NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Paediatric guidance

Painful sickle cell crisis

NOTE
This guidance is not intended to be interpreted or used as a standard of care. Standards of care are determined based on the full range of clinical information available for an individual case and may change as scientific knowledge, technology, and clinical practice evolve. Following the recommendations in this guidance does not guarantee a successful outcome in every situation, nor does it encompass all appropriate methods of care or exclude other acceptable approaches that may achieve the same results. Clinical judgement remains the responsibility of the healthcare professional(s) involved in the patient's care. Clinicians must also be mindful of the requirements of patient autonomy and patient-centred care, which may involve a patient opting (after discussion of the available treatment options) for a treatment which is not the treatment recommended in the guidance. Where treatment involves deviation from national guidance, or local guidance derived from them, this should be fully documented in the patient's records at the time the decision is made.

This document has been prepared by NHS Public Services Delivery Scotland (PSD Scotland) on behalf of the Scottish Paediatric & Adult Haemoglobinopathies network (SPAH). Accountable to Scottish Government, PSD Scotland works at the heart of the health service providing national strategic services to the rest of NHS Scotland and other public sector organisations to help them deliver their services more efficiently and effectively. SPAH is a collaboration of stakeholders involved in the care of patients with a haemoglobinopathy, who are supported by an NSS Programme Team to drive improvement across the care pathway.

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Author	S Baird, Consultant Paediatric Haematologist NHS Lothian
Stakeholders involved	Consultant Paediatric Haematologists Consultant Paediatricians Haematology Nurse Specialists Consultant Paediatric Radiologist
Pharmacy Review	N/A
Methodology used	Evidence-based literature review and engagement with clinical experts within the field of Paediatric Haematology and review of UK based guidelines.
Rationale	The network was established to support and develop haemoglobinopathy services throughout Scotland in improving standards of clinical care for patients. The network supports the delivery of evidence based, patient centred care through the development and implementation of clinical guidelines, care pathways and information resources utilising a once for Scotland approach. Document first created in 2015. To aid healthcare professionals in the acute management of pain in patients with sickle cell disease.
Scope	Haematology specialists and general paediatricians including hospital staff managing children with sickle cell disease and Emergency medicine hospital staff in paediatric and adult hospitals.
Approval process	The guideline was approved by the SPAH Paediatric Guideline and Protocol subgroup on 10 March 2026. This subgroup has the authority to develop, review and endorse guidelines. This has been agreed by the SPAH Steering Group and noted in its Terms of Reference. See appendix for list of Paediatric Guideline and Protocol subgroup members.

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Background

Acute painful vaso-occlusive 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. Most uncomplicated 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.

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
- some patients may have an individualised care plan which should be used

General management includes:

- **reassurance** that the patient's pain will be relieved as soon as possible
- **warmth** and establishing a position of maximum comfort
- **prompt analgesia** including assessment of prior analgesia at home
- **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 clinical reassessment**

Consider complex presentations

- Fever greater than 38 – consider active infection
- Chest pain, respiratory signs or symptoms including SaO2 less than 95% - **Acute chest syndrome guidance**

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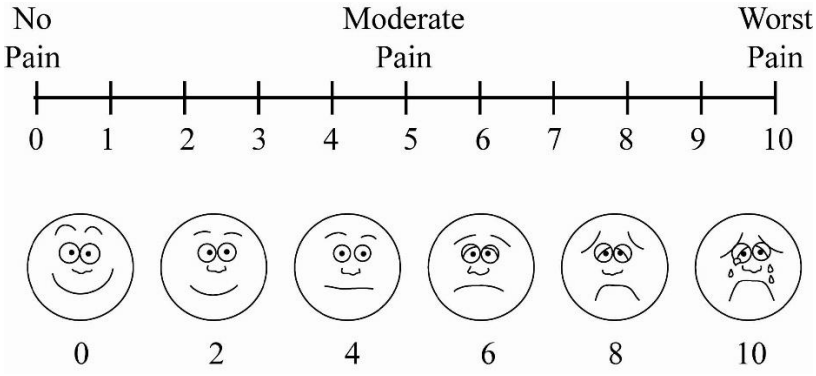
- Abdominal pain consider girdle syndrome, hepatic or splenic sequestrations – **Management of acute abdominal pain in children with Sickle Cell disease**
- Neurological symptoms or signs or severe headache – consider stroke **Stroke and other CNS complications**
- Priapism – persistent painful unwanted penile erection **Management of priapism in sickle cell disease**

Analgesia

Pain needs to be addressed urgently. **Analgesia should be administered within 30 minutes of arrival** and aim for pain control within 60 minutes. There is evidence that this reduces hospitalisation rates.

