brand is FEIBA. Dose is 75 IU/kg.

Factor IX (9) prothrombin complex concentrates (PCCs) - plasma product

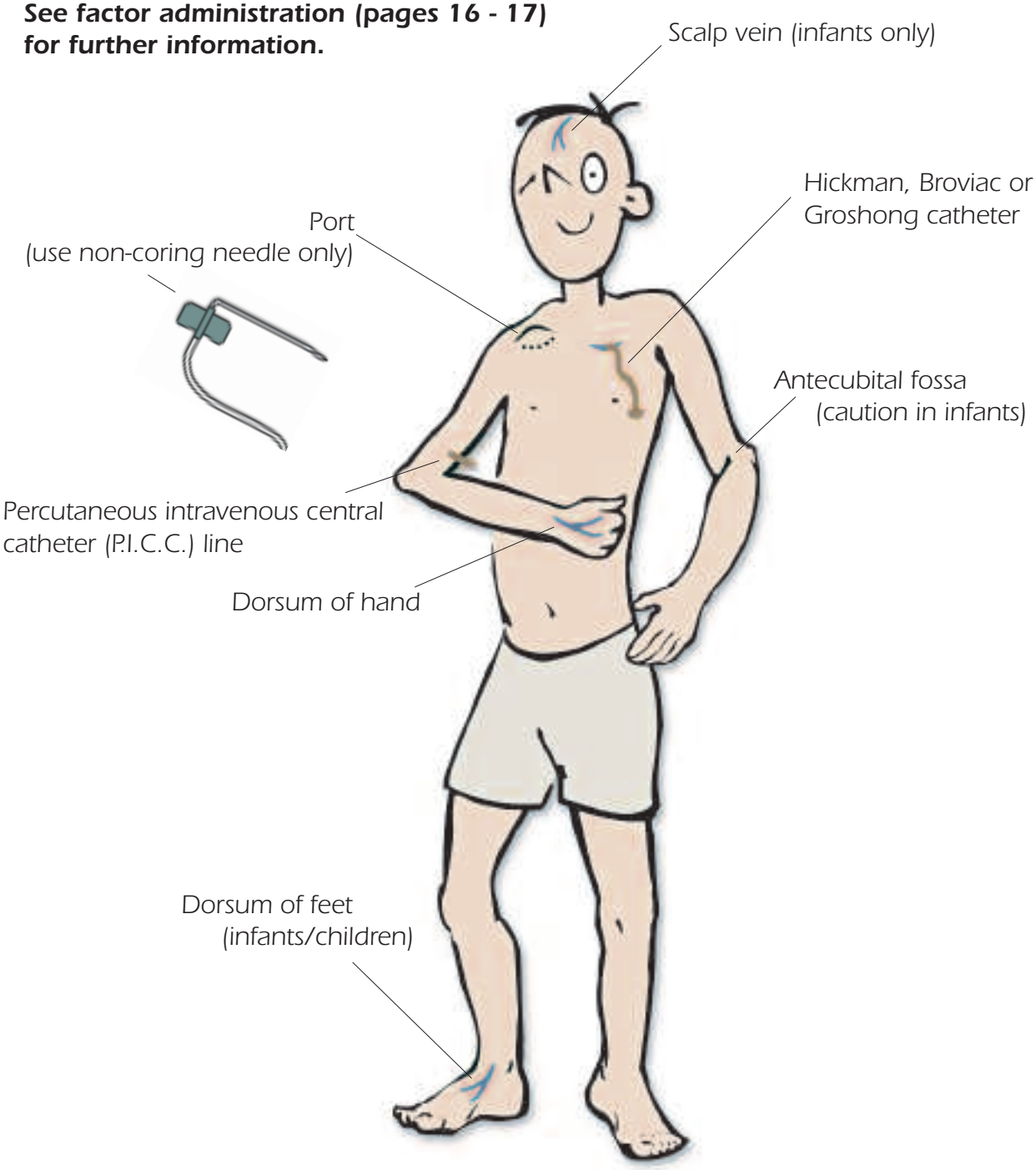
Also contains some other clotting factors and small amounts of activated coagulation factors. Used to treat Hemophilia B and some patients with Factor VIII or IX inhibitors.

Do not use the affected or injured limb for venous access.

No jugular or femoral sticks except in life-saving situations.

Use 25g or 23g "winged" needles. After the needlestick, apply pressure and bandage.

See factor administration (pages 16 - 17) for further information.



Intravenous access for a person with hemophilia is basically the same as for any patient. The use of 23 gauge or 25 gauge "winged" needles is preferable, especially for children.

