Emergency Room Care

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While all acute bleeding episodes are considered serious and emergent, there are five major sites of serious bleeding in hemophilia that threaten life, limb, or function. They are intracranial or spinal cord bleeds, throat bleeds, intra-abdominal bleeds, limb compartment syndromes (e.g. thigh, calf, forearm, upper arm) and ocular bleeds. All of these areas are characterized by (1) bleeding into an enclosed space, (2) compression of vital tissue, and (3) potential loss of life, limb, or function. Since bleeding episodes also cause anxiety and fear in patients and families, the nursing interventions of communication, reassurance, education, and support will help them through these crises.

TREATMENT REGIMENS IN HEMOPHILIA

Hemophilia is managed by either on-demand therapy or prophylaxis. In on-demand therapy, factor concentrate infusion therapy is initiated at the onset of a bleeding episode. In prophylaxis therapy, factor concentrate is infused on a regular basis to prevent bleeding episodes from occurring. Prophylaxis is now the mainstay of treatment regimen for children with severe hemophilia.

With the introduction of safe recombinant factor concentrates in the mid-1990's, children with severe hemophilia were increasingly placed on prophylactic regimens. These same children are now adults, and many choose to stay on prophylaxis if the resources are available to them. Many older adults are also opting for a routine prophylaxis regimen to decrease bleeding episodes, even though they may have joint damage from years of hemophilia-related bleeding (See Chapter 6).

Individuals with hemophilia treated with factor concentrates prior to 1985 may be infected with HIV and/or hepatitis B or C. These infections and the various therapeutic agents used in their treatment may contribute to the severity of bleeding episodes (e.g., individuals taking protease inhibitor drugs may experience an increase in bleeding episodes or prolonged bleeding). Medications used to control chronic pain from arthritic joints also need to be considered in assessing and treating acute bleeds (e.g., individuals taking non-steroidal anti-inflammatory drugs may experience prolonged bleeding episodes).

FUNDAMENTAL PRINCIPLES OF CARE FOR ANY LIFE- OR LIMB-THREATENING BLEED

1. Obtain history of the event.
2. Obtain history of hemophilia including type, severity, inhibitor status, last factor replacement time and dose, current medications, and associated medical history (e.g., HIV, hepatitis B or C infection).

3. Take primary survey and assess injury or bleeding site.
4. Infuse factor replacement prior to conducting further diagnostic studies.

TREATING EMERGENT BLEEDS

When calculating factor doses and timing for the treatment of life- and limb-threatening bleeds, the healthcare provider must take into consideration the patient's baseline factor level, routine treatment plan, access route, and time of last dose. Normal factor levels range from 50% to 150%; severe hemophilia level is less than 1%, moderate hemophilia level is up to 5%, and mild hemophilia levels are 6-49%. The individual's factor level may vary from normal to mild, as opposed to baseline severe, depending on the time from the last factor infusion. Patients on prophylactic regimens should receive a replacement dose of factor unless they received a 100% concentrate dose in the last three to four hours prior to the trauma or bleeding episode.

Inhibitor patients who use prothrombin complex concentrates (PCC) or activated prothrombin complex concentrates (aPCC) and have had a recent infusion (within three to four hours) should have careful consideration in the determination of their factor replacement needs. Prothrombin complex and activated prothrombin complex concentrates contain factors II, VII, IX, and X. The half-life of each factor differs, ranging from 2 hours for factor VII to 72 hours for factor II. Frequent and repeated doses of these concentrates allow accumulation of the longer-lived factors, predisposing the patient to a thromboembolic state. Disseminated intravascular coagulation and intramural cardiac thrombus have been reported as a result of too frequent or too rapid administration of these concentrates. (17) Thombotic events may also be precipitated with the concurrent use of PCC or APCC's and recombinant factor VIIa. Concurrent systemic use of Amicar, which inhibits fibrinolysis, may also predispose the patient to thrombus formation. (23)

CENTRAL NERVOUS SYSTEM BLEEDS

Intracranial or spinal cord bleeding can occur spontaneously or as the result of trauma in individuals with bleeding disorders. Intracranial hemorrhage remains a leading cause of death among patients with hemophilia. Such hemorrhages can occur in newborns, children, and adults of all ages. Common bleeding sites are subdural, epidural, subarachnoid, intra-ventricular, and intra-cerebral spaces. Brainstem or multiple site bleeds are less frequent. Patients with central nervous system bleeds do not always show external symptoms, making these cases potentially difficult to evaluate. When signs such as a scalp laceration or hematoma are present, the patient should be treated immediately with a major dose (100% dose) of factor replacement therapy, undergo a CT scan of the head, and be evaluated and observed in the hospital (4). The provider should not rely solely on bruises or signs and symptoms of central nervous system (CNS) bleeding, which may only develop hours or even days after the initial trauma. The trauma history should trigger the initial treatment.