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

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



Some patients may have an individual care plan which should be followed if available.

Ibuprofen or other Non-Steroidal Anti-inflammatory Agents (NSAIDs) and Paracetamol may have synergistic effects and **should both be prescribed regularly** (unless contraindicated) in addition to any opiate analgesia that is needed.

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. Clonidine can also be used as an opioid sparing.

De-escalation of parenteral opioids to regular oral opioids should be considered when the patient hasn't needed boluses for 12 hours or frequency has reduced to less than 4 hourly.

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Adjunctive non pharmacologic approaches include heat pads, acupuncture, massage, and psychological techniques such as relaxation techniques, self-hypnosis, and distraction. (Cold packs should NOT be used).

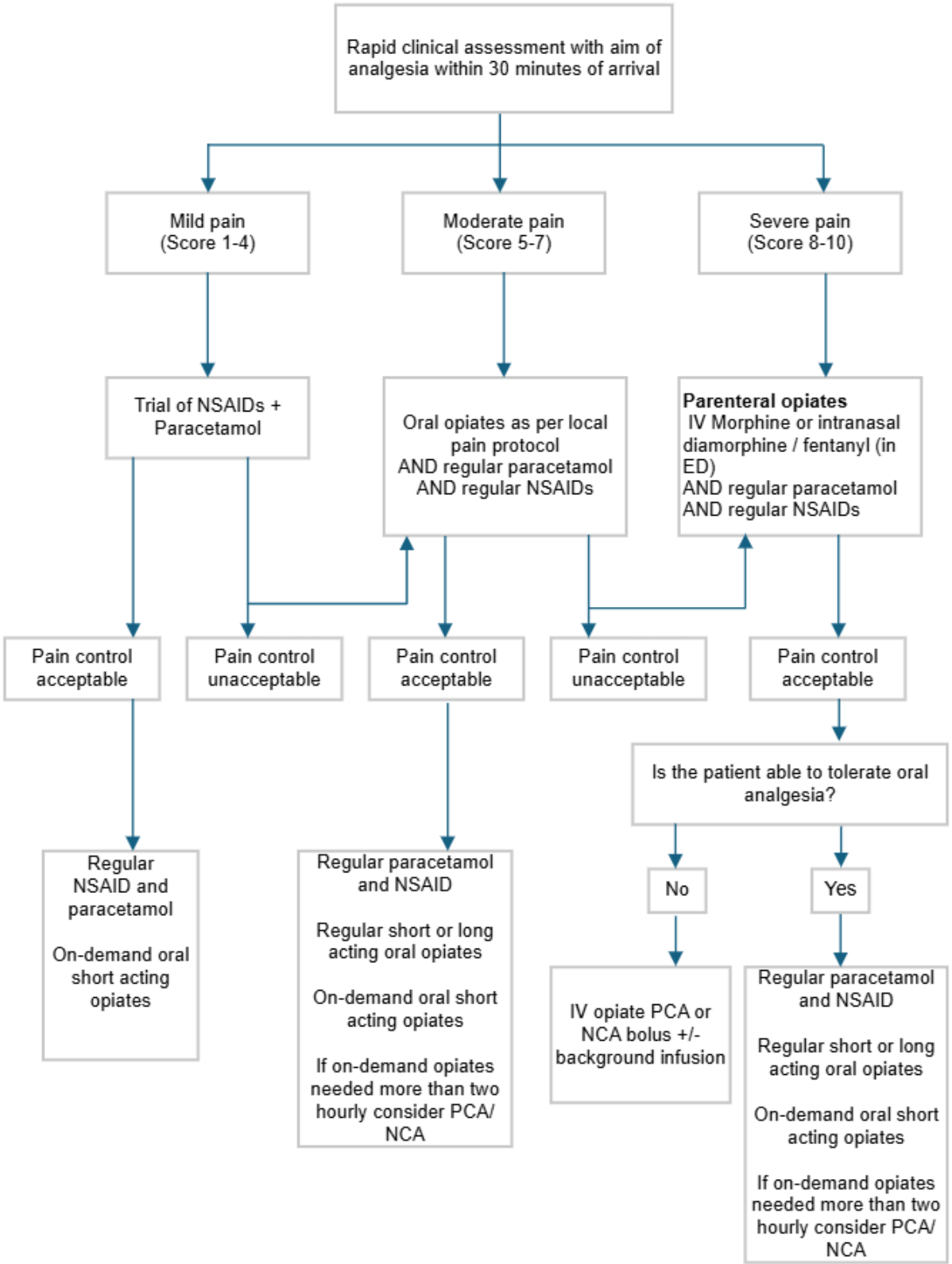
Monitoring of patients on analgesia

- 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.

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FOR DOSES OF DRUGS PLEASE REFER TO LOCAL PAIN PROTOCOLS



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Communication and escalation

Prompt discussion with the haematology or admitting team is indicated for severe pain, PEWS greater than=4 or in complex presentations as patients can deteriorate rapidly.

Other supportive management

Fluids:

Dehydration due to poor intake or increased fluid losses occurs more commonly in children with sickle cell disease due to impairment of renal concentrating ability and may aggravate red cell sickling due to increased blood viscosity. However, 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.

IV access 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 can drink the required amount, hydration can be given orally. (As an alternative, consider a nasogastric tube in an alert patient.

Oxygen:

There is no evidence of benefit in a uncomplicated painful episode but may be given at the lowest flow rate if requested by the patient. The patient's oxygen saturation (SaO_2) should be monitored by pulse oximetry with regular readings *on air* (minimum 4 hourly)

