Scottish Paediatric
& Adult Haemoglobinopathy
Managed Clinical Network

NHS SCOTLAND

MANAGEMENT GUIDELINES FOR ADULT PATIENTS WITH SICKLE CELL DISEASE

Approved August 2012
Review August 2014

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GUIDELINE FOR THE USE OF HYDROXYCARBAMIDE IN ADULTS WITH SICKLE CELL DISORDERS

Hydroxycarbamide has been shown in a large randomised-controlled study to decrease the frequency of painful vaso-occulusive crises and of chest crises in adults with homozygous sickle cell disease. It is not yet known whether it will lessen the risk of other sickle-related complications. There is early evidence to indicate a survival benefit, probably because of a reduction in fatal chest crises in patients taking hydroxycarbamide.

Side effects include bone marrow suppression, gastro-intestinal disturbances, and increased skin and nail pigmentation. It is potentially teratogenic and there is concern about an increased risk of malignancy, but this risk remains theoretical and has not been confirmed with follow-up of patients on the drug to date.

Indications for use

- >3 admissions with painful crises in the previous 12 months, or
- >1 admission with painful crisis in the previous 12 months, and are symptomatic in the community such that lifestyle is affected, or
- >1 life threatening complications of the disease, such as acute chest syndrome
- other indications (such as secondary stroke prevention, pulmonary hypertension, prevention of renal disease) are controversial and should be discussed with consultant colleagues within the network

Exclusions

- Pregnancy or not practicing active contraception (if sexually active)
- Active hepatitis
- Hb<6g/dl, WCC<1x10^9/l, Plts<100x10^9/l Prior to commencing drug

Requirements prior to starting therapy

- The benefits and hazards of using hydroxycarbamide should be considered for each patient individually, and discussed
- Ensure that the patient is willing to attend regularly to monitor blood counts
- Discuss the possibility of infertility with male patients. Offer sperm banking
- It is important to discuss the possible (small but not quantifiable) risk of malignancy

Baseline investigations

- FBC and reticulocytes
- HbF%
- U+Es, LFTs, Urate, LDH

Monitoring

- Weekly FBC for first 4 weeks
- Fortnightly FBC for next 8 weeks
- Monthly FBC thereafter if counts stable
- 3 monthly U+Es, LFTs, Urate, LDH and HBF%

Regimen details:

- Commence at 15mg/kg/day orally to nearest 500mg
- If there is a good clinical response continue on this dose (Minimal effective dose)
- If clinical response is sub-optimal, increase by 2.5mg/kg/day every 8 weeks until limited by toxicity

Toxicity

- Neutrophils $< 1.5 \times 10^9/l$
- Platelets $< 80 \times 10^9/l$
- Retics $< 10 \times 10^9/l$
- Haemoglobin <3g/dl from baseline

- If any of the above problems with FBC encountered, stop hydroxycarbamide, until full blood count has recovered
- Restart at 2.5mg/kg/day (or 1 capsule – 500mg) lower. This is the maximum tolerated dose (MTD)

Caution

- If there is a significant rise in Hb (>11g/dl in HbSS) stop the hydroxycarbamide and consider venesection
- If there is a downwards trend in FBC parameters, increase frequency of monitoring
- Use with caution in renal impairment: start at a lower dose and increment more cautiously
- If Creatinine Clearance < 60ml/min, commence at 50% dose (7.5mg/kg/day)

Toxicities

Common: Bone marrow suppression and cytopenias.
Hyperpigmentation of nails and skin
Nausea and vomiting
Diarrhoea
Skin rash

Uncommon: Alopecia
Teratogenicity
Leg ulcers
Decreased sperm count and function
Low risk of second malignancy

Dose modifications

Neutrophils		**Platelets**	**Hydroxycarbamide**
$\geq 1.5 \times 10^9/L$	&	$\geq 80 \times 10^9/L$	100% dose
$< 1.5 \times 10^9/L$	or	$< 80 \times 10^9/L$	Stop treatment and recheck FBC until N>1.5 and Plt >80. Restart treatment at 2.5mg/kg/day or 500mg daily lower.

