NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric Sickle Cell Disease Acute Anaemia

NOTE
This guideline is not intended to be construed or to serve as a standard of care. Standards of care are determined on the basis of all clinical data available for an individual case and are subject to change as scientific knowledge and technology advance and patterns of care evolve. Adherence to guideline recommendations will not ensure a successful outcome in every case, nor should they be construed as including all proper methods of care or excluding other acceptable methods of care aimed at the same results. The ultimate judgement must be made by the appropriate healthcare professional(s) responsible for clinical decisions regarding a particular clinical procedure or treatment plan. This judgement should only be arrived at following discussion of the options with the patient, covering the diagnostic and treatment choices available. It is advised, however, that significant departures from the national guideline or any local guidelines derived from it should be fully documented in the patient's case notes at the time the relevant decision is taken.

Splenic Sequestration

Splenic sequestration is more common in infants and young children (< 3 years old).
It may be precipitated by fever, dehydration or hypoxia. Rapid sequestration of red cells can lead to sudden anaemia and even death from hypoxic cardiac failure with pulmonary oedema. In some patients it may have a more insidious onset and can be recurrent.

Symptoms

- Abdominal pain (pulling legs up to abdomen)
- Abdominal distension
- Sudden collapse

Signs

- Rapidly enlarging spleen (may or may not be painful)
- Pallor, shock (tachycardia, hypotension, tachypnoea)
- +/- Fever due to associated sepsis

Investigations

- FBC and retics (raised in sequestration, cf absent in aplastic crisis)
- Blood cultures and other infection screen, as clinically indicated
- Erythrovirus serology (differential diagnosis is aplastic crisis)
- URGENT cross match

Management

- Assess the need for volume expansion but crystalloid should be used with caution as it may exacerbate cardiac failure.
- Emergency top-up transfusion, if necessary with uncross-matched O Rh -ve blood
- Broad spectrum antibiotics e.g. Tazocin (or as per local antibiotic policy)
- Before discharge, teach parents to recognise the symptoms and to detect an increase in spleen size
- Consider a regular transfusion regime for 2-3 months
- Consider splenectomy if recurrent (> 1 episode)

Hepatic Sequestration

Symptoms

- Right hypochondrial pain, abdominal distension
- +/- Fever due to associated sepsis

Signs

- Enlarging tender liver, increasing jaundice
- Collapse/shock is less common than with splenic sequestration

Investigations

- FBC and retics (raised in sequestration, cf absent in aplastic crisis)
- LFTs (bilirubin may be very high)
- Exclude gallstones/cholestasis by ultrasound
- Blood cultures and other infection screen, as clinically indicated
- URGENT cross match

Management

- May need urgent top-up transfusion.
- IV piperacillin-tazobactam or other broad spectrum antibiotic as per local antibiotic policy.
- If the patient becomes tachypnoeic, or develops chest signs, then check oxygen saturation and treat as for acute chest syndrome.

Aplastic Crisis

A temporary red cell aplasia caused by erythrovirus/parvovirus B19 can lead to a sudden severe worsening of the patient's anaemia. A viral prodromal illness may have occurred, but classical *erythema infectiosum* ('slapped cheek syndrome') is uncommon.

The main differential diagnosis is splenic sequestration. Aplastic crisis may affect multiple members of a family concurrently or consecutively.

Diagnosis

- Rapidly falling Hb
- Reticulocytopenia (but retics may be increased if in early recovery phase)
- Erythrovirus IgM present

Investigations

- Exclude hepatic or splenic sequestration
- FBC, reticulocytes, B12 and folate
- Parvovirus serology +/- PCR
- Urgent cross-match

Management

- Urgent red cell transfusion may be necessary (if Hb < 60g/l)
- Early follow-up until unsupported recovery of Hb and reticulocytes is documented.
- Spontaneous recovery is heralded by return of nucleated RBCs and reticulocytes to peripheral blood.
- Reassure. Recurrence does not normally occur, as immunity is lifelong.