PATHOPHYSIOLOGY

The brain and spinal cord are encased in bone with little room for expansion in the brain, spinal cord, or meningeal membranes once bleeding begins. Compression of the brain tissue, nerves, or spinal cord occurs quickly with beginning signs of CNS irritation. The bleeding site will determine the clinical symptoms of the patient. Presenting signs of the intracranial bleed vary

from mild to severe and may also be confused with other problems such as viral illness. Such symptoms include nausea, irritability, or headache and indicate increasing intracranial pressure.

Symptom-free intervals may occur in subarachnoid, epidural, and intracerebral hemorrhages and may be delayed 24 hours to one week after the trauma. Later symptoms of CNS bleeding include stiff neck or back, confusion, changes in mentation, slurred or confused speech, ataxia, blurred or double vision, nystagmus, unequal pupil size, muscle weakness or paralysis, parasthesia or numbness, and seizures. These clinical presentations indicate a significant accumulation of blood within the skull, with resulting brain compression and damage.

Intraspinal hemorrhage is less frequent than intracranial bleeding. One review showed trauma as a causative factor in only one third of the cases; 75% of hematomas were extramedullary, and 25% were intramedullary. (4) A history of trauma followed by progressive pain and neurologic changes, such as parasthesia or numbness in a limb or motor weakness, leads to a diagnosis of intraspinal hemorrhage. Symptoms of acute neck or back pain or trauma should also suggest this diagnosis. Treatment with factor concentrate should begin immediately, with neurological consultation and follow-up care. Prompt treatment will prevent paralysis and neurological sequelae as well as an extensive rehabilitation period. Factor administration should occur as soon as the diagnosis is suspected, before a diagnostic workup and prior to any invasive procedure, such as lumbar puncture. Any traumatic injury which could potentially precipitate an intraspinal hemorrhage should be treated with a major dose of factor, even if the patient is not symptomatic.

EVALUATION AND TREATMENT

Hemophilia patients presenting with a history of significant head trauma or sustained headache greater than four hours (3, 22) should receive an immediate infusion of factor concentrate to raise the factor level to at least 100% (i.e., 50-60 IU/kg FVIII concentrate or 100-110 IU/kg FIX concentrate). A complete neurological evaluation should be performed as a baseline and be followed with regular reassessment and monitoring for signs of increased intracranial pressure.

Laboratory evaluation: If stat labs are available, blood should be drawn for CBC and platelet count. If the patient has known or suspected liver disease, a prothrombin time (PT) should be ordered. If the timing and the amount of the patient's last infusion is known, a factor level assay may be useful in providing infused factor recovery information. A factor inhibitor screen will provide baseline inhibitor information, especially if the patient faces large and prolonged exposure to factor concentrate. These samples should be obtained prior to administering factor concentrate. However, these results will not be immediately available; treatment should not be delayed waiting for these results.

Treatment: Administration of factor concentrate should not be delayed or postponed in order to obtain lab work when there is potential brain hemorrhage. CT scans or MRI should be obtained. Airway maintenance, IV access, drug therapy, and cervical stabilization (if indicated by diagnosis) should be initiated for patients presenting with any neurological deficit. Factor replacement to 100% should always be given as the first intervention; it should be given empirically, prior to obtaining lab testing or diagnostic imaging.

