PAEDIATRIC SICKLE CELL DISEASE INPATIENT MANAGEMENT

Version 3.0

Purpose:	To advise and inform hospital staff on the responsibilities in caring for paediatric sickle cell patients in the inpatient setting.
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This is a Controlled Document. Staff must refer to the Intranet version of this document to confirm the most up to date version of this guideline. If older versions are in circulation, they must be either returned to the author above or destroyed.

VERSION CONTROL SHEET

Version	Date	Author	Status	Comment
1	June 18	O Wilkey	Published	
2	July 19	O Wilkey	Published	
3	Dec 2020	O Wilkey/A Samanta	Published	Minor admin change

CHANGES & ALTERATIONS TO CURRENT GUIDELINES (For revised guidelines only)

Use this section to identify the changes you have made to the guideline.
Refer to specific paragraphs and page numbers.

Sickle cell in patient management : Page 12 – Changes to referral pathway for boys presenting with priapism, to be referred to UCLH if not resolved by 4 hours.

Clinical Guideline Approval Sheet

Clinical Guideline Title **Paediatric Sickle Cell Disease Inpatient Management**

Version of Clinical Guideline **3**

Speciality **Paediatrics**

Approved in departmental/divisional meeting

Paediatric Guideline Committee

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PAEDIATRIC SICKLE CELL DISEASE: INPATIENT MANAGEMENT

1. CLINICAL RESPONSIBILITY

All Paediatric Haematology patients are the joint responsibility of:

Paediatric Team: Dr Wilkey, Admitting Consultant, Paediatric Registrar for the wards, Paediatric SHOs, and Nurses.
All children are seen by a consultant Paediatrician within 12 hours and are reviewed daily by the haematology team.
Haematology Team: Dr Kreuk, Dr Marilyn Roberts-Harewood and Ward Haematology Registrars. Please contact Dr Wilkey or Dr Mensah regarding any queries.

Notes Please use CIPP for recent clinic letters and notes are on EDMS.

2. PAEDIATRIC PASSPORT

(Fast-Track Access to Children's Wards):
All children with sickle cell disease have pink Paediatric Passports and can be admitted straight to the ward after discussion with the nursing staff, or sent home for follow-up the next day by the Home Care Nurses. (Nurse on call uses structured questionnaire)**.**
Children brought by ambulance will be taken to A & E and should be seen by the paediatric medical team as a matter of urgency as they may have a life-threatening complication.
Admitting Specialist trainee must copy clinical details inside the Passport/Patient Held Records and incorporate into admission notes.

3. GENERAL PRINCIPLES

3.1 History

Remember that the parent knows the child better than we do.
How ill is the child? Is this incident different / worse than usual?
Remember that some patients get dangerously ill quickly e.g. Pneumococcal infection, Chest syndrome.
Check the problems list on the Passport /old notes. .

3.2 Examination

All children MUST have observation on admission and before discharge.
Especially chest, abdomen, and site of pain.
Compare size of liver and spleen with previous notes. (Measure with tape)
NB Most children with sickle are jaundiced, anaemic and have a cardiac flow murmur.
Examine for evidence of infection such as

- core temperature out of normal range;
- focal signs of infection;
- abnormal heart rate (raised), blood pressure (low or raised) or respiratory rate (raised);
- chills or rigors;
- raised or very low white blood cell count; and
- prolonged capillary refill time

3.3 Investigations

- In all cases a blood gas which will give you a haemoglobin before discharge , In most cases: **FBC**, reticulocytes, U&E, CRP, and group and save serum for cross match
- If recent blood transfusion: non-sickling Hb
- If fever is present or any other clinical evidence of infection, then collect Blood culture, MSU, Throat Swab, stool culture before starting empirical IV antibiotics
- If breathless or chest signs or low O2 saturation: CXR; consider blood gases (see chest syndrome)
- Save serum for serology (e.g. parvovirus, mycoplasma) - send if clinically indicated
- If suspected gallstones: direct bilirubin and amylase; check if abdominal U/S done yet.
- Also if clinically indicated e.g. other X-ray, LP, osmolality etc.

3.4 Diagnostic difficulties

Sickle cell disease is the great mimic, e.g.:

- Bone infarction or osteomyelitis?
- Abdominal crisis or appendicitis/cholecystitis NB: frequently constipated (due to infarctive crisis and opiate therapy)
- Pulmonary crisis or pneumonia?
- Cerebral infarction or meningitis?
- Sickle cell crisis is the more common cause of these, but if in doubt, investigate/treat for both as
- appropriate.

4. PAIN

Please read this section in conjunction with the pain chart, pain management flow chart and pain assessment chart (see appendices 1-4)

Never underestimate the pain experienced by patients. Many children by the time of presentation will already have been taking Paracetamol and NSAIDs and therefore may require opiate therapy.

