Chapter 2: **Comprehensive Care of Hemophilia**

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RECOMMENDATIONS

2.2 | Comprehensive care

Recommendation 2.2.1

For people with hemophilia, the WFH recommends coordinated delivery of comprehensive care by a multidisciplinary team of healthcare professionals with expertise and experience in hemophilia.

• REMARK: The core members of the comprehensive care team should consist of a medical director, nurse coordinator, musculoskeletal specialists, medical laboratory specialist, psychosocial specialist, and the patient and family caregivers. The roles assumed by the core team members may differ at different centres depending on the availability and expertise of trained staff and the organization of services within the centre. **CB**

Recommendation 2.2.2

For people with hemophilia, the WFH recommends availability of and access to:

◦ appropriate emergency care at all times;

◦ a coagulation laboratory capable of performing clotting factor assays and inhibitor testing;

◦ appropriate clotting factor concentrates (CFCs), either plasma-derived or recombinant, as well as other hemostatic agents such as desmopressin (DDAVP), emicizumab, and antifibrinolytics;

◦ safe blood components such as fresh frozen plasma (FFP) and cryoprecipitate that have been adequately screened, tested, and/or virus-inactivated if CFCs are not available;

◦ casting and/or splinting for immobilization and mobility/support aids, as needed;

◦ other specialists to address specific medical and health-related issues that some individuals may encounter, as needed. **CB**

Recommendation 2.2.3

For all patients with hemophilia, the WFH suggests the preparation of written clinical management protocols to ensure continuity of care in the event of changes in clinic personnel. **CB**

Recommendation 2.2.4

For people with hemophilia, the WFH recommends a multidisciplinary checkup including hematologic, musculoskeletal, and quality-of-life assessments by the core comprehensive care team members at least yearly (every 6 months for children).

• REMARK: Smaller centres and family physicians can provide primary care and management of some complications of hemophilia, in frequent consultation with the hemophilia comprehensive care centre, especially for patients who live a long distance from the nearest hemophilia treatment centre.

Recommendation 2.2.5

For all patients with hemophilia, the WFH recommends systematic data collection in patient registries, where possible, to inform allocation of resources, support improvement of care delivery services, and promote collaboration among centres in sharing data and conducting research. **CB**

Recommendation 2.2.6

The WFH recommends that adequate education be provided to people with hemophilia, their family members, and other caregivers to enable self-management and sufficient understanding of the disease for prevention of bleeds and related complications and for life planning. **CB**

Recommendation 2.2.7

For people with hemophilia and their families, the WFH recommends promotion and/or facilitation of educational and recreational activities in collaboration with patient organizations, to provide them with opportunities to discover new interests and capabilities and build a support network with diverse members of the hemophilia community. **CB**

2.3 | Fitness and physical activity

Recommendation 2.3.1

For people with hemophilia, the WFH recommends promotion of regular physical activity and fitness, with special attention on bone health maintenance, muscle strengthening, coordination, physical functioning, healthy body weight, and positive self-esteem. **CB**

Recommendation 2.3.2

For people with hemophilia, the WFH recommends promotion of non-contact sports. High-contact and collision sports and high-velocity activities should be avoided unless the individual is on a prophylactic regimen that is adequate to cover such activities and is properly educated on the potential risks and other required protective measures.

• REMARK: The choice of sports activities should take into consideration the individual's physical condition and ability, preferences and interests, local customs, and available resources. **CB**

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Recommendation 2.3.3

For people with hemophilia, the WFH recommends consultation with a physical therapist or other musculoskeletal specialist before engaging in sports and physical activities to discuss their appropriateness specific to the individual's condition and their requirement for particular physical skills and/or protective gear. CB

2.4 | Adjunctive management

Recommendation 2.4.1

For people with hemophilia with a muscle or joint bleed, the WFH recommends following the PRICE principles (protection, rest, ice, compression, and elevation) in addition to increasing factor level. CB

Recommendation 2.4.2

For people with hemophilia recovering from a joint or muscle bleed, the WFH recommends gradual re-initiation of physical activities under the supervision of a physical therapist with experience in hemophilia to assess resumption of normal motor development and coordination.