Nursing Care

I. Primary Assessment
   A. Obtain history
      1. Time of onset of symptoms
      2. Presentation: headache, seizures, etc. Were they spontaneous or the result of trauma?
      3. Precipitating symptoms: trauma, loss of consciousness, trauma with symptoms.
      4. Baseline factor level, time and dose of last infusion, inhibitor history, aspirin (ASA) or non-steroidal anti-inflammatory drug (NSAID) use, current medications, and allergies, including factor concentrate allergy history.
   B. Observe patient
      1. Determine level of consciousness. This is a quick screening survey.
      2. Observe airway and circulatory states.
      3. Obtain vital signs.
   C. Initiate Treatment
      1. Secure patent airway.
      2. Obtain IV access and blood for lab studies.
      3. Infuse with factor concentrate to raise factor level to 100%.
   D. Obtain hematology and neurology consults
   E. Obtain further evaluation, such as x-rays and CT scans.

II. Secondary Assessment
   A. Neurological Assessment
      1. Conscious: awake, aware, and ability to interact.
      2. Orientation: person, place, and time.
      3. Language: rhythm and clarity of speech, word formation, and sentence structure.
      4. Memory: short term since may be amnesic with a bleed, and long term.
      5. Attention span: alert or in need of stimulation.
   B. Physical exam
      1. Look for evidence of head injury: hematoma, ecchymosis, frank bleeding, swelling.
      2. Examine the rest of the body: look for hematomas, ecchymosis, or other evidence of bleeding or trauma.
      3. Assess skin integrity and warmth.
      4. Check neurovascular integrity of all extremities.
   C. Evaluate motor response:
      1. Assess ability to move extremities spontaneously and on command;
      2. Localized motor response versus decerebrate or decorticate posturing;
      3. Evaluate deep tendon reflexes.
   D. Assess motor strength.
      1. Check equality in all extremities.
   E. Assess cranial nerve function:
      1. Check cough and gag reflex (nerves IX and X).

2. Eyelid and facial movement (nerves IX and VII).
3. Pupillary reflex (nerves II and III).
4. Eye movement to all four quadrants (nerves III, IV, and VI).
5. Tongue motion (nerve XII).
6. Hearing (nerve VIII).
7. Smell (nerve I).
8. Head turning and shoulder shrug (nerve XI).

F. Obtain in-depth history of patient or parent/family
1. This event
2. Past and current medical history.

G. Evaluate the patient/family assessment of the situation.
1. Provide ongoing communication about the medical status and plan of care.
2. Provide for physical and emotional support for family members.

III. Patient Care and Observation

A. Reassess patient frequently and
1. Watch for signs of increasing intracranial pressure, such as change in metal status, eye opening response, response to pain, or pupillary changes.

B. Raise head of bed 30° to decrease intracranial pressure.
C. Prevent IV fluid overload
D. Provide a calm, quiet environment.
E. Explain the need for continual observation and factor infusion to maintain levels recommended by the physician.

PATIENT AND FAMILY EDUCATION

Patients and families must be made aware of the potential outcomes and sequelae of any intracranial or CNS bleed. Head injuries can be difficult to evaluate, especially if no symptoms are present. Any suspected bleed, whether it be a witnessed or a reported incident of trauma and whether or not symptoms are present, should always be treated with factor concentrate first, followed by diagnostic tests. Headache should never be medicated with aspirin or NSAIDS as they inhibit platelet function.

Every person with hemophilia and members of their family should be well versed in the signs and symptoms of CNS bleeding. This information should be given to parents when a child is newly diagnosed with hemophilia, and bleeding disorders care providers should provide additional education on an ongoing basis. Stressing prevention, such as the use of bike helmets, care seats, and seat belts; recognition of symptoms; and immediate medical intervention provide parents with an action plan of care. Reassurance to the family is essential, as intracranial/CNS bleeding is a very frightening event. Patients and families should be taught to assume that any head injury is life- threatening and to treat such a bleeding episode immediately, raising the patient's factor level to 100% prior to going to an emergency department if possible or else immediately on admission to an emergency department. Hemophilia nurses and hemophilia treatment centers are best able to assess and provide for the patient's and family's level of understanding and educational needs.

PHARYNGEAL BLEEDS

The pharynx is divided into three parts: the nasopharynx (nose and soft palate), the oropharynx (the back of the mouth and upper throat), and the laryngopharynx (epiglottis to the larynx). This is a densely vascular area, from the mouth to the great vessels and muscles in the neck. Bleeding can occur in any of these areas.

