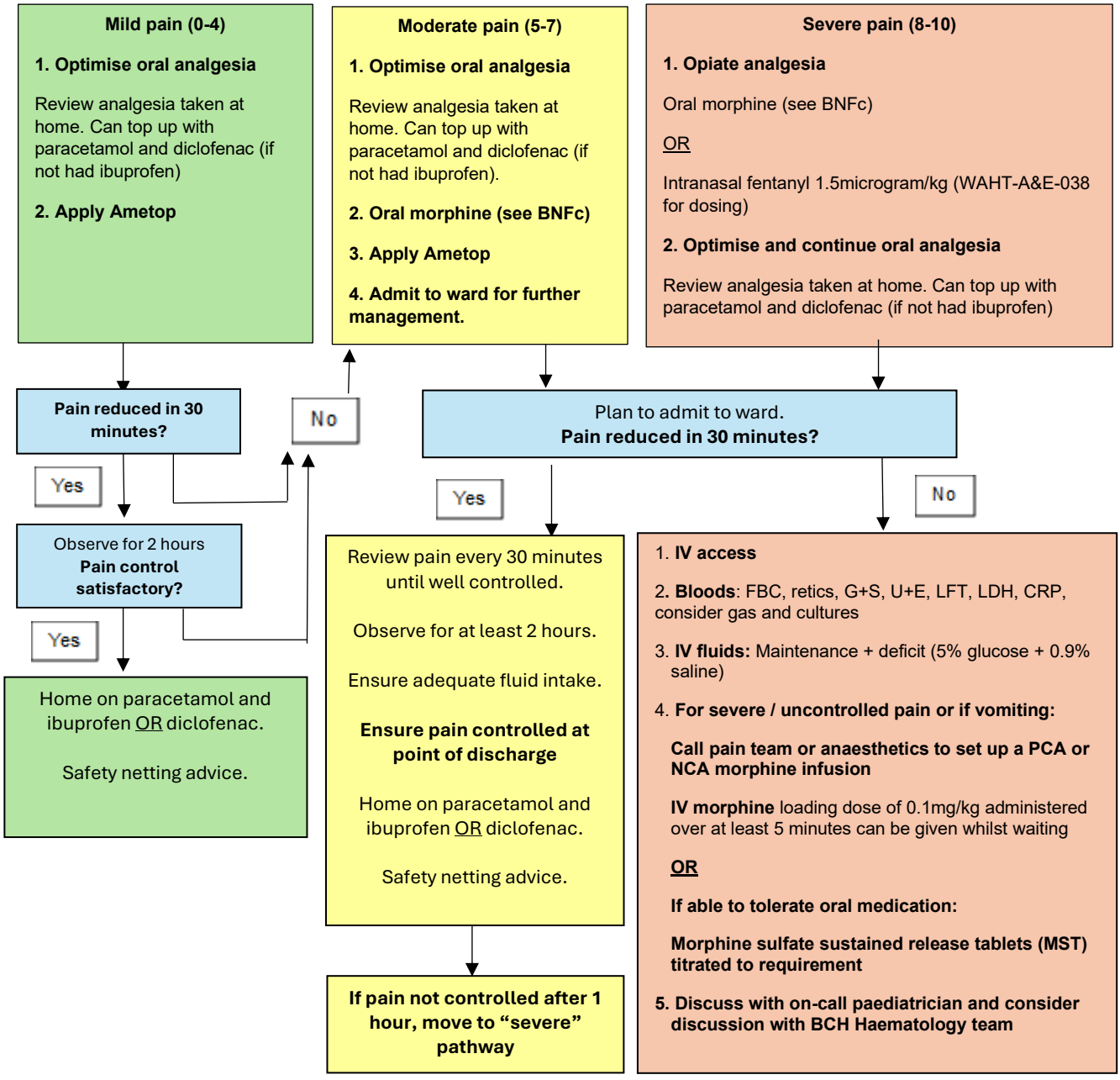
Flowchart for the initial triage and management of Sickle Cell Disease patients <18 years presenting to PAU / CED with a vaso-occlusive crisis

Triage / admitting nurse: If analgesia is required that you are unable to PGD, please hand over to nurse-in-charge to request prescription from a doctor. Analgesia should be given within 30 minutes of arrival.

All SCD patients seen in ED should be discussed with the paediatric consultant / registrar on call via bleep 676, even if you think they do not need admission.



