Scottish Paediatric & Adult Haemoglobinopathy Network

Paediatric guideline - Painful Sickle Cell Crisis (Assessment and management)

Acute painful sickle cell episodes (also known as painful crises) are caused by blockage of the small blood vessels by sickled red cells. Pain can fluctuate in both intensity and duration, may be severe and is often underestimated by healthcare professionals. The majority of painful episodes are managed at home, with patients usually seeking hospital care only if the pain is uncontrolled or they have no access to analgesia. Patients who require admission may remain in hospital for several days.

The primary goal in the management of an acute painful sickle cell episode is to achieve effective pain control both promptly and safely. Otherwise, management is supportive unless there are indications for transfusion. The aim of treatment is to break the cycle of: sickling, hypoxia and acidosis - all exacerbated by dehydration.

TREAT AN ACUTE PAINFUL SICKLE CELL EPISODE AS AN ACUTE MEDICAL EMERGENCY.

Throughout an acute painful sickle cell episode, regard the patient (and/or their carer) as an expert in their condition, listen to their views and discuss with them:

- the planned treatment regimen for the episode
- treatment received during previous episodes
- any concerns they may have about the current episode
- any psychological and/or social support they may need.

GENERAL MANAGEMENT INCLUDES:

- **reassurance** that the patient's pain will be relieved as soon as possible
- **warmth** and establishing a position of maximum comfort
- **analgesia** including assessment of analgesia taken prior
- **hydration** which may be oral or IV depending on clinical assessment
- **intravenous access** if required for fluids, analgesia, antibiotics
- **identification and treatment of infection**
- **regular observations and reassessment**

For children with Chest pain or respiratory symptoms or signs consider acute chest syndrome

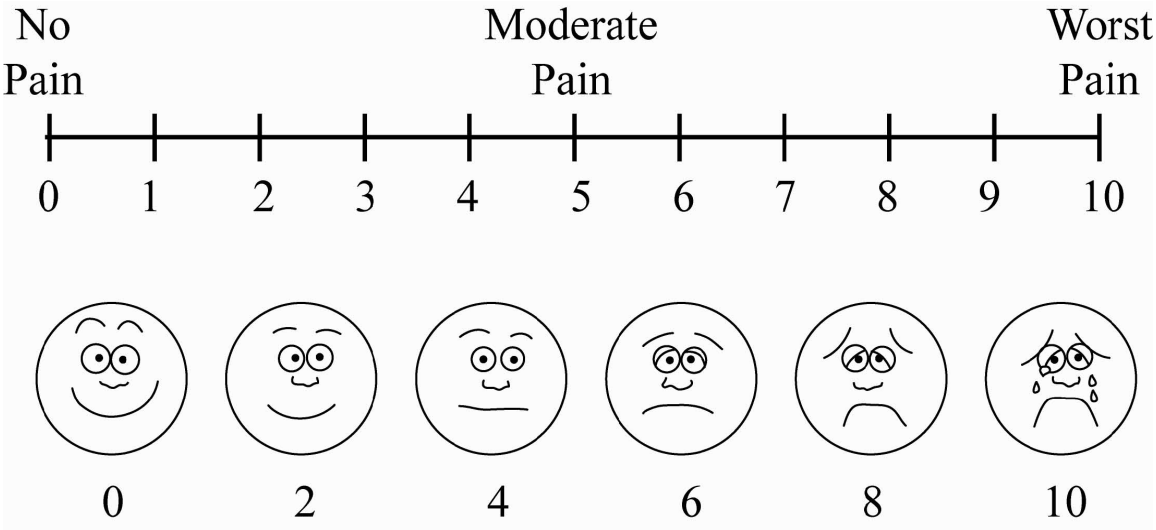
Abdominal pain consider girdle syndrome, hepatic or splenic sequestrations

Analgesia

Pain is the commonest cause of hospital admission and needs to be addressed urgently. **Analgesia should be administered within 30 minutes of arrival** and aim for pain control within 60 minutes.

Pain assessment should include the use of a validated age-appropriate **pain assessment score**.

In children, the Visual analogue score (below) is preferred:



Non-Steroidal Anti-inflammatory Agents (NSAIDs) and Paracetamol may have **synergistic** effects and should be prescribed regularly in addition to opiate analgesia.

Consider patient or nurse-controlled analgesia if repeated bolus doses of a strong opioid are needed within 2 hours used in accordance with locally agreed protocols.

De-escalation of parenteral opioids to regular oral opioids should be considered when the patient hasn't needed boluses for 10 -12 hours.

