Sickle Cell - Paediatric Full Clinical Guideline - Joint Burton & Derby

Reference no.: CH CLIN G 91/ Aug 20/v005

Purpose

To improve and standardise care of children presenting with sickle cell crises according to recently published evidence and guidance.

Aim & Scope

This guideline covers the management of children presenting with sickle cell to the CED, day case or on the ward across both sites of Burton and Derby.

Background

Prevalence: Sickle cell disease is the most common genetic disorder in England, affecting more than 1 in 2000 live births. It causes high levels of morbidity, and still significant rates of mortality (1-2%), in childhood. It should now be detected in most cases at routine neonatal screening.

Cause: Normal adult haemoglobin (Hb A) consists of 2x α + 2x β chains. Hb S results from a single amino acid substitution on the beta chain and is less soluble. When deoxygenated, Hb S undergoes polymerization, leading to sickle-shaped cells. This results in endothelial damage and blockage of blood vessels, causing vaso-occlusive events. Children who are homozygous for Hb S (SS) have the most severe disease; those with an S/C or S/β-thalassaemia genotype are also affected. Heterozygotes are usually asymptomatic, except in high risk situations where there is less oxygen then normal, such as general anaesthesia.

Hyposplenism: these children are also at higher risk of severe infection from encapsulated bacteria (e.g. Pneumococcus, Streptococcus, HiB, Salmonella spp.). They should therefore receive regular Pneumovax immunisation following normal course of vaccinations, and annual influenza vaccination, and start long- term prophylactic penicillin before the age of 3 months.

Regional Network: Queens Medical Centre in Nottingham is the local regional centre for Derby patients. Birmingham Children's Hospital is the regional centre for Burton patients. These hospitals provides advice and expertise, accepts transfers of serious or difficult cases. QMC offers annual review of all children in the region in a joint clinic in derby with Dr Claire Weights. Birmingham Hospital provide all annual reviews for Burton patients. Transcranial Doppler screening is offered for all children with SS disease every year at the QMC for Derby patients and Birmingham for Burton patients. Derby is part of the East Midlands Sickle Cell and Thalassaemia Network.

Acute Presentations

Type	Pathogenesis	Symptoms + Signs
Bony crisis -Most common reason for admission	Vaso-occlusion within bone / bone marrow vasculature	Acute pain +/- swelling in any area, mild fever may be present
Acute chest syndrome -Major cause of mortality	Infarction, infection	Chest pain, fever, cough +/- wheeze, respiratory distress, hypoxia, chest signs, CXR abnormalities (signs of infection, infarction or sequestration)
Abdominal crisis	Ischemia / infarction leading to ileus	Abdo pain, fever, vomiting, distension, guarding, fluid levels in bowel loops on AXR
Acute sequestration - usually spleen, rarely liver	Blockage of venous drainage	Rapid increase in spleen (or liver) size, abdo pain, symptomatic anaemia, hypovolaemic shock
CNS event - TIA, stroke	Ischemia / infarction / haemorrhage	Acute neurological deficit, seizure, headache, raised intracranial pressure.
Priapism	Blockage of venous drainage	Pain NEEDS URGENT TX
Aplastic crisis	Usually caused by infection with Parvovirus B19	Symptomatic anaemia - pallor, shortness of breath, fatigue. Also signs of Parvovirus B19 infection may be present (fever, red cheeks). Low Hb with absent reticulocytes

Other complications

Eye complications - proliferate retinopathy, haemorrhage, orbital infarction – urgent ophthalmology review required

Airway hyperreactivity, and longer term risk of pulmonary hypertension

Osteomyelitis – causative organisms include salmonella

Cholecystitis - gallstones secondary to high rate of haemolysis

DVT (very rare)

Acute myocardial infarction (very rare)

Always take note if a patient tells you their pain is not typical – this may require further investigation

Management

Please contact Dr Claire Weights to make aware of admission for all Derby patients, and Dr Mansoor Ahmed for all Burton patients.

General

1. O2
2. Warmth
3. Fluids - 150% maintenance: oral / NG if very well, otherwise IV.
   (Avoid cannulation of lower limbs if possible – risk of ulceration).
4. Analgesia – should be started <15 minutes from arrival, and pain controlled <30 minutes from arrival
   Consult each child's individual pain management plan (Notes, with patient, ICM or folder in CED)

⇒ If Severe -usually require oral or IV morphine / Intranasal diamorphine** for rapid control of pain. Once controlled manage with oral morphine if possible, otherwise PCA / NCA.