• REMARK: For children with hemophilia recovering from a joint or muscle bleed, the physical therapist and family caregiver should remain in close contact to discuss and decide on the appropriate sports and activities for the child's progressive rehabilitation. CB

Recommendation 2.4.3

For people with hemophilia with established hemophilic arthropathy or after recovery from musculoskeletal bleeding, the WFH recommends physical therapy and rehabilitation activities. CB

Recommendation 2.4.4

For people with hemophilia, the WFH recommends the use of antifibrinolytic drugs (e.g., tranexamic acid, epsilon aminocaproic acid [EACA]) alone or as adjuvant treatment, particularly in controlling mucosal bleeds and for invasive dental procedures. CB

2.5 | Home therapy

Recommendation 2.5.1

Patients (or caregivers of children) with hemophilia should be taught how to manage their care at home and be able to demonstrate understanding of how to recognize bleeds and the ability to infuse or self-infuse, with monitoring of venous access skills over the patient's lifetime. CB

Recommendation 2.5.2

For patients with hemophilia, a detailed record of all treatments administered (reason, batch number, number of units, etc.) should be documented and used to personalize treatment plans. CB

Recommendation 2.5.3

For children with hemophilia, central venous access devices could be considered to facilitate early access to bleed treatment and prophylaxis. CB

2.6 | Pain management

Recommendation 2.6.1

For people with hemophilia with acute or chronic pain, the WFH recommends the use of age-appropriate pain assessment tools to determine the cause and guide appropriate management. CB

Recommendation 2.6.2

For people with hemophilia with venous access pain, discomfort or anxiety, the WFH recommends the application of a local anesthetic spray or cream at the site of venous access. CB

Recommendation 2.6.3

For people with hemophilia with acute pain due to a joint or muscle bleed, the WFH recommends immediate administration of clotting factor concentrates to stop bleeding, pain medication, and adjunctive measures such as immobilization, compression, and splinting to minimize pain, if appropriate. CB

Recommendation 2.6.4

For patients with hemophilia and postoperative pain, the WFH advises proportionate management of postoperative pain in coordination with the anesthesiologist or pain specialist. CB

Recommendation 2.6.5

For patients with hemophilia and postoperative pain, the WFH recommends analgesia similar to that used in patients without hemophilia including, as appropriate, the use of intravenous morphine or other narcotic analgesics, followed by an oral opioid (e.g., tramadol, codeine, hydrocodone, etc.) and paracetamol/acetaminophen as pain decreases.

• REMARK: With the exception of selective COX-2 inhibitors, NSAIDs should not be used in patients with hemophilia.

• REMARK: The intramuscular route for administration of analgesia is not advised. CB

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Recommendation 2.6.6
For people with hemophilia and chronic hemophilic arthropathy in need of pain management, the WFH recommends functional training and adaptations alongside appropriate analgesics. CB

Recommendation 2.6.7
For people with hemophilia and chronic hemophilic arthropathy, the WFH recommends education on pain management including the use of complementary pain management techniques (e.g., meditation, distraction, mindfulness, or music therapy). CB

Recommendation 2.6.8
For children and adults with hemophilia with pain due to chronic hemophilic arthropathy, the WFH recommends the use of paracetamol/acetaminophen, selective COX-2 inhibitors, tramadol, or morphine, and avoidance of other NSAIDs. Codeine may be used for children over 12 years of age but is contraindicated in younger children.
• REMARK: Prolonged use of these medications may have risks of dependence or addiction, as well as organ damage, and must be carefully monitored.
• REMARK: People with persistent pain should be referred to a specialized pain management team. CB

Recommendation 2.6.9
For patients with hemophilia with disabling pain from chronic hemophilic arthropathy, the WFH recommends referral to an orthopedic specialist for consideration of orthopedic surgery. CB

Recommendation 2.6.10
For children and adults with hemophilia, the WFH recommends interim management of dental or orofacial pain according to a proportionate approach for pain relief and referral to a dental care professional for assessment. CB

2.7 | Dental care management

Recommendation 2.7.1
For children and adults with hemophilia, the WFH recommends provisions for access to regular preventive dental and oral health care as part of comprehensive hemophilia care. CB

Recommendation 2.7.2
For children with hemophilia, the WFH recommends referral to a designated dental care centre at the time of the first tooth eruption (around 6 months of age) or by age 1 in order to reduce the complications, morbidity, costs, and health and psychosocial impacts associated with oral diseases in people with hemophilia. CB

Recommendation 2.7.3
For adults with hemophilia, the WFH recommends facilitating access to appropriate adult dental services and procedures, with regular dental assessments throughout their lives to monitor and safeguard oral health using evidence-based and personalized preventive dental protocols. CB

Recommendation 2.7.4
For people with hemophilia, the WFH recommends preventive dental and oral care as a priority to ensure optimal oral health and hygiene to prevent periodontal disease and dental caries, which predispose to gum bleeding, dental pain, tooth loss, chewing difficulties, and social impacts. CB

