Guidelines on the Management of a Child with Sickle Cell Disease and worsening Anaemia

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Aim

This guideline describes the management of children in Hospital with sickle cell disease who have symptomatic anaemia or a significant fall in haemoglobin below their baseline level. It applies to patients with sickle cell disease who are currently under the care of the Paediatric Haematology team or have been admitted to a paediatric ward. It is mainly written as a tool for the medical team managing these patients, but any member of the multidisciplinary team may find it useful.

This guideline provides advice on the assessment and management of children with sickle cell disease who are found to have low haemoglobin, more than 20g/l below their steady state, or less than 50g/l. This principally involves two conditions: acute sequestration and transient red cell aplasia due to Parvovirus B19 infection. Severe anaemia may occur as part of an episode of acute pain or acute chest syndrome but will be dealt with in guidelines specifically dealing with these problems.

Background

Steady-state haemoglobin varies widely in children with sickle cell disease, from about 50 - 110g/l, and the blood count needs to be interpreted in the light of the known steady state which will typically be available on EPR. Patients with HbSC disease and some types of HbS/β^{+}thalassaemia typically have higher baseline haemoglobin levels, >100g/l. If there is no record of steady-state haemoglobin, the parents or child may know the normal level. Haemoglobin less than 50g/l is likely to be significant. Similarly spleen and liver size need to be compared to the steady-state size, which should be available in the notes and clinic letters on EPR. The spleen is palpable in some children up to the ages of 5 years years, and typically becomes smaller after that. Some children have persistent splenomegaly or hepatomegaly, and clinical findings are difficult to interpret unless the size has been recorded previously when the child was well.

History and Examination

Severe anaemia should be suspected in children with any of the following symptoms and signs: pain, dyspnoea, tiredness, lethargy, fever, pallor, increased jaundice, splenomegaly, hepatomegaly, tachycardia, tachypnoea. History and examination should elucidate these features in all children with sickle cell disease. Severe anaemia can be present in a relatively well child, who does not have pain. Children may have had finger-prick haemoglobin measured in A&E, (venous gas or haemocue) and any low readings should be confirmed by formal laboratory testing.

Investigations

The following tests should be performed in children with suspected anaemia:

-FBC
-Reticulocyte count
-Blood group & save
-Renal function
-Liver function tests and ALT.

Interpretation of Investigations

Haemoglobin should be considered low if it is more than 20g/l less than the steady-state for that child, or <50g/l.

Low Hb, reticulocytes > 200 x 10^9/l, spleen larger than in steady state (or palpable if previous spleen size unknown)
- probable splenic sequestration.

Low Hb, reticulocyte > 200x10^9/l, liver larger than steady state, no increased splenomegaly
- **probable hepatic sequestration (very rare).**

Low Hb, reticulocytes < 100 x 10^9/l
- **probable red cell aplasia due to Parvovirus B19 infection**.

Low Hb, reticulocytes > 200 x 10^9/l, spleen unchanged from steady state
- **probable Increased haemolysis in acute episode**

Other results interpreted depending on clinical situation, previous findings.

Acute Splenic Sequestration
Typically occurs under 3 years of age, but can occur in older children who have persistent palpable splenomegaly i.e. those with HbS/β thalassaemia, HbSS with α thalassaemia. It is often precipitated by infection.

Symptoms: collapse, shock, fever, abdominal pain, lethargy
Signs: Splenomegaly (often increasing rapidly in size over a few hours), pallor, tachycardia, tachypnoea, hypotension.

Investigations: as above plus blood cultures.

Initial Management:

- If shocked, resuscitate with intravenous fluids and emergency blood – Group O RhD negative according to APLS algorithm. Contact HDU.
- If not shocked, await results, cross-matched blood. If the spleen is enlarging or Hb < 50g/l, blood transfusion will usually be necessary. The maximum volume to be given in a single transfusion should be no more than 20ml/kg to avoid fluid overload, and then reassess by checking the FBC. More than one transfusion may be required; the Hb should not increase > 100g/l due to the risk of hyperviscosity.
- Start intravenous antibiotics – cefuroxime and adjust antibiotics according to blood culture results. If the patient has a known allergy to penicillin, ciprofloxacin (4mg/kg 12 hourly intravenously) and gentamicin should be given. Please seek local microbiology advise.
- Antibiotics should be continued until the child is clinically better and may be adjusted depending on the results of blood cultures. Typically the child will be discharged on their

normal penicillin prophylaxis, although a course of oral antibiotics may be given, depending on the clinical situation and the results of blood cultures.
- Monitor and record spleen size 6-hourly.
- Repeat FBC 12 hourly until stable, or more frequently if clinical deterioration.
- Treat other complications e.g. pain according to other protocols.
- Ultrasound of the abdomen may be helpful but should not delay resuscitation and transfusion

Further Management

Patients should be kept in hospital until pain and other symptoms have resolved, and the spleen size is stable or reducing and the haemoglobin has been stable for 24 hours.