5. Non-opioids

(If pain score1)

Paracetamol:

Child 1 month–6 years: 20–30mg/kg as a single dose then 15–20mg/kg every 4–6 hours; max. 75 mg/kg daily in divided doses

Child 6–18 years 20–30mg/kg (max. 1 g) as a single dose then 15–20mg/kg every 4–6 hours; max. 75 mg/kg (max. 4 g) daily in divided doses

And

Ibuprofen:

Child 1–3 months 5 mg/kg 3–4 times daily

Child 3–6 months 50 mg 3 times daily; max. 30 mg/kg daily in 3–4 divided doses

Child 6 months–1 year 50 mg 3–4 times daily; max. 30 mg/kg daily in 3–4 divided doses

Child 1–4 years 100 mg 3 times daily; max. 30 mg/kg daily in 3–4 divided doses

Child 4–7 years 150 mg 3 times daily; max. 30 mg/kg daily in 3–4 divided doses

Child 7–10 years 200 mg 3 times daily; max. 30 mg/kg (max. 2.4 g) daily in 3–4 divided doses

Child 10–12 years 300 mg 3 times daily; max. 30 mg/kg (max. 2.4 g) daily in 3–4 divided doses

Child 12–18 years initially 300–400 mg 3–4 times daily; increased if necessary to max. 600 mg 4 times daily; maintenance dose of 200–400 mg 3 times daily may be adequate

Or

Naproxen:

Child 6 months – 18 years: 5mg/kg (max. 500mg) every 12 hours

NB: No licensed liquid is available, round doses to 250mg or 500mg where possible.

5.1 Non pharmacological intervention

- Reassurance
- Warmth
- Massage
- Distraction techniques

5.2 Opioids

NB - stat doses must be given as soon as possible <u>if pain score 5-7</u>

Dihydrocodeine (weak opioid):

Child 1–4 years 500 micrograms/kg every 4–6 hours

Child 4–12 years 0.5–1 mg/kg (max. 30 mg) every 4–6 hours

Child 12–18 years 30 mg every 4–6 hours

Paracetamol and Ibuprofen should be used in addition to morphine but stop Dihydrocodeine, then Oramorph can be given after 3- 4 hours

Morphine (moderately strong opioid):
Loading dose
Morphine sulphate oral solution 10mg/5ml (Oramorph)
Child 1–3 months initially 50–100 micrograms/kg every 4 hours, adjusted according to response
Child 3–6 months 100–150 micrograms/kg every 4 hours, adjusted according to response
Child 6–12 months 200 micrograms/kg every 4 hours, adjusted according to response
Child 1–2 years initially 200–300 micrograms/kg every 4 hours, adjusted according to response
Child 2–12 years initially 200–300 micrograms/kg (max. 10 mg) every 4 hours, adjusted according to response
Child 12–18 years initially 5–10 mg every 4 hours, adjusted according to response
If more than 3 doses of Oramorph required start:

Continuation doses
Morphine slow release 1mg/kg (rounded up to nearest 5mg) 12 hourly, (maximum 70mg per dose)

PLUS
Breakthrough pain, especially likely to be needed in first 24 hours
Oramorph as outlined above every 4-6 hours as necessary

Review at 24 hours
If more than 3 doses of Oramorph/day still needed, may increase MST to 1.5mg/kg 12hourly (maximum 70mg per dose).

Review at 48 hours
If more than 3 doses of Oramorph/day were needed, may increase MST to 1.75mg/kg 12hourly (maximum 70mg per dose) or change to IV opiates.

Discontinuation of MST MR:

MST MR should be stopped when patients is only requiring less than 2 doses of Oromorph daily If patient is on 1mg/kg BD, then the morphine can be stopped without weaning. However if patient is on a dose >1mg/kg BD, the dose should be reduced on a daily basis by 0.5mg/kg until 1mg/kg is reached after which morphine can be stopped safely.

IV Morphine doses at 0.1-0.2mg/kg 4 – 6 hourly (see table below for guide), or commence PCA contact pain team

Examples of dose ranges:

		Stat dose		Breakthrough
Age	**Weight/kg**	Oramorph oral 4 – 6 hourly	Morphine IV 4 – 6 hourly	
3-6m	5	500-750mcg	500 - 750mcg	500-750mcg
6-12m	10	2mg	1mg	2mg
	15	3mg	1.5mg	3mg
1-2 y	20	4 – 6mg	2mg	4 – 6mg
3-12 y	25	5 – 7mg	2.5mg	5 – 7mg
	30	5 – 9 mg	3mg	5 – 9 mg
	35	5 – 10mg	3.5mg	5 – 10mg
	40	5 – 10 mg	4mg	5 – 10 mg
13-18 y	50	5 – 10mg	5mg	5 – 10mg

NB: All children receiving opiates will become **constipated**.

