Haemophilia and Other Bleeding Disorders - Emergency Management - Full Clinical Guideline

Reference no.: CG-HAEM/2023/012

1. Introduction

In the UK, care for patients with haemophilia and related bleeding disorders is coordinated via a network of designated Haemophilia Centres (HC) or Haemophilia Comprehensive Care Centres (HCCC). All HCs and HCCCs are involved with the provision of emergency care. Registered patients have open and direct access to the haemophilia centre during the normal working day. However, arrangements for the provision of care in the evenings, at night and at weekends varies between centres. The experience of patients attempting to access emergency care is also variable. Many patients report poor experiences when dealt with in the Emergency setting

2. Aim and Purpose

To enable individuals with bleeding disorders who present in an emergency to receive timely and appropriate management.

3. Definitions, Keywords

Haemophilia A - Congenital deficiency of clotting factor VIII; Haemophilia B - Congenital deficiency of clotting factor IX. Severe haemophilia is associated with spontaneous bleeding into joints, muscles and organs e.g. the brain. Even minor interventions can cause bleeding if the bleeding disorder is not treated beforehand.

Clotting Factor Concentrate – this is given therapeutically to replace the clotting factor that is missing or low. CFC's come as vials of lyophiliyzed powder with diluent and a transfer mechanism to transfer the diluent into the vial of CFC. All CFC's are on the Medusa database used by the Trust.

Desmopressin. This is a medication which stimulates the release of Factor VIII and von Willebrands factor from their stores in the endothelial cells of blood vessels. Desmopressin can be used to treat mild haemophilia A and von Willebrands disease.

4. Main body of Guidelines

Triage

1) Individuals with bleeding disorders should be given a triage category of very urgent (category 2) because delays in administering factor concentrate treatment can significantly affect morbidity and mortality in these individuals with bleeding disorders.

If treatment is required the maximum time to delivery of treatment should not exceed 30 minutes.

2) Contact the on-call consultant haematologist (consultant or Registrar); however, this should not delay giving clotting factor replacement to the patient.

Assessment

1. Treatment for a suspected bleeding episode is based on clinical history. Physical exam findings may be normal in the early phases of most haemophilic bleeds. Spontaneous bleeding is common in individuals with severe disease (factor levels <1%). When in doubt, administer clotting factor replacement therapy immediately.
2. Treatment decisions should be based on the ***suspicion*** of a bleeding-related problem, not the documentation of one.
3. If the patient or the parent of a patient suspects that occult bleeding is occurring, administer clotting factor replacement, as below. Patients or their parents may have their own individualized treatment plan,and/or their own supply of clotting factor concentrate. If so, use that.

Clotting factor stocks

There are 2 emergency stocks of clotting factor: CED resus (in the fridge in the room at the back of resus), and on the 5th floor in the blood issue room next door to blood bank (in the small fridge directly opposite the door with a combination lock – code 2244, then swivel lever up).

The main stock of clotting factor is in pharmacy stores (contact the on-call pharmacist).

Diagnostic Studies

1. Clotting factor replacement therapy should be given before any diagnostics studies (X-rays, Ct scans etc.) are performed to evaluate a suspected bleeding problem, especially in the case of head trauma or suspected intracranial haemorrhage. For routine joint bleeding, no radiographic studies are indicated.
2. For patients with haemophilia who have illnesses or disorders that necessitate an invasive procedure (e.g. lumbar puncture, arterial blood gas, arterial line, central venous line arthrocentesis, etc.- this is not an exhaustive list) or surgery, factor replacement therapy to 100% must be administered before the planned procedure or surgery.
3. For an individual with known haemophilia, routine laboratory studies (PT, PTT, factor levels), are not indicated in the treatment of a routine bleeding episode unless requested by the patient's haematologist. The clinical severity of a patient's haemophilia is gauged by his or her baseline clotting factor level, a value that remains fairly constant throughout that person's life.
4. Von Willebrands disease: VW levels can increase with age. However in an emergency use the last known baseline to guide treatment.

Indications for Factor Replacement Therapy

1. Suspected bleeding into a joint or muscle.
2. Any significant injury to the head, neck, mouth or eyes or evidence of bleeding in these areas.
3. Any new or unusual headache, particularly one following trauma.
4. Severe pain or swelling at any site.
5. All open wounds requiring surgical closure, wound adhesive, or steri-strips.
6. History of an accident or trauma that might result in internal bleeding.
7. Any invasive procedure or surgery.
8. Heavy or persistent bleeding from any site.
9. Gastrointestinal bleeding.
10. Acute fractures, dislocations and sprains.

Treatment

All patients should carry a bleeding disorders card which states their usual treatment (i.e. which clotting factor or desmopressin). This information will also be found on ecasenotein Lorenzo and haematology clinic letters.

Haemophilia A: Give recombinant FVIII.e.g. Advate, Refacto or Kogenate. The dose of factor VIII is **50 units/kg**. This should result in a factor VIII level of 80-100%. (Cryoprecipate and fresh frozen plasma are no longer recommended for treatment of individuals with haemophilia A).

Mild Haemophilia A with non-Life or limb threatening bleeding: Individuals with mild haemophilia A with factor VIII greater than 15% who are experiencing non-life or limb threatening bleeding may respond to desmopressin if they have been shown to respond to this treatment previously. The dose of desmopressin is 0.3 micrograms/kg subcutaneous or IV.