Parents should be warned that recurrent episodes can occur and the symptoms explained. They should be shown how to feel the spleen and know to feel it if the child develops symptoms. They should know to bring the child to hospital urgently if symptoms recur or the spleen is thought to be enlarging.

The child should be seen in outpatients in 5 to 7 days. If episodes recur, splenectomy may be appropriate; recurrent episodes usually require splenectomy.

Hepatic Sequestration

Hepatic sequestration can occur at any age, and is less well defined than splenic sequestration. In general, acute enlargement of the liver accompanied by falling haemoglobin constitutes hepatic sequestration, and treatment is similar to that given for splenic sequestration. Rapid enlargement and shock are less common, but deteriorating liver function and very rarely acute liver failure can occur.

Symptoms: abdominal pain (right upper quadrant), abdominal distension, lethargy, malaise

Signs: Enlarging tender hepatomegaly, increasing jaundice.

Investigations: as above with blood cultures, clotting screen. Ultrasound of the abdomen may be helpful but should not delay resuscitation and transfusion.

Initial Management:

- If shocked, resuscitate with intravenous fluids and emergency blood – Group O RhD negative.
- If not shocked, await results, cross-matched blood. If the liver is enlarging or Hb < 50g/l, blood transfusion will usually be necessary, to increase Hb to about 100g/l. The maximum volume to be given in a single transfusion should be no more than 20ml/kg to avoid fluid overload, and then reassess by checking the FBC. More than one transfusion may be required; the Hb should not increase > 100g/l due to the risk of hyperviscosity.
- If the patient is in severe abdominal pain and clinically deteriorating, an exchange transfusion should be considered.

- Start intravenous antibiotics – cefuroxime and adjust antibiotics according to blood culture results. If the patient has a known allergy to penicillin, ciprofloxacin (4mg/kg 12 hourly intravenously) and gentamicin should be given.
- Monitor and record liver size 6 hourly.
- Repeat FBC, LFT's and INR 12 hourly until stable, or more frequently if clinical deterioration.
- If liver failure occurs, discuss with on-call liver team.
- Treat other complications e.g. pain according to protocols.

Further Management

If liver remains enlarged or liver function tests markedly deranged, arrange further investigations following discussion with hepatologists.

Acute Anaemia with Reticulocytopenia

The combination of low haemoglobin with low reticulocyte count is called acute red cell aplasia. This is nearly always caused by Parvovirus (Erythrovirus) B19 infection, although other viruses, such as influenza, may cause this to a lesser extent. Parvovirus B19 infection may also cause a reduction in platelet and white cell count, although this is usually not clinically significant. Thrombocytopenia (Platelets < 50 x 10^9/l) or neutropenia (Neutrophils < 1.0 x 10^9/l) are not typical and should be further investigated.

Symptoms: lethargy, tiredness. Parvovirus may also cause high fevers, abdominal pain, 'Slapped-cheek syndrome', Fifth Disease, arthritis, nephritic syndrome, nephrotic syndrome.

Signs: Pallor, lymphadenopathy, tachycardia, tachypnoea, fever (jaundice is usually absent), red cheeks.

Investigations: FBC, reticulocyte count, Parvovirus B19 serology, Group and Save/Cross match, Urea and electrolytes, liver function tests and ALT, urine albumin: creatinine ratio. Parvovirus DNA levels may be useful if there is diagnostic uncertainty, particularly in immunosuppressed children or it there has been recent blood transfusion.

Acute Management:

- If shocked, resuscitate with intravenous fluids and emergency blood – Group O RhD negative according to APLS algorithm.
- If not shocked, await results, cross-matched blood and transfuse to a target haemoglobin of 80-100g/l.
- Start intravenous antibiotics if febrile.
- Repeat FBC, reticulocytes daily.
- Treat complications e.g. pain according to other protocols.
- If Parvovirus infection is thought likely (anaemia and reticulocytopenia), the child should be reverse-barrier nursed in a side-room. Particular care should be taken to keep the child isolated from patients who might be immunosuppressed. Clinical and ward staff who are, or might be, pregnant should avoid contact with the child.
- The family should be warned that other family members/contacts with haemolytic anaemias might also develop significant anaemia, and blood tests arranged if appropriate. Similarly, pregnant relatives should be warned to avoid the patient.

Further Management

Once the haemoglobin has been corrected by transfusion and the child is stable, the child can be discharged before the reticulocyte count has recovered. A follow-up appointment should be made for 3-5 days later to document blood count recovery.

Any family member with sickle cell disease should be advised to contact their teams

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South Thames Sickle Cell & Thalassaemia Network

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Additional contacts can be found on the STSTN website (www.ststn.co.uk)

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