

Anesthesia recommendations for **Hemophilia A**

Disease name: Hemophilia A

ICD 10: D66

ORPHAcode: 98878

Synonyms: Classic hemophilia, Factor VIII deficiency

Brief disease summary: Hemophilia A is a rare inherited X-linked recessive bleeding disorder, resulting from factor VIII (FVIII) deficiency and characterized by intra-articular and intramuscular bleeding. Numerous different mutations can cause hemophilia A. Due to differences in the gene involved (and the subsequent resulting protein), patients with hemophilia (PWH) have varying levels of FVIII clotting activity [1]. Individuals with less than 1% FVIII clotting activity are classified as having 'severe' hemophilia, those with 1-5% as 'moderate', and those with between 5-40% as mild. Most severe hemophilia patients require regular supplementation with intravenous recombinant or plasma-derived FVIII concentrate (prophylaxis). When surgical procedures are needed in hemophiliac patients, perioperative management should be planned with a multidisciplinary team [2].

Diagnosis may be incorrect; if uncertainty exists, the diagnosis should be re-evaluated.

Every patient is unique; individual circumstances must always guide clinical care.

Medicine is in progress; new clinical knowledge may not be yet reflected in this recommendation.



Recommendations are not rules or laws; they provide a framework to support clinical decision-making. Although this recommendation has passed a structured review process, it does not meet the formal criteria of a guideline.

Translations may not always reflect the most recent updates of the English version.



Find more information on the disease, its centers of reference and patient organizations on Orphanet: www.orpha.net

Emergency information

A	AIRWAY / ANESTHETIC TECHNIQUE	General anesthesia: all intravenous and volatile anesthetics can be used. Difficult intubation is rare. There is no increase in risk of aspiration. Regional or neuraxial anesthesia are best avoided. Local anesthetic infiltration with lidocaine is very commonly used for minor procedures after adequate factor replacement therapy. Patients with a severe coagulation disorder require specific preparatory measures, such as administration of FVIII. Consultation and coordination with the patient's hematologist or hemophilia treatment center are essential.
B	BLOOD PRODUCTS (COAGULATION)	Patients with hemophilia A must be provided FVIII concentrates. If a patient with hemophilia A and inhibitors is receiving emicizumab prophylaxis, treatment should be switched to recombinant factor VIIa (rFVIIa) or an activated prothrombin complex concentrate (aPCC), such as Factor Eight Inhibitor Bypassing Agent (FEIBA). Prophylactic tranexamic acid may be useful. Routine coagulation profile should include activated partial thromboplastin time (aPTT), prothrombin time (PT), fibrinogen activity (Clauss assay), plasma FVIII concentration, and testing for FVIII inhibitors. aPTT and PT, while useful in the initial evaluation of a patient with suspected hemophilia, are generally irrelevant tests once the diagnosis of hemophilia has been established.
C	CIRCULATION	There is no increased risk of cardiac or hemodynamic problems. There is a need to provide sufficient FVIII trough levels at the time of antithrombotic treatment in people with hemophilia A without inhibitors.
D	DRUGS	NSAIDs are best avoided because of the potential risk of hemorrhagic complications. Multimodal analgesia should be used to enhance synergistic effects, reduce opioid requirements, and minimize side effects.
E	EQUIPMENT	Regular monitoring of plasma FVIII levels in the postoperative period until the risk of hemorrhagic complications is over.

Additional disease information

None.

Typical surgery and procedures

Orthopedic surgery: hemarthrosis involving hip and shoulder joints (ball & socket joints) is uncommon in hemophilia. A diagnosis of hip hemarthrosis is often a misdiagnosis of an iliopsoas muscle bleed [1].

Total knee replacement or total elbow replacement, and ankle arthrodesis (all hinged joints) are the most common hemophilia-related surgical interventions. Surgery for non-hemophilia-related conditions (e.g., adenotonsillectomy, hernia repair, appendectomy, and cholecystectomy) is also commonly performed in PWH [2].

Type of anesthesia

General anesthesia is the best choice: all intravenous and volatile anesthetics can be used.

Regional or neuraxial anesthesia are best avoided as there is a high risk of hemorrhagic complications, such as epidural hematoma. Local anesthetic infiltration with lidocaine is very commonly used for minor procedures such as dental extractions in PWH, after adequate factor replacement therapy [3,4].