Recommendation 2.7.5
For all people with hemophilia, the WFH recommends education on the importance of good oral hygiene to prevent dental problems and complications, including instructions for twice-daily brushing of the teeth using a soft- or medium-texture toothbrush and fluoridated toothpaste to remove plaque deposits; the toothpaste should not be rinsed away but rather retained ("spit, but don't rinse") after brushing to maximize fluoride benefit.
• REMARK: The use of dental floss or interdental brushes should be encouraged to ensure complete plaque removal.
• REMARK: Individuals with elbow or shoulder restrictions may benefit from modified or electric toothbrushes and flossing aids. CB

Recommendation 2.7.6
For children with hemophilia 6 years of age and younger, the WFH recommends parental/caregiver supervision of toothbrushing. CB

Recommendation 2.7.7
For patients with hemophilia, the WFH recommends that dental extraction or other invasive procedures within the oral cavity (e.g., dental implantation, periodontal surgery, or gum biopsy) be performed only with a personalized plan for hemostasis management in consultation with a hematologist. CB

Recommendation 2.7.8
For patients with hemophilia, the WFH recommends the use of systemic or topical tranexamic acid or epsilon aminocaproic acid (EACA) as adjunct treatment in the management of dental interventions pre- and postoperatively, to reduce the need for factor replacement therapy. CB

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Recommendation 2.7.9
For patients with hemophilia requiring dental extractions, the WFH recommends local hemostatic measures. Typical procedures include wound suture, topical use of antifibrinolytics, oxidized cellulose, and fibrin sealant, applied as appropriate.
• REMARK: Patients should be advised to maintain a soft diet and undertake careful brushing around the wound site for a minimum of 3-5 days postoperatively to avoid disturbing the clot and wound healing within the tooth socket. CB

Recommendation 2.7.10
For patients with hemophilia, the WFH recommends appropriate local anesthesia for dental treatments as an essential part of pain and anxiety management. Most dental injections pose a low risk for patients with hemophilia when delivered by a dental care professional using local anesthesia with a vasoconstrictor, and when the agent is delivered slowly with a single-use fine-gauge needle. CB

Recommendation 2.7.11
For patients with hemophilia requiring higher-risk intramuscular oral injections commonly associated with the provision of surgical dentistry (such as inferior alveolar dental block [IDB], superior alveolar nerve block, or injections in the floor of the mouth or vascular lingual tissues), the WFH recommends systemic hemostatic measures preoperatively to avoid the risk of hematoma. These measures should be established in consultation with the hematologist.
• REMARK: The availability and effectiveness of alternative low risk routes of local anesthetic delivery (such as intraligamentary single-tooth anesthesia, or buccal infiltration injections with 4% articaine) are effective alternatives to IDB and permit dental procedures in primary and permanent mandibular molar teeth. CB

Recommendation 2.7.12
For patients with hemophilia, the WFH recommends the use of antifibrinolytic agents as effective adjunct treatment in the management of dental hygiene therapies that facilitates access to regular dental care delivered by a dental hygienist. CB

Recommendation 2.7.13
In patients with hemophilia, the WFH asserts that the presence of blood-borne infections does not affect the safety of dental treatment as stringent universal cross-infection procedures are now mandatory across all disciplines of dentistry and recommends the provision of full dental services regardless of infectivity or immunological status. CB

2.8 | Transition from pediatric to adult care

Recommendation 2.8.1
Children and adolescents with hemophilia should be supported with ongoing education and skills development, including the ability to self-infuse and other self-efficacy skills, to gain necessary hemophilia knowledge for self-management of their condition before they make the transition from pediatric to adult care.
• REMARK: The comprehensive care team should support young patients and their families through the transition period. When possible, the first visit should be performed by both the pediatric and adult hematologists. CB

Recommendation 2.8.2
For adolescents with hemophilia on prophylaxis, the WFH recommends individual education and training, ideally from a hemophilia nurse coordinator, to ensure adequate knowledge of hemophilia, and to support prophylaxis adherence and self-care management. This should include understanding measurements of adherence, as well as factors and risks that can lead to changes in bleeding rates. CB

Recommendation 2.8.3
For adolescents 12-18 years of age with hemophilia, the WFH recommends age-specific hemophilia camps to foster peer group support and develop their self-infusion skills and understanding of the importance of adherence to treatment. CB

CB, consensus based; CFCs, clotting factor concentrates, DDAVP, desmopressin; EACA, epsilon aminocaproic acid; FFP, fresh frozen plasma; IDB, inferior alveolar dental block; NSAIDs, nonsteroidal anti- inflammatory drugs; PRICE, protection, rest, ice, compression, and elevation.

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For more information on the WFH Guidelines for the Management of Hemophilia, visit www.WFH.org/TGResourceHub
THE WFH GUIDELINES FOR THE MANAGEMENT OF HEMOPHILIA 3RD EDITION