Management of an Acute Painful Crisis in Sickle Cell Disease

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Adults with Sickle Cell Disease presenting with an acute painful sickle cell episode should be treated as an acute medical emergency.

Background : Sickle cell disease affects up to 15000 people in the UK (ref). Many of these patients will present acutely to a variety of hospitals in the country and may not be known already to staff at the hospital.

Sickle cell disease (either HbSS or compound heterozygote states Hb SC, HbSβ Thal or other compound states) commonly gives rise to acute painful crises . The pain may occur in any part of the body but is commonly in the limbs. Pain in the chest or abdomen should prompt consideration of alternative causes of pain in those sites (eg MI, acute appendicitis). Sickle pain is very severe usually requiring opiate analgesia, may have a pain score of up to 10/10 and it is usually clear to the patient that this is their 'usual' sickle pain (ie patients with sickle cell disease are the experts in their painful crises).

Patients with sickle cell disease often have a chronic anaemia of 60-100g/L which is normal for them. Blood transfusion is **NOT** routinely indicated and in fact may exacerbate a crisis. **Blood transfusions should only be used after advised by the Haematologist.**

The pathogenesis of sickle cell pain is thought to be due to deoxygenated sickle Hb forming large polymers which causes red cells to become deformed and show irreversible sickling leading to small vessel obstruction. Triggering factors are cold, dehydration and infection but often no specific trigger is found.

Common Presentations

- severe pain
- pyrexia, tachycardia, tachypnoea, hypotension
- increased pallor, breathlessness, exhaustion
- chest pain, consolidation on CXR, SaO_2 <94% on air
- abdominal pain or distension, diarrhea, vomiting

- severe thoracic/back pain
- headache, drowsiness, CVA,TIA or any abnormal CNS signs

Arrangements for Admission at RDH (adults)

Patients are advised to contact the Haematology Clinical Nurse Specialist on mobile via the switch board at Royal Derby Hospital during the working hours and on-call Haematologist out of hours. The patient will be told where they can be seen [Haematology wards or Medical Assessment Unit (MAU)]. Patients via ambulance, self presentation and some patients not known to the service would be admitted to A+E /MAU. In all cases the on-call Haematologist must be informed by the Haematology CNS during the working hours and by the A&E/MAU during the out of hours.

If seen on MAU and requiring admission for longer than 18 hours, patients will normally have to be moved to another ward: Haematology wards or another available bed.

Assessment and Initial Management of Patient with Sickle Cell Disease

The general guide for these patients is to speedily assess them – review the pain and observations immediately. Generally these patients get admitted as the pain is uncontrolled with Home treatment.

PAIN: If the pain is typical sickle cell pain uncontrolled with home analgesics and observations are stable and there are no unusual features then analgesia should be given as below. **Analgesia must be administered within 30 minutes of admission (NICE recommendation).**

- Check previous drug charts for reference or
- Give the combination of analgesics for adequate and quicker pain control. escalate rapidly if severe pain.
    - REGULAR Paracetamol 1g Four times a day - IV if necessary initially) then Oral. Plus
    - REGULAR Ibuprofen 400mg Three times a day (if not contraindicated). Plus
    - Morphine 5-10mg (0.15mg/kg) IV - IV Morphine 1mg per ml in a 10ml syringe 5mg initial bolus titrating up to a dose of 20mg with a review every 5 minutes. Titrate until pain controlled Commence and record frequent observations
    - Patients with tolerance may need a higher starting dose and a higher total dose**.** Pethidine should **NOT** be used.
    - IV morphine should be given by a trained nurse or a doctor managing the acute admission.

- **Adequate analgesia should be achieved by 60 minutes. This will reduce the requirement for morphine and avoid the need for PCA.**
- Once pain controlled observations must be done HOURLY for first 6 hrs and then 4hourly.
- Inform the Acute Pain Team Nurses early via bleep if the sickle pain is not controlled with i.v. or s.c. morphine by 60mins and they will review and advise appropriately.
- For persistant pain after 90min of i.v Morphine, contact the ITU outreach team/ITU on-call for PCA **after discussion with the on-call Haematologist.**
- Laxatives, antiemetics and anti-pyretics should be prescribed usually on a PRN basis

Investigations

Patients presenting with sickle cell crisis require the following investigations

- FBC and reticulocyte count
- Group and save
- U&E and LFT (may be obstructive if gall stones)
- CXR
- Blood cultures, MSU
- ABG if < 95% on air
- XR is indicated if a patient develops localised bone pain and fever as osteomyelitis is a recognised and potentially serious complication of sickle cell disease.