Necessary additional preoperative testing (beside standard care)

Hemophilia A is a bleeding disorder; therefore, its diagnosis requires evaluation of relevant laboratory tests.

Hemophilia A should be considered in children presenting with unexplained spontaneous bleeding (after ruling out child abuse) or delayed bleeding following surgery [5,6]. The disease is caused by a de novo mutation in 30% of cases; therefore, a positive family history may be absent. Moreover, hemophilia A has also been reported in girls (1.7% of patients with hemophilia A in the USA).

Routine laboratory tests of blood coagulation: a prolonged aPTT is observed and FVIII and von Willebrand factor assays should be performed to evaluate for hemophilia. The severity of the defect is classified according to the circulating FVIII levels. Individuals with less than 1% FVIII clotting activity are classified as having 'severe' hemophilia (50%), those with 1-5% as 'moderate' (10-20%), and those with between 5-40% as "mild" (30-40%). The latter may remain clinically asymptomatic until they face a hemostatic challenge (trauma, surgery) [1].

Thromboelastography (TEG) and thromboelastometry have no role in the day-to-day diagnosis or management of uncomplicated hemophilia. They are used in a limited number of centers to guide the choice or use of bypassing agents (FEIBA or rFVIIa) in hemophilia patients with inhibitors. Otherwise, TEG and other global hemostatic assays have no established role in the diagnosis or routine management of hemophilia. In cases of major bleeding, however, they may help guide the replacement of other coagulation factors (e.g., fibrinogen), as in patients without hemophilia [4,7].

Routine coagulation profile should include aPTT, PT, fibrinogen activity (Clauss assay), plasma FVIII concentration, and testing for FVIII inhibitors. aPTT and PT, while useful in the initial evaluation of a patient with suspected hemophilia, are generally irrelevant tests once the diagnosis of hemophilia has been established [1].

The patient should have been vaccinated against hepatitis A and B. Hepatitis C virus (HCV) has been the major cause of liver disease in hemophilia. Blood samples must be obtained for ALT, AST, HCV antibody, HCV genotype, HIV antibody, platelet count, hemoglobin. HCV and HIV viral loads are only required if patients have tested positive for the respective antibody [8].

Particular preparation for airway management

Our own retrospective data showed a very rare case of difficult intubation.

There is no increase in risk of aspiration as compared with non-hemophilic patients.

Particular preparation for transfusion or administration of blood products

Hemophilia A is a disorder caused by missing or defective FVIII, a clotting protein, so there is a high risk of bleeding if perioperative factor replacement is inadequate. There may be a higher requirement for blood products during surgery. Patients with hemophilia A must be provided FVIII concentrates. Hemophilia patients have no excess requirements for fresh frozen plasma (FFP) or packed red cells compared to non-hemophilia patients, provided perioperative FVIII replacement has been adequate. Cryoprecipitate (which contains FVIII, von Willebrand factor and fibrinogen) is used in the treatment of hemophilia A only when and where FVIII concentrates are not available. These blood products (FFP, packed red blood cells, and cryoprecipitate) need only be available on standby and should be administered as clinically indicated. With adequate preparation, they are usually not required [1,2].

Patients with hemophilia A and inhibitors must be treated with rFVIIa or FEIBA. rFVIIa or FEIBA are licensed for the on-demand treatment of bleeding episodes and the prevention of bleeding in surgery or invasive procedures in patients with congenital hemophilia with inhibitors [10].

The prophylactic administration of tranexamic acid may be useful.

Particular preparation for anticoagulation

Patients with mild hemophilia A can maintain near-normal or even normal FVIII levels postoperatively as a result of the acute-phase response to surgery. Such patients are as prone to postoperative venous thrombosis as patients without hemophilia.

Particular precautions for positioning, transportation and mobilization

Some patients may have difficulty with mobility associated with hemarthroses. It depends on the degree of arthropathy. Some mild forms may be clinically silent [1,4].

Interactions of chronic disease and anesthesia medications

The use of narcotic analgesics by PWH with chronic pain syndrome (as a result of hemophilic arthropathy) can sometimes complicate postoperative analgesia [4]. Analysis of our own retrospective data showed paracetamol and opiates to be both highly effective and safe on the first postoperative day following highly traumatic surgical procedures in PWH. Patient-controlled analgesia is highly effective in postoperative analgesia [1,9].