Renal Impairment : Use with caution. If Creatinine Clearance < 60ml/min, commence at 50% dose (7.5mg/kg/day)

Hepatic Impairment: Use with caution

References: Charache S, Terrin ML, Moore RD et al. Effect of hydroxycarbamide on the frequency of painful crises in sickle cell anaemia. Multicentre Study of Hydroxyurea. N Engl J Med 1995: 332: 1317-1322

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.

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.

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

Discussion with the local on call haematology team is recommended, urgently, if patient has chest or abdominal signs, neurological signs, priapism or shock.

Principals of management

- Effective analgaesia
- Hydration
- 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

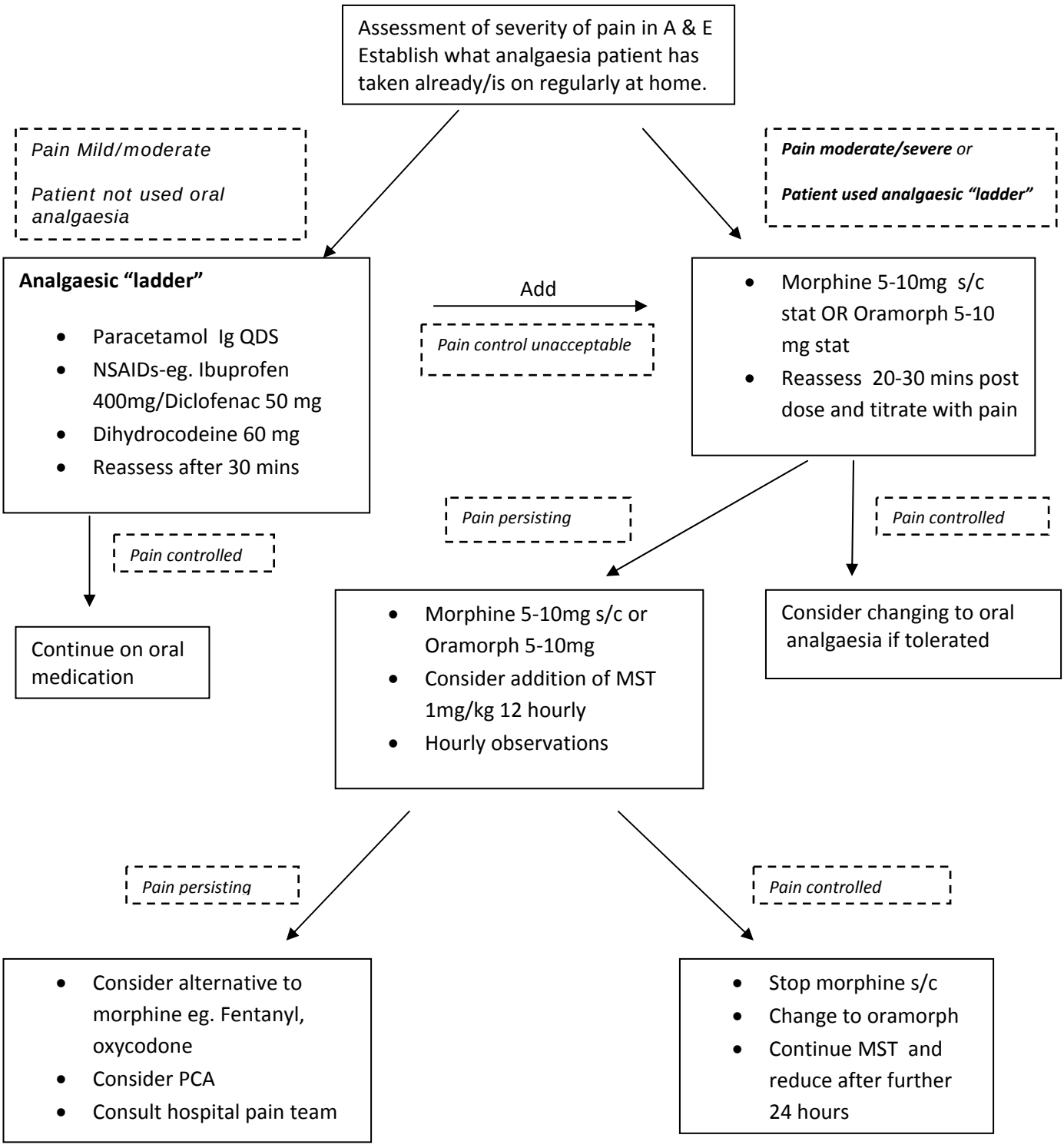
Additional Investigations

- If there are chest signs or temperature >38^{o}: – Chest X-ray
- If O_2 sats on air < 94% – Arterial gases on air
- 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.

Analgaesia

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

Some patients will have individualised pain protocols which should be referred to if available, otherwise follow chart below.



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

Consider additional therapies:

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

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 0xygen 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). 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

Reference: Management of Acute painful crises in sickle cell disease BCSH GUIDELINE 2003

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.

Principles of management

The Haematology Team should be informed as far in advance of elective surgery as possible. Preparation for surgery will usually require AT LEAST a weeks notice.

Patients should be managed jointly between haematology, surgery and anaesthetics. The anaesthetic team should be made fully aware of the patients needs.

An individual management plan should be in place prior to surgery which takes into account the patients particular risk factors and the type of procedure planned. This should be circulated to all relevant individuals prior to an elective surgical procedure.

If a period in HDU/ICU is anticipated the necessary arrangements should be made in advance.

Sickle patients should be placed near the beginning of the theatre list to minimise time fasting and reduce likelihood of cancellation.

Ensure the patient is well informed and involved in their management plan when possible.

Fluid balance must be managed with great care in sickle patients as they dehydrate easily due to impaired renal concentrating ability.

