

PREGNANCIES AT RISK FOR NEWBORNS WITH HEMOPHILIA CLINICAL PRACTICE GUIDELINE

This clinical guideline has been developed to recommend appropriate *perinatal* management of mothers and newborns who are at risk for hemophilia. Please direct any questions to the Maine Hemophilia & Thrombosis Center, 207-396-7565 (for pediatric patients) or 207-396-7683 (for adult patients).

Inherited hemophilia most often occurs in the male offspring of maternal carriers of Factor VIII and Factor IX deficiency. In approximately 30% of new hemophilia patients, there is no family history because the hemophilia is the result of a new mutation.

INTRACRANIAL HEMORRHAGE (ICH)

The risk of ICH in vaginally-delivered, term newborns with hemophilia is low. However, if an ICH occurs, it can be very serious. Many of the signs and symptoms of ICH are not obvious at the time of delivery or in the first few days of life. The following may increase the risk of ICH in newborns:

Cephalopelvic Disproportion
Prolonged, Difficult Labor
Vacuum- or Forceps-Assisted Delivery

Primiparous Mother
Precipitous Delivery

RECOMMENDATIONS

Each pregnancy, mother, and newborn must be managed individually. The following recommendations should be considered to decrease the risk of bleeding during labor, delivery, and in the newborn period:

Internal scalp monitoring should be avoided.

Vacuum devices and forceps should be avoided.

Heelsticks and **venipunctures** should have pressure dressings placed immediately and be monitored for several hours for delayed bleeding. **Femoral and jugular blood sampling should be avoided** to prevent hematoma formation and compression of vital structures. **Arterial punctures should be avoided.**

Intramuscular injections should be performed with attention to the following recommendations:

Use a fine-gauge needle (23 gauge or smaller caliber)

Apply firm pressure to the site, without rubbing, for at least 2 minutes after the injection

Notify the patient/caregiver of the risk of hematoma development at the injection site

Provide anticipatory guidance for reasons to call the physician or Maine Hemophilia & Thrombosis Center, including adverse reactions, such as: hematoma, fever, warmth, redness.

For pain/fever relief, avoid aspirin and NSAIDS (such as ibuprofen, naproxen, etc.) due to the risk of bleeding. Acetaminophen is a safe alternative in patients without liver disease/risk of liver disease.

Circumcision should be discouraged to prevent the potential of prolonged bleeding. Contact the Maine Hemophilia & Thrombosis Center (207.396.7565) if the family insists on circumcision.

RADIOGRAPHIC STUDIES

A **non-sedated MRI scan of the brain** is routine in the first 24 hours of life. Because the risk of a subdural hematoma is significant in newborns with hemophilia, head ultrasound is not the ideal study in these patients.

Algorithms are not intended to replace providers' clinical judgment or to establish a single protocol. Some clinical problems may not be adequately addressed in this guideline. As always, clinicians are urged to document management strategies.

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LABORATORY STUDIES

- A **cord blood sample** from newborns with a suspected diagnosis of hemophilia is an acceptable source of blood for diagnostic studies if:
 1. The specimen is collected properly (as outlined below)

AND

2. The specimen is obtained *immediately* after birth.

CAUTION: Cord blood testing most accurately diagnoses newborns with severe Hemophilia A (Factor VIII deficiency). Cord blood testing is less accurate in newborns with mild or moderate Hemophilia A and all severities of Hemophilia B (Factor IX deficiency). The gestational age of the newborn and the type of delivery (vaginal vs Caesarian section) may affect coagulation studies and Factor levels. Therefore, venous sampling may be necessary in the newborn period. For patients with suspected hemophilia, **femoral and jugular blood sampling should be avoided** to prevent hematoma formation and compression of vital structures. **Arterial punctures should be avoided.**

Any **abnormal Factor level** should be discussed with a hematologist at the Maine Hemophilia & Thrombosis Center, 207-396-7565. Results of hemophilia testing should be provided to families by an experienced hemophilia center.

CORD BLOOD COLLECTION PROCEDURE OF PATIENTS WITH SUSPECTED HEMOPHILIA

Materials: 20-ml plastic syringe with an 18- or 19-gauge needle attached
Two or three 3 ml blue top tubes (3.2% Sodium Citrate). Each tube requires 2.7 ml of blood.
Sterile saline and 70% alcohol prep pad.

Method:

1. **Prepare** the tube and syringe prior to delivery and have supplies available in the delivery room. Pre-label the sample-collection tubes (including mother's name, hospital name, date, and "CORD BLOOD").
2. Immediately after delivery, **clamp** a 10-20 cm section of the umbilical cord. Handle the cord gently. Aggressive manipulation activates coagulation.
3. **Wipe** the surface of the cord with sterile saline and, then, alcohol.
4. Identify the placental end of the cord. Insert the needle into the placental side of the umbilical vein, avoiding contamination with Wharton's jelly. (Wharton's jelly activates coagulation; contamination makes interpretation of the results difficult.)
5. Collect a minimum of 6-9 ml of cord blood for coagulation studies and Factor levels. Place 2.7 ml of unclotted blood into each blue top tube. (Because cord blood clots easily, collecting multiple tubes increases the potential for analyzing an acceptable sample.) The tubes contain approximately 0.3 ml of anticoagulant. Gently mix the tubes by inverting them 4-5 times. Do not shake the tubes vigorously. Record the time the sample was collected.
6. Send the blood sample to the laboratory *as soon as possible* (but absolutely not later than two hours after collection). Check to assure no clots have formed in the tubes. If clots are noted, discard the sample.
7. Spin the unclotted blood sample at approximately 3,000 RPMs for 20 minutes at 4 degrees C. Remove the plasma and freeze in plastic tubes in 0.5 ml aliquots. Store samples at -20 degrees C (or lower if possible).
8. Send the tubes on dry ice as soon as possible to:
 - NorDx Coagulation Laboratories at Maine Medical Center; 22 Bramhall St.; Portland, ME 04102
 - Call 207-662-4029 to notify the lab that a specimen is arriving or to discuss questions about collecting cord blood, specimen handling, and specimen shipping to NorDx at MMC.