Prescribe **Paediatric Movicol**
Under-1 year: HALF sachet to ONE daily, adjusted according to response
1– 6 years: ONE sachet daily, adjusted according to response 6–12 years: TWO sachet daily, adjusted according to response

Prescribed **Movicol (Adult)**
>12 years: ONE to TWO sachets daily, adjusted according to response

OR

Prescribe **Lactulose** if Movicol not tolerated
1 month–1 year 2.5mL twice daily, adjusted according to response
1–5 years 2.5–10mL twice daily, adjusted according to response
5–18 years 5–20mL twice daily, adjusted according to response

Especially if on oral opiate, prescribe **antiemetic** e.g. **Cyclizine** IV/PO:
1 month–6 years 0.5–1 mg/kg up to 3 times daily; max. Single dose 25 mg
6–12 years 25 mg up to 3 times daily
12–18 years 50 mg up to 3 times daily

Some patients **itch** with IV morphine and may need antihistamines:

Chlorpheniramine oral:
1 month–2 years 1 mg twice daily
2–6 years 1 mg every 4–6 hours, max. 6 mg daily
6–12 years 2 mg every 4–6 hours, max. 12 mg daily
12–18 years 4 mg every 4–6 hours, max. 24 mg daily

Or
Hydoxyzine oral:
Child 6 months–6 years initially 5–15 mg at night, increased if necessary to 50 mg daily in 3–4 divided doses

Child 6–18 years
Body-weight under 40kg: initially 15–25 mg at night, increased if necessary maximum dose of 2mg/kg daily in divided doses
Body weight over 40kg: initially 15-25 mg at night, increased if necessary to 50-100 mg daily in 3–4 divided doses

Prescribe **Naloxone** on PRN section whenever an opioid is prescribed:

By intravenous injection ***(****doses can be given by SC or IM routes but only if IV route is not feasible; IV administration has more rapid onset of action.)*

1 month–12 years 100 micrograms/kg (max. 2 mg); if no response, repeat at intervals of 1 minute to a total max. 2 mg, then review diagnosis; further doses may be required if respiratory function deteriorates

12–18 years 400 micrograms; if no response after 1 minute, give 800 micrograms, and if still no response after another 1 minute, repeat dose of 800 micrograms; if still no response, give 2 mg (4 mg may be required in a seriously poisoned patient), then review diagnosis; further doses may be required if respiratory function deteriorates

During crises change prophylactic Pencillin-V to the treatment dose unless temperature is greater than 38.0°C then IV antibiotics should be started immediately (See infections, section 6.0)

5.3 Intravenous analgesia

Consider IV analgesia if oral therapy is refused (IV rarely needed) or vomiting.

Morphine sulphate injection over 5 minutes

Child 1–6 months initially 100 micrograms/kg every 6 hours, adjusted according to response
Child 6 months–12 years initially 100 micrograms/kg every 4 hours, adjusted according to response
Child 12–18 years initially 5 mg every 4 hours, adjusted according to response

Reduce dose if respiratory depression. Reassess after 1 hour if pain still not controlled prescribe a second dose stat. Then continue as doses and frequencies described above.

If Patient Controlled Analgesia (for over 7 years of age) or Nurse Controlled Analgesia (for less than 7 years of age) is required, contact the pain team.

6. FLUID REPLACEMENT

If vomiting or unable to drink required daily amount, give IV as 5% Dextrose /0.9% sodium chloride. Add 10mmols of potassium chloride (KCl) if passing urine. Pre-mixed bags of both fluids are stocked in children's A&E and Rainbow ward.

Rationale: Many patients with sickle cell disorders have reduced tubular concentrating ability. Continued fluid loss without adequate replacement causes a reduction in plasma volume with an increased blood viscosity and aggravation of sickling.

Minimum Requirements over 24 hours:
100 ml/Kg for the first 10 Kg body weight
50 ml/Kg for the next 10kg body weight
20ml/kg thereafter

Example:
A child weighs 26 Kg:

100 ml X 10Kg=1000 ml
50 ml X 10Kg= 500 ml
20 ml X 6Kg= 120 ml

Minimum requirement: 1620 ml

Fluids may be given orally or intravenously. Use standard (5% dextrose + 0.9% sodium chloride) re-hydration solution. Review the need for potassium.

NB:

- Children with sickle cell disease need individualized fluid regimes. They are often dry and will need additional fluids; conversely over zealous fluid replacement may make the situation worse by precipitating cardiac failure.
- The oral route should be used whenever possible but children with severe pain who are not settling, or who have abdominal symptoms should receive intravenous re-hydration.
- Intravenous therapy should be stopped once the patient is stable and pain is controlled.
- Adequate oral intake should be documented