These patients may have a history of chronic pain and be relatively insensitive to opiate analgesia. This should be taken into account when prescribing in the post-operative period.

Avoid factors which may precipitate the development of a sickle crisis;

Hypoxia
Dehydration
Acidosis
Cold
Pain

Day case surgery may not be suitable for this patient group due to the above considerations.

Pre-operative Care

Ensure Haematology Team is made aware of admission as soon as possible.

Blood transfusion is not always indicated prior to surgery (see advice below). Where indicated blood transfusion will be arranged by the haematology team and arranged during the week before surgery for elective procedures. **Do not transfuse without prior discussion with Haematology**.

In addition to FBC, HbS level and routine bloods, a sample for Group and Save, antibody screening and red cell phenotyping (if not performed previously) should be sent at the pre-op assessment visit and BTS informed of the date and nature of the planned procedure.

Admit the day before surgery if possible.

Hydration

Maintenance IV fluids should be commenced the evening before surgery unless the patient is drinking freely.
IV fluids must be started if nil by mouth for >2 hours.
Maintenance fluids may need to run at a higher rate than usual to avoid dehydration. Monitor fluid balance closely.

Thromboprophylaxis

This should be considered for all procedures, particularly for
major surgery or if patient will be immobile for >24 hours post procedure.
Follow local guidelines on VTE prophylaxis.

Oxygenation

Avoidance of hypoxia is vital to prevent sickling and tissue ischemia.
Document baseline O_2 saturation (may have pre-existing cardio-respiratory disease)
Consider use of incentive spirometry.
Oxygen saturation should be monitored continuously once premedication given.
Consider giving supplemental oxygenen from the time of premedication.
Hyper oxygenate at induction of anaesthesia.

Temperature Regulation

Hypothermia can trigger peripheral stasis and sickling. Attention should be paid to keeping the patient normothermic during surgery. This may require the use of warmed intravenous fluids, warm air blankets and adjustment to the ambient temperature in theatre. Ensure the anaesthetic team is made aware of these requirements in advance.

Infection Management

These patients are functionally hyposplenic and are therefore at increased risk of perioperative infection. If additional antibiotic prophylaxis is needed this should be detailed in the patients management plan.
Elective surgery should not proceed in the presence of active infection as this will greatly increase the risk of serious sickle related complications.

