HAEMATOLOGY EMERGENCIES IN THE EMERGENCY DEPARTMENT - CHW

PRACTICE GUIDELINE ©

DOCUMENT SUMMARY/KEY POINTS

- Refer to the CHW Paediatric Handbook for further details of all topics in this guideline.
- Contact the Haematologist on-call as early as possible to discuss the patient's management.
- **Sickle Cell Disease**: The most common presentation in children with sickle cell disease is painful vaso-occlusive crisis (acute pain crisis) but they may also present with life-threatening complication including sepsis, acute splenic sequestration, acute chest syndrome and stroke.
- **Haemophilia**: Patients presenting with even relatively minor trauma should receive factor prior to being sent for X-ray or any other imaging as haemorrhage can be life-threatening.
- **Platelet disorders**: Patients with severe thrombocytopenia or platelet function disorders may occasionally present with serious bleeding complications.
- **Neutropenia**: Patients with severe neutropenia due to bone marrow failure (eg aplastic anaemia, inherited bone marrow failure syndromes) are at risk of sepsis and should be managed urgently if they present with fever
- **Acute severe anaemias**: acute splenic sequestration in sickle cell disease and acute intravascular haemolysis may present with life-threatening anaemia.

Approved by:	SCHN Policy, Procedure and Guideline Committee	
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CHANGE SUMMARY

- Changes made: addition of platelet disorders, chronic neutropenia and acute severe anaemias.

READ ACKNOWLEDGEMENT

- ED clinical staff (medical officers and nurses) should read and acknowledge they understand the contents of this document.

1 Sickle Cell Disease

The most common presentation in children with sickle cell disease is painful vaso-occlusive crisis (acute pain crisis). They may also present with acute splenic sequestration, acute chest syndrome, stroke and bacterial sepsis.

- The following symptoms are indications for emergency care:
  - Temp > 38°C
  - Pain inadequately relieved by home measures
  - Respiratory symptoms (cough, shortness of breath, chest pain, hypoxia)
  - Abdominal pain, distension and/or acute splenic enlargement
  - **Any** neurologic symptom or sign – even transient
  - Significant increase in pallor and/or fatigue/lethargy
  - Priapism episode lasting > 2 hours

Consider sickle cell disease in new patients presenting with anaemia and the above features, if they are from risk ethnicities e.g. Middle-eastern, African and Indian.

Management:

- Notify the Haematologist on-call within 1 hour of reviewing the patient.
- Obtain FBC, reticulocyte count, group and hold, blood culture (if febrile), EUC/LFT and blood gas
- If sodium is low, discuss with a senior prior to starting IV fluids
- Commence0.45% sodium chloride + 5% glucose OR 0.9% sodium chloride; run at 1½ x maintenance rate (see: intravenous fluid management – CHW practice guideline).
- In patients needing IV analgesia, establish a morphine infusion; PCA for patients > 10 years and continuous infusion for patients < 10 years. ***All patients needing morphine infusions, including PCA, should have a background infusion rate of 10 – 20 microg/kg/hour.***
- Patients needing IV analgesia must not be sent to the ward before the infusion is started.
- Consider intranasal Fentanyl while awaiting IV opioids.
- Arrange a Pain Team (or Anaesthetic Registrar for afterhours/weekends) consultation for ongoing care on ward.

2 Haemophilia

- Patients presenting with even relatively minor trauma should receive factor **prior** to being sent for X-ray or any other imaging as haemorrhage can be life- threatening.

- Contact the Haematologist on-call within 1 hour of review in ED.
- Patients presenting with bleeds in limbs (i.e. joints or muscle) usually do not require imaging. Discuss with the Haematologist before arranging imaging. Factor should be given before the patient leaves ED for the Radiology Department.
- All haemophilia patients (particularly young children) presenting with unusual symptoms, including those that do not appear to be related to bleeding must be discussed with the Haematologist on-call.
- Consider line sepsis in patient with CVAD.

3 Platelet disorders

Patients with the following may present with severe or life-threatening bleeding

- Immune thrombocytopenia (ITP)
- Congenital platelet function disorders
- Bone marrow failure/aplastic anaemia

Patients may present with

- Intra-cranial bleeding, usually following trauma but this is rare
- Pelvic bleeding (ruptured ovarian cyst) or menorrhagia in adolescent girls
- Severe epistaxis

- Notify the Haematologist on-call within 1 hour of reviewing patient.
- Management is dependent on the bleeding complication and the underlying disease.

4 Aplastic Anaemia

- Patients presenting with febrile neutropenia should be managed urgently in the same way as Oncology patients with neutropenia and fever (see: Oncology/Transplant Patient – Fever or Suspected Sepsis – Initial Management).
- Thrombocytopenic patients may present with serious bleeding complications.
- Notify the Haematologist on-call within 1 hour of reviewing patient.
- Consider line sepsis in patients with CVAD even if not neutropenic.

5 Chronic neutropenia

- Patients with severe congenital neutropenia (an inherited bone marrow failure syndrome) presenting **unwell** with fevers and neutrophil counts of < 0.2 are managed as above (see: Oncology/Transplant Patient – Fever or Suspected Sepsis – Initial Management).

- Notify the Haematologist on-call within 1 hour of reviewing patient.
- The risk of sepsis in children with other forms of neutropenia is low and most do not need hospital care during febrile illnesses. Discuss management with the Haematologist on-call.

6 Severe anaemia

Life-threatening anaemia may occur in the following

- Acute splenic sequestration in sickle cell disease
- Acute blood loss
- Acute haemolysis, especially intravascular haemolysis e.g. acute oxidant haemolysis in G6PD deficiency or paroxysmal cold haemoglobinuria (PCH)
- Acute decompensation in patient with chronic anaemia, usually related to infection

- Notify the Haematologist on-call within 1 hour of reviewing patient.
- Urgent blood transfusion is often required. Un-crossmatched O neg packed cells may sometimes be required.
- The onset of iron deficiency anaemia is chronic and patients generally do not require blood transfusions even when anaemia is severe, and the majority can be managed as outpatients. Occasional patient may experience acute decompensation during infections and may require blood transfusions.

7 β-Thalassemia

- If a patient presents with fever, discuss the patient with the Haematologist on-call before discharge
- Patients with β-Thalassemia receiving desferrioxamine chelation therapy are more prone to bacterial sepsis, especially with Yersinia spp.

8 Related information

- Transfusion of Blood and Blood Components – CHW
  http://chw.schn.health.nsw.gov.au/o/documents/policies/policies/2007-8092.pdf
- Medication Management and Handling – CHW:
  http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2006-8232.pdf
- Oncology/Transplant Patient – Fever or Suspected Sepsis – Initial Management
  http://chw.schn.health.nsw.gov.au/o/documents/policies/guidelines/2016-9000.pdf
- CHW Paediatric Handbook (Chapter 5 – Haematology)
  http://chw.schn.health.nsw.gov.au/ou/medicine/resources/chw_handbook_2nd_edition.pdf