Monitoring

Plasma sodium, potassium, urea and/or creatinine should be measured at baseline and at least once a day. Consider measuring every 4-6 hours if an abnormal reading is found. This should definitely be done if the plasma

sodium is below 130mmol/L. Check plasma electrolytes immediately if clinical features suggest hyponatraemia is developing. Symptoms include increased headaches, vomiting, nausea, irritability, and altered levels of consciousness, seizures and apnoea.
Ensure patient is weighed prior to fluid therapy and daily thereafter losses continues. On-going losses and oral intake should be recorded using a fluid balance chart and fluid requirements assessed every four hours.
BEWARE inappropriate Anti diuretic hormone (ADH) secretion - monitor blood U&E and urine output and weight daily

7. FEVER AND INFECTION

Pneumococcus is the most dangerous but the patients are also susceptible to infections caused by other organisms such as Haemophilus Influenza, Meningococcus, Staphylococcus and Salmonella

BEWARE: Children should have had Prevenar and Pneumovax (also HIB) and be on prophylactic Penicillin BUT

- There may be poor adherence
- Prophylaxis does not eliminate all infection with organisms such as Pneumococcus. Resistant organisms can appear
- Remember: Children brought straight to A & E could have pneumococcal or salmonella septicaemia if very unwell
- Gram-negative bacteria is also important
- Always look for the focus of infection (blood, lungs, urine, and stool) and then treat appropriately
- Collect blood cultures, urine, and throat swabs, mycoplasma titres before starting antibiotics [but if child unwell and cannot pass urine – do not wait for specimen. start antibiotics anyway].
- If children present in crises with fever temperature ≥ 38, and are relatively well (monitor for evidence of sepsis such as temperature, WBC, CRP, and clinical signs of sepsis before considering the possibility of viral infection) mixed viral and bacterial infection may occur. Collect microbiological investigation, start empirical antibiotic therapy and review microbiological investigation with 48h.

Ceftriaxone IV
Child 1 month–12 years
Body-weight under 50 kg 50 mg/kg once daily; up to 80 mg/kg daily in severe infections and meningitis; doses of 50 mg/kg and over by intravenous infusion only
Body-weight 50 kg and over dose as for child 12–18 years
Child 12–18 years 1 g daily; 2–4 g daily in severe infections and meningitis; intramuscular doses over 1 g divided between more than one site; single intravenous doses above 1 g by intravenous infusion only.

Given by intravenous injection over 2–4 minutes (up to doses of 50mg/kg), or by intravenous infusion over at least 30 minutes (doses greater than 50mg/kg), or by deep intramuscular injection (see BNFC or page 11).

- If severe penicillin allergy, consider combination of teicoplanin + clarithromycin /azithromycin + ciprofloxacin OR contact consultant microbiologist.
- **If diarrhoea present, (see section for salmonella infection)**
- Reassess at 48 hours; if child well and if blood, stool and urine cultures are negative, consider stopping IV and change to oral **Augmentin**
- **If clinical evidence of severe sepsis or if failed to respond to ceftriaxone then start empirical meropenem + teicoplanin and discuss with consultant microbiologist**

Add oral **Clarithromycin** if suspect mycoplasma, -chest infection or chest syndrome.
Oral Clarithromycin

Child 1 month–12 years
Body-weight under 8 kg 7.5 mg/kg twice daily
Body-weight 8–11 kg 62.5 mg twice daily
Body-weight 12–19 kg 125 mg twice daily
Body-weight 20–29 kg 187.5 mg twice daily
Body-weight 30–40 kg 250 mg twice daily
Child 12–18 years 250 mg twice daily, increased if necessary in severe infections to 500 mg twice daily; usual duration 7–14 days

If chest signs are present, complete course of 7 days total.
If Mycoplasma positive, complete 10 days total.

8. PRIAPISM

Many patients are not aware this is a complication of sickle cell disease or are reluctant or embarrassed to discuss it. Parents and children should be informed about this and enquiries made as outpatients.

Stuttering priapism - symptoms lasting for more than 30 minutes and up to 4 hours.

Fulminant priapism- Duration lasting > 4hours need urgent intervention.

This is an emergency. If not completely resolved **promptly** there is risk of permanent loss of erectile function.

Discuss urgently with Consultant Paediatrician and Haematologist.

Treatment
Analgesia- you may have to give a stat dose of IV/IM Morphine.
Hydration
General measures i.e. attempt to urinate

Etilefrine (Not licensed in children and adolescents, doses are from Evelina formulary, 2015 edition)
http://cms.ubqo.com/public/d2595446-ce3c-47ff-9dcc-63167d9f4b80

Liquid:
< 2 years: 1mg - 2.5mg 3 times a day PO
2 - 6 years: 2.5mg - 5mg 3 times a day PO
> 6 years: 5mg - 10mg 3 times a day PO

Tablets:
All ages: 0.5mg/kg once daily or 0.25mg/kg twice a day
Doses up to 50mg daily have been used in patients > 6 years

- Etilefrine should be used with caution in patients with diabetes mellitus, hypercalcaemia, hypokalaemia, severe impairment of renal function and cor pulmonale.
- Etilefrine may induce palpitations, restlessness, sleeplessness, diaphoresis, dizziness and gastrointestinal symptoms. Symptoms of angina, tachycardia, ventricular arrhythmias and hypertensive episodes associated with headaches and tremor may occur.
- 1ml of Etilefrine oral solution 7.5mg in 1ml contains 30% alcohol.
- Etilefrine should be taken with a small amount of liquid before a meal.
- Etilefrine should not be taken late in the afternoon or in the early evening (e.g. 4pm) as its stimulating effect may cause difficulties with sleeping.