Intra-operative Care

Hydration
Volume status must be monitored closely throughout the procedure and hypovolaemia avoided at all costs through the prompt use of volume replacement. Again these patients are more prone to dehydration due to impaired renal concentrating ability.

Positioning
As prolonged venous stasis can lead to sickling, the patient should be repositioned periodically where possible during long procedures.

Tourniquet
These are contra indicated in patients with Sickle Cell disease and relatively contra indicated in Sickle Cell Trait

Oxygenation
Pulse oximetry should be monitored continuously throughout the procedure.

Temperature Regulation
Ensure the patient remains normothermic. Warmed IV fluids are particularly important during long procedures.

Cell Salvage
Intra-operative cell salvage is contra indicated in patients with Sickle Cell Disease.

Post-op Care

Hydration
Continue IV fluids until the patient is able to tolerate a sufficient oral intake
Avoid fluid overload post-op as this may increase the risk of complications in high risk patients.

Analgesia
Prescribe post-op analgesia as per local guidelines. Sickle patients with a history of chronic pain may be *relatively opiate resistant* and require higher doses for adequate analgesia. There is a high incidence of painful crises in the post-op period (see separate guideline).

Oxygenation
Pulse oximetry should continue for at least 24 hours or until stable.
Maintain saturations >95% or above baseline whichever is higher.
Continue incentive spirometry, encourage patient to take regular deep breaths (every 10minutes) if not available.

Temperature regulation
Ensure the patient is normothermic until able to regulate body temperature independently.

Infection Management
If patient develops a fever (>38.0°c) ensure blood cultures are taken and start intravenous antibiotics as per local antimicrobial policy. (NB hyposplenic)
Check cannula and line sites daily for signs of infection.
Monitor for symptoms such as productive cough, shivering or myalgia.

Encourage early mobilisation with physiotherapy input if appropriate.

Should the patient develop new symptoms suggestive of a sickle related complication, such as a painful crisis, chest crisis or CVA the haematology Team MUST be informed immediately.

Blood Transfusion Guidance

Transfusion Prior to Elective Surgery
Patients undergoing major surgery and those with a history of previous sickle related complications are at higher risk in the perioperative period. Conversely over aggressive transfusional support may unnecessarily expose low risk patients to transfusion related hazards. Transfusion should therefore be considered dependant on patient factors and the nature of surgery.

1. Low Risk
Even low risk patients undergoing minor procedures under general anaesthetic with a low risk of complications require discussion with Haematology. **Aim for pre-op Hb >7g/dl. Top up transfusion should be considered.**

2. Intermediate Risk
Patients' with a previous history of chest crisis (or other co-morbidity) or patients' undergoing Intermediate risk surgery e.g. tonsillectomy, splenectomy or cholecystectomy.
Top-up transfusion aiming for Hb of 10-11g/dl (irrespective of HbS level)

3. High Risk
High-risk procedures e.g. thoracic, major upper abdominal surgery, neurosurgery or previous serious sickle related complications eg severe chest crisis, CVA.
Aim for pre-op HbS % of <30. This will almost certainly require exchange transfusion (see separarate guideline). Hb should be kept below 11g/dl.

In patients with a high baseline Hb (>9.5g/dl), top-up transfusion should be used with caution to avoid hyperviscosity. Exchange transfusion is likely to be needed for all but low-risk procedures. Particular care is needed in patients with HbSC disease as they are more likely to develop hyperviscosity related complications with top-up transfusion.

Blood should be sickle negative and matched for Rh and Kell antigens as a minimum.

These principles are also applicable to patients undergoing **emergency surgery** if time permits. As a minimum these patients should have suitable cross-matched blood available should an emergency exchange procedure be needed (see separate guideline). Haematology Team and Blood Transfusion Laboratory should be informed on admission if surgery is deemed likely.

ACUTE CHEST SYNDROME IN SICKLE CELL DISEASE

Acute chest syndrome is the leading cause of mortality in adults with HbSS.
• CONTACT HAEMATOLOGY REGISTRAR ON CALL as soon as suspected

Acute Chest syndrome can develop rapidly and clinicians should have a high index of suspicion.