If admitting the patient, please prescribe:

1. Naloxone PRN if on IV morphine or large doses of oral morphine
2. Lactulose or Movicol/Laxido PRN
3. Ondansetron PRN
4. Continue REGULAR paracetamol and diclofenac/ibuprofen
5. Consider omeprazole for gastric protection
6. For breakthrough pain:

   Oral morphine (as per BNFc) 4 hourly PRN
7. Write up patient's usual folic acid and prophylactic antibiotic (usually Penicillin V).
8. Hydroxycarbamide to continue unless neutrophils <1.0 or platelets <100

Further information provided in full Sickle Cell guideline available on the intranet

FURTHER TREATMENT POINTS

High risk patients are:

- Children under 2 years (Low threshold for admission in children under 5 years)
- Acute sequestration (splenic or hepatic)
- Abnormal neurology
- Severe anaemia (>20g/L below baseline)
- Respiratory disease requiring O2 / any chest or respiratory signs
- Acute priapism > 2 hours
- Pyrexia > 38C

Investigations in Patients with Sickle Cell Disease

1st line Investigations

- FBC
- Reticulocyte count
- Group & Save
- U&Es, LFTs, LDH

Additional investigations may be indicated

- CXR (if high fever, chest signs or abdominal signs)
- Venous / capillary blood gas
- AXR & amylase (if abdominal signs)
- Appropriate microbiological specimens (if pyrexial)
- Full respiratory viral panel
- Erythrovirus serology (in aplastic crisis)

Antibiotics in Patients with Sickle Cell Disease

- SCD children are at risk of sepsis due to functional hyposplenism
- Always look for a focus of infection (blood, lungs, bones, meninges, urinary tract etc) and treat appropriately.
- If no obvious focus, collect blood, urine and swabs for bacteriology before starting antibiotics as follows:
  - Any child with a high fever (>38° C), a sequestration syndrome, or who is clinically unwell should receive **IV co-amoxiclav** or **IV ceftriaxone** (refer to paediatric antimicrobial guideline)
  - All other patients should continue their prophylactic Penicillin V.
  - If in doubt, it is better to give antibiotics, particularly in children <5 years.
  - Chest infections should be treated with **co-amoxiclav + clarithromycin**, as atypical infections are often found in patients with SCD.
  - Ensure fever free for 24-48 hours prior to discharge

Management of Acute Splenic Sequestration

Liaise with on-call paediatrician and/or BCH Haematology team.

- Transfuse blood without delay (use unmatched O-neg in extremis)
- Do not infuse other fluids while awaiting blood as this will exacerbate heart failure.
- IV ceftriaxone or co-amoxiclav

Acute Splenic Sequestration

- Rapid sequestration of red cells lead to sudden anaemia and can result in death from hypoxic cardiac failure with pulmonary oedema
- Characterised by sudden onset of tachypnoea, pallor, abdominal pain and splenic enlargement
- May be precipitated by fever, dehydration, hypoxia
- Hepatic sequestration is less common than splenic sequestration in children but presents with a similar clinical picture and investigated and treated in the same way.

Acute Chest Syndrome

- Characterised by "T-shirt" distribution of pain and signs of lung consolidation – often bilateral and starting at the bases, with high fever, tachycardia, tachypnoea.
- Physical signs may precede CXR changes up to 12 hours. However, CXR changes can also precede signs.
- Falling Hb without evidence of splenic or hepatic sequestration is an indication for CXR.
- Consolidation in the upper or middle lobes without basal changes is suggestive of chest infection rather than acute chest syndrome.

Management of Acute Chest Syndrome

Discuss early with on call paediatric consultant, and on call BCH Haematologist. Most patients will need transfer to BCH / PICU for exchange transfusion.

- Transfusion if Hb falling or deteriorating clinical condition.
- Analgesia
- IV ceftriaxone + clarithromycin
- Oxygen
- IV fluids with careful fluid balance monitoring
- Aggressive physiotherapy

OTHER POTENTIALLY LIFE-THREATENING CONDITIONS

Pneumococcal & Haemophilus Septicaemia/ Meningitis

- Suspect in any sick SCD patient with fever
- Treat with broad spectrum IV antibiotics without waiting for culture results (IV co-amoxiclav or ceftriaxone)

Malaria

- Needs urgent anti-malarial therapy appropriate to the region of infection - IV artesunate (or artemether with lumefantrine if less severe) for falciparum, chloroquine if non-falciparum – discuss with paediatric infectious diseases team if unsure.

Cerebral Infarction

- Sickling within the cerebral vessels. Note that minor symptoms may precede a stroke.
- An indication for urgent exchange transfusion.
- Liaise immediately with BCH Haematology and KIDS for urgent transfer.

Aplastic Crisis

- Onset of profound anaemia over 1 – 3 days without sequestration.
- Due to transient erythrovovirus-induced marrow hypoplasia.
- If low reticulocyte count, transfuse
- Immunity appears to be lifelong

Priapism

- 2 main patterns:
  - Stuttering (repeated painful erections lasting < 4 hours)
  - Fulminant (lasting > 4 hours)
- Liaise urgently with BCH Urology

Management of Priapism

Discuss with BCH Haematology & Urology urgently regarding further management.

- Initially get patient to relax, urinate if possible (sometimes warm bath will help). If simple measures have failed then:
- IV fluids & catheterisation
- Analgesia

Further information

- Full Sickle Cell Guideline available via the intranet