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SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

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Management of Acute Painful Crisis in adults with 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 based on 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.

Introduction

Painful crisis (vaso-occlusive crisis) is the commonest manifestation of sickle cell disease requiring hospital assessment and admission. Crises can be precipitated by many things including cold, infection, hypoxia and dehydration. Vaso-occlusion within the bone marrow vasculature leads to bone infarction, which in turn results in the release of inflammatory mediators that activate afferent nerve fibres and cause pain. Pain can be extremely severe and should be addressed urgently.

Patients should be triaged as high priority, with first dose of analgesia within 30 minutes, and the Haematology team should be contacted.

Management is supportive (i.e. conservative) unless there are additional complications. The aim of treatment is to provide symptomatic relief, whilst breaking the vicious cycle of sickling, hypoxia, acidosis and tissue damage.

Principals of clinical assessment

- How severe is the pain and what analgesia is required?
- Is there evidence of concurrent infection/sepsis?
- Is there evidence of chest crisis?
- Are there features of other severe sickle complications? E.g. stroke, acute severe anaemia (>20g/l below baseline), priapism, splenic enlargement

Principals of management

1. Timely, safe, effective analgesia SEE FLOWCHART BELOW
2. Hydration
3. Oxygenation
4. Warmth
5. Awareness of complications requiring urgent specialist intervention
6. Antimicrobials (prophylactic or therapeutic)
7. Thromboprophylaxis

Routine investigations

- FBC, reticulocytes, U&E, LFT, LDH, Ferritin
- Group & Save
  Specify "Sickle Cell Disease". Request full red cell phenotype if new patient.
- Diagnostic Haemoglobinopathy screen in new patients

If febrile or clinical features of infection

- Blood cultures/Throat swab/Urine culture/Sputum/Stool/Wound swabs etc
- Chest Xray

MANAGEMENT: Commence appropriate antibiotics e.g. If no clear source Co-amoxiclav (Unless penicillin allergic) for 5 days.

If respiratory symptoms/signs, fever, chest pain or oxygen saturations <95% on air. Consider co-existing ACUTE CHEST CRISIS = leading cause of mortality in SCD

- Chest Xray
- Arterial blood gas

MANAGEMENT: Discuss any concerns urgently with Haematology on call (24/7) as urgent intervention may be indicated. Highlight to HDU/ITU teams. Refer to SPAH acute chest syndrome guideline.