NASOPHARYNGEAL BLEEDS

Nasopharyngeal bleeds, usually epistaxis, pose little threat unless the patient loses large volumes of blood. A small child may experience significant blood loss over the course of several days. Swallowing blood from a slow but unrelenting site of bleeding in the nose may produce severe anemia to the point where the child may be in need of a transfusion of packed red blood cells or whole blood. Because the child may be swallowing the blood, the parent may not be aware of the large cumulative amount of blood loss. Pallor, listlessness, tachycardia, and clammy skin should initiate an evaluation for anemia. Any report by a parent of frequent nose bleeding or uncontrolled nose bleeding should initiate an evaluation.

Treatment: Pressure on the nose for 15 minutes while in a sitting position, with head slightly forward, will usually stop the bleeding as well as prevent aspiration or swallowing of blood that can lead to nausea and vomiting. Nasal packing or topical agents may be used if necessary. Cauterization and factor infusion may also be indicated to control bleeding. Adjunctive treatment with aminocaproic acid (Amicar) or transexamic acid may be useful. Amicar inhibits fibrinolysis, so the clot is maintained to tamponade the bleeding site. Dosing regimen for Amicar elixir is 100mg/ kg up to a maximum dose of 3-4g every 6 hours for 5-7 days for mild bleeds or 10-14 days for more severe or prolonged bleeds. For pediatric patients the recommended oral dose is 50-100mg/kg (not to exceed 4 gms) every 6 hours for 5-7 days or up to 10-14 days following dental extraction. (6) For oral bleeds and extractions it is recommended to swish the Amicar before swallowing it because Amicar elixir also has a topical effect as well as an internal effect. Duration of therapy varies with the bleeding episode (21).

MOUTH, OROPHARYNGEAL, AND RETROPHARYNGEAL BLEEDS

These can lead to acute respiratory emergencies. Bleeding episodes may be spontaneous or induced by surgery, infection, or trauma. Mouth bleeds from small cuts, gums, or tooth loss usually are not a problem unless the bleeding continues to the point of volume depletion. General measures, such as avoiding straws and hot fluids and using cold, moist household air, as well as adjunctive therapy with Amicar in conjunction with factor concentrates provide good hemostasis. Bleeding at the base of the tongue, bleeding into a dissecting tract caused by dental infection or anesthetic injection, and bleeding from a lacerated cheek or tongue may cause pooling of blood in the oropharynx, potentially obstructing the airway (1).

Retropharyngeal bleeding may occur following an infection such as pharyngitis or peritonsillar abscess or may result from a hematoma secondary to throat trauma. Airway patency is of major concern, as these conditions can easily lead to obstruction.

Evaluation: Patients should be assessed for ability to swallow saliva and receive a lateral neck x-ray to check for the presence of a mass.

Treatment: Replacement therapy with factor concentrate to raise factor level to 100% will prevent further bleeding and obstruction. Admission, observation, and continued factor therapy should be arranged.

Nursing Care

I. Primary Assessment
- **A.** Obtain history
  - **1.** Recent illness.
  - **2.** Fever.
  - **3.** Sore throat.
  - **4.** Trauma.
  - **5.** Dental work.
- **B.** Observe patient
  - **1.** Airway patency.
  - **2.** Ability to swallow saliva.
  - **3.** Respiratory effort.
  - **4.** Color.
  - **5.** Condition of throat.
- **C.** Initiate Interventions:
  - **1.** Obtain IV access and blood for lab studies
  - **2.** Infuse with factor concentrate to raise factor levels to 100%.
  - **3.** Have patient sit upright and lean forward.
  - **4.** Administer antipyretic or analgesic as indicated or ordered.
  - **5.** Administer Amicar if indicated.
  - **6.** Obtain lateral neck x-ray to assess presence of mass.
  - **7.** Obtain ENT consultation.

II. Secondary Assessment
- **A.** Assess oxygen status
- **B.** Obtain in-depth history
- **C.** Perform in-depth physical exam:
  - **1.** Fever.
  - **2.** Pain.
  - **3.** Circulatory status.
  - **4.** Hydration.

III. Patient support
- **A.** Provide psychosocial support
  - **1.** Take a calm, relaxed, reassuring approach.
  - **2.** Provide information on medical interventions, plan of care, and patient condition.
  - **3.** Explain need for admission for observation and ongoing replacement therapy.
  - **4.** Include family members in explanation and care plan.

