Aktiv Druck & Verlag GmbH | ISSN 0170 - 5334 I 02330

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63. Jahrgang | März 2022

A&I

ANÄSTHESIOLOGIE & INTENSIVMEDIZIN

Offizielles Organ: Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V. (DGAI)
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Deutsche Akademie für Anästhesiologische Fortbildung e.V. (DAAF)
Organ: Deutsche Interdisziplinäre Vereinigung für Intensiv- und Notfallmedizin e.V. (DIVI)

Erdheim-Chester disease

Haemophilia A

orphanaesthesia

a project of the German Society
of Anaesthesiology and Intensive Care Medicine

SUPPLEMENT NR. 4 I 2022

OrphanAnesthesia –

ein krankheitsübergreifendes Projekt des Wissenschaftlichen Arbeitskreises Kinderanästhesie der Deutschen Gesellschaft für Anästhesiologie und Intensivmedizin e.V.

Ziel des Projektes ist die Veröffentlichung von Handlungsempfehlungen zur anästhesiologischen Betreuung von Patienten mit seltenen Erkrankungen. Damit will Orphan Anesthesia einen wichtigen Beitrag zur Erhöhung der Patientensicherheit leisten.

Patienten mit seltenen Erkrankungen benötigen für verschiedene diagnostische oder therapeutische Prozeduren eine anästhesiologische Betreuung, die mit einem erhöhten Risiko für anästhesieassoziierte Komplikationen einhergehen. Weil diese Erkrankungen selten auftreten, können Anästhesisten damit keine Erfahrungen gesammelt haben, so dass für die Planung der Narkose die Einholung weiterer Information unerlässlich ist. Durch vorhandene spezifische Informationen kann die Inzidenz von mit der Narkose assoziierten Komplikationen gesenkt werden. Zur Verfügung stehendes Wissen schafft Sicherheit im Prozess der Patientenversorgung.

Die Handlungsempfehlungen von OrphanAnesthesia sind standardisiert und durchlaufen nach ihrer Erstellung einen Peer-Review-Prozess, an dem ein Anästhesist sowie ein weiterer Krankheitsexperte (z.B. Pädiater oder Neurologe) beteiligt sind. Das Projekt ist international ausgerichtet, so dass die Handlungsempfehlungen grundsätzlich in englischer Sprache veröffentlicht werden.

Ab Heft 5/2014 werden im monatlichen Rhythmus je zwei Handlungsempfehlungen als Supplement der A&I unter www.ai-online.info veröffentlicht. Als Bestandteil der A&I sind die Handlungsempfehlungen damit auch zitierfähig. Sonderdrucke können gegen Entgelt bestellt werden.

Bisher in A&I publizierte Handlungsempfehlungen finden Sie unter:

www.ai-online.info/Orphsuppl
www.orphananesthesia.eu

OrphanAnesthesia –

a project of the Scientific Working Group of Paediatric Anaesthesia of the German Society of Anaesthesiology and Intensive Care Medicine

The target of OrphanAnesthesia is the publication of anaesthesia recommendations for patients suffering from rare diseases in order to improve patients' safety. When it comes to the management of patients with rare diseases, there are only sparse evidence-based facts and even far less knowledge in the anaesthetic outcome. OrphanAnesthesia would like to merge this knowledge based on scientific publications and proven experience of specialists making it available for physicians worldwide free of charge.

All OrphanAnesthesia recommendations are standardized and need to pass a peer review process. They are being reviewed by at least one anaesthesiologist and another disease expert (e.g. paediatrician or neurologist) involved in the treatment of this group of patients.

The project OrphanAnesthesia is internationally oriented. Thus all recommendations will be published in English.

Starting with issue 5/2014, we'll publish the OrphanAnesthesia recommenations as a monthly supplement of A&I (Anästhesiologie & Intensivmedizin). Thus they can be accessed and downloaded via www.ai-online.info. As being part of the journal, the recommendations will be quotable. Reprints can be ordered for payment.

