NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

Acute anaemia in sickle cell disease

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.

Acute anaemia is a rare but important complication in sickle cell disease. The differential diagnosis is wide and can include common causes of anaemia such as blood loss. Causes of anaemia common in sickle cell disease include aplastic crises, sequestration crises (more common in childhood) and delayed transfusion reactions.

The appropriateness of transfusion of sickle cell patients should always be discussed with the haematologist on call, with the exception of acute blood loss or life threatening anaemia. NB blood bank should be alerted to the sickle cell disease status of patient in order that appropriate blood products meeting special requirements are issued.

Aplastic Crisis

More common in childhood. Usual presentation is with a viral prodrome and onset of fatigue, breathlessness, and pallor. Anaemia is caused by Erythrovirus /parvovirus B19 infection.

The Hb is often far below baseline (typically 30-60g/l) with reticulocytopenia. Parvovirus serology IgM +/- parvovirus DNA should be checked to confirm. Management is supportive with blood transfusion.

Recovery usually occurs within 5-7 days of presentation and is accompanied by a reticulocytosis and the development of IgG antibodies which give lifelong immunity and prevent recurrence.

Sequestration crisis

More common in childhood. Sequestration (pooling of blood in an organ) can occur especially in children with HbSS. In contrast to aplastic crises, the reticulocyte count is elevated in sequestration crises.

Splenic sequestration is a major cause of acute anaemia in children but less common in adults with HbSS who usually have small, autoinfarcted spleens. Splenic sequestration, while still rare, may occur more commonly in patients with HbSC disease and milder sickle phenotypes.

Management

- Fluid resuscitation if hypovolaemic
- +/- Antibiotics as per sickle pain crisis protocol
- Top-up transfusion if significant fall in Hb
- If transfused set lower target Hb to prevent hyperviscosity on reversal of pooling
- Monitor FBC closely as splenomegaly regresses and consider venesection if haematocrit > 0.36
- If splenic sequestration recurs consider splenectomy

Acute hepatic sequestration is a rare presentation in adults but should be considered in an adult presenting with acute anaemia and tender, enlarging hepatomegaly and increasing jaundice (predominantly conjugated bilirubin).

Management

- Fluid resuscitation if hypovolaemic
- Consider broad spectrum antibiotics e.g. cefuroxime 1.5g tds iv and metronidazole 500mg tds iv
- Top-up or exchange transfusion
- If transfused set lower target Hb to prevent hyperviscosity on reversal of pooling
- Monitor FBC closely as hepatomegaly regresses and consider venesection if haematocrit > 0.36

Delayed haemolytic transfusion reaction (DHTR)

Often presents with acute bone pain, anaemia and increasing jaundice 5-14 days post transfusion.

Fall in Hb is associated with rise in bilirubin and LDH and positive DAT due to the development of new red cell allo-antibodies.

Management is supportive with analgesia and fluids. If fall in Hb is significant and new allo-Abs detected, then transfusion with Ag negative blood is appropriate.

If fall in Hb is to a level below pre-transfusion Hb and the reticulocyte count is low then consider hyperhaemolytic transfusion reaction and avoid transfusion. Transfusion under cover of IVIgs and steroids may be necessary if anaemia is life-threatening.

See hyperhaemolysis guideline.