Pain management after first 90 minutes

Time from 1st review	Reason	Action	Comments
0-90 mins	Immediate control acute pain Supportive measures	1. Paracetamol can be given IV in acute phase 2. Ibuprofen if no contraindication to NSAIDs 3. Morphine 0.15 mg/kg i/v and titrate until pain controlled and commence frequent observations 4. Reassess pain and side-effects of morphine regularly Prochlorperazine or cyclizine, Chlorpheniramine, Lactulose + Senna - should be prescribed as PRN 5. Enoxaparin 40mg sub-cutaneous for thromboprphylaxis 6. Intravenous fluids: 2-3 litres over 24hours 7. Oxygen – maintain O2 satuartions >95% at all times	Can give sub-cutaneously to avoid delays **All these action measures are very essential upfront**
>90 mins	Continued intermittent analgesia-administered by staff	Morphine 0.15 mg/kg i/v or s/c every 2-4 hours. Extra doses if pain persistent (50% of given dose) after 1 hour. Observations atleast HOURLY for the first 6 hours **OR**	
>90mins	PCA (patient controlled analgesia)	Follow the Trust guidelines on PCA Monitor the pain Record the pain score and sedation score on the Acute Pain Chart	**Avoid the delays in starting PCA**
Every 30 mins then every 1 hr	Monitor	Pain, sedation, vital signs, respiratory rate until pain controlled and stable then every 1 hour	

Sedation assessment

Patient	Sedation score	Action
Awake and alert	0	Continue with regular observations
Drowsy, easy to rouse	1	If respiratory rate <10/min, give 40% oxygen + inform the ITU outreach team/ on-call ITU doctor
Very drowsy, difficult to rouse	2	If respiratory rate <6/min, give 100% Oxygen and Naloxone. Hold opioids. Inform the outreach team.
Normal sleep, easy to rouse	S	Continue with regular observations

Alternative analgesics to consider

- Oxycodone, Ketamine , Methadone are alternatives but their use should always be discussed with a consultant.

Alteration of dose of analgesia

Requirements for analgesia needs to be reconsidered on a daily basis at least – when pain is well controlled aim to reduce dose by small amounts and switch to oral equivalents when patient is clearly improving (pain score /chart may be helpful)

Oxygen

- There is no good evidence for this being used routinely in all cases of painful crisis
- Its use should be dictated by the clinical situation and oxygen saturations:
- If SaO_2 <95% on air, give O_2 by face mask
- Check arterial gases if SaO_2 on air is <95%
- Monitor SaO_2 while patient is on supplementary oxygen aiming to keep O_2 level > 98%
- If arterial pO_2 (PaO_2) <10.7kPa use mask giving 35% inspired oxygen
- If arterial PaO_2 < 9.3kPa seek additional help – generally by involving HDU outreach team

Thromboprophylaxis

- Sickle cell disorders are associated with an increased thrombotic potential
- All patients who are admitted with a severe sickling crisis who are immobile should be commenced on low molecular weight heparin prophylaxis
- Do not use TED stockings

Antibiotics

- If the patient is febrile or has a history suggestive of an infective cause of the sickle crisis they should be commenced on antibiotics
- Co-amoxiclav is the antibiotic of choice in most cases unless penicillin allergic.
- Most patients are on oral penicillin maintenance which can be stopped and restarted when the course of Co-Amoxiclav has been completed.

Transfusions

- Patients with sickle cell disease often have a chronic anaemia of 6-10 g/dl which is normal for them.
- Hb S has a lower oxygen affinity than Hb A so tissue oxygenation is better than expected

- It is useful to check the patient's steady state Hb level (eg from an OPD appointment) when reviewing their blood count.
- The Hb may fall 1-2 g during a sickle crisis but blood transfusion is **NOT** routinely indicated and in fact may exacerbate a crisis

Blood transfusions should only be used after advised by the Haematologist if there are symptoms of severe anaemia, or if the Hb has fallen > 2g and is below 5 g/dl.

- Check HbS% before transfusion.
- The transfusion should aim to return the Hb to the steady state level and the blood should be matched for Rhesus and Kell antigens.

Exchange Transfusion

- The aim of an exchange transfusion is to replace the sickle blood with HbAA
- Exchange transfusion should be discussed with a Consultant Haematologist.
- The indications for urgent exchange transfusion are for
    - Sickle chest crisis
    - Cerebral sickling
    - Multiorgan failure
- Red cell exchange is preferably done using the cell separator via the stem cell nurses at Haematology Day Case Unit, Nottingham City Hospital. This is discussed directly as Consultant to Consultant referral with Dr Sarah Donohue at Nottingham City Hospital.
- Manual exchange transfusion may be necessary and performed in an emergency
- Routine elective exchange transfusion may be done for recurrent severe crises or pre-operatively
- Blood should be Rhesus and Kell genotyped for the patient and Blood Bank should be contacted as soon as an exchange transfusion is planned
- Hb A and S levels should be sent to the Haematology laboratory after manual exchange transfusion.
- A Hb S level of < 30% and Hb of 90gm(<100)/L is the goal of this therapy

References

Guidelines for the management of the acute painful crisis in sickle cell disease. Rees et al, British Journal of Haematology 2003, 120: 744-752.

Sickle cell acute painful episode: management of an acute painful sickle cell episode in hospital, NICE clinical guideline 143, June 2012

Adopted from East Midlands Network Guideline, Nottingham City Hospital.

Trust guidelines on IV and Sub-cutaneous morphine; and PCA, Royal Derby Hospital.

Documentation Controls

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