Find a survey of the recommendations published until now on:

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orphananesthesia

Anaesthesia recommendations for

Haemophilia A

Disease name: Haemophilia A

ICD 10: D66

Synonyms: Classic haemophilia or Factor VIII deficiency

Disease summary: Haemophilia A is a rare, inherited X-linked recessive bleeding disorder, resulting from factor VIII deficiency and characterised by intra-articular and intramuscular bleeding. There are numerous different mutations which cause haemophilia A. Due to differences in the gene involved (and the subsequent resulting protein), patients with haemophilia (PWH) have varying levels of factor VIII clotting activity. Individuals with less than 1 % FVIII clotting activity are classified as having 'severe' haemophilia, those with 1–5 % as 'moderate', and those with between 5–40 % as mild. Most severe haemophilia patients require regular supplementation with intravenous recombinant or plasma derived factor VIII concentrate (prophylaxis).

Medicine is in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnosis is wrong

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

▶ **Citation:** Levchenko O: Haemophilia A. Anästh Intensivmed 2022;63:S106–S112.
DOI: 10.19224/ai2022.S106

Typical surgery

Orthopaedic surgery: Haemarthrosis involving hip and shoulder joints (ball & socket joints) is uncommon in haemophilia. A 'diagnosis' of hip haemarthrosis is often a misdiagnosis of an iliopsoas muscle bleed.

Total knee replacement or total elbow replacement, and ankle arthrodesis (all hinged joints) are the most common haemophilia related surgical interventions. Surgery for non-haemophilia related conditions, (e.g. hernia repair, appendectomy, cholecystectomy) are also, not uncommonly, performed in PWH.

Type of anaesthesia

General anaesthesia has to be administered as total intravenous anaesthesia. All intravenous and volatile anaesthetics can be used.

Regional or neuraxial anaesthesia are best avoided as there is a high risk of haemorrhagic complications, such as epidural haematoma. Local anaesthetic infiltration with lignocaine is very commonly used for minor procedures such as dental extractions in PWH, after adequate factor replacement therapy.

Necessary additional pre-operative testing (beside standard care)

Haemophilia A is a blood disorder and therefore diagnosis of the condition requires the evaluation of relevant blood tests.

Routine laboratory tests of blood coagulation, FVIII and von Willebrand factor assays should be performed for evaluating the presence of haemophilia. Thromboelastography and thromboelastometry have no role in the day-to-day diagnosis or management of uncomplicated haemophilia. They are used in a limited number of centres to guide the choice or use of bypassing agents (FEIBA or recombinant FVIIa) in haemophilia patients with inhibitors. Otherwise, TEGs and other global haemostatic assays have no recognised role in either the diagnosis or the routine management of haemophilia.

Routine coagulation profile, including activated partial thromboplastin time (aPTT), Prothrombin time, fibrinogen activity (Clauss assay), plasma factor VIII (FVIII) concentration and factor VIII inhibitors. APTT and PT, while useful in the initial evaluation of a patient with suspected haemophilia, are generally irrelevant tests once the diagnosis of haemophilia has been established.

Hepatitis C virus (HCV) has been the major cause of liver disease in haemophilia. Blood samples must be obtained for ALT, AST, HCV antibody, HCV genotype, HIV antibody, haemoglobin analyses and platelet counts. HCV and HIV viral loads will only be required if patients have tested positive for the respective antibody.

Particular preparation for airway management

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

There is no increase in risk of aspiration.

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Particular preparation for transfusion or administration of blood products

Haemophilia A is a disorder caused by missing or defective factor VIII, a clotting protein, so there will be a high risk of bleeding if peri-operative factor replacement is inadequate. There may be a higher requirement for blood products during surgery.

Patients with haemophilia A must be provided with factor VIII (FVIII) concentrates. Haemophilia patients have no excess requirements for fresh frozen plasma (FFP) or packed red cells compared to non-haemophilia patients, provided peri-operative FVIII replacement has been adequate. Cryoprecipitate, (which contains FVIII, von Willebrand factor and fibrinogen) is used in the treatment of haemophilia A only when and where FVIII concentrates are not available. It is only necessary to have these blood products (FFP, packed red blood cells, cryoprecipitate) on standby, and they are to be used only as and when needed. With adequate preparation, they are usually not required.

Haemophilia A inhibitor patients must be treated with recombinant FVIIa or FEIBA (activated prothrombin complex concentrate). Recombinant FVIIa or FEIBA (factor eight inhibitor bypassing agent) are licensed for the on-demand treatment of bleeding episodes and the prevention of bleeding in surgery or invasive procedures in patients with congenital haemophilia with inhibitors.