Patient and Family Education

Patients and families must be aware of the hazardous nature of pharyngeal bleeds and the need for immediate factor therapy. Signs and symptoms must be clearly presented and reinforced. Specific educational needs include:

1. Why bleeds in this area are problematic.
2. Observe for symptoms, such as difficulty breathing, neck swelling, inability to swallow, change in voice, choking, discoloration.
3. Identify causes of bleeds such as infection or trauma, severe coughing, or vomiting.
4. Immediately raise factor level to 100%.
5. Seek professional care if any symptoms occur.
6. Never medicate pain with aspirin, ibuprofen or other NSAID products.

ABDOMINAL BLEEDS

Acute abdominal pain in a patient with hemophilia may have many origins, such as GI tract bleeding, hematomas (spontaneous or trauma induced), pseudotumors, iliopsoas bleed, or retroperitoneal bleeding. Because of its size, the peritoneal cavity can hold significant amounts of blood with little external evidence of the sequestered blood volume. Any abdominal pain that is associated with hip or groin pain or is accompanied by inability to bear weight on a leg or inability to lie flat may indicate an iliopsoas bleed. While this area is not part of the abdomen per se, large amounts of blood can accumulate in this muscle group in the retroperitoneal space, causing intense pain and significant blood loss.

SYMPTOMS, LABORATORY EVALUATION, TREATMENT

The primary presenting symptom of an abdominal bleed is usually pain, although bleeding into the gastrointestinal tract may not produce pain. Patients may also complain of dizziness, light headedness, shortness of breath, nausea, vomiting, or rectal bleeding. Clinical evidence of volume sequestration in the peritoneal cavity include tachycardia, increased respiratory rate, orthostatic blood pressure changes, referred pain to the shoulder or scapula, lowered blood pressure, pallor, clammy skin, hematemesis, or rectal bleeding.

Laboratory values include decreased hemoglobin and hematocrit levels, altered oxygen levels, changes in white count if infection or inflammation is present, and altered values for organ-specific tests, such as increased amylase in pancreatitis.

Treatment: Treat empirically with factor concentrate to 100% as a first intervention. Do not wait for lab results and do not perform procedures before factor is given.

Nursing Assessment

I. Primary Assessment
   A. Obtain history
      1. Onset and location of symptoms.
      2. Precipitating factors: such as trauma, vomiting or other symptoms of illness.
   B. Observe patient

1. General appearance.
2. Skin color.
3. Check skin: warm, dry, or cold and clammy
4. Note appearance, body language or position, and patient's description of pain on a scale of one (mild) to 10 (severe) or other pain scales deemed appropriate for assessment.

C. Obtain vital signs: blood pressure, pulse, respiratory rate and temperature
    1. Take blood pressure, pulse, and respiratory rate lying, sitting and standing to determine if the patient is orthostatic.

D. Establish IV access and blood for lab studies
E. Infuse patient to raise factor level to 100%.
F. If abdominal bleeding is suspected, order abdominal CT scan or ultrasound.

II. Secondary Assessment

A. Obtain focused history of complaint
    1. Pain assessment: location, quality, intensity, onset, duration, aggravating or alleviating factors.
    2. Associated symptoms: vomiting, diarrhea, constipation, hematemesis, dark stools, rectal bleeding.
    3. Nutrition: ability/desire to eat, time and content of last food intake.
    4. Assess history of past abdominal bleeds.

B. Physical exam
    1. Inspect abdomen: ecchymosis, swelling, pulsation, and contour.
    2. Auscultate all abdominal quadrants and epigastric area for presence of bowel sounds.
    3. Percuss all abdominal quadrants.
    4. Palpate abdomen: assess for areas of tenderness or rebound.
    5. Check groin area for tenderness, hematomas.
    6. Check range of motion to evaluate presence of hip or inguinal bleeds.

C. Obtain laboratory and x-ray tests:
    1. Obtain baseline CBC, platelet count, PT, electrolytes, amylase, lipase, urinanalysis.
    2. Check factor and inhibitor levels if unknown or if recently treated.

III. Patient care and observation

A. Infuse fluids to maintain hydration and volume status.
B. Monitor vital signs.
C. Observe patient for changes in pain.
D. Medicate patient for pain control after evaluation has been completed.
    1. Aspirin and NSAID products should not be used.
E. Explain need for admission for observation, factor replacement, and pain control to patient and family.
F. Provide psychosocial support to patient and family.

