

THE TREATMENT OF HEMOPHILIA BLEEDING WITH LIMITED RESOURCES

Revised Edition

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Table of Contents

Introduction.....	1
Clinical Situations with External Blood Loss.....	1
Minor and Recurrent Joint Bleeds (Hemarthroses).....	2
Special Notes	3
In Conclusion	3
Minimal Replacement Therapy	3
Exercises for the Knee	4
Exercises for the Ankle.....	6
Exercises for the Elbow	9

The Treatment of Hemophilia Bleeding with Limited Resources

Shelby L. Dietrich

Introduction

Although the fundamental treatment of hemophilia bleeding problems is replacement therapy of the missing coagulation factor, nevertheless in many places and on many occasions adequate replacement therapy (clotting factor concentrates, fresh frozen plasma, cryoprecipitate) is not available, but proper and prompt use of conservative measures that decrease or control the bleeding and prevent future damage may be employed.

This article will describe those measures with specific application to hemophilia-related common bleeding problems. It is helpful to recall that bleeding in hemophilia is generally slow, with continuing ooze because of poor clot formation; this principle enables one to apply conservative measures, stop the bleeding, and permit tissue repair. In essence one can "buy time."

The goals of treatment are identical whether clotting factor replacement therapy is available or not, to minimize permanent damage, minimize suffering and pain, permit tissue healing, and restore function. If even minimal clotting factor replacement therapy is available (cryoprecipitate which contains Factor VIII only, fresh frozen plasma which contains both factor VIII and factor IX, or specific factor concentrates), then this therapy should be administered as promptly as possible when the bleeding episode begins or is recognized. Early use of replacement therapy, even in minimal amounts, produces better hemostasis than delayed use.

Clinical Situations with External Blood Loss

External blood loss results from:

- lacerations or abrasions of the skin;

- oral mucosal bleeding from dental problems, from the mucosa of the mouth and from cuts or scrapes to the tongue;
- nose bleeds (epistaxes);
- gastrointestinal bleeding; and
- kidney bleeding.

General measures which can be applied in most of these situations excepting GI and GU bleeds are the use of firm, constant pressure, elevation of the head when applicable, and application of ice.

- a) In general, superficial skin lacerations which in a normal individual would require suturing or use of sterile adhesive strips may be treated in the same manner in a person with hemophilia. Significant lacerations should be immediately sutured, a dose of replacement therapy given (if available), and ice, pressure, and elevation applied consistently for as long as possible. In general, skin lacerations which in a normal individual would require suturing or use of sterile gauze strips may be treated in the same manner in a person with hemophilia. Ice should not be applied directly to the skin; ice packs must be wrapped in a thick towel since ice burns and skin damage can occur with prolonged application. Ice is effective in stopping bleeding when used **promptly** and early but does not assist in the resolution of old hematomas. In toddlers and small children, small scalp lacerations are often the site of formation of exuberant clots; if such a situation occurs with repeated oozing and enlargement of clots, gentle removal of the clot should be undertaken in order to approximate the skin edges, permit application of pressure, and allow healing.

