HEMATOLOGY · BLEEDING DISORDERS

# Hemophilia

An X-linked recessive bleeding disorder defined by deficiency of clotting factor VIII (Hemophilia A) or factor IX (Hemophilia B). Prolonged bleeding into joints, muscles, and soft tissue defines its clinical signature.

NGN NCLEX 2026 | NCJMM ALIGNED | HIGH-YIELD | PEDS + ADULT

**SOURCE TRANSPARENCY**

Hemophilia content was not present in your uploaded personal notes for this project. This infographic is built from standard NCLEX references: *Saunders Comprehensive Review for the NCLEX-RN Examination (2026)*, *ATI RN Adult Medical Surgical Nursing*, *Lippincott Illustrated Reviews: Pharmacology*, the *National Hemophilia Foundation* and *World Federation of Hemophilia* 2020 treatment guidelines, and current FDA prescribing information for factor concentrates.

1

## Definition & Overview

- **Inherited bleeding disorder** — deficiency or absence of a clotting factor in the intrinsic pathway.
- **Hemophilia A** (classic, ~80% of cases) — **Factor VIII** deficiency.
- **Hemophilia B** (Christmas disease) — **Factor IX** deficiency.
- **X-linked recessive** — passed from carrier mothers to sons; daughters usually carriers.
- **Almost exclusively males** are clinically affected.
- Severity is categorized by factor activity: **severe <1%, moderate 1–5%, mild 5–40%**.

2

## Pathophysiology

**STEP 1** Gene mutation on the X chromosome → deficient/absent Factor VIII or IX

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**STEP 2** Injury occurs → platelets form initial plug (primary hemostasis intact)

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**STEP 3** Intrinsic coagulation cascade fails — no stable fibrin clot forms

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**STEP 4** Prolonged / recurrent bleeding into joints, muscles, soft tissue, CNS

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**STEP 5** Repeated joint bleeds → synovial damage → chronic hemophilic arthropathy

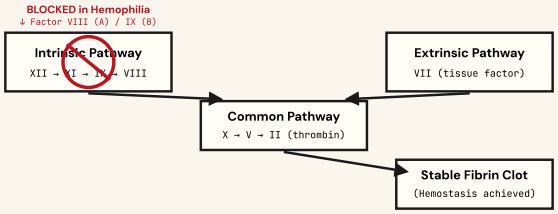


FIG 1 · Intrinsic coagulation cascade is blocked → PTT prolonged, PT normal

3

# Signs & Symptoms

### MILD / EARLY PRESENTATION

- Easy bruising / large hematomas after minor trauma
- Prolonged bleeding after circumcision, dental work, or minor cuts
- Recurrent epistaxis (nosebleeds)
- Bleeding after immunizations or venipuncture
- Prolonged PTT (intrinsic pathway); PT & platelets normal
- Bleeding may be delayed after injury (not immediate)

### SEVERE / LATE PRESENTATION

- **Hemarthrosis** — warm, swollen, stiff, painful joint (knee, ankle, elbow most common)
- Spontaneous bleeding without trauma (severe disease)
- Deep muscle hematomas → compartment syndrome
- GI bleeding — melena, hematemesis
- Hematuria
- Chronic hemophilic arthropathy (joint deformity)

**PRIORITY RED FLAG — INTRACRANIAL HEMORRHAGE:** headache, vomiting, lethargy, irritability, seizure, or altered LOC after ANY head trauma (or spontaneously in severe disease). This is the #1 cause of death in hemophilia. **Headache after a fall = ICH until proven otherwise.**

**AIRWAY ALERT:** bleeding into the neck, throat, or tongue can rapidly compromise the airway. Assess for hoarseness, stridor, dysphagia, or neck swelling.

4

# Priority Nursing Interventions · ABC + SAFETY

### A · AIRWAY / B · BREATHING / C · CIRCULATION

- **Assess airway first** if oropharyngeal, neck, or tongue bleeding suspected — prepare for emergency airway management.
- **Monitor for shock** — tachycardia, hypotension, restlessness, pallor, ↓ urine output (early signs of hypovolemia).
- **Neuro checks q1–4 h** after any head trauma — LOC, pupils, motor strength.

### ACUTE BLEED MANAGEMENT

- **Administer factor replacement IV** (Factor VIII or IX concentrate) — give BEFORE imaging, BEFORE procedures, and at the first sign of bleeding.
- **Apply direct pressure × 10–15 minutes** to external bleeding sites.
- **RICE for hemarthrosis** — Rest, Ice, Compression, Elevation; immobilize the joint briefly, then gentle ROM.
- **Avoid aspiration of bleeding joints** unless ordered — increases infection risk.