Patient and Family Education

Abdominal pain can have many etiologies. In patients with bleeding disorders, all episodes of abdominal pain should be treated as potential abdominal bleeds until ruled out. Symptoms of

gastrointestinal bleeding include bright red/tar-colored stools or coffee-colored/bright emesis. The presence of any of these symptoms requires medical treatment and evaluation. Retroperitoneal bleeding may present as flank pain and may not be trauma-induced. Hematuria and fever may or may not be present. Severe, continued flank pain needs medical evaluation. Aspirin and ibuprofen products for pain control should always be avoided, as should Pepto-Bismol liquid or tablets, which contain aspirin, due to their platelet effect. Trauma to the abdomen may be difficult to evaluate. A child who receives a blow to the abdomen, especially one that is hard enough to cause bruising or crying in the child, should receive factor replacement. Do not wait for signs or symptoms of bleeding, treat first.

COMPARTMENT SYNDROMES

The second most common type of hemorrhage in hemophilia is muscle hemorrhage. Common sites of muscle hemorrhage are iliopsoas, quadriceps, and gastrocnemius muscles and the common muscles of the flexor surface of the forearm. Bleeding may be precipitated by trauma or fractures or may be spontaneous. Compartment syndrome is an increase in the pressure within a muscle sheath following musculoskeletal trauma. It can occur up to 6 days post-trauma, but usually occurs between 12 hours and three days after the trauma. The increased pressure can compress blood vessels and compromise circulation in the already injured extremity. (15)

Bleeding into the large muscle compartments of the upper and lower arms, thigh, and calf can cause tissue compression and lead to nerve compression, tissue death, and loss of function or the limb for a person with hemophilia. Amounts of blood loss are often underestimated but are reported to be 1 to 2 liters in the upper arm muscle, 0.5 to 1 liter in the forearm, 1to 2 liters in the thigh, and 0.5 to 1.5 liters in the calf.

SYMPTOMS AND TREATMENT

Early symptoms of muscle hemorrhage include tingling and warmth, followed by increasing pain, swelling, and muscle spasm. Late signs include pallor, paresthesia, and decreased capillary refill. Pulselessness of an extremity occurs as a very late sign.

Treatment: Treatment for muscle hemorrhage consists of factor replacement, immobilization, ice, elevation, and analgesia. Severe case of muscle hemorrhage and compartment syndrome may require surgical decompression by fasciotomy. The hematologist should always be consulted before fasciotomy is performed. This procedure can often be avoided with aggressive factor therapy. Hemophilia patients who receive fasciotomy will need to be treated as other surgical cases and receive prolonged post-surgery factor management. Always treat with factor as the first intervention and prior to any invasive procedures, including pressure monitoring.

Complications of muscle hemorrhage can range from stiffness, muscle atrophy, decreased flexibility, destruction of the muscle fiber with replacement by non-elastic fibrous tissue, joint contractures, and in extreme cases limb loss.

Nursing Care of Compartment Syndrome

I. Primary Assessment

A. Obtain history
    1. Ascertain if bleed is spontaneous or trauma induced.
    2. Time of onset of symptoms.
B. Physical Assessment.
    1. Color.
    2. Skin temperature and tenseness.
    3. Range of motion (joints above and below area).
    4. Limb girth.
    5. Capillary refill, pulse presence.
    6. Sensory and motor function, including gait if lower extremity is involved.
    7. Degree of muscle response and activity.
    8. Pain.

II. Intervention

A. Factor replacement
    1. Raise factor level to 100% at first symptoms or if history of trauma
    2. On-going factor replacement therapy and admission are warranted in extensive hemorrhage.
B. Other interventions.
    1. Splint area to prevent movement and re-injury.
    2. Ice.
    3. Elevate.
    4. Provide on-going assessment of the muscle bleed.
    5. Pain medication.
    6. Provide psychosocial support and explanation to patient and family.

Patient and Family Education

Patients and families of young children require education to be able to distinguish bleeding in a muscle from bleeding under the skin. Inability or unwillingness to use an affected area is a major indicator that muscle bleeding is occurring. Early recognition and treatment of muscle hemorrhages can prevent compartment syndrome and long-term complications, reduce pain, and allow a quick return to normal function and activity.