- b) A retained friable berry-like clot in the mouth impedes healing and allows slow bleeding to continue. Continuing ooze from loose deciduous (baby) teeth is a nuisance and occasionally the cause of significant blood loss. The loose tooth should be gently but expeditiously extracted and pressure and ice applied to the bleeding site for several hours. The loose tooth acts as a foreign body and irritant to the gingiva.
- c) Continued oozing from tongue lacerations can ultimately result in significant blood loss, and if such bleeding continues, the child's hemoglobin should be checked, and if significant anemia has occurred, transfusion of blood (as fresh as possible) is indicated. This situation is particularly dangerous in infants and small children who may suffer considerable blood loss in proportion to their total circulating blood volume with progressive anemia resulting in cardiovascular compromise. Diet for children with bleeding from the oral cavity should consist of cold or chilled and soft foods. Use of straws should be prohibited to avoid dislodging the formed clot. Excessive use of the nursing bottle is also discouraged, since the nipple of the bottle may dislodge a clot.
- d) Significant epistaxis (nose bleed) is rare in the person with hemophilia compared to the person with von Willebrand Disease and can usually be successfully treated with rest with the head slightly elevated (to 30%), ice to the nose, and if necessary, packing of the anterior nasal cavities; packing, however, must be done with care. Gentle pressure should be applied to the nares. Children often swallow blood from nose bleeds and later vomit the blood; they can have significant blood loss in this fashion. Ingested and digested blood may cause black ("tarry") stools. In small children, hemoglobin and hematocrit should be measured frequently in the face of ongoing ooze or bleeding.
- e) Bleeding from the gastrointestinal tract, whether upper or lower, indicates the need for medical consultation; blood in the urine or hematuria, on the other hand, can be treated with watchful waiting and a high fluid intake (150-200 ml/hr) to prevent clot obstruction in the ureters. Even without replacement therapy a daily dose of prednisone for three to five days is often helpful in stopping GU bleeding.
- f) Local/topical treatment: hemostasis of minor lacerations and abrasions in the mouth, including the tongue and gingiva, may be aided by application of topical thrombin preparations or gauze soaked in dilute epinephrine solutions (1-10,000 aqueous epinephrine). Topical hemostatic preparations can also be applied to skin abrasions.

Minor and Recurrent Joint Bleeds (Hemarthroses)

Minor and recurrent joint bleeds are the hallmark of hemophilia A and B and ultimately lead to significant morbidity, impairment of function, and joint damage. Prompt recognition and treatment of joint bleeds is essential to prevent these long-term complications. Basic conservative principles of treatment are: rest in the position of least pain, application of ice, and elevation and immobilization. In the upper extremity, bleeds in the wrist and elbow should be treated with a sling. If cryoprecipitate or fresh frozen plasma is available, administer one dose when the bleed is recognized and a second dose in 24 hours. Whenever necessary, analgesia should be used, acetaminophen or paracetamol, avoiding the use of aspirin-containing compounds. When pain, stiffness, and limitation of range of motion subsequent to knee bleeds have subsided, it is critical to begin or resume strengthening exercises to the muscles protecting the knee. Strong muscles around the knee can prevent recurrent hemorrhages and chronic synovitis. Active isometric resistive exercise is possible without the use of replacement therapy. Similarly, gentle active exercise and range of motion should be undertaken when elbow bleeds subside. The use of high-top boots or tennis shoes for recurrent ankle bleeds, particularly in toddlers and children, is helpful.

Special Notes

- a) Bleeding after inadvertent circumcision: Continue the use of topical pressure as long as feasible. **Do not suture or cauterize!**
- b) "Sore throat" and tonsillitis with or without cough: Tonsils and adenoids reach their maximum size during early childhood, and respiratory infections are frequent. Inflamed tonsils can precipitate peritonsillar bleeding with serious results. Similarly, continued coughing can cause pharyngeal and tracheal bleeding. Antibiotics should be employed freely together with a cool, liquid diet until the child recovers. If necessary to suppress cough, a codeine-containing cough medicine is efficacious. If a child has increasing difficulty speaking or swallowing, suspect or rule out pharyngeal, laryngeal, or tracheal bleeding.
- c) Awareness of possibility of significant blood loss within the retroperitoneum and thoracic and abdominal musculature: A significant amount of blood leading to anemia and even shock can be lost within the large muscles of the thorax and back and in the retroperitoneal space. Unexplained pallor, weakness, or lethargy and lower abdominal pain may lead to the diagnosis of such problems and medical intervention.
- d) Intramuscular deep hematomas of the calf and forearm: These are potentially dangerous because of pressure on nerves and blood vessels, and must be treated with bed rest and elevation until the swelling subsides.

In Conclusion

- a) **The "do's" of conservative treatment are:**
 - For extremity bleeds, do rest in a position of elevation where feasible, apply ice, and resume normal activity when pain and stiffness disappear with special attention to restoration of muscle strength.