Or

Ephedrine (Not licensed in children)
2-18 years: 15mg -30mg daily PO

If priapism is still present, immediate cross-match for possible exchange
1. Proceed to immediate exchange transfusion and continue exchanging until improvement or non-sickling Hb >70%
2. Discuss urgently with Urology registrar at UCL and Paediatric team, if not completely resolved by 4 hours and agree whether when the boy might need to be transferred as treatment may include aspiration/intracavernous administration of Etilefrine at tertiary centre.

9. BONE and JOINT PAIN

Usually due to ischemia/infarction. X-rays are usually not necessary
However, patients with sickle cell disease can develop osteomyelitis and it can be difficult to differentiate
Please do not aspirate joint

If child is febrile, start IV **Ceftriaxone** (contact consultant microbiologist if allergic to Penicillin)

Child 1 month–12 years
Body-weight under 50 kg 50 mg/kg once daily; up to 80 mg/kg daily in severe infections and meningitis; doses of 50 mg/kg and over by intravenous infusion only
Body-weight 50 kg and over dose as for child 12–18 years

Child 12–18 years 1 g daily; 2–4 g daily in severe infections and meningitis; intramuscular doses over 1 g divided between more than one site; single intravenous doses above 1 g by intravenous infusion only

Given by intravenous injection over 2–4 minutes (up to doses of 50mg/kg), or by intravenous infusion over at least 30 minutes (doses greater than 50mg/kg), or by deep intramuscular injection (see below and BNFC)

For IM administration of Ceftriaxone:

- A 1g ceftriaxone vial should be dissolved in 3.5ml of 1% Lidocaine Injection BP. The solution should be administered by deep intramuscular injection into a relatively large muscle. Dosages greater than 1g should be divided and injected at more than one site.
- As the solvent used is lidocaine, the resulting solution should NEVER be administered intravenously.

If child is febrile with swinging temperatures and fails to respond to antibiotics after 48 -72hr:
Repeat blood cultures, ESR, CRP, and consider CT or MRI; X-ray changes of osteomyelitis may take 10-14 days to become apparent.

Review and switch to oral antibiotics as soon as possible or discontinue/change over according to sensitivities where appropriate. Referrals for Osteomyelitis are to at Great Ormond Street. (See surgical pathway

For Avascular necrosis of the hip these patients should be referred to Orthopaedic surgeon at RNOH /UCH

10. EYE PROBLEMS AND ACUTE VISUAL DISTURBANCES

Sickle retinopathy -changes in the retina due to vascular damage caused by SCD, which are grouped into non-proliferative and proliferative. Infarction of the peripheral retina results in the proliferation of fragile, thin-walled blood vessels 'sea fans' at high risk of bleeding, neovascularisation. The normal age of onset is adolescence and after.

Proliferative Sickle Retinopathy: Staging criteria

Stage 1:	Peripheral arteriolar occlusions
Stage 2:	Peripheral arteriolar-venular anatomises
Stage 3:	Neovascular and fibrous proliferations
Stage 4:	Vitreous haemorrhage
Stage 5:	Retinal detachment

An initial ophthalmology review should be performed at age 16.
For those patients with retinopathy or those on regular desferrioxamine, annual review is recommended

1. Vitreous Haemorrhage and Retinal Detachment

More common in SC and S/BetaThalassaemia, (especially in pregnancy). Surgical treatment should not be undertaken without prior exchange transfusion.

2. Hyphema

Usually due to blunt trauma. It may lead to raised intra-ocular pressure and thus to retinal vessel occlusion with blindness. Refer to Eye Clinic IMMEDIATELY. If surgery is required, exchange transfusion . (See surgical pathway

For all acute visual disturbances refer urgently to the ophthalmology or eye casualty. All Haemoglobinopathy with eye complications must be discussed with the paediatric and haematology team .

11. RENAL COMPLICATIONS

Haematuria -
Microscopic is not uncommon in sickle cell disease
Frank haematuria is usually due to renal papillary necrosis, infection, rarely stone.

