NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Scottish Paediatric & Adult Haemoglobinopathy Network

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 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.

Introduction

The painful crisis is the commonest manifestation of sickle cell disease requiring hospital assessment and admission. The pain can be extremely severe and should be addressed urgently, with patients triaged as high priority and contact should be made with the on-call Haematology team.

Management is supportive (i.e. conservative) unless there are indications for exchange transfusion.

- Acute cerebro-vascular event
- Acute chest syndrome
- Multi-organ failure

The aim of treatment is to break the vicious cycle of sickling, hypoxia and acidosis leading to more sickling — all exacerbated by dehydration.

Prompt treatment of painful crises can reduce suffering and prevent further sickle related complications. **Analgaesia should be given within 30 minutes of the patient presenting with frequent reassessment (every 30-60 minutes).**

Principals of management

- Effective analgaesia
- Hydration (oral or IV fluid)
- Oxygenation
- Antimicrobials – prophylactic or therapeutic if pyrexial
- Ongoing assessment of analgaesic efficacy

Assessment

Routine Investigation (*Urgent requests)

- FBC, reticulocytes *
- Group & screen (state on form that patient has Sickle Cell Disease. Request full red cell phenotype if new patient)
- Urea, creatinine electrolytes *
- LFT's, LDH
- Baseline pulse oximetry ON AIR

Haemoglobin electrophoresis **in NEW patients only**

If indicated

- Blood cultures
- Viral serology
- Urine dipstick + MSU
- Throat swab

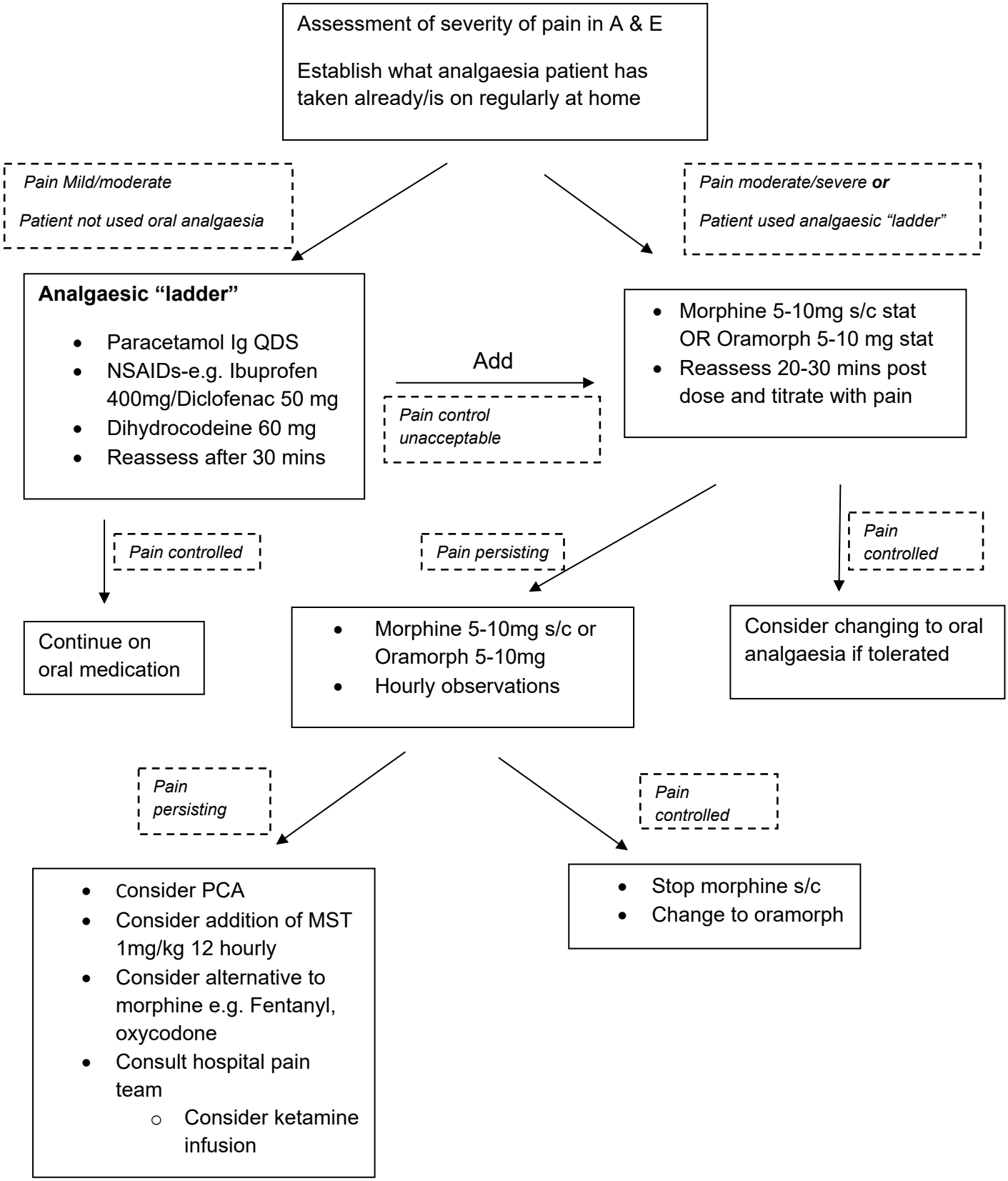
Additional Investigations

- If there are chest signs or temperature >38°: – Chest X-ray
- If O_2 sats on air < 94% – Arterial gases on air
- If there are abdominal signs: – Chest X-Ray, Abdominal X-ray and amylase
- Appropriate microbiological specimens (sputum, stool, wound, etc.)
- **Note:** Patients on Desferrioxamine (DFO), admitted with diarrhoea/abdominal pain, should have blood and stool screened for *Yersinia* and the DFO stopped.

