PAEDIATRIC ACUTE CARE GUIDELINE	
Haemophilia	
Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

This document should be read in conjunction with this DISCLAIMER
http://kidshealthwa.com/about/disclaimer/

Haemophilia

Also see guideline Management of a Child with a Bleeding Disorder

Background

- Most children with severe Haemophilia are treated with prophylactic factor replacement starting between age 1 – 2 years. This is given intravenously, one to three times weekly for severe Haemophilia A, twice weekly for severe haemophilia B. Recombinant factor (Advate or Xyntha – factor VIII; Benefix – factor IX) is the treatment of choice.
- Supplies of recombinant factor VIII and factor IX are kept in the fridge in Transfusion Medicine (Haematology laboratory).

- **All presentations to the Emergency Department should be discussed with the on-call Haematology/Oncology Fellow or Haematologist**.
- Please refer to Haematology Transfusion Medicine Protocols for further information.
- Mild Haemophilia is not usually treated with prophylactic factor replacement but can still be a cause of severe bleeding problems and may require urgent factor replacement therapy or DDAVP.

Management

Haemophilia A: Factor VIII deficiency

- This is a sex-linked condition due to factor VIII deficiency. Severe cases have <2% factor

VIII, moderate cases 2-5% and mild >5%.

Prophylactic treatment

- Generally started in severe cases over 12 months of age
- The main reason for prophylactic treatment is the prevention of haemarthroses, and this becomes more likely as the child starts walking, usually around 12 months of age.
- Despite prophylactic treatment, occasionally these patients may present with spontaneous joint bleeding and may require additional factor therapy.
- Approximately 10% of patients with Haemophilia A may develop inhibitors to treatment – usually this occurs in the first ten treatments
    - Treatment of patients with inhibitors is difficult and may require bypassing agents such as factor VIIa.

Mucosal bleeding is a common problem in young children and can be treated with antifibrinolytic therapy using Tranexamic Acid.

- The dose is 15-20mg/kg/dose given TDS orally. Tranexamic Acid comes as 500 mg tablets. For children:
    - < 25 kg give half a tablet TDS
    - 25-35 kg give one tablet TDS
    - 35-50kg give one and a half tablets TDS
    - > 50kg give two tables TDS
- Advice about specifics of treatment should be sought from a Clinical Haematologist

Head injury

- Treatment should be regarding as **emergency** and a dose of factor VIII 50 Units/kg IV given urgently
- For minor knocks to the head, such as contact with furniture or doors, factor treatment does not need to be administered
- Admit to hospital if there is any suspicion of concussion or persistent headache or vomiting

Haemophilia B (Christmas Disease): Factor IX deficiency

This is a sex-linked condition with factor IX deficiency. The presentation is identical to haemophilia A with similar factor levels.

- The treatment of choice is recombinant Factor IX (Benefix)
- Most patients with severe factor deficiency are treated with prophylactic infusions, given twice weekly
- Inhibitors to factor IX are less common but can present with allergic manifestations

Head injury

- The same principles apply as for haemophilia A – give factor IX 50 units/kg IV urgently.

References

External Review: Catherine Cole (Paediatric and Adolescent Oncologist/Haematologist) – July 2015

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