Investigations
MSU for MC&S;
Renal and bladder USS

Urinary tract infections – are common.
This should be treated promptly like any other child with a UTI (See UTI protocol)

Enuresis
Common due to hyposthenuria , consumption of large fluid intake and possible UTI
Investigations
Urine for MC&S
Star charts
If still not improving refer to local enuresis clinic for bed or body alarms
May need DDAVP for a short time

12. POTENTIALLY LIFE THREATENING CONDITIONS

12.1 Chest Syndrome

Multifactorial. Including sequestration in the Lungs
Often associated with mycoplasma infection or precipitated by chest infection
Progressive pulmonary infiltration, which is potentially fatal
Look out for breathlessness, crackles on auscultation, and falling saturations in air
Abnormal CXR especially if progressive, Falling Hb
It may be difficult to differentiate between sickle chest and a chest infection, and both may be present.

MANAGE URGENTLY
Inform Consultant Haematologist and Paediatrician at an early stage, if pO2 [oximeter] is 5% or more below the steady state level for that child [see clinic notes].
Check old notes to assess severity of previous crises.
Xmatch blood in case needed
Give humidified oxygen by mask or nasal prongs

If pneumonia or chest infection, consider the possibility of viral infection such as influenza as well as bacterial infections.
IV Ceftriaxone (Contact consultant microbiologist if allergic to Penicillin)

1 month–12 years
Body-weight under 50 kg 50 mg/kg once daily; up to 80 mg/kg daily in severe infections and meningitis; doses of 50 mg/kg and over by intravenous infusion only
Body-weight 50 kg and over dose as for child 12–18 years

12–18 years 1 g daily; 2–4 g daily in severe infections and meningitis; intramuscular doses over 1 g divided between more than one site; single intravenous doses above 1 g by intravenous infusion only

Give by intravenous injection over 2–4 minutes (up to doses of 50mg/kg), or by intravenous infusion over at least 30 minutes (doses greater than 50mg/kg), or by deep intramuscular injection (see BNFC or page 11)

Add clarithromycin for mycoplasma :

Oral Clarithromycin
Child 1 month–12 years
Body-weight under 8 kg 7.5 mg/kg twice daily
Body-weight 8–11 kg 62.5 mg twice daily
Body-weight 12–19 kg 125 mg twice daily
Body-weight 20–29 kg 187.5 mg twice daily
Body-weight 30–40 kg 250 mg twice daily
Child 12–18 years 250 mg twice daily, increased if necessary in severe infections to 500 mg twice daily; usual duration 7–14 days

- Monitor by pulse oximeter etc
- Blood gases if saturations less than apparent clinical severity
- IV fluids
- Chest physiotherapy by incentive spirometry
- Consider nebulised bronchodilators
- Re-examine chest clinically at least three times in 24 hours and re-X-ray if signs increasing

Plan exchange transfusion (see separate protocol) if: **increasing tachypnoea or chest signs and failure to improve**
and/or falling saturations or saturations <90% in air
and/or pO2 < 8KpA)
and/or deteriorating CO2

Consider transfer to ITU, contact anaesthetist , inform Consultant Paediatrician and haematology team .

12.2 Stroke/Neurological events

May present with fits, hemiplegia or severe headache. Differential diagnosis = meningitis
Discuss urgently with Consultant Paediatrician and Haematologist. May need urgent CT and LP.
Managed by urgent exchange transfusion [without waiting for imaging, if there is any delay], possible transfer to ITU.
Will need MRI, MRA, Transcranial Dopplers, Sleep study to be arranged at GOS.

The child will need to start a regular transfusion programme, usually 3-4 weekly exchange or top-up transfusions depending on the pre-transfusion Hb and HbA%, aiming to maintain HbA > 70% and Hb not > 120 g/L.

Fits can occur after strokes
Treat with anticonvulsant for immediate management. Refer for EEG at GOS as will be seen by Neurology team there. Inform Consultant Paediatrician and Haematologist . Will need further imaging to be arranged

12.3 Severe Anaemia

1.1.1 Splenic (or hepatic) sequestration

Suspect if Hb very low and spleen (or liver) larger than previously documented.
Hb usually drops by more than 20g/L from steady state
Often associated with infection, including parvovirus, making diagnosis difficult

Investigations
Hb, Retics, crossmatch, blood cultures, parvovirus and other viral serology,
May need urgent blood transfusion to raise Hb to 80-100g/L, if Hb dropped more than 20g/dl from steady state
Consider giving antibiotics **Ceftriaxone IV** *(contact consultant microbiologist if allergic to Penicillin)*

Child 1 month–12 years
Body-weight under 50 kg 50 mg/kg once daily; up to 80 mg/kg daily in severe infections and meningitis; doses of 50 mg/kg and over by intravenous infusion only
Body-weight 50 kg and over dose as for child 12–18 years

Child 12–18 years 1 g daily; 2–4 g daily in severe infections and meningitis; intramuscular doses over 1 g divided between more than one site; single intravenous doses above 1 g by intravenous infusion only

Given by intravenous injection over 2–4 minutes (up to doses of 50mg/kg), or by intravenous infusion over at least 30 minutes (doses greater than 50mg/kg), or by deep intramuscular injection (see BNFC or page 11).