If SaO_2 is persistently less than 95% in a child with normal baseline SaO_2, prescribe oxygen and exclude emerging acute chest syndrome (ACS). Inform consultant if deteriorating respiratory condition.

Physiotherapy:

All children, particularly those with chest, back or abdominal pain and those requiring opiate analgesia should be referred to **physiotherapy** for incentive spirometry or other measures as appropriate for age.

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Thromboprophylaxis:

Consider LMWH prophylaxis in those 13yrs of age and above or have other risk factors.

Other drugs:

Consider the need for laxatives, antiemetics and antipruritic.

Antibiotics:

Infection is a common precipitating factor of painful or other types of sickle crises. These children are immunocompromised due to functional asplenia or hyposplenia and have an increased susceptibility to infection, with encapsulated bacteria such as *pneumococcus, Neisseria, Haemophilus influenzae*– 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.**

If there is evidence of infection with fever or local symptoms or signs of infection, then broad spectrum antibiotics as per local policy or antibiotics targeted to the likely source of infection should be prescribed.

References

RCEM Best Practice Guideline -Management of Acute Presentations of Sickle Cell Disease. (2024). Available at: https://rcem.ac.uk/wp-content/uploads/2024/03/RCEM_BPG_Sickle-Cell-Disease_v3.2.pdf.

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Appendix – Paediatric guidance subgroup membership

Name:		Job Title:	Organisation:
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Shahzya	Chaudhury	Consultant Paediatric Haematologist	NHS GG&C
Sarah	Clarke	Non-Malignant Paediatric Haematology Consultant	NHS GG&C
Emma	Cockburn	Consultant Paediatrician	NHS Tayside
Cheryl	Hill	Non-Malignant Haematology Nurse Specialist	NHS GG&C
Kirsten	Husselbee	Consultant Paediatrician	NHS Tayside
Jennifer	Milne	Paediatric Haematology Nurse	NHS Tayside
Lucy	Paterson	Clinical Nurse Specialist - Benign Haematology	NHS Lothian
Fernando	Pinto	Consultant Paediatric Haematologist	NHS GG&C
Rosina	Shujaat	Consultant Paediatric Haematologist	NHS Lothian
Gordon	Taylor	Consultant Paediatric Haematologist	NHS Grampian
Sandhya	Vaidyanathan	Consultant Paediatrician	NHS Grampian
Sasi	Yadavali	Specialty Doctor in paediatric Haematology	NHS GG&C

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