Sites for access

Sites to consider for a peripheral IV include:

- dorsum of the hands
- antecubital fossa (caution in infants due to the risk of compartment syndrome)
- dorsum of feet (infants and children)
- scalp veins (infants only)

Groin and neck veins are contraindicated except in life-threatening situations.

The patient's IV ***should not*** be started in the affected limb. The injured area should be minimally manipulated, if at all.

Venous access device

Some venous access devices currently used in hemophilia care include:

Port or peripheral port - access with non-coring needle as per your institution's procedure
External central catheter - Hickman, Broviac, or Groshong; access per your institution's procedure
P.I.C.C. line - access per your institution's procedure

Heparin flush

It is recommended to do a final flush with heparin for any venous access device (except for Groshong's which are only flushed with normal saline). Check with the parent or the patient's institution for the amount of heparin flush to use. If these options are not available, use your institution's procedure for the amount and concentration of heparin. This small amount of heparin will not harm the patient (remember - you've just given him factor). The access device needs to stay patent; this is accomplished with the heparin.

The patient on "prophylaxis"

Some patients with hemophilia now receive "prophylactic" doses of factor replacement on an every-other-day or 2-3 times a week routine schedule. This should be asked of the patient. These patients usually have venous access devices for infusions. So, always check to see if the patient has a venous access device before starting a *peripheral* infusion.

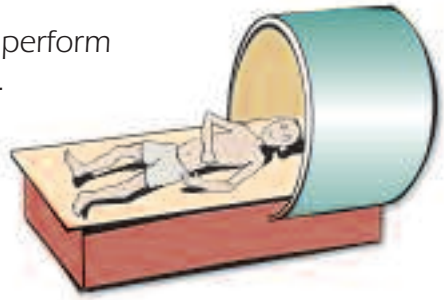
X-rays and lab are not indicated for a joint or muscle bleed.

Head injury

(see page 5)

First give a major dose of factor* . . .

. . . then perform a CT scan.



Fracture

First give a major dose of factor* . . .

. . . then apply the cast.

Discharge Instructions

Patient should follow-up with the hemophilia treatment center or his hematologist the next day.

Head injury: Discharge with routine post head injury instructions (patient to follow-up for two weeks instead of 48 hours).

Sutures: Remind the patient he will need factor for suture removal.

 **Dose chart inside front cover*

In general, patients with hemophilia who are experiencing an acute bleeding episode need factor only. Other procedures should not be done unless there is another clinical indication for the study. In any situation, the infusion of *factor should never be delayed* if any bleeding is suspected. Delaying the infusion simply increases bleeding that will result in greater morbidity.

Laboratory studies

If the only complaint is an acute joint or muscle bleed, no laboratory studies are necessary. If GI, abdominal, large muscle, or oral cavity bleeding is suspected and has potentially been extensive, a CBC may be indicated to determine if the individual is anemic. Factor levels and inhibitor levels are not necessary for treatment in an acute emergency setting. ***Factor should not be delayed for laboratory studies to be drawn or completed.***

X-rays and other radiological studies

Give factor first, then decide if a radiological study is indicated. Remember that a swollen joint or extremity is usually the result of internal bleeding, not a fracture. X-rays of the joint can be used to document a joint bleed, but are generally not useful in detecting early onset bleeds (and that is when treatment is optimal). The patient will be aware of joint bleeding before radiological changes are evident.

A CT of the head (see page 5) is necessary when dealing with a potential intracranial hemorrhage. It can document location and extent of bleeding and help direct further treatment. First give a major factor dose.*

Fractures

Give a major dose of factor* replacement, then x-ray and set the bone.

Lacerations and sutures

Sutures and staples should be used as on any other patient. If the laceration is significant enough to require sutures, the patient should first receive a routine dose of factor* then the procedure. Contact the patient's hematologist for follow-up factor infusion instructions. For removal of sutures, a routine dose of factor* is usually needed.