Long-term use of paracetamol should be avoided in PWH with hepatitis C (hepatotoxicity) [11]. NSAIDs are best avoided because of the potential risk of hemorrhagic complications [4]. Given a number of limitations on the use of perioperative analgesia in PWH, wound analgesia (with ropivacaine, using a system for anesthetizing surgical wounds) may be one of the components of multimodal analgesia in this category of patients [12].

Anesthetic procedure

Patients with severe and moderate hemophilia A must be treated with FVIII concentrates before induction of anesthesia, because there is a high risk of hemorrhagic complications, such as hematoma of the epiglottis and trauma-induced bleeding from the upper respiratory tract. Most mild hemophilia patients do not necessarily require FVIII concentrates before intubation or surgery. Most mild hemophilia patients respond very well to desmopressin (DDAVP), which can increase FVIII (and von Willebrand factor) levels three- to fivefold above baseline. DDAVP is the hemostatic agent of choice for mild hemophilia, not FVIII concentrate. Drug selection will also depend on the extent and type (invasiveness and hemorrhagic risk) of surgery [1].

Patients with hemophilia A and inhibitors must be treated with rFVIIa or FEIBA before intubation [10].

Particular or additional monitoring

The heart rate variability-based Analgesia Nociception Index (ANI) has been proposed to reflect different levels of acute pain [13].

Possible complications

Hemophilia A is a disorder caused by missing or defective FVIII, a clotting protein, so there is a high risk of bleeding. Hemorrhagic complications may occur if perioperative hemostatic preparation is inadequate or is poorly planned.

Postoperative care

The degree of postoperative monitoring is dependent on the surgical procedure and preoperative condition of the patient [1,3,4].

It is essential to exclude the presence of inhibitors before surgery (at most 1 week before surgery) and to ascertain that FVIII levels are at the desired level for the particular type of surgery. aPTT and other routine coagulation tests are not clinically helpful in a patient with a

known diagnosis of hemophilia. In the postoperative period, FVIII levels must be maintained at levels advised by the hematologist or hemophilia center where the patient is registered. Requirements for other blood products should be no different from those in patients without hemophilia and should be administered purely based on clinical circumstances. It is necessary only to have such products on standby and used only as and when required. Patients with hemophilia A and inhibitors will similarly require bypassing agents (rFVIIa or FEIBA) in the postoperative period, as advised by the responsible hematologist or hemophilia center [10].

Patients should be monitored for potential complications, such as periprosthetic joint infection and recurrent hemarthrosis [1].

Disease-related acute problems and effect on anesthesia and recovery

Patients may develop circulating inhibitor antibodies against FVIII following repeated exposure to FVIII concentrates [1].

Ambulatory anesthesia

Ambulatory anesthesia for surgery should be avoided in patients with severe and moderate hemophilia A because of the high risk of bleeding [14].

Obstetrical anesthesia

Cases of hemophilia in women are extremely rare. A female carrier of hemophilia A may have reduced FVIII levels. However, FVIII levels typically increase during pregnancy. This can make it difficult to identify women who have hemophilia A during pregnancy if they have not already been diagnosed. There is an increased risk of postpartum bleeding as FVIII levels decline toward baseline after delivery [15].