1.1.2 Parvovirus aplasia

Suspect if Hb very low and reticulocyte count not raised or inappropriately low without Splenic or Hepatic enlargement. Hb usually drops by more than 20g/L from steady state

Investigations
HB, Retics, crossmatch, Parvovirus serology
May need urgent blood transfusion to raise Hb to 80-100g/L if Hb drops by more than 20g/dl from steady state

Remember cross-infection; should be isolated and other family members with sickle cell should also be checked

12.4 Abdominal pain and girdle syndrome

Abdominal pain -This may be due to mesenteric vasocclusion, hepatic or splenic sequestration, any cause of acute abdomen, gallstones and/or constipation. With fever and increased jaundice consider cholecyctitis.
Biliary colic with increased jaundice may be due to gall stones
Girdle syndrome due to mesenteric sickling may present with vomiting and abdominal distension

Management
IV fluids (as above) and analgesia (as above)
N-G tube on free drainage if continuous vomiting and abdominal distension
Measure abdominal girth (around the umbilicus)
Abdominal X-ray if bowel sounds absent- ask for surgical opinion.
Abdominal x-ray often shows dilated bowel loops

If fever and jaundice worsening consider- acute cholecystitis and / or biliary obstruction will need to add Metronidazole to antibiotics. Request urgent abdominal USS to determine whether or not there are gallstones +/- dilated common bile duct. If biliary obstruction is found, refer Great Ormond Street Hospital for ERCP. (See surgical pathway)

12.5 Malaria (see NMUH Malaria protocol)

12.6 Salmonella

Salmonella can cause potentially life- or limb-threatening infections in children with sickle cell disease, including septicaemia, multifocal osteomyelitis, liver failure or multi-organ failure. If Salmonella is strongly suspected from the clinical history, Start IV ceftriaxone empirically 50-80mg/kg once daily(maximum 4gm) then discuss appropriate antibiotic therapy with the microbiologist and Consultant Paediatrician. If Salmonella species is isolated from any child, they require IV antibiotic therapy irrespective of whether they are a febrile or well. The use of oral antibiotics should be discussed with the microbiologist and Consultant Paediatrician. On discharge, the patient will require early clinical review on Day assessment unit and blood test monitoring to ensure their adequate response to therapy.

PLEASE NOTE :
ALL CHILDREN ADMITTED OR SEEN IN PAU SHOULD HAVE A BASELINE FBC OR BLOOD GAS TO CHECK HAEMOGLOBIN LEVEL

APPENDIX 1A

PAEDIATRIC PAIN ASSESSMENT TOOL CHART

- Please document which Pain Assessment Tool(s) are being used.
- If Sickle Patient, Remember to use **THE SICKLE CELL CRISIS IN CHILDREN PAIN ASSESSMENT AND INITIAL ANALGESIA CHART,** together with the Tool Chart and the Interventions/Evaluations Chart. (Score pain on admission, then at 30 mins and 1 hour)
- Document all treatment methods, management and interventions given/used.
- Pain Team contact details; #6439 or 0208 887 2629

FACES SELF REPORT TOOL

Suggested age group 4 years and over. (Adapted from Wong and Baker, 1988)

FLACC BEHAVIOURAL TOOL

Suggested age group 2months to 7 years, any child with a profound learning disability (adapted from Merkel et al, 1997)

categories	Score 0	Score 1	Score 2
FACE	No expression or smile.	Occasional grimace / frown, disinterested.	Frequent to constant quivering chin, clenched
LEGS	Normal or relaxed	Uneasy/restless/tense	Kicking/or drawn up
ACTIVITY	Quiet/moves easily	Squirming/shifting	Arched/rigid/jerking
CRY	No cry/settled	Moans or whimpers	Crying/sobs or screams
Consolability	Content/relaxed	Reassured/distractible	Hard to console/comfort
Each of the 5 categories; are scored from 0 – 2 which results in a total score of 0 - 10			

VAS SELF REPORT NUMBER LINE

Suggested for use with older children (Adapted from Wong and Baker, 1988)

0	1	2	3	4	5	6	7	8	9	10
NO HURT		HURTS A BIT		HURTS MORE		HURTS A BIT MORE		HURTS LOTS		HURTS +++

1. Assess pain severity and chart overleaf Pain Scale used: FACES/ FLACC/VAS

Site(s) of pain: _____ Onset of pain: _____

2. Documents analgesia administered at home

Drug	Date & Time	Drug	Date & Time

3. Immediate Analgesia

Administer the following analgesics ONCE only (based on last recorded weight)

