Chapter 10: **Musculoskeletal Complications**

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RECOMMENDATIONS

10.2 | Synovitis

Recommendation 10.2.1

For people with hemophilia, the WFH recommends regular physical assessment of the synovial condition after every bleed, preferably using suitable imaging techniques such as ultrasound until the situation is controlled, as clinical assessment alone is inadequate to detect early synovitis. **CB**

Recommendation 10.2.2

For patients with hemophilia who have chronic synovitis and no access to regular prophylaxis, the WFH recommends nonsurgical treatment, including short-term prophylaxis for 6-8 weeks to control bleeding; physical therapy to improve muscle strength and joint function; and selective COX-2 inhibitors to reduce pain and inflammation.

- REMARK: Physical therapy with individualized goals and exercises based on the patient's functional level should start slowly with increasing progression of weight-bearing activities.
- REMARK: For patients with acute pain and recurrent bleeding, bracing may stabilize the affected joint and limit motion, but caution is advised as prolonged immobilization leads to muscle weakness, so isometric exercises even within bracing are advised.
- REMARK: If unresponsive to nonsurgical interventions, treatment should be escalated to treat the synovitis directly, by the treatment intervention of the local expert. **CB**

Recommendation 10.2.3

For patients with hemophilia who have chronic synovitis (characterized only by minimal pain and loss of range of motion) the WFH recommends consultation with an experienced musculoskeletal specialist in a hemophilia treatment center. **CB**

Recommendation 10.2.4

For patients with hemophilia who have unresolved chronic synovitis, the WFH recommends nonsurgical synovectomy as a first-line treatment option using radioisotope synovectomy with a pure beta emitter (phosphorus-32, yttrium-90, rhenium-186, or rhenium-188). One dose of CFC per dose of isotope should be used.

- REMARK: Choice of isotope depends on the joint being injected and isotope availability.
- REMARK: The joint should be immobilized for at least 24 hours, followed by progressive rehabilitation for restoration of strength and function.

  REMARK: When radioisotopes are not available, chemical synoviorthesis with either rifampicin or oxytetracycline chlorhydrate (once weekly injection for 5-6 weeks) is an alternative, accompanied by one dose of CFC per treatment, a local anesthetic, and oral analgesics. **CB**

Recommendation 10.2.5

For patients with hemophilia who have chronic synovitis that no longer responds to nonoperative interventions, the WFH recommends surgical synovectomy (preferably arthroscopic, not open) only by an experienced team in a hemophilia treatment centre. **CB**

10.3 | Hemophilic arthropathy

Recommendation 10.3.1

For the prevention and treatment of chronic hemophilic arthropathy in people with hemophilia, the WFH recommends a combination of regular replacement therapy to reduce frequency of bleeding and physical therapy aimed at preserving muscle strength and functional ability. Physical therapy may be done with or without factor coverage, depending on availability and the patient's response to therapy. **CB**

Recommendation 10.3.2

For the prevention and treatment of the sequelae of joint arthropathy in people with hemophilia, the WFH recommends nonsurgical measures such as bracing, orthotics, mobility aids, and serial casting and traction devices to aid in the correction of flexion contractures. This may be done with or without factor coverage. **CB**

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Recommendation 10.3.3

For patients with hemophilia with chronic hemophilic arthropathy for whom nonsurgical measures have failed to provide satisfactory pain relief and improved function, the WFH recommends consultation with an orthopedic specialist on surgical intervention options which may include:

- synovectomy and joint debridement;
- arthroscopy to release intra-articular adhesions and correct impingement;
- extra-articular soft tissue release to treat contractures;
- osteotomy to correct angular deformity;
- arthrodesis (of the ankle);
- joint replacement in end-stage arthritis.

- REMARK: Adequate resources including a sufficient supply of CFCs or other appropriate hemostatic agents (e.g., bypassing agents for patients with inhibitors) and postoperative rehabilitation services must be available to increase the likelihood of success for any surgical procedure. **CB**

10.4 | Muscle hemorrhage

Recommendation 10.4.1

All hemophilia patients with muscle bleeds should be given clotting factor replacement therapy immediately and, where applicable, be observed for neurovascular complications associated with the bleed. **CB**

Recommendation 10.4.2

For all hemophilia patients with muscle bleeds, the WFH recommends detailed clinical assessment, grading, and monitoring of pain according to the WHO pain scale, as muscle bleed pain may be an early indicator of reversible neurovascular and tissue damage.

- REMARK: While many pain assessment scales exist, use of the WHO pain scale is preferred because it is a simple and universal tool that permits uniform measurement of pain in people with hemophilia and generates comparable population-level outcome data important to advancing hemophilia treatment and research. **CB**

Recommendation 10.4.3

In hemophilia patients with muscle bleeds with evidence of compartment syndrome and neurovascular compromise, a fasciotomy is required within 12 hours from the time of onset of symptoms before irreversible damage sets in due to tissue necrosis. **CB**

10.5 | Pseudotumours

Recommendation 10.5.1

For hemophilia patients with soft tissue bleeding and signs of a possible pseudotumour, the WFH recommends clinical assessment and radiological confirmation using X-ray, ultrasound, and magnetic resonance imaging, as appropriate.

