orphanaesthesia

Anaesthesia recommendations for patients suffering from

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 characterized 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 in progress

Perhaps new knowledge

Every patient is unique

Perhaps the diagnostic is wrong

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

Typical surgery

Orthopaedic surgery: Haemarthrosis involving hip and shouder joints (ball & socket joints) is uncommon in haemophilia. A 'diagnosis' of hip haemathrosis 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, appendicectomy, 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 hematoma. 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 diagnostic procedures (preoperative)

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 presence of hemophilia. 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 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. The 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, platelet count, haemoglobin. HCV and HIV viral loads are only required if patients have tested positive for the respective antibody.

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.

Particular preparation for transfusion or administration of blood products

Hemophilia A is a disorder caused by missing or defective factor VIII, a clotting protein, so there is 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 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 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 (**F**actor **E**ight **I**nhibitor **B**ypassing **A**gent) 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 near 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, transport or mobilisation

Some patients may have difficulty with mobility associated with hemarthroses. It depends on degree of athropathy. Some mild forms may be clinically silent.

Probable interaction between anaesthetic agents and patient's long-term medication

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

Long-term use of paracetamol should be avoided in PWH with Hepatitis C (hepatotoxicity). NSAIDs are best avoided because of the potential risk of hemorrhagic complications.

Anaesthesiologic procedure

Patients with severe and moderate hemophilia A must be treated with factor VIII (FVIII) concentrates before intubation, because there is a high risk of haemorrhagic complications, such as hematoma of the epiglottis and trauma induced bleeding from the 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 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 heart rate variability based analgesia nociception index (ANI) has been proposed to reflect different levels of acute pain.

Possible complications

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

Postoperative care

Degree of postoperative monitoring is dependent on the surgical procedure and preoperative condition of the patient.

It is essential to exclude 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 haemophilia.

In the postoperative period, FVIII levels must be maintained at levels advised by the haematologist or Haemophilia Centre under who the patient is registered. Other blood products requirements should be no different from that of non-haemophilia patients, and should be dictated purely by clinical circumstances. It is necessary only to have such products on standby, and 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.

Information about emergency-like situations / Differential diagnostics

caused by the illness to give a tool to distinguish between a side effect of the anaesthetic procedure and a manifestation of the diseases, e.g.:

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

Ambulatory anaesthesia

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

Obstetrical anaesthesia

All PWH are men.

Literature and internet links

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Last date of modification: December 2016

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