- Do be aware of the possibility and potential of significant blood loss where oozing is continuous over several days in small children.
 - Do be extremely observant of a toddler's level of activity and do encourage children and adolescents to report the first subjective symptom or objective sign of joint bleeding in order to promptly administer conservative measure.
 - Do observe the child's body appearance closely during daily bath; look for asymmetry and swelling.
 - Do suture lacerations.
 - Do exercise gradually after joint bleeds subside; learn isometric exercises. Strong muscles protect joints.
- b) Be aware of deep muscle hemorrhages in the forearm and calf which can cause nerve entrapment and neurovascular compromise and ultimate tissue death.
 - c) **The "don'ts" of conservative treatment are:**
 - Do **not** cauterize!
 - Do not apply constricting circular bandages or wraps.
 - Do not scold a child for having a bleed.
 - Do not circumcise if a family history of bleeding is known.
 - Do not apply ice to the skin without a protective towel or cloth.

Minimal Replacement Therapy

- a) Fresh frozen plasma contains factor VIII and factor IX, 250 units in 250 ml of plasma. To avoid fluid overload, administer one bag every 8 to 12 hours for hemophilia A. One bag every 24 hours may be adequate for hemophilia B. **Reduce** volume for infants and children.
- b) Cryoprecipitate: one bag usually contains 80-100 units of factor VIII only.

Aims:**Muscle Groups
Being Exercised:****Exercise 1**

With your leg lying straight, tighten the Quadriceps muscle, causing the knee cap to move up.

Relax - then REPEAT 10-15 times.

Exercise 2

With the knee over a roll (e.g. a bottle wrapped in a towel), lift the foot up and straighten the knee. Lower slowly.

Relax - then REPEAT 10-15 times.

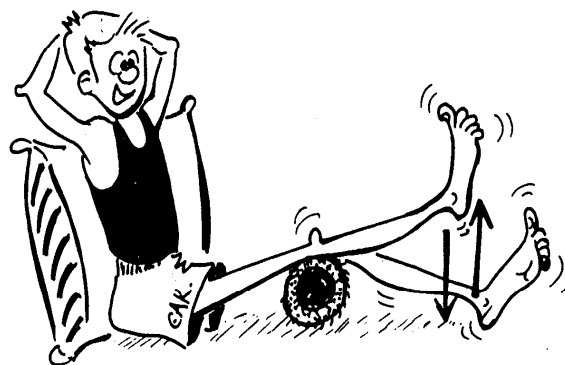
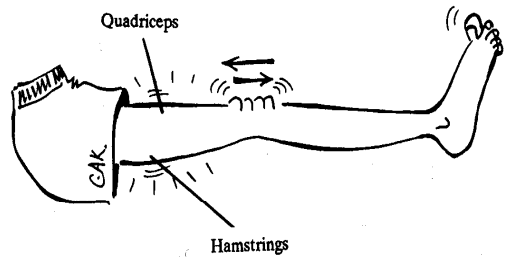
EXERCISES FOR THE KNEE

Good Quadriceps control (this is the muscle which straightens the knee and stops the knee collapsing when walking and running)

Full range of bending and straightening

**Quadriceps
Hamstrings**

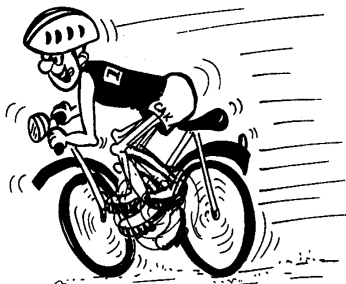
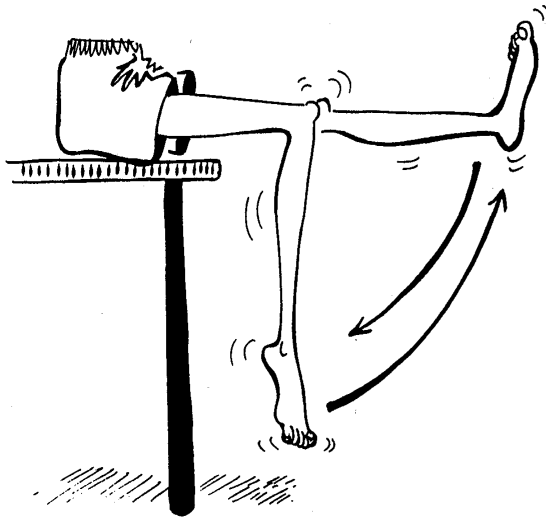
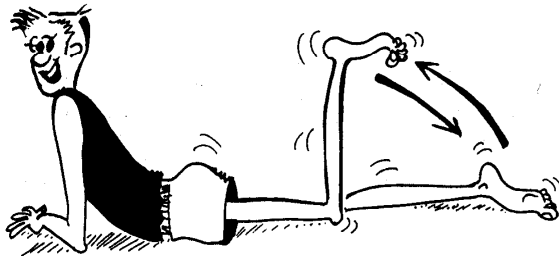
If your child is keen to be up but you're concerned that the knee is still a bit weak, you can protect it by letting him/her walk with the support of the knee splint bandaged on. Persist with this until the muscle strength and control are back to normal.