Analgesia

Aim is to achieve safe, effective analgesia whilst avoiding IV opiates if possible.

Patients with end stage renal failure, consider alternative opiate e.g. Fentanyl
Some patients will have individualised pain protocols which should be referred to if available, otherwise follow chart below.


Assessment of severity of pain in A & E
Establish what analgaesia patient has taken already/is on regularly at home
Pain Mild/moderate
Patient not used oral analgaesia
Pain moderate/severe or
Patient used analgaesic "ladder"
Analgaesic "ladder"
Paracetamol Ig QDS
NSAIDs-e.g. Ibuprofen 400mg/Diclofenac 50 mg
Dihydrocodeine 60 mg
Reassess after 30 mins
Add
Pain control unacceptable
Morphine 5-10mg s/c stat OR Oramorph 5-10 mg stat
Reassess 20-30 mins post dose and titrate with pain
Pain controlled
Pain persisting
Pain controlled
Continue on oral medication
Morphine 5-10mg s/c or Oramorph 5-10mg
Hourly observations
Consider changing to oral analgaesia if tolerated
Pain persisting
Pain controlled
Consider PCA
Consider addition of MST 1mg/kg 12 hourly
Consider alternative to morphine e.g. Fentanyl, oxycodone
Consult hospital pain team
Consider ketamine infusion
Stop morphine s/c
Change to oramorph

Note

- Paracetamol and NSAIDS should be used in addition to opiates as required, as they have a synergistic effect
- All patients will have different analgaesic requirements and many know what they have required to achieve pain relief in the past. Analgaesia should be titrated with pain
- Patients should be monitored every 30 mins until pain is controlled and patient is stabilised and every 2 hours thereafter
- Monitoring must include pain, sedation, vital signs, respiratory rate, O2 Saturation
- Naloxone should be available for reversal of sedation and/or respiratory depression (RR<12/min)
- Pethidine is not recommended because of risk of seizures at high doses
- There is no role for the use of corticosteroids in acute pain management

Consider additional therapies:

- Antipruritics: Hydroxyzine 25 mg bd po
- Antiemetics: e.g. Cyclizine 50 mg tds
- Laxatives if opioid analgesia is to continue
- Folic acid 5mg od
- Prophylactic Low Molecular Weight Heparin
- Prophylactic antibiotics (usually penicillin V 250mg bd)
- Physiotherapy - **LINK**

Fluids

- Adequate fluid intake is essential.
- Patients should be encouraged to drink at least 3 litres of water-based fluids per 24 hours.
- Every patient must have a fluid balance chart which should be completed by the nursing staff or by the patient (if able).
- Intravenous or ng fluids may be required if the patient is unable to tolerate oral fluids

Oxygen

- Oxygen saturations on air should be monitored regularly
- Many patients have a symptomatic benefit from Oxygen therapy, and it should be prescribed and be available whatever the oxygen saturations (even if >98%) if the patient requests
- Oxygen saturations on air should be >94%
- If oxygen saturations on air <94% Call haematologist
- Check Arterial Blood Gases (ABGs) on air
- Administer humidified oxygen at 2-4 L/min by mask or nasal cannulae

- Increase frequency of observations to hourly or more frequently if clinical picture dictates
- Arterial Blood Gases: Consider a diagnosis of Acute Chest Syndrome if worsening hypoxia

- **Remember that excessive use of opioids can cause respiratory suppression**. (Naloxone is occasionally required).

Antimicrobials

- If apyrexial continue prophylactic antibiotics: **Penicillin V** 250 mg bd po (**Erythromycin** 250mg bd po if allergic)

- If temperature greater than 38°C, undertake blood cultures/septic screen and commence Co-amoxiclav (Unless penicillin allergic) for 5 days. Prophylactic antibiotics should be stopped

- If patient is on Hydroxycarbamide (Hydroxyurea), check FBC urgently and stop the Hydroxycarbamide if the platelet count <100x10^9/l, reticulocytes <100x10^9/l or neutrophils <1x10^9/l

Consider:

- Pneumococcal sepsis (especially if not taking prophylaxis and not vaccinated)
- Gram negative sepsis
- Lower respiratory tract infection
- Urinary tract infection
- Osteomyelitis
- Malaria if travelled recently
- Parvovirus B19 if low reticulocyte count
- *Yersinia* if on DFO and have diarrhoea

References

NICE guidance "Sickle cell disease: managing acute painful episodes in hospital (2012) NICE guideline CG143
https://www.nice.org.uk/guidance/cg143/evidence/full-guideline-pdf-186634333
American Society of Haematology 2020 guidelines for sickle cell disease management of acute and chronic pain
https://doi.org/10.1182/bloodadvances.2020001851

Acknowledgements

Thank you to the Haematology Departments of the Royal London Hospital, Manchester Royal Infirmary and St. Thomas's Hospital for allowing reference and adaptation of their guidelines for the management of Adults with Sickle Cell Disease.