Invasive procedures

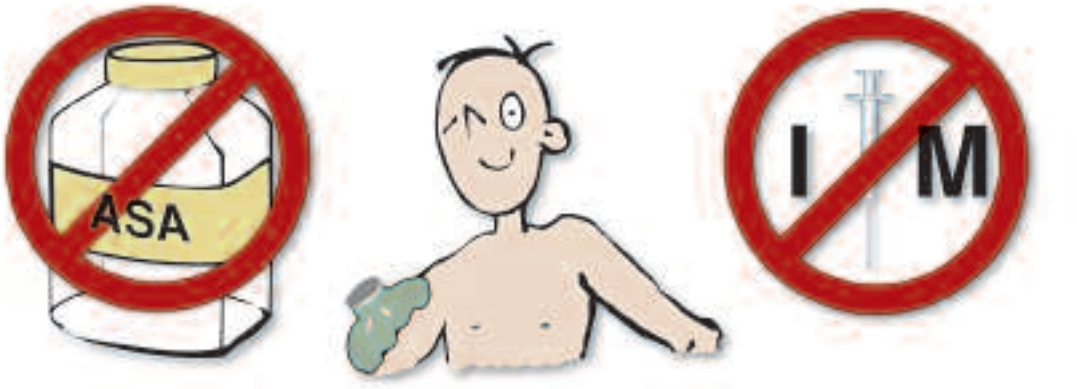
Invasive procedures should be performed as clinically indicated, i.e., lumbar puncture with symptoms of meningitis. However, a major dose of factor* should be given ***before*** the procedure begins.

Arterial sticks and venipunctures

Do not do arterial sticks unless no other option is available. If an arterial stick must be done, then a major factor dose* ***must*** be given first.

Venipuncture (see page 19) may be done at any location; hands are generally excellent and no factor treatment is necessary. Avoid "digging" for deep veins. Apply pressure for several minutes after the puncture.

Other medications



Pressure and ice pack after injections

Routine medications

Patients with hemophilia can receive routine medications (e.g. pain medications, antibiotics, etc.) that do not interfere with clotting function. Avoid non-steroidal anti-inflammatories (NSAIDS), ASA and any product with aspirin-related ingredients (e.g. Pepto-Bismol, Excedrin, Percodan).

Medications for fever or pain

Acetaminophen can be given for fever or pain. Narcotics/opioids are often recommended to control the pain experienced by a patient with hemophilia. ***Avoid giving intramuscular injections of antibiotics, pain medications, or immunizations because of the possibility of causing a muscle bleed.***

Routes of administration

Medications which can be given PO, SC, or IV are preferred. Routine immunizations and tetanus toxoid ***may be given subcutaneously***. If the rabies vaccination series is needed, an experienced hematologist (preferably the patient's) should be contacted to arrange factor infusions prior to and after the injections in order to prevent internal bleeding.

For any needle stick, pressure and an ice pack afterward will minimize soft tissue or muscle bleeding.

Caution

Some patients with hemophilia may have liver disease from hepatitis. Use caution when prescribing drugs that may cause liver toxicity. Other patients may be on other therapies for hemophilia-related complications such as HIV or hepatitis. Be aware of potential serious drug interactions.

Trauma / emergencies

Many different emergencies/traumas may occur to persons with hemophilia, just as to others. The more common are:

- Animal bites
- Burns
- Falls
- Fractures (see page 21)
- Gunshot wounds
- Motor vehicle accidents
- Myocardial infarctions
- Ocular injuries
- Puncture wounds
- Dislocated joints

Treatment

A major dose of factor* should be infused as soon as possible (before any test, x-rays, debriding, sutures, etc.).

Contact the hemophilia treatment center or the patient's hematologist due to the complexity of managing inhibitors.

High Responding Inhibitors

Products used to treat bleeding:

Activated PCC
e.g. FEIBA

Factor IX complex concentrates (PCC)
Used for Factor VIII (8) or IX (9) patient e.g. Profilnine, Bebulin

Recombinant F VIIa (7) (activated)
e.g. NovoSeven

Low Responding Inhibitors

Factor product and dose is variable.
Dose may be 2-3x routine range.*

 **Dose chart inside front cover*

Definition

An inhibitor is an antibody that some individuals with hemophilia develop against factor VIII (8) or IX (9). These antibodies neutralize the factor procoagulant activity, thus counteracting the desired effect of an infusion of factor concentrate.

How inhibitors are measured

Labs in the U.S. express the presence of an inhibitor in terms of Bethesda units (BU). One Bethesda unit is the amount of antibody that destroys half of the factor VIII in an equal mixture of normal and patient plasma in two hours.

Low responding inhibitor

Measures less then 10 BU.

High responding inhibitor

Measures greater than 10 BU. An infusion of factor concentrate further stimulates the inhibitor antibodies, causing a rise in BUs.

When to suspect an inhibitor

Suspect an inhibitor if bleeding doesn't stop after several infusions of factor concentrates.

Call the patient's hematologist if the inhibitor is known or suspected before attempting treatment. Be sure to also ask the patient and family if they have been told the patient has an inhibitor.

Treatment of inhibitors

Inhibitor management is difficult for the experienced hematologist. Contact the patient's hematologist or hemophilia treatment center when these patients present in the emergency department for treatment. Bleeding in an inhibitor patient can quickly lead to serious life- or limb-threatening complications without expert management.

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