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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

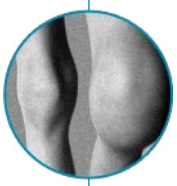
RECURRENT JOINT BLEEDS CAUSE PROGRESSIVE JOINT DAMAGE:

HEMARTHROSIS

Failure to manage acute synovitis results in recurrent hemarthroses and subclinical bleeds and chronic synovitis

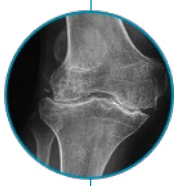
EXTENDED EROSIONS
of the articular surface

Acute SYNOVITIS
Synovium becomes inflamed, hyperemic, friable.



Chronic SYNOVITIS
With repeated bleeding, synovium becomes chronically inflamed and hypertrophic.

Chronic HEMOPHILIA ARTHROPATHY
This final stage of joint destruction often manifests during the second decade of life.



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.

This educational material was made possible through the support of the Hemophilia Alliance
For more information on the WFH Guidelines for the Management of Hemophilia, visit www.WFH.org/TGResourceHub