Protocol for Urgent Care of Patients With Congenital Bleeding Disorders in Accident & Emergency Centers

Scope: Delay in treatment of bleeding complications of hemophilia A or B significantly affects morbidity and mortality and dramatically increases the cost of care. Patients with hemophilia A or B referred for treatment to Accident & Emergency Centers require urgent initial therapeutic interventions.

I. Bleed complications – A&E and Paediatric Medical Ward Guidelines

A. Background.

1. Bleeding is related to severity of deficiency. **Severe Hemophilia** factor activity <1%; **moderate hemophilia** – factor activity 1-5%; **mild hemophilia** – factor activity >5%.

2. Bleeding that threatens life, limb, or function includes: intracranial, spinal cord, throat, intra-abdominal, limb compartments, and ocular.

B. Treatment decisions should be based on a **suspicion** of bleeding-related problems, **not on documentation alone**.

C. **Respect patient/parent experience**.

D. Triage patient as an “urgent care” need. Consultation with haematologist is useful, but should not delay intervention in treatment with specific clotting factor.

E. **Immediately treat with factor replacement for:**

1. **Suspected** bleed – joint or muscle.
2. New/unusual **headache**, particularly following trauma.
3. Pain or swelling – any site.
4. Wounds requiring suture
5. History of accident or trauma that **may** result in internal bleeding.
6. Prior to **any** invasive (i.e., spinal tap) or surgical procedure.
7. GI bleeding.
8. Fractures.
9. Any heavy or persistent bleeding.
F. Treatment

**Hemophilia A** (no known inhibitor): 50 units per kg Factor VIII
Intravenous over 3 minutes

**Hemophilia B** (no known inhibitor): 100-120 units per kg Factor IX
Intravenous over 3 minutes

**Hemophilia A or B with inhibitor:** Contact the paediatric haematologist on-call immediately, see Item H below.

**Other rare coagulation protein deficiencies:** FFP 20 ml per kg infused over 1 hour maximum

*Round to nearest whole vial – Factor products are very expensive and should never be wasted*

1. Use a small gauge needle with a butterfly needle if available. Infuse factor as quickly as possible. Avoid repeat sticks.

2. Factor replacement should occur within **45 minutes of arrival to Accident & Emergency.** After arrival and triage, *this is the first intervention.*

3. CT, diagnostic testing, detailed examinations should occur only following *factor replacement.*

4. No IM shots; for routine & chronic joint bleeds, no radiographic studies are necessary; for routine bleeding episodes, PT, PTT and factor assays are *not necessary.*

5. No Aspirin or NSAIDs (e.g. Brufen) should be given to these patients.
G. Follow-up care.

1. For life-threatening bleeds admit for observation and further therapy.

2. For routine joint/muscle bleeding – treat with factor concentrate, and rest, ice, nonweight bearing instructions, splint or ace-wraps, elevation, and follow-up within 12-24 hours with Hemophilia Treatment Center.

H. Hemophilia patients with inhibitors.

1. Options include (where available):
   - Factor 7a: 90-180 micrograms/kg IVP. Repeat dose at 90 micrograms/kg at 2-hour intervals x 2.
   - Feiba 75-100 units/kg IVP. May repeat in 12 hours

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References:
3. Clinical Practice Guidelines, Texas Children’s Cancer Center & Hematology Service