Key Features *(patient may not have all of these)*
• O2 Sats on air <94% or PaO2 <8kPa
• Fever, Cough, Chest pain
• Respiratory distress/hypoxemia
• New opacity on chest x-ray
• Worsening anaemia
• Bilirubin stained sputum
• Antecedent painful crisis

Symptoms and Signs
• Hypoxia
• Pain in chest wall, upper abdomen, and/or thoracic spine.
• Signs of lung consolidation; usually bilateral and, generally, starting at the bases (can progress in hours).
• Fever
• Tachypnoea
• Tachycardia
• Shortness of breath
• Cough may be a late symptom
• Physical signs often precede X-ray changes

Special investigations
• Arterial blood gases on air
• Chest X-ray
• Blood cultures, sputum cultures, respiratory atypical serology (including Chlamydia, legionella, mycoplasma).
• Group and save – please state patient has 'Sickle cell disease' on form
• Full blood count and reticulocytes initially and then daily until improving (compare patients baseline values)
• Renal and liver function tests

Initial Management
• Call Haematology Registrar/Consultant
• IV fluids – with close monitoring of fluid balance
• IV Co-amoxiclav 1.2g tds (plus Clarithromycin 500mg bd if atypical pneumonia suspected), for 5-7 days (consult local antibiotic policy if penicillin allergic)
• Monitor pO2 on air, pulse, respiratory rate, arterial blood gases, Hb
• Oxygen therapy to increase oxygen saturations >95%
• Bronchodilators if there is evidence of wheeze or reversible airways disease, or a history of asthma.

- PaO2 Room Air
  < 10.0 kPa - Oxygen, and monitor oxygen saturations on air, hourly
  < 8 kPa – give oxygen and consider transfusion (top up or exchange)
- Discuss transfer to HDU with HDU team if:
  Worsening hypoxia or developing hypercapnia
  Patient is deteriorating/tiring

Additional supportive management

- Incentive spirometry – 10 deep breaths 1-2 hourly when awake
- Consider positive pressure ventilation (nasal CPAP or mask BiPAP) for patients with poor respiratory effort or reduced ventilation
- Analgaesia (as per painful crisis)
- Consider one dose of frusemide 0.5-1mg/kg if signs of fluid overload
- Prophylatic Low Molecular Weight Heparin

Transfusion

N.B. Sickle cell patients should ideally have Rh and Kell matched sickle negative blood

- **Simple transfusion** for moderately severe illness, but do not transfuse acutely to >10g/dl, or take the haematocrit above 30%
- If the patient's Haemoglobin has fallen to <6.0 g/dl consider initial top up transfusion.

- **Exchange blood transfusion** can be done by the automated or manual method. (See separate guideline)
  For both types of exchange transfusion:
  - Cross match 8 units of blood.
  - Ensure adequate venous access.
  Consider femoral line insertion if peripheral access is poor. If femoral line is inserted, use a vascath double lumen for automated exchanges.
- When the exchange transfusion is completed check Haemoglobin and HbS% level. The results of these and the patient's clinical condition will help ascertain whether a further exchange is necessary. Aim HbS%<30%