Straight Leg Raises:

Lie on a bed or floor with your legs straight and feet pulled up towards your head. Raise your leg with the knee as straight as possible until your foot is approximately 15 inches off the floor. Lower slowly.

Relax - then REPEAT, alternating legs, 10 times.



Exercise 3

Exercise 4

Lying on your tummy, bend and straighten your knee.

Relax - then REPEAT 15-20 times.

Exercise 5

Sitting over the edge of a bed or table, straighten and bend your knee.

Relax - then REPEAT 10-15 times

Exercises 2 to 5 can be made harder by tying a small weight to the ankle, such as a packet of rice or a sandbag

Exercise 6

Bike-riding

Aims:**Muscle Groups
Being Exercised:****EXERCISES FOR THE ANKLE**

A stable ankle which doesn't give way or cause pain when walking. Don't let the calf muscle tighten. This restricts the amount you can move the ankle up. If it becomes tight and the heel is unable to reach the ground, this will affect walking and balance.

Dorsiflexors
Calf Muscle

Invertors and Evertors

(These move the ankle in and out respectively.)

The ability to pull the foot up (usually to 10 or 15 degrees past the neutral position) is most often affected after a bleed into the ankle. It is important that this movement be regained, or normal walking may be impaired.

Exercise 1

Sitting on a bed or table, roll your toes right up and then point them right down.

**Exercise 2**

Sitting on the floor with your legs straight, make circles with your ankles and toes - clockwise and anticlockwise.





Exercise 3

Turn your feet in - heels and soles facing each other, and then out.

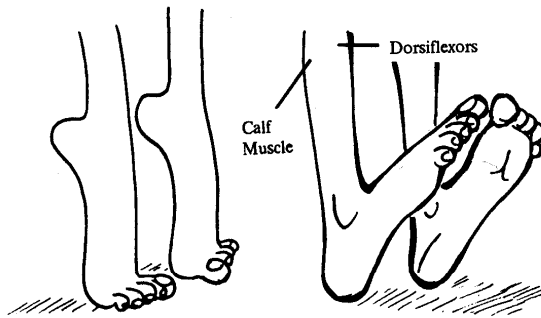
Exercise 4

Exercises (1) and (3) can be made harder by doing the same movements against the resistance of a bike inner tube. This is hooked under the leg of a table or bed. Then hook your foot underneath the tube. The greater the stretch on the tube, the harder the muscles have to work.



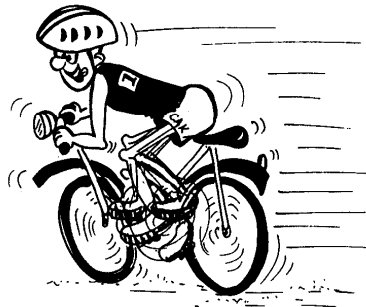
Exercise 5

Walking on heels and walking on toes



Exercise 6

Bike-riding.