### SAFETY / PREVENTION OF BLEEDING

- **NO IM injections** — use SQ, oral, or IV routes; if IM is unavoidable, give factor first and apply prolonged pressure.
- **NO aspirin, NSAIDs, or anticoagulants** — use acetaminophen for pain.
- Soft-bristle toothbrush, electric razor, no rectal temperatures or rectal suppositories.
- Pad the crib / bed rails for young children; avoid contact sports.
- Hold pressure on venipuncture sites **at least 5–10 minutes**.

### MONITORING & DIAGNOSTICS

- **PTT** — prolonged (intrinsic pathway); **PT, INR, platelets, bleeding time** all normal.
- **Factor VIII or IX assay** — confirms diagnosis and severity.
- **CBC** — H&H for occult or chronic blood loss.
- Genetic testing for carrier identification.

5

# Pharmacology

| DRUG / CLASS | MECHANISM | KEY SIDE EFFECTS | NURSING CONSIDERATIONS |
|---|---|---|---|
| **Factor VIII concentrate** (recombinant or plasma-derived) *Hemophilia A* | Replaces deficient Factor VIII to restore intrinsic clotting | Allergic reaction, inhibitor (antibody) formation, transient flushing | IV only. Reconstitute gently — do NOT shake. Give at first sign of bleed. May be self-infused at home. |
| **Factor IX concentrate** (recombinant) *Hemophilia B* | Replaces deficient Factor IX | Thrombosis risk, inhibitor formation, hypersensitivity | IV only. Longer half-life than F-VIII → less frequent dosing. Monitor for DVT/PE. |
| **Desmopressin (DDAVP)** Synthetic ADH analog | Stimulates release of stored Factor VIII and vWF from endothelium | Hyponatremia, water retention, flushing, headache, ↑ BP | Only for **MILD Hemophilia A** — NOT effective for Hemophilia B. Restrict fluids; monitor sodium. IV, SQ, or intranasal. |
| **Aminocaproic acid (Amicar)** Antifibrinolytic | Inhibits plasminogen activation → stabilizes formed clots | Thrombosis, hypotension, GI upset, myopathy | Especially useful for oral / dental / mucosal bleeds. Do NOT use with DIC or hematuria (risk of clot in urinary tract). |
| **Emicizumab (Hemlibra)** Monoclonal antibody | Bridges Factor IXa and X to mimic Factor VIII function | Injection-site reaction, thrombotic microangiopathy when combined with bypassing agents | SQ injection for **prophylaxis** of Hemophilia A (with or without inhibitors). Reduces bleed frequency dramatically. |
| **Acetaminophen** Non-opioid analgesic | Central COX inhibition; no platelet effect | Hepatotoxicity (dose-dependent) | **Preferred** pain reliever in hemophilia. AVOID aspirin, ibuprofen, naproxen, ketorolac. |

6

# NCLEX Test Traps

✗ WRONG ANSWER

"Give ibuprofen for joint pain in a child with hemophilia."

✓ RIGHT ANSWER

Give **acetaminophen**. NSAIDs and aspirin impair platelets and worsen bleeding.

✗ WRONG ANSWER

"Give IM vitamin K — it's a clotting factor."

✓ RIGHT ANSWER

**NO IM injections.** Hemophilia bleeding is NOT a vitamin K deficiency — it is a factor VIII/IX deficiency. Vitamin K does not correct it.

✗ WRONG ANSWER

"Child has a headache after a fall — give Tylenol and reassess in 4 hours."

✓ RIGHT ANSWER

Headache + head trauma = **intracranial bleed until proven otherwise**. Give factor IMMEDIATELY, notify HCP, prepare for CT.

✗ WRONG ANSWER

"Hemophilia A and Hemophilia B require the same treatment."

✓ RIGHT ANSWER

A = Factor VIII; B = Factor IX. **DDAVP works ONLY in mild Hemophilia A.** It does NOT raise Factor IX.

✗ WRONG ANSWER

"PT will be prolonged in hemophilia."

✓ RIGHT ANSWER

**PTT** is prolonged (intrinsic pathway). PT, INR, platelets, and bleeding time are normal. Both start with P → PTT = Prolonged.

✗ WRONG ANSWER

"Hemophilia and von Willebrand disease are the same."

✓ RIGHT ANSWER

vWD affects BOTH sexes (autosomal), involves vWF + mild ↓ Factor VIII, and presents with mucosal bleeding (nose, gums, menorrhagia). Hemophilia presents with deep joint/muscle bleeds.

✗ WRONG ANSWER

"Apply heat to the bleeding joint to reduce pain."