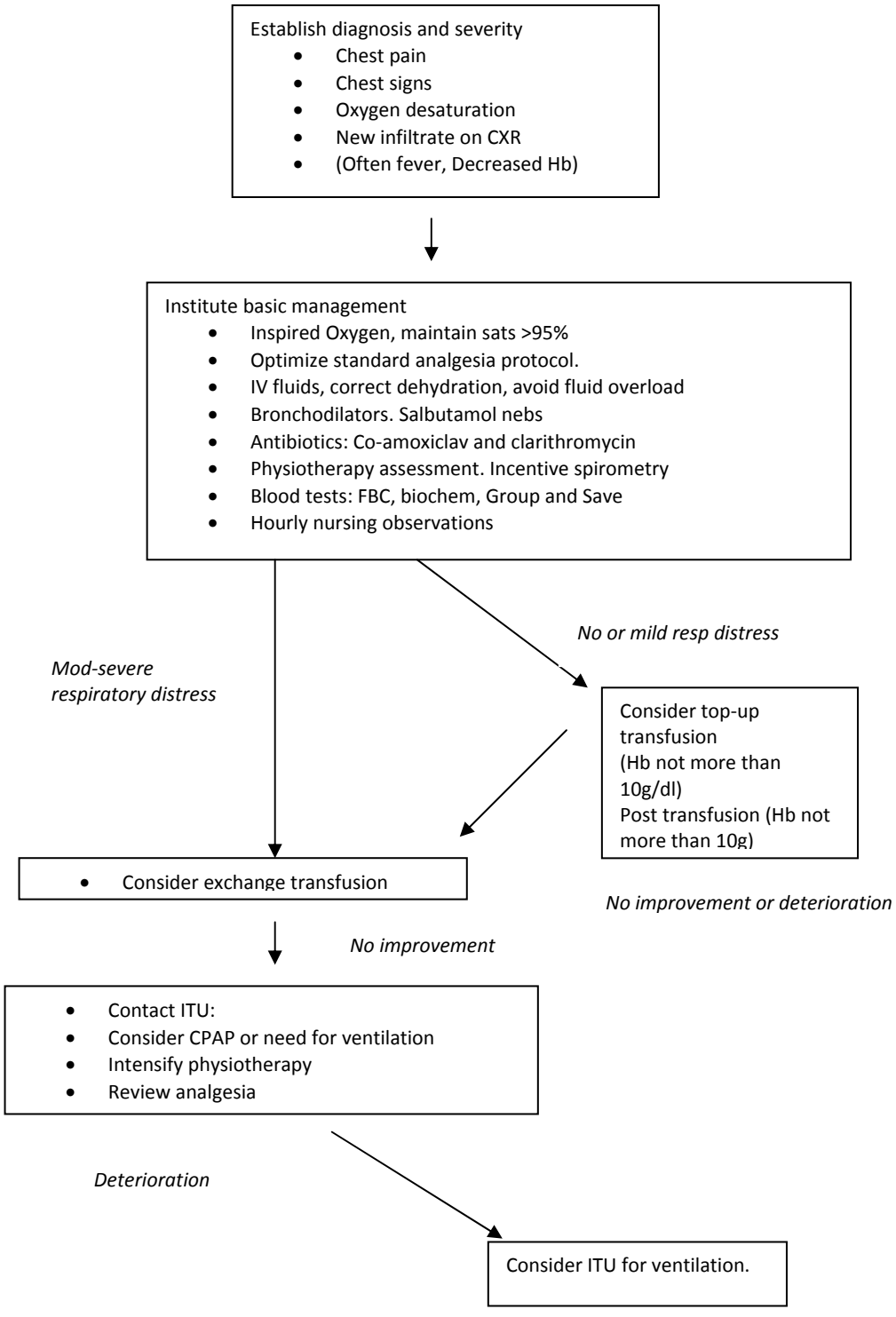
Monitoring

- Ensure that vital signs are taken and documented hourly (more frequently if patients condition is deteriorating)
- Continuous saturation readings.
- Record $Sa0_2$ in air and if <92% ABG's
- Inform haematology team urgently if respirations increase or $Sa0_2$ falls
- Record temperature 2-4 hourly and if >38°c take cultures and review Antibiotics
- Maintain a strict fluid balance

Discharge Criteria – only in consultation with Haematologist

- Improved pulmonary symptoms and adequate oxygenation on room air
- Afebrile >24hours and negative cultures if applicable
- Stable/rising haemoglobin
- Tolerating adequate oral fluids and able to take medications orally
- Adequate pain relief on oral analgaesia

MANAGEMENT OF ACUTE SICKLE CHEST SYNDROME



GUIDELINE FOR RED CELL EXCHANGE TRANSFUSION IN SICKLE CELL DISEASE

Most patients with sickle cell anaemia are relatively asymptomatic despite baseline Hb concentrations between 5-12g/dl as HbS is a low-affinity haemoglobin and oxygen delivery to tissues is enhanced. **Chronic steady state anaemia alone is not an indication for transfusion.** Top-up transfusion increases whole blood viscosity and may aggravate sickling. Top up transfusion is not indicated for uncomplicated sickle crises (see separate guideline on perioperative management).