Date	**Drug (amount per dose)**	**Dose**	**Route**	**Dose**	**Doctor**	**Time given**	**Nurse**	**Nurse**
	Paracetamol (20mg/kg)		PO	ONE dose to be given STAT				
	Ibuprofen (10mg/kg)		PO					
	Naproxen 5mg/kg		PO					
	Dihydrocodeine 1–4 years 0.5mg/kg 4–12 years 0.5–1 mg/kg (max. 30 mg) 12–18 years 30 mg		PO					
	Oramorph 1–3 months 0.05-0.1 mg/kg 3–6 months 0.1-0.15 mg /kg 6–12 months 0.2mg/kg 1–2 years 0.2-0.3 mg/kg 2–12 years 0.2-0.3 mg/kg (max. 10 mg) 12–18 years 5–10 mg		PO					

If required:

	Naloxone (10mcg/kg)		IM	PRN				

4. Repeat pain assessment in 30 minutes and plot on chart overleaf

(Child to be encouraged to assess their own pain where possible)

5. Subsequent pain assessment to be carried out with TPR observations and recorded overleaf.

(Initially hourly then reduce to 2 – 4 hourly once pain is better controlled)

6. Additional nursing observations

DATE & TIME	

APPENDIX 2

PAEDIATRIC PAIN ASSESSMENT INTERVENTIONS / EVALUATIONS RECORDS CHART

NAME_____HOSPITAL NO_____

DOB_____WEIGHT_____ALLERGIES_____ _____

PAIN ASSESSMENT TOOL(S) BEING USED_____

KEY: **SS** Sedation Score (0 – 5). **HR** Heart Rate, **RR** Respiratory Rate (SS Score only if on opiate).
SICKLE PATIENT TO BE SCORED AT 3O MINS, AT 1 HOUR THEN AS REQUIRED (1 – 4 hourly).

DATE	TIME	PAIN ASSESSMENT SCORE			INTERVENTIONS/EVALUATIONS				SIG
		CHILD	PARENT	NURSE	HR	RR	SS		
	Pain at start								
	Pain at 30 mins								
	Pain At 1 hour								

THIS CHART TO BE USED WITH PAIN CHART NO 1 AND SICKLE INITIAL ANALGESIA PAIN CHART AS NEEDED. PAIN TEAM NO # 6439

PAEDIATRIC PAIN ASSESSMENT INTERVENTIONS / EVALUATIONS RECORDS CHART

DATE	TIME	PAIN ASSESSMENT SCORE			OBSERVATIONS			INTERVENTIONS/EVALUATIONS	SIG
		CHILD	PARENT	NURSE	HR	RR	SS		

This chart is to be used with pain chart 1 and sickle pain assessment/analgesia initial chart

APPENDIX 3

Prescribing Guidelines in Paediatric Sickle Cell Pain

Reassess

Reasse

FAST-TRACK FOR PAEDIATRIC SICKLE CELL PATIENTS
A guide for ward and PAU Nurses

General considerations
All Sickle cell patients should have a Pink **Paediatric Passport** instructing them that the nurse in charge will be able to advise them over the phone.

All Sickle cell patients have clinic letters that can be obtained via CIP portal on any computer

When parents phone up, concerned about their child with sickle cell disease, the child may or may not need to be brought to the hospital promptly for further assessment and/or admission.

The following questions should help you to decide whether to advise the parent to bring the child to Accident and emergency if severely unwell, the ward right away or not.

Please fill in the child's details and tick answers to the following questions

Name

D.o.b. **Hospital number**

Ask -

"WHAT IS THE PROBLEM? "

"ARE YOU RINGING FOR ADVICE"

"DO YOU THINK YOUR CHILD DEFINITELY NEEDS TO COME IN?"

The child needs to be brought up promptly if any of the following:

	yes	**no**
Fever >38 "feels burning hot" etc	( )	( )
Markedly pale, more than usual	( )	( )
Pain in the CHEST and/or difficulty in breathing	( )	( )
Pain in the ABDOMEN if severe	( )	( )
Severe diarrhoea with vomiting, can't keep fluids down	( )	( )
Child seems MORE UNWELL or DIFFERENT than on previous occasions	( )	( )
Priapism (a prolonged erection lasting longer than 30mins)	( )	( )

The child may NOT need to be brought to hospital immediately if:

	yes	no
Miserable, off his/her food	( )	( )
Runny nose, cough, other symptoms of viral URTI	( )	( )
Pains in the limbs adequately controlled by usual analgesia	( )	( )

CHECK: That they have given a lot of fluids
: That they have given / continue to give Paracetamol 4 hourly
: That they have tried Paracetamol, Ibruprofen and Dihydrocodiene if they have any
: A warm bath may be worth a try while analgesia takes effect

PLAN
a) Ward nurse to phone back (phone number.....) and check on child's progress at ..... (time)

or

b) Child should be brought up promptly for further assessment, and please inform them where to go

NB: before signing off say: "If he/she gets worse or you are worried at all, please phone again"

Advice given:

Action taken:

Signed.....
Date.....Time.....

August 2002 Mary Rossiter Updated by Olu Wilkey May 2004, Updated Jan 201,Updated Sept 2015 , Updated Jan 2019, updated June 2020