Education about muscle hemorrhage can be integrated into joint hemorrhage education. Attaining good muscle strength for prevention of injury and increasing the integrity of muscle tissue are two important messages to convey. Families and patients should be aware that compartment syndromes due to injuries to the forearm and lower extremities are common. Factor infusion at the earliest signs of bleeding or after trauma, followed by rest, ice, compression or immobilization, and elevation is recommended. Once bleeding is controlled, physical therapy and rehabilitation are important for full return of function. Patients or parents should be encouraged to seek medical evaluation any time symptoms such as numbness or color change occur, if the usual methods for home treatment are ineffective, or if they have any questions or concerns about a particular bleeding episode. Pain medication containing aspirin or ibuprofen should be avoided, and treatment with other non-steroidal anti-inflammatory drugs should be used only after consultation with a hematologist.

INTRAOCULAR BLEEDING

Eye hemorrhages can occur spontaneously or after trauma. A diagnosis of hemophilia neither precludes nor precipitates common eye disorders such as glaucoma, cataract, astigmatism, or myopia, but there is an increased risk of bleeding following eye trauma in patients with hemophilia. A person's baseline factor level is the most significant element in susceptibility to an ocular bleed. Bleeding may also occur as a result of another condition, such as CMV retinitis.

SYMPTOMS AND TREATMENT

In hemophilia, trauma is the most common cause of eye injury. Blunt trauma occurs when the eye is hit with and object such as a baseball, snowball, or fist. Penetration injuries occur from thrown objects, such as darts, or by falling on an object, such as a pencil or stick. Spontaneous bleeding can occur in hemophilia, although it is rare. Symptoms include pain and decreased vision.

Treatment: Initiate factor infusion to 100% correction as soon as symptoms are noticed or trauma occurs. Due to the nature and location of the eye, the usual adjunctive methods of bleeding control are ineffective with the exception of ice to help control swelling. Any episode of ocular bleeding should have urgent ophthalmological evaluation and follow-up.

Nursing Care of Intraocular Bleeds

I. Primary Assessment
   A. Obtain history
      1. Description of trauma and presence of foreign body vs. spontaneous onset.
      2. History of precipitating events.
      3. Onset and duration of symptoms.
   B. Note physical appearance: eye swelling, ecchymosis, etc.

II. Intervention
   A. Provide calm atmosphere and reassurance.
   B. Administer factor to level of 100%
   C. Put eye at rest by patching, applying ice bad, and darkening room.
   D. Initiate ophthalmology consult.
   E. Provide emotional support to patient and family. There is often fear of permanent visual loss.

III. Secondary Assessment
   A. Focused history including other medical diagnoses, history of past bleeds, or eye complications.
   B. Pain Assessment
      1. Quality
      2. Duration
      3. Radiation
   C. Visual Changes
      1. Diplopia

2. Blurred or lost vision
3. Photophobia
4. Visual field acuity
5. Itching, tears, discharge

Patient and Family Education

Trauma prevention one of the best and easiest ways to avoid ocular bleeds. Patients should wear goggles during any activity that may involve flying objects, such as a baseball, or when using mechanical equipment. Children should not run while carrying objects such as pencils, sticks, or arrows. Early recognition of the symptoms of eye problems, such as pain around the eye, swelling, discoloration, double or blurred vision, or other visual change, and the need for immediate infusion should be stressed. Any trauma to the eye should be reported to the treatment center immediately so treatment can be initiated. Eye pain should be not treated with aspirin or ibuprofen products. Families should be aware of the need for an ophthalmology consult (not optometry) if eye symptoms occur.

CONCLUSION

All episodes of bleeding in hazardous areas require immediate assessment, intervention, and treatment. Such situations, or the threat of such an occurrence, generate high levels of anxiety and fear of damage or loss in patients and families. Visits to the emergency department produce added stress. Hemophilia nurses can help parents and individuals learn advocacy skills and supply tips for dealing with emergent situations, such as consulting with their hemophilia treatment center first and then calling ahead to the emergency department. Parents and persons with hemophilia are knowledgeable about the management of their disorder, and their input should always be sought and heeded. Whenever possible, the individual with hemophilia should present to an emergency department which is associated with a hemophilia treatment center. Emergency departments associated with HTCs are more likely to have familiarity with treating hemophilia patients and will generally know to consult the hemophilia treatment center team.

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