References

1. Srivastava A, Santagostino E, Dougall A, et al. WFH Guidelines for the Management of Hemophilia, 3rd edition. *Haemophilia*. 2020; 26 Suppl 6:1-158. doi:10.1111/hae.14046
2. Hanley J. Musculoskeletal Working Party of the UKHCDO. Guidelines for the management of acute joint bleeds and chronic synovitis in haemophilia: a United Kingdom Haemophilia Centre Doctors' Organisation (UKHCDO) guideline. / J. Hanley, A. McKernan, M.D. Creagh [et al.] // *Haemophilia*. – 2017. – P. 511–520
3. Flores RP, Bagatini A, Santos AT, Gomes CR, Fernandes MS, Molon RP. Hemophilia and anesthesia. *Rev Bras Anesthesiol*. 2004; 54(6):865-871. doi:10.1590/s0034-70942004000600017
4. Levchenko OK, Shulutko EM, Zorenko VY, Galstyan GM. The specific features of pain syndrome in patients with hemophilia. *Ter Arkh*. 2016; 88(7):84-88. doi:10.17116/terarkh201688784-88
5. Tagaya A, Miyajima M, Ito M, Nakamura L et al. Spontaneous spinal epidural hematoma in an infant with undiagnosed hemophilia A. *Pediatrics International* 2010; 52: 296-8.
6. Sumie M, Toyama K, Morokawa K, Yokoo-Matsuoka T et al. Inadvertent bleeding in an infant after inguinal hernioplasty leads to diagnosis of hemophilia B. *J Anesth* 2012; 26: 399-400.
7. Klamroth R, Ay C, De Moerloose P, et al. Applicability of the European Society of Cardiology Guidelines on the management of acute coronary syndromes to older people with haemophilia A - A modified Delphi consensus by the ADVANCE Working Group. *Haemophilia*. 2023; 29(1):21-32. doi:10.1111/hae.14674
8. Makris M, Baglin T, Dusheiko G, et al. Guidelines on the diagnosis, management and prevention of hepatitis in haemophilia. *Haemophilia*. 2001;7(4):339-345. doi:10.1046/j.1365-2516.2001.00527.x
9. Mahagna AA, Annunziata S, Torriani C, et al. Perioperative Pain Management in Hemophilic Patient Undergoing Orthopedic Surgery: A Narrative Review. *Healthcare (Basel)*. 2024;12(19):2007. Published 2024 Oct 8. doi:10.3390/healthcare12192007
10. Santagostino E, Escobar M, Ozelo M, Solimeno L, Arkhammar P, Lee HY, Rosu G, Giangrande P Blood Rev. Recombinant activated factor VII in the treatment of bleeds and for the prevention of surgery-related bleeding in congenital haemophilia with inhibitors. 2015 Jun;29 Suppl 1: S9-18.
11. VanVeen JJ, Gleeson DC, Makris M. Paracetamol/acetaminophen usage in haemophilia: more caution needed? *Haemophilia* 2008;14(3):434-435
12. Levchenko OK, Polyanskaya TU, Zorenko VY Galstyan GM. Wound analgesia in a patient with hemophilia in a highly traumatic operation. *Ter Arkh*. 2018; 90(7):82-85. doi:10.26442/terarkh201689082-85
13. Levchenko OK Berseneva EA The features of pain diagnostics in patients with blood system diseases. *The Bulletin of Contemporary Clinical Medicine*. 2018; 11(3):65-76. doi:10.20969/VSKM.2018.11(3).65-76
14. Nanayakkara L, Yahaya N, Parreira M, Bajkin B. Dental management of people with complex or rare inherited bleeding disorders. *Haemophilia*. 2024;30 Suppl 3:128-134. doi:10.1111/hae.15005
15. Vipra Sharma, MD1 Aysha Khalid, MD1 Alice J. Cohen, MD1 Management of Pregnancy in a Patient with Severe Hemophilia Type A. *American Journal of Perinatology Reports* 2013; 3: 29-32

Date last modified: **September 2026**

This recommendation was prepared in 2016 by:

Author(s)

Olga Levchenko, Reanimatologist, Anesthesiologist, Therapist, Hematologist, Healthcare organizer, National Research Center for Hematology, Moscow, Russia; Head of the Medical Accreditation and Simulation Center, National Medical Hematology Research Center, Moscow, Russia

levchenkokp@rambler.ru

Disclosure: The author(s) has no financial or other competing interest to disclose. This recommendation was unfunded.

This recommendation was reviewed by:

Reviewer(s)

Gennady M. Galstyan, Anesthesiologist, National Research Center for Hematology, Moscow, Russian Federation, Russia

Patrick Mensah, Hemophilia Center, Leicester Royal Infirmary, Leicester, United Kingdom

Patrick.Mensah@uhl-tr.nhs.uk

Disclosure: The reviewer(s) have no financial or other competing interest to disclose.

Update and revision (2026)

Olga Levchenko, Reanimatologist, Anesthesiologist, Therapist, Hematologist, Healthcare organizer, National Research Center for Hematology, Moscow, Russia; Head of the Medical Accreditation and Simulation Center, National Medical Hematology Research Center, Moscow, Russia

levchenkokp@rambler.ru

Gennady M. Galstyan, Anesthesiologist, Head of the Intensive Care Department of the National Research Center for Hematology, Moscow, Russia

Reviewer

Francis Veyckemans, Retired pediatric anesthesiologist and honorary professor

UCLouvain Medical School, Belgium

veyckemansf@gmail.com

Editorial Review

Christine Gaik, Anesthesiologist, Department of Anesthesiology and Intensive Care Medicine, University Hospital Giessen and Marburg, Campus Marburg and Philipps University of Marburg, Germany

gaikc@med.uni-marburg.de
