

Chapter 10

MUSCULOSKELETAL COMPLICATIONS

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HEMOPHILIA IS CHARACTERIZED BY ACUTE BLEEDS



>80% of acute bleeds in hemophilia occur in specific joints.
(most commonly the ankle, knee, and elbow)

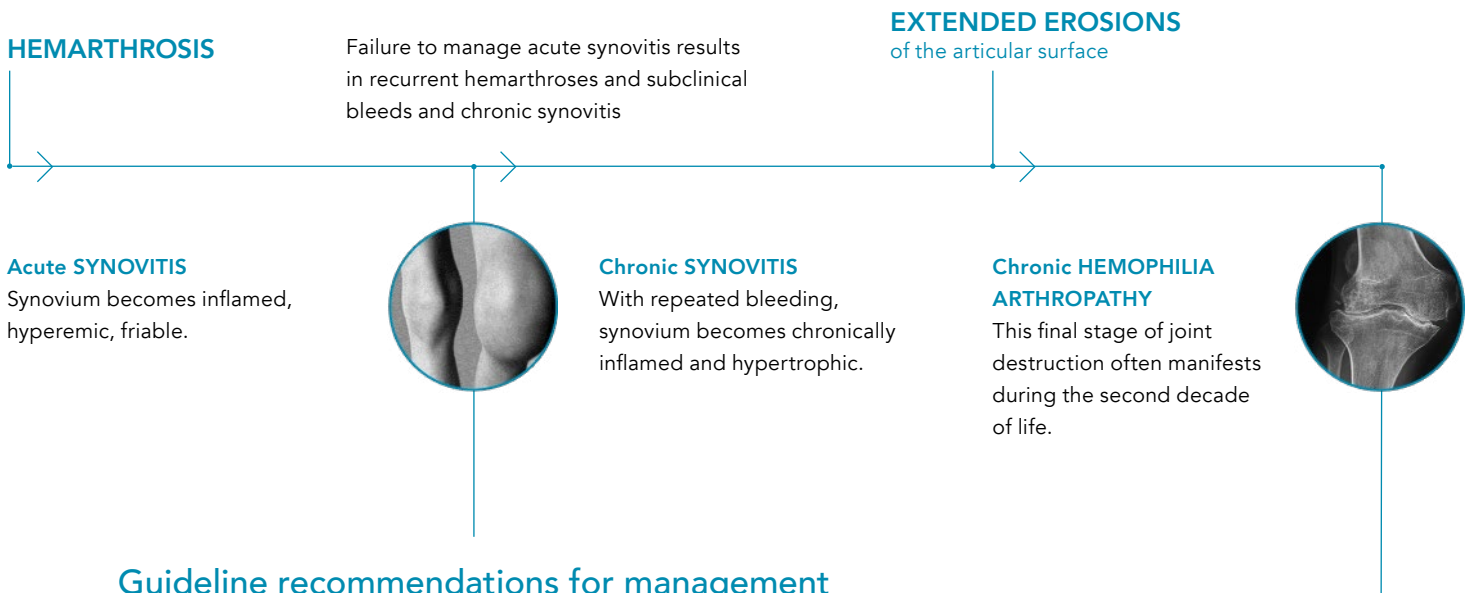


Standard of care to prevent bleeding
PROPHYLAXIS

Complete functional recovery generally requires

CFC REPLACEMENT THERAPY & PHYSICAL THERAPY

RECURRENT JOINT BLEEDS CAUSE PROGRESSIVE JOINT DAMAGE:



Guideline recommendations for management

- Clinical assessment alone is inadequate to detect early synovitis, therefore ultrasound (preferred) or MRI is advised.
- Synovial condition should be reassessed after every bleed and until fully rehabilitated.

Nonsurgical options for chronic synovitis:

- CFCs or other hemostatic coverage (e.g., bypassing agents for patients with inhibitors) and physical therapy.
- For patients with no access to regular prophylaxis, short-term prophylaxis (6-8 weeks) is recommended

- **Consult** with an experienced musculoskeletal specialist in a hemophilia treatment centre.
- **Nonsurgical synovectomy** is a first-line treatment option for unresolved chronic synovitis.

- **Combine** regular replacement therapy and physical therapy.
- If nonsurgical measures fail, **consult with an orthopedic specialist** on surgical intervention options.

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OTHER MUSCULOSKELETAL COMPLICATIONS OF HEMOPHILIA INCLUDE:

Muscle hemorrhage

Bleeding into a muscle, determined clinically and/or by imaging studies

- **Treat immediately** with clotting factor replacement therapy.
- **Assess pain frequently** for early indication of reversible damage.
- **Monitor continuously** for possible compartment syndrome (fasciotomy within 12h is associated with improved patient outcomes).

Inadequate treatment can lead to **compartment syndrome** with secondary neurovascular and tendon damage, muscle contracture, and necrosis.

Pseudotumours

Rare complication of inadequately treated soft tissue bleeds consisting of progressive cystic swelling of muscle and/or bone

- **Assess and serially follow up using ultrasound** (CT and MRI for more detail and accuracy).
- **Small early pseudotumours:** Treat with 6 to 8 weeks of clotting factor replacement therapy; repeat evaluation after 4 to 6 months.
- **Large pseudotumours:** Treat by surgical excision followed by close monitoring and long-term prophylaxis.

Joint replacement

- Consider **only if not responsive** to nonsurgical or other surgical treatments.
- Start postoperative physical therapy as soon as possible.

Surgical considerations:

- Meticulous hemostasis is critical for procedure success.
- Usually, no need for deep vein thrombosis prophylaxis unless very high plasma levels are maintained during the postoperative period.
- Antibiotic-loaded cement should be used in all cases where cement fixation is performed.

Fractures

Patients with hemophilic arthropathy may be at risk for fractures around a joint with significant loss of motion and in osteoporotic bones.

- **Treat immediately** with clotting factor concentrates or other hemostatic agents. (Maintain **factor levels ≥ 50 IU/dL** for at least a week.)
- Consider **external fixators** for open or infected fractures; **use splints** over full casts to prevent compartment syndrome.
- Avoid prolonged immobilization.

Orthopedic surgery

Simultaneous or staggered multiple-site elective procedures may:

- Expediate recovery of gait and overall function
- Allow for judicious use of factor replacement therapy or other hemostatic agents

- **Control blood oozing** with lignocaine/lidocaine and/or bupivacaine + an adrenaline and fibrin sealant/spray.
- Postoperative continuous infusion of factor replacement therapy is preferred.
- Both **pre- and postoperative** physical therapy is needed for optimal outcomes.

Psychosocial impacts

- **Develop tailored interventions** to help individuals adapt to pain and functional impairment and develop coping strategies.
- **Promote** support networks, peer mentoring, and group educational opportunities.