Particular preparation for anticoagulation

Patients with mild haemophilia A can sustain nearly normal or even normal levels of FVIII post-operatively as a result of the acute-phase response to surgery. Such patients are as prone to post-operative venous thrombosis as non-haemophilia patients.

Particular precautions for positioning, transportation and mobilisation

Some patients may have difficulty with mobility associated with haemarthroses. This depends on the degree of arthropathy. Some mild forms may be clinically silent.

Interactions of chronic disease and anaesthesia medications

The use of narcotic analgesics by PWH due to an existing chronic pain syndrome (as a result of haemophilic arthropathy) can sometimes complicate post-operative analgesia. An analysis of our own retrospective data showed paracetamol and trimeperidine to be both highly effective and safe on the first post-operative day following high-trauma surgical procedures in patients with haemophilia.

The long-term use of paracetamol should be avoided in PWH with hepatitis C (hepatotoxicity).

NSAIDs are best avoided because of the potential risk of haemorrhagic complications.

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Anaesthetic procedure

Patients with severe and moderate haemophilia A must be treated with factor VIII (FVIII) concentrates before intubation, because there is a high risk of haemorrhagic complications, such as haematoma of the epiglottis and trauma-induced bleeding from the upper respiratory tract. The most mild-haemophilia patients do not necessarily require FVIII concentrates before intubation or surgery. Most mild-haemophilia patients respond very well to desmopressin (DDAVP), which can raise FVIII (and von Willebrand factor) levels 3-5 times over baseline levels. DDAVP is the haemostatic agent of choice for mild haemophilia, ***not*** FVIII concentrate. Drug selection will also depend on the volume and type of surgery.

Haemophilia A patients with inhibitors must be treated with recombinant FVIIa or FEIBA before intubation.

Particular or additional monitoring

In case of opioid-tolerant PWH, ANI is very useful.

The analgesia nociception index (ANI) based on heart rate variability has been proposed to reflect various levels of acute pain.

Possible complications

Haemophilia A is a disorder caused by missing or defective factor VIII, a clotting protein, so there is a high risk of bleeding. Haemorrhagic complications may occur if peri-operative haemostatic preparation is inadequate or is poorly planned.

Post-operative care

The degree of post-operative monitoring depends on the surgical procedure and pre-operative condition of the patient.

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

In the post-operative period, FVIII levels must be maintained at levels advised by the haematologist or haemophilia centre where the patient is registered. Other blood product requirements should not differ from those of non-haemophilia patients, and should be dictated purely by clinical circumstances. It is necessary only to have such products on standby, and they should be used only as and when required. Haemophilia A patients with inhibitors will similarly require bypassing agents (recombinant FVIIa or FEIBA) in the post-operative period, as advised by the responsible haematologist or haemophilia centre.

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Disease-related acute problems and effect on anaesthesia and recovery

Development of "inhibitor" antibodies against factor VIII due to frequent infusions.

Ambulatory anaesthesia

Ambulatory anaesthesia should be avoided in patients with severe and moderate haemophilia A because of the high risk of bleeding.

Obstetrical anaesthesia

All PWH are men.

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Date last modified: **December 2016**

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

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Disclosure The reviewers have no financial or other competing interest to disclose.

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Herausgeber

DGAI
Deutsche Gesellschaft für Anästhesiologie und Intensivmedizin e.V.
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Berufsverband Deutscher Anästhesisten e.V.
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Schriftleitung
Präsident/in der Herausgeberverbände
Gesamtschriftleiter/Editor-in-Chief:
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Verlag & Druckerei

Aktiv Druck & Verlag GmbH
An der Lohwiese 36 |
97500 Ebelsbach | Deutschland
www.aktiv-druck.de

Geschäftsführung
Wolfgang Schröder | Jan Schröder | Nadja Schwarz
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Verlagsrepräsentanz
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Herstellung | Gestaltung
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Titelbild
Gestaltung: Klaus Steigner
Paumgartnerstraße 28 | 90429 Nürnberg
E-Mail: mazyblue@klaus-steigner.de
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Erscheinungsweise 2022
Der 63. Jahrgang erscheint jeweils zum Monatsanfang, Heft 7/8 als Doppelausgabe.

Bezugspreise (inkl. Versandkosten):

- **Einzelhefte** 30,- €
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Europa (ohne Schweiz) (inkl. 7 % MwSt.)	258,- €
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