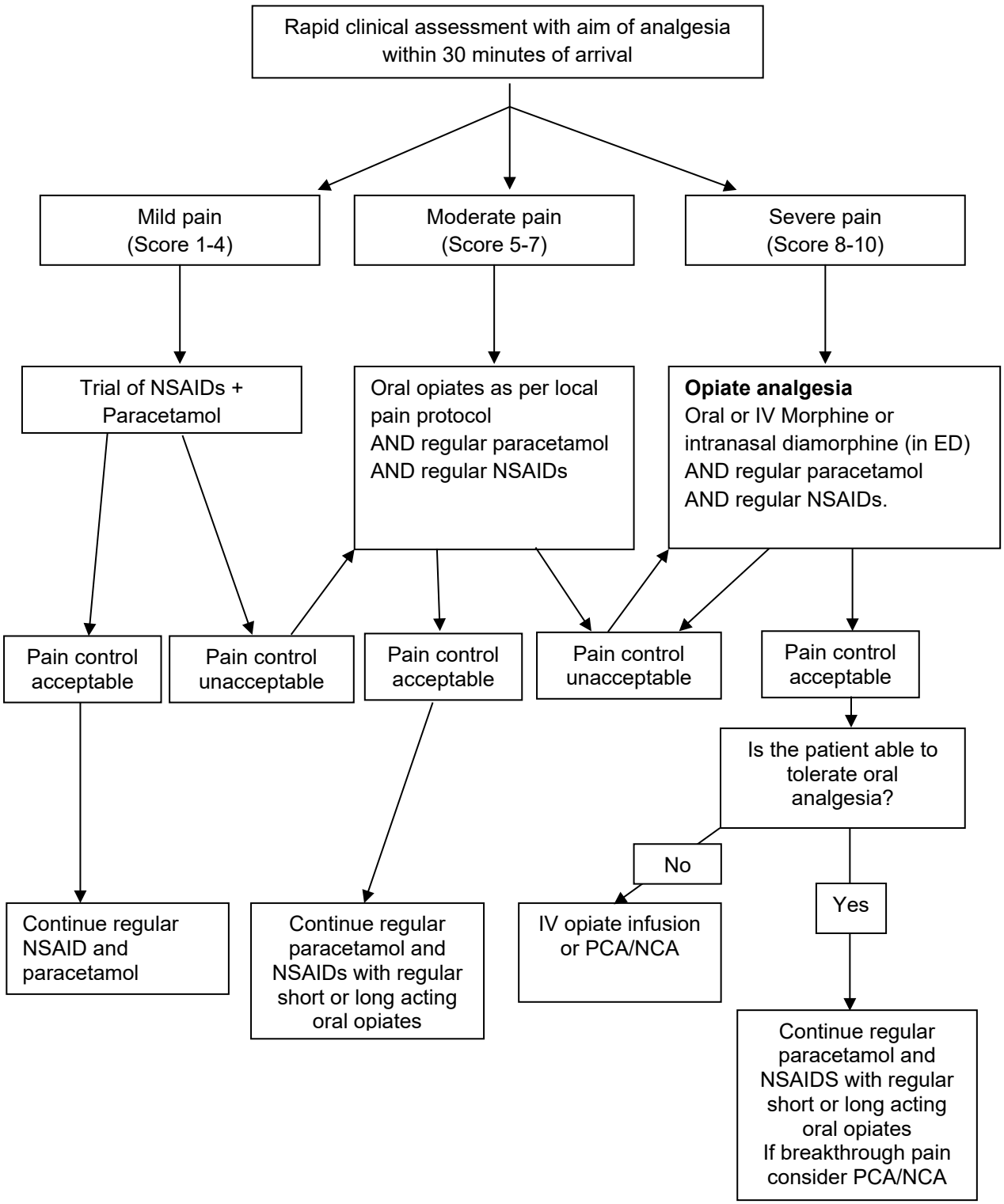
Adjunctive non pharmacologic approaches include heat pads, acupuncture, massage, and psychological techniques such as relaxation techniques, self-hypnosis, and distraction.

Monitoring of patients on analgesia

- Severity of pain (use validated pain score)
- Response to analgesia – using pain score, assessment of behaviour, body language, vital signs and questions
- Sedation level
- Pulse, BP, temperature
- Respiratory rate and oxygen saturation in air

These observations should be performed at least every 30 minutes until pain has been controlled and observations are stable, then according to local pain management protocols or at least every 2 hours whilst the patient is on opiate analgesia. If the respiratory rate falls below 10/minute then any opiate infusion should be discontinued and consider the use of naloxone.

FOR DOSES OF DRUGS PLEASE REFER TO LOCAL PAIN PROTOCOLS



OTHER SUPPORTIVE MANAGEMENT

Fluids:
Dehydration occurs readily in children with sickle cell disease due to impairment of renal concentrating ability and may aggravate to sickling due to increased blood viscosity. Diarrhoea and vomiting are thus of particular concern. Patients may also have cardiac or respiratory compromise and so fluid overload must also be avoided.

Careful assessment of individual fluid status, administration of an appropriate hydration regimen and close monitoring of fluid balance is therefore imperative.

An IV line should be established whenever parenteral opiates have been given, or if the patient is not taking oral fluids well. In the less ill patient who is able to drink the required amount, hydration can be given orally. As an alternative, consider a nasogastric tube in an alert patient.

Oxygen:
This is of doubtful use if the patient has only limb pain, but may be given if requested by the patient. The patient's oxygen saturation (SaO2) should be monitored by pulse oximetry with regular readings *on air* (minimum 4 hourly)

If SaO2 remains <95% in a child with normal baseline SaO2, prescribe oxygen and exclude emerging acute chest syndrome (ACS). Inform consultant if deteriorating respiratory condition.

Physiotherapy:
All children with acute chest or back pain and those requiring opiate analgesia should be referred to physiotherapy with a view to incentive spirometry or other measures as appropriate for age.

Thromboprophylaxis:
Consider LMWH prophylaxis in those 13yrs of age and above.

Other drugs
Consider the need for laxatives, antiemetics and antipuritics.

Antibiotics:
Infection is a common precipitating factor of painful or other types of sickle crises. These children are immunocompromised. Functional asplenia or hyposplenia occurs, irrespective of spleen size, resulting in an increased susceptibility to infection, in particular with capsulated organisms such as *pneumococcus Neisseria, Haemophilus influenzae* and *salmonella* – all of which can cause life-threatening sepsis.

In uncomplicated painful crisis without specific evidence of infection

- **increase prophylactic Phenoxymethylpenicillin (Penicillin V) to 4 times per day**
- **If penicillin allergic, use erythromycin 4 times per day.**

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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.