✓ RIGHT ANSWER

**ICE** — heat causes vasodilation and worsens bleeding. Remember **RICE**: Rest, Ice, Compression, Elevation.

✗ WRONG ANSWER

"Wait until bleeding starts to give factor."

✓ RIGHT ANSWER

Factor is given **prophylactically before dental work, surgery, or after known trauma** — not after bleeding becomes uncontrolled.

7

## Patient & Family Education

- **Medical alert bracelet** at all times — identifies type and severity.
- **Avoid contact sports** — football, wrestling, hockey, boxing, rugby.
- **Safe activities** — swimming, golf, biking with helmet, walking.
- Use soft-bristle toothbrush, electric razor (no straight razors).
- **Avoid aspirin, NSAIDs, ginkgo, garlic supplements, vitamin E, fish oil** — all interfere with platelet function.
- Recognize early signs of a bleed: tingling, warmth, or "bubbling" in a joint BEFORE visible swelling.
- Home factor infusion training — proper reconstitution, IV technique, storage.
- Routine dental hygiene to prevent gum bleeding; **inform dentist before procedures**.
- **Genetic counseling** for carrier sisters and for family planning.
- Inform ALL HCPs (surgeons, anesthesia, ED staff) BEFORE any procedure.
- Childproof the home: padded corners, helmets when learning to walk, no toys with sharp edges.
- Keep an updated factor supply at home and when traveling.

8

## Memory Boosters

### A = 8

Hemophili**a A** → Factor **VIII (8)**. Alphabetical order matches numerical: A before B, 8 before 9.

### B = 9

Hemophilia **B** = Factor **IX (9)**. Also called Christmas disease — "B" for "Birthday" / Christmas.

### Boys Bleed

X-linked recessive → almost exclusively **males** are affected; mothers are carriers.

### PTT = Prolonged

Both start with **P**. The intrinsic pathway (PTT) is the broken one. PT and platelets stay normal.

### RICE

**R**est · **I**ce · **C**ompression · **E**levation — for every joint bleed. Never heat.

### No IM, No ASA

NO **I**ntra**M**uscular injections. NO **A**spirin / NSAIDs. Use acetaminophen instead.

### "Hot Joint = Bleed"

**W**arm · **S**wollen · **S**tiff · **S**ore = hemarthrosis until proven otherwise.

### Headache → CT

Any headache, vomiting, or altered LOC after head trauma in a hemophiliac = **treat as ICH**. Give factor → CT → notify HCP.

9

# NCJMM Clinical Judgment Scenario · 6-Step Framework

CLINICAL SCENARIO

*A 7-year-old boy with severe Hemophilia A is brought to the ED by his mother. He fell off his bicycle 1 hour ago and struck his head on the curb (helmet was loose). On arrival: HR 124, BP 96/58, RR 22, $SpO_2$ 98%, T 37.0°C. He reports a "really bad" headache and his right knee is "throbbing." Mother notes he seems sleepier than usual. His right knee is visibly swollen, warm, and he refuses to bear weight.*

STEP 1

## Recognize Cues

Severe Hemophilia A · head trauma · headache · increased sleepiness · tachycardia (HR 124) · hemarthrosis (swollen, warm, painful knee, refusing to bear weight) · loose helmet at time of injury.

STEP 2

## Analyze Cues

Headache + altered mentation + head trauma in a severe hemophiliac = **intracranial hemorrhage** (life-threatening, #1 cause of death). Warm/swollen/painful knee = **hemarthrosis**. Tachycardia may reflect pain, blood loss, or both.

STEP 3

## Prioritize Hypotheses

**#1 Intracranial hemorrhage** (airway/neuro/life threat — ABC). **#2 Acute hemarthrosis** of right knee. **#3 Pain control**. ICH is the immediate priority — Maslow + ABC.

STEP 4

## Generate Solutions

Administer Factor VIII concentrate IV **STAT** (before imaging). Establish large-bore IV access (NOT IM). Stat CT head. Neuro checks q15min. RICE to right knee. Acetaminophen for pain — NO NSAIDs/aspirin. Notify HCP and hematology.

STEP 5

## Take Action

Give Factor VIII IV first. Continuous cardiac & $SpO_2$ monitoring. Q15min neuro & vital signs. Elevate & ice the right knee. Keep NPO pending imaging/possible OR. Provide emotional support to child and parent. Document baseline and trend.

STEP 6

## Evaluate Outcomes

Improved or stable LOC, HR < 110, BP stable, no new neuro deficits, CT negative or treated bleed resolving, decreased knee swelling/pain, controlled bleeding, age-appropriate behavior returning. Reinforce helmet safety and home factor protocol at discharge.