⇒ Prescribe paracetamol + NSAID as well as opioids (**avoid pethidine**)

⇒ Prescribe regular laxative and PRN antiemetic if commenced on opioids

5. Check: FBC (including reticulocyte count), U+E, CRP, LFTs, Hemoglobin electrophoresis for HbS levels, cultures of blood / other sites as appropriate
5. Treat infection with antibiotics if suspected

** Guideline for Intranasal Diamorphine (ref CH PH C07) can be found the intranet, in the Children's Clinical and operational Policy and Guideline folders and there is a copy in available in CED.

Specific / Additional

Bony crisis - Consider osteomyelitis (including salmonella) if persistent pyrexia, positive blood cultures, high CRP (may be secondary to infarction, similar symptoms)

Acute chest syndrome - Urgent CXR, blood gas, broad spectrum antibiotics (IV cefuroxime + IV clarithromycin [or oral erythromycin if well enough]), consider trial of bronchodilators, incentive spirometry. **If severe will require urgent exchange transfusion- discuss with regional center if suspected.** If persistent pain consider checking cardiac enzymes.

Abdominal crisis - Check LFTs, abdo USS to look for gallstones + splenic size.
Surgical review unless mild symptoms without guarding.
If on iron chelation therapy, and presenting with abdominal pain and diarrhoea, consider Yersinia infection.

Sequestration – **Urgent transfusion and discuss with regional center** (not dependent on Hb level), needs careful monitoring. Avoid surgery during acute episode but consider splenectomy at a later date.

CNS event - Urgent CT brain, **urgent exchange transfusion if confirmed**.
Involve neurosurgeons if haemorrhagic event

Priapism – NEEDS URGENT TREATMENT.
Early surgical review. May require aspiration and irrigation with etilefrine / ephedrine. ***D/W Paediatric Consultant and Surgical Registrar and involve tertiary centre early.***

Aplastic crisis - Check Parvovirus B19 serology. See 'Transfusion' below.

Pyrexia - Culture blood and any other suspected sites of infection. Consider malaria films, particularly if travelled to at risk region <1 year ago.

Transfusion - Consider if: symptomatic anaemia, Hb<5g/dl with significant drop from normal level (except in sequestration), particularly if absent reticulocyte response.
Blood must be Rh and Kell antigen matched, leucocyte depleted and CMV negative if child is CMV negative.
Aim for Hb of 10g/dl.

Other Considerations

1. Continue regular folic acid and prophylactic penicillin.
2. Check vaccination status.
3. Involve the Regional Haemoglobinopathy Specialist Nurse, Hazel Marriott (07812 268407) for Derby patients. Involve tertiary centres if needed.
4. Physiotherapy for acute chest or back pain, chest infections, difficulty mobilising once pain controlled.
5. If on hydroxyurea, stop during acute crisis if: neutropenic (neutrophils <1.5l)/ thrombocytopenic (<100)/ low reticulocyte count (<100).
6. Ensure follow-up appointment in place with Dr Claire Weights (CLWSS) for Derby patients or Dr Mansoor Ahmed for Burton patients

References

1. 'Sickle cell disease in children, standards for clinical care'. https://www.sicklecellsociety.org/wp-content/uploads/2019/11/SCD-in-Childhood_Final-version-1.pdf November 2019
2. Sickle Cell Disease: managing acute painful episodes in hospital. Nice Guidelines, published June 2012, reviewed 2016. https://www.nice.org.uk/guidance/cg143
3. Sickle Cell Acute Management – Nottingham Guidelines
4. National confidential enquiry into patient outcome and death – May 2008 – 'a sickle crisis?'. Full report available via NCEPOD website: http://www.ncepod.org.uk/newsletters.htm
5. Dick MC. Standards for the management of sickle cell disease in children. Arch Dis Child Educ Pract Ed, 2008;**93:**169-176.
6. Shord SS et al. Evaluation of opioid induced nausea and vomiting in sickle cell disease. Am J Hematol, 2008 Mar;**83(3):**196-9
7. Guidelines for the management of the acute painful crisis in sickle cell disease. Prepared on behalf of the British Committee for Standards in Haematology General Haematology Task Force by the Sickle Cell Working Party. British Journal of Haematology, 2003;**120:**744-52

Documentation Controls

Development of Guideline:	Dr C Weights
Consultation with:	Dr Simone Stokely (Consultant Paediatric Haemotologist QMC),Liz Taylor (Pain Specialist), Lisa Taylor & Liz Bedford (Paediatric Pharmacists), Dr Khare (Consultant Microbiologist), Dr Elliot (Consultant Paediatric Radiologist)
Approved By:	*Paediatric Business Unit, Guidelines Group, Women's and Children's Division, 27th August 2020.*
Review Date:	August 2023 – Extended to May 2025
Key Contact:	Dr C Weights