- REMARK: While ultrasound is useful for serial assessment of a soft tissue pseudotumour, MRI provides more detailed information prior to surgical intervention.
- REMARK: A CT scan or CT angiogram may be indicated, especially for a large pseudotumour and/or pre-operative planning. **CB**

Recommendation 10.5.2

For patients with hemophilia who have developed small early pseudotumours (prior to acquiring a pseudocapsule) and have no access to regular prophylaxis, the WFH recommends a short course (6-8 weeks) of clotting factor replacement therapy with possible continuation of therapy if serial ultrasound evaluations indicate that the pseudotumour is shrinking, with repeat evaluation after 4-6 months. **CB**

Recommendation 10.5.3

For patients with hemophilia who have developed large pseudotumours, the WFH recommends surgical excision of the pseudotumour with the pseudocapsule, performed only by a surgical team with experience in hemophilia, in a hemophilia treatment centre wherever possible, followed by close monitoring and long-term prophylaxis to prevent recurrence of bleeding. **CB**

- REMARK: Fluctuations in factor levels during the first postoperative year may increase the likelihood of bleed recurrence. Therefore, close monitoring and optimal correction of factor levels are of paramount importance. **CB**

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10.6 | Fractures

Recommendation 10.6.1

For people with hemophilia who incur fractures, the WFH recommends immediate treatment with clotting factor concentrates or other hemostatic agents, and continued treatment to maintain sufficiently high factor levels for bleed control for a week or longer, depending on the likelihood of bleeding due to fracture site or stability. Subsequently, lower factor levels may be maintained for 10-14 days to prevent soft tissue bleeding while the fracture becomes stabilized. Clinical monitoring is paramount due to the risk of compartment syndrome. **CB**

Recommendation 10.6.2

For people with hemophilia who incur fractures, the WFH recommends splints over full casts to avoid compartment syndrome (especially in the early stages), and external fixators for open or infected fractures. **CB**

Recommendation 10.6.3

For people with hemophilia who incur fractures, the WFH recommends avoiding prolonged immobilization and advises supervised physical therapy and rehabilitation as soon as the fracture is stabilized to restore range of motion, muscle strength, and function. **CB**

10.7 | Orthopedic surgery in hemophilia

Recommendation 10.7.1

For patients with hemophilia requiring orthopedic surgery, especially in cases where oozing is present at closure as well as dead space or cavities, the WFH suggests the use of local coagulation enhancers and wound infiltration with local anesthetic agents (lignocaine/lidocaine and/or bupivacaine) with an adrenaline and fibrin sealant or spray to control blood oozing when operating in extensive surgical fields. **CB**

Recommendation 10.7.2

For patients with hemophilia requiring orthopedic surgery, the WFH recommends factor replacement therapy and close pain control and monitoring, with higher doses of analgesics in the immediate postoperative period. **CB**

Recommendation 10.7.3

For patients with hemophilia in the postoperative period following orthopedic surgery, the WFH recommends gradual rehabilitation by a physical therapist experienced in hemophilia management. **CB**

10.8 | Joint replacement

Recommendation 10.8.1

For patients with hemophilia, the WFH recommends joint replacement only in cases of established hemophilic arthropathy that is not responsive to nonsurgical or other surgical treatments, and that is accompanied by associated pain, functional impairment, and loss of participation in activities of daily living.

- REMARK: Perioperatively, tranexamic acid and fibrin sealants may be used to reduce blood loss.
- REMARK: Physical therapy should ideally start on the day of surgery with early mobilization and appropriately progressive exercises to regain motion and muscle strength. **CB**

10.9 | Psychosocial impacts of musculoskeletal complications

Recommendation 10.9.1

For patients with hemophilia who have chronic musculoskeletal pain or functional limitations, the WFH recommends psychosocial interventions tailored to meet the specific needs of each individual based on their physical, emotional, social, educational, and cultural circumstances. **CB**

Recommendation 10.9.2

For patients with hemophilia who have chronic musculoskeletal pain or functional limitations, the WFH recommends specific individualized psychosocial assessments and intervention strategies aimed at achieving better quality of life, including psychosocial counselling, educational and employment counselling, and financial planning. **CB**

Recommendation 10.9.3

For patients with hemophilia who have chronic musculoskeletal pain or functional limitations, the WFH recommends the promotion of support networks, peer mentoring, and group educational opportunities to support their ability to cope with musculoskeletal complications, reduce social isolation, and strengthen resilience. **CB**

CB: Consensus based; CFC, clotting factor concentrate; MRI, Magnetic resonance imaging; CT, computerized tomography.

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