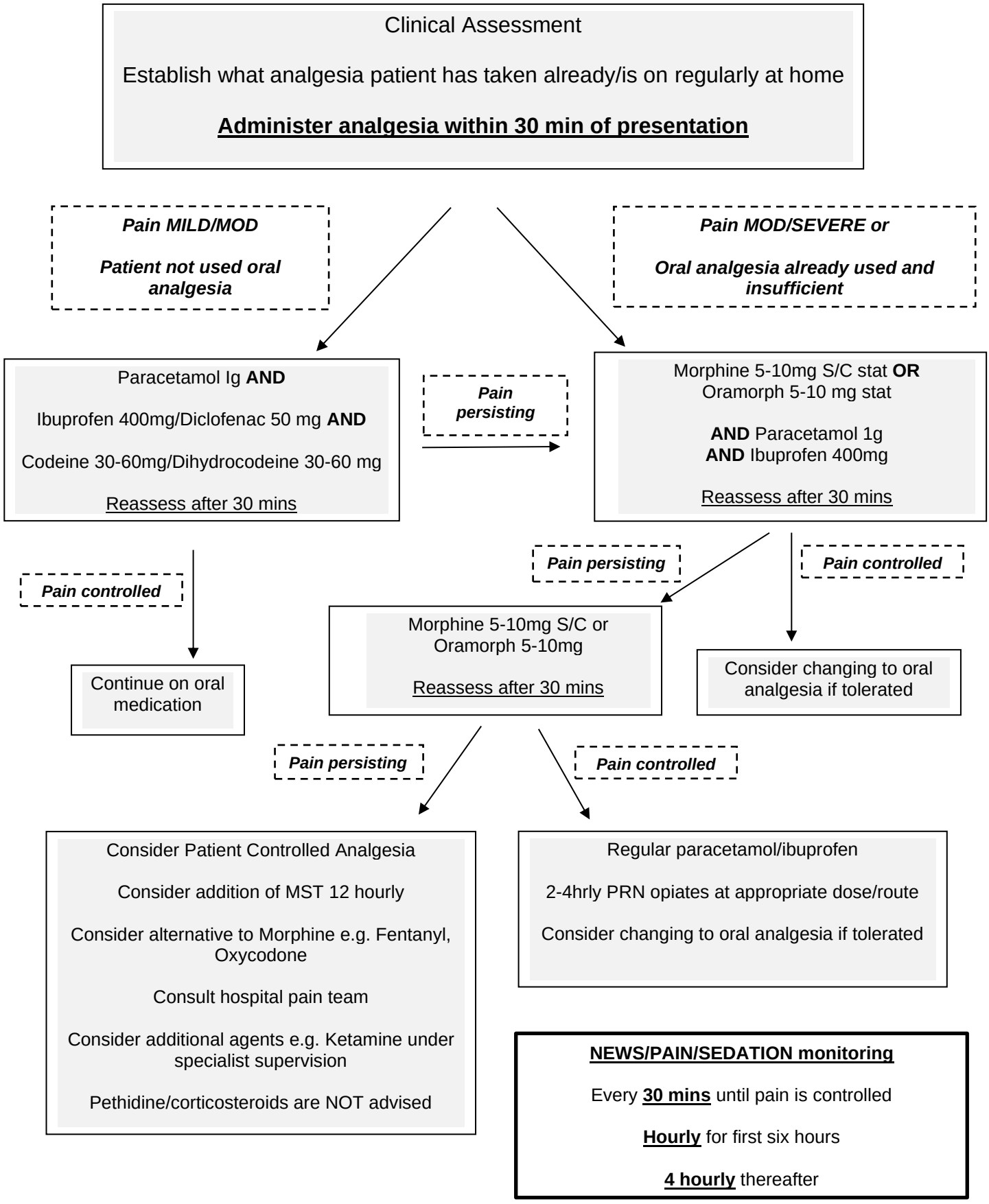
1. Analgesia

Some patients have individualised pain protocols which should be followed; otherwise **follow chart below**, assuming normal renal function.

For patients with renal impairment, consider dose reduction/alternative agents e.g. oxycodone or fentanyl.

Naloxone should be available for reversal of sedation and/or respiratory depression

Consider supportive medications:
Folic acid 5md OD PO
Antipruritics: Hydroxyzine 25 mg BD PO PRN
Antiemetics: e.g. Cyclizine 50 mg TDS PO PRN (avoid IV cyclizine)
Laxatives (if opioid analgesia is to continue)


Clinical Assessment
Establish what analgesia patient has taken already/is on regularly at home
Administer analgesia within 30 min of presentation
Pain MILD/MOD
Patient not used oral analgesia
Pain MOD/SEVERE or
Oral analgesia already used and insufficient
Paracetamol Ig AND
Ibuprofen 400mg/Diclofenac 50 mg AND
Codeine 30-60mg/Dihydrocodeine 30-60 mg
Reassess after 30 mins
Pain persisting
Morphine 5-10mg S/C stat OR
Oramorph 5-10 mg stat
AND Paracetamol 1g
AND Ibuprofen 400mg
Reassess after 30 mins
Pain controlled
Continue on oral medication
Pain persisting
Pain controlled
Morphine 5-10mg S/C or
Oramorph 5-10mg
Reassess after 30 mins
Consider changing to oral analgesia if tolerated
Pain persisting
Pain controlled
Consider Patient Controlled Analgesia
Consider addition of MST 12 hourly
Consider alternative to Morphine e.g. Fentanyl, Oxycodone
Consult hospital pain team
Consider additional agents e.g. Ketamine under specialist supervision
Pethidine/corticosteroids are NOT advised
Regular paracetamol/ibuprofen
2-4hrly PRN opiates at appropriate dose/route
Consider changing to oral analgesia if tolerated
NEWS/PAIN/SEDATION monitoring
Every 30 mins until pain is controlled
Hourly for first six hours
4 hourly thereafter

2. **Hydration**

- Adequate fluid intake is essential, aim for around 3L per 24 hours
- A fluid balance chart should be completed by nursing staff or patient (if able).
- Intravenous/NG fluids may be required if oral intake is inadequate

3. **Oxygenation**

- Some patients have a symptomatic benefit from oxygen therapy. This should be prescribed/available if the patient requests, even if not hypoxic
- If oxygenation deteriorates or saturations <95% on air, contact the on call haematologist urgently and consider chest crisis as above

4. **Warmth**

- Patients should be provided with extra blankets if requested
- Patients may have symptomatic benefit from warm packs

5. **Awareness of complications requiring urgent specialist intervention**

It is out with the scope of this document to cover all major complications of SCD; contact Haematology urgently if a patient with SCD is acutely unwell. Ensure valid transfusion samples are in the laboratory as transfusion is sometimes required.
Indications for/risks of transfusion in SCD differ significantly from the general population and should <u>always</u> be guided by specialist Haematology input

Below are some specific clinical emergencies to be aware of:

- Sepsis (related to hyposplenism)
- Acute Chest Syndrome (chest signs/symptoms, hypoxia, pulmonary infiltrates)
- Priapism (painful, unwanted erection)
- Stroke, at any age (ischaemic/haemorrhagic)

6. **Antimicrobials**

- Patients with SCD are hyposplenic and at risk of severe sepsis
- If febrile, seek to identify source of infection and start appropriate antibiotics
- If apyrexial continue prophylactic antibiotics e.g. Penicillin V/Erythromycin
- If patient is on Hydroxycarbamide (Hydroxyurea), check FBC urgently for neutropenia

7. **Thromboprophylaxis**

- Patients with SCD are at higher risk for venous thromboembolism and should be considered for thromboprophylaxis according to local protocols

References:

Sickle cell disease: managing acute painful episodes in hospital (2012) NICE guideline CG143
Overview | Sic
https://ashpublications.org/bloodadvances/article/4/12/2656/460974/American-Society-of-Hematology-2020-guidelines-forkle cell disease: managing acute painful episodes in hospital | Guidance | NICE

American Society of Haematology 2020 guidelines for sickle cell disease management of acute and chronic pain
American Societ
https://ashpublications.org/bloodadvances/article/4/12/2656/460974/American-Society-of-Hematology-2020-guidelines-fory of Hematology 2020 guidelines for sickle cell disease: management of acute and chronic pain | Blood Advances | American Society of Hematology (ashpublications.org)

Sickle Cell Society Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK
Standards-f https://www.sicklecellsociety.org/wp-content/uploads/2018/05/Standards-for-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdfor-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdf (sicklecellsociety.org)

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