PAEDIATRIC ACUTE CARE GUIDELINE	
Bleeding and Clotting Disorders	
Scope (Staff):	All Emergency Department Clinicians
Scope (Area):	Emergency Department

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Bleeding and Clotting Disorders

Also see guideline Management of a Child with a Bleeding Disorder

Background

Common presentations of bleeding disorders are:

- Excessive bruising and bleeding after trauma
- Recurrent epistaxis and mucosal bleeding
- Bleeding after operations and dental extractions
- Spontaneous haemarthroses only in severe factor deficiency

Assessment

History and Examination

- Try to determine whether this is an **acquired** or **inherited** bleeding problem, and whether this is a **platelet disorder** or a **coagulation deficiency**
- How long have symptoms been present, and is the process local or general?

- **Platelet problems** usually present with mucosal and skin bleeding whereas **coagulation defects** present with deep muscle haematomas, haemarthroses, but also have skin bruising.
- Be alert to the possibility of non-accidental injury, bleeding disorder associated with hepatic and renal disease and, rarely, connective tissue problems.

Investigations

- Full blood count, including platelets and blood film
- Standard coagulation profile, including prothrombin time, APPT and fibrinogen
- If there is a family history of von Willebrand's disease (VWD) or a strong suspicion clinically, factor VIII studies including Ristocetin cofactor and von Willebrand factor antigen should be performed ("Von Willebrand's screen")

Common Diagnosis

- Haemophilia
- von Willebrand's Disease
- Immune Thrombocytopaenic Purpura

Diagnosis	Description	Screening tests
Haemophilia A (Factor VIII deficiency)	A sex linked condition but approximately 30% of new cases have no family history. Can be mild, moderate or severe depending on factor levels.	Prolonged APTT which corrects with normal plasma. All other tests normal.
Haemophilia B (Factor IX deficiency)	Similar to Haemophilia A with mild, moderate and severe cases.	Prolonged APTT which completely corrects with normal plasma. All other tests normal.
von Willebrand's disease	A common mild bleeding disorder usually presenting with bruising and mucosal bleeding. Menorrhagia and post-partum haemorrhage are common problems in females. Inherited as an autosomal dominant in most cases.	Most common form is **type 1** where there is a quantitative deficiency, i.e. reduced factor VIII coagulant, Ristocetin cofactor, and von Willebrand factor antigen. In **type 2** disorders there is a quantitative abnormality with reduced Ristocetin cofactor relative to a von Willebrand factor antigen. A rare form is the severe **type 3** disorder, where patients are homozygotes and levels of factor VIII coagulant, Ristocetin cofactor, and von Willebrand factor antigen are all markedly reduced.
Immune Thrombocytopaenic Purpura	A common disorder in children. ITP is the most common cause of thrombocytopaenia in childhood.	
Two common causes of prolonged APTT **not** associated with bleeding are: • Factor XII deficiency and • A lupus-like anticoagulant which is usually a transient post-viral phenomenon, prolongs the APTT and is not corrected by mixing "50:50" with normal plasma		

References

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

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