Haemophilia B

The treatment of choice for individuals with haemophilia B (factor IX deficiency) is recombinant factor IX (Benefix) or else the patient's product of choice. Plasma-derived concentrate is a suitable alternative in an emergency situation when recombinant Factor IX is not available. (Cryoprecipate and fresh frozen plasma are no longer recommended for treatment of individuals with haemophilia B).

When bleeding is severe, the appropriate dose of factor IX is **100-120 units/kg**. This should result in a factor IX level of 80-100%.

Haemophilia B does not respond to desmopressin.

Von Willebrands Disease (VWD)

Most people with VWD have type 1 VW which responds to desmopressin – 0.3 micrograms/kg (capped at 25 micrograms) IV in 50 mls N/Saline over 30 minutes or subcutaneously. Peak response by 60 – 90 minutes. Partially fluid restrict for the following 24 hours (drink when thirsty, 1 L per 24 hours).

A minority of people with VWD have type 2 VW which may need clotting factor concentrate (Wilate)

1. If a patient with haemophilia or the parent of a patient with haemophilia brings clotting factor with them to the emergency department, allow them to use it. They should be permitted to reconstitute the product and administer it whenever possible. Individuals with bleeding disorders are encouraged to have an emergency dose of factor concentrate or desmopressin in their home and to take it with them when they travel. In those situations where a patient does not bring their own clotting factor concentrate, factor from the emergency stocks or pharmacy stores must be used (and made available within 30 minutes of the patients arrival). In order to expedite care, emergency physicians should order/obtain *unreconstituted* factor from the emergency stocks or pharmacy and reconstitute the product in the emergency department

2. **How to and who can administer the clotting factor concentrate**? ***All hospital staff who have received training in the administration of intravenous drugs may administer clotting factor concentrates. Details can be found on Medusa.***

All clotting factor concentrates administered in hospital must be prescribed on the patient's prescription chart (onLorenzo). It is essential that the decision to prescribe, the batch number, the reason for giving and the dose calculation is recorded in the patient's notes. Family members who have received appropriate training can administer concentrates. Older children and adults may have learned to administer their own treatment. If they or their family wish to administer their own treatment they should be allowed to do so.

3. Factor replacement must be administered intravenously by IV push over 1-2 minutes. The factor dose should be ordered as "up to the closest vial contents." The full content of each reconstituted vial should be infused, since a moderate excess of factor concentrate will not create a hypercoagulable state but will prolong the therapeutic level of the product administered; thus, it is prudent to "round up."

4) For individuals with inhibitors (antibodies to factor VIII or IX), treatment decisions may be more complicated. The care of inhibitor patients should be urgently discussed with a haematologist. If an individual with an inhibitor presents in a life- or limb-threatening scenario, the safest immediate action is to prescribe recombinant factor VIIa (rFVIIa) at a dose of 90 mcg/kg or activated prothrombin complex concentrates (FEIBA) at 75-100 units/kg.* The patient or family can also provide information on response to these therapeutic bypassing agents.

* Note: In factor IX patients with a history of inhibitors and anaphylaxis, do not give factor IX-containing products unless the bleeding is life-threatening.

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5. When treating an individual with mild haemophilia A who is responsive to desmopressin, the dose and prior responsiveness are usually known. The dose of desmopressin is 0.3mcg/kg (capped at 25 micrograms) subcutaneously or else intravenously in 50 ml normal saline over 30 minutes. It may also be administered as an ultra- concentrated nasal spray "Octim"" at a dose of 1 spray in one nostril for individuals <50 kg and 1 spray in each nostril for individuals >50 kg. Partially fluid restrict for 24 hours (drink when thirsty, 1 L per 24 hours). Peak effect 60 – 90 minutes.

6. The most experienced IV therapist or phlebotomist should perform any venipuncture. Traumatic venipunctures and repeated needle sticks cause painful hematomas that may limit further IV access.

7. In any suspected bleeding emergency in which the clotting factor level of an individual with haemophilia is unknown, the factor level should be assumed to be 0%.

8. Intramuscular injections should be avoided. If they must be given, factor replacement therapy must precede the injection. Parenteral agents should be given intravenously or subcutaneously. Tetanus immunizations may be administered subcutaneously.

9. In situations in which patients are hemodynamically stable and are not requiring volume replacement, the smallest gauge needle should be utilized for obtaining IV access (25g butterfly needles in young infants, 23g butterfly needles in older children and adults).
10. Tourniquets should not be applied tightly to extremities because they may cause bleeding.

11. Aspirin and aspirin-containing products are contraindicated in individuals with haemophilia. Paracetomol and/or codeine may be used for analgesia. Non-steroidal anti-inflammatory (NSAID) drugs may be carefully administered to select patients,

such as individuals with chronic arthritic pain who are not actively bleeding or being treated for a recent bleeding problem.

12. If an individual with haemophilia is bleeding and requires transportation to another Trust for definitive care, all efforts should be made to replace the deficient clotting factor before transport.

5. References

Emergency and out of hours care for patients with bleeding disorders – Standards of care for assessment and treatment. John Hanley , Mary Mathias , Emma Franklin , Chris Harrington , Oliver Chapman , Kate Talks and Stephanie Smith on behalf of the UK Haemophilia Centre Doctors Organisation (UKHCDO) 2009.

GUIDELINES FOR EMERGENCY DEPARTMENT MANAGEMENT OF INDIVIDUALS WITH HEMOPHILIA AND OTHER BLEEDING DISORDERS. *Medical and Scientific Advisory Council (MASAC) of the National Hemophilia Foundation (NHF) August /September, 2017.*

6. Documentation Controls

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7. Appendices