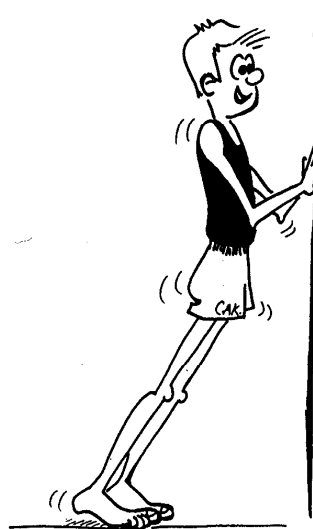
Exercise 7**Exercise 8**

Wobble board activities

NOTE:

If the calf muscle becomes tight, try some calf stretches. Stand about 12-18" from a wall or support. Keep your feet and heels flat on the ground and move your hips towards the wall. Stretch the calf muscles. Hold for 2-3 minutes.

Balancing on one leg (e.g. kicking a ball)



Aims:**Muscle Groups
Being Exercised:****EXERCISES FOR THE ELBOW**

**Full range of movement and muscle power
in the elbow and forearm**

**Biceps
Triceps
Pronators
Supinators**

After a joint bleed, the ability to straighten the elbow is often affected, along with the ability to turn the forearm so that the palm of the hand faces upwards (pronation/supination).

If the elbow is bent, do not force it out straight, as this will cause damage to the joint. Aim for active muscle control. (Your child may need serial splinting to restore elbow extension - see your Physiotherapist.)

Exercise 1**Exercise 2****Exercise 3**

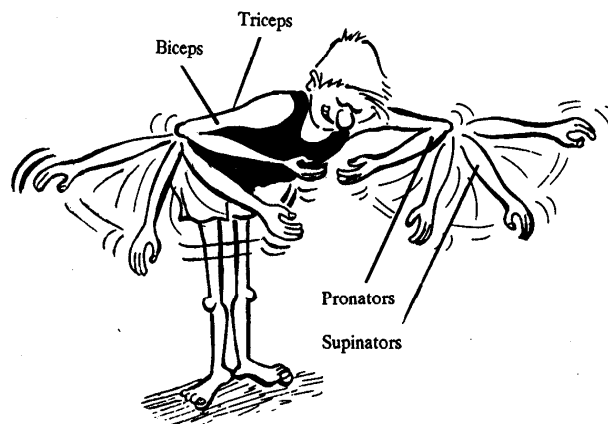
(1) Bend over, keeping your back straight, with elbows out from your sides and hands on your chest.

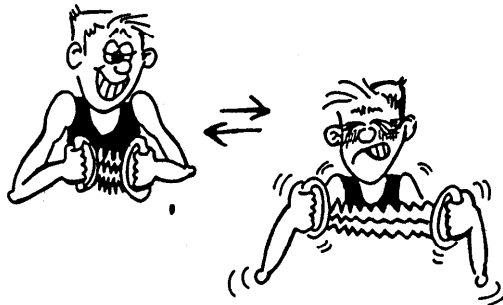
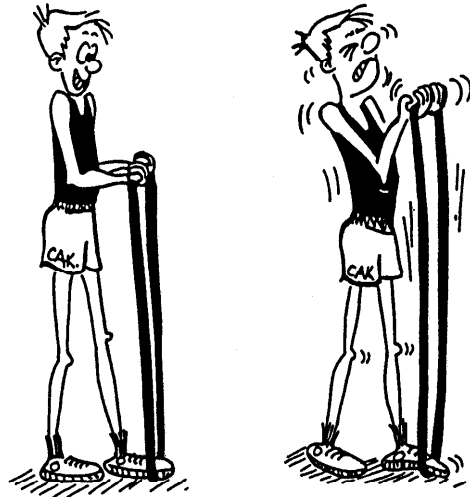
(2) Keep the part of the arm above the elbow still.

(3) Now straighten your arms without moving your shoulders, then bring your hands back to your chest. Isolate the movement so that you are just moving your hands and forearms.

Bend and straighten the elbow.

With your elbow bent, turn the forearm so that your hand faces up, and then down.





Balloon Tennis or Nerf (foam) Ball Cricket.

Exercise 4

Using a small weight, inner tube or spring, copy the activities in the drawings.

Exercise 5

Extracted from *Physiotherapy in Haemophilia - Exercises to do at Home* by Genny Dwyer and Alicia Hosking with the kind permission of the Educational Resource Centre of Adelaide Medical Centre for Women and Children (Adelaide Children's Hospital Campus) North Adelaide, South Australia 5006.