Exchange transfusion is a potentially life saving procedure that allows correction of anaemia *without* increasing blood viscosity and may improve tissue oxygenation whilst reducing microvascular sickling.

The aim of exchange transfusion is to lower the HbS level to 30% or less while keeping the Haemoglobin close to 10g/dl. C*linical benefit may be seen even with a partial manual exchange*.

Prior to embarking on an exchange procedure the case must be discussed with the Consultant Haematologist on call.

Indications for Exchange Transfusion

Acute cerebro-vascular event
Acute chest syndrome
Multi-organ failure
Preoperatively in selected cases

Manual Red Cell Exchange Transfusion

1. Background

This procedure should only be performed by suitably qualified staff under the supervision of the responsible Consultant Haematologist.

Staff should be familiar with the procedure and management of associated complications.

In some units automated exchange using an apheresis machine may be available. (Check local policy).

However manual exchange should not be delayed if it is clinically indicated.

Adult patients may require insertion of a femoral or internal jugular line but those with good peripheral access can be managed with one or preferably two large bore IV cannulae.

Trust policies for safe blood transfusion must be adhered to at all times.

2. Patient work up

Consent

Ensure the patient is consented for the exchange procedure. Often verbal consent alone will be practical when the patient is seriously ill.

Aspects which should be specifically mentioned include:

- Access (peripheral vs vascath)
- Vasovagal episodes
- Blood transfusion reactions and alloimmunisation
- Transfusion related infection

Baseline Blood tests

- HbS percentage
- Urea and electrolytes
- Calcium
- Magnesium
- Liver function tests
- Virology – HIV, hepatitis B and C
- Ferritin levels, glucose, thyroid and endocrine function if appropriate (not relevant in emergency setting)

Inform Transfusion Laboratory of planned exchange and check for history of previous transfusion reactions or allo-antibodies. Make them aware of degree of urgency of exchange.

Cross match 6-8 units of packed red cells (depending on size of patient). These should be;

- Sickle negative
- Phenotypically matched (Rh and Kell as minimum),
- Ideally less than 5days old (not essential in an emergency)
- Nb patients should have full red cell phenotype performed if not already known and not had recent transfusion

N.B. It may be several hours before compatible blood is available for patients with allo-antibodies

Patient Assessment

Prior to the procedure, check:
Baseline observations (blood pressure, heart rate, temperature and oxygen saturations)
Patient weight (estimate in an emergency when accurate measurement not feasible).

3. Equipment and supplies

- Baxter infusion pump
- Blood giving set
- Blood warmer
- 500mls normal saline (several packs)
- Packed red cells (amount dependant on patient's size and condition, usually 6-8 units)
- Dynamap and tympanic thermometer
- Recent blood results including; haemoglobin, haematocrit, and HbS percentage
- Calcium gluconate in case of hypocalcaemia or hyperkalaemia

Equipment for Peripheral Access/Venesection

- Sterile gloves
- 16g Kimal access needle x1
- 18g or 20g cannula
- Chloraprep/skin preparation
- 4x10ml syringes
- 20mls of normal saline for flush
- Tegaderm dressing to secure cannula
- Mepore tape to secure kimal needle
- Gauze swabs
- Consent form (if applicable)
- Blood collection form, observation chart and nursing documentation
- Large sharps box
- Weighing scales (to assist calculation of volume venesected)
- 3 way tap (useful in patients with limited peripheral access)
- Sterile bungs (to allow repeated access of large bore venesection pack needle into 3 way tap extension set)
- Venesection packs

Patients requiring a central line

Follow local line insertion protocol

4. Method

Perform the venepuncture for the access (venesection) line and return lines. Ensure access on both sides is secured and both lines are flushed with normal saline prior to commencing the procedure.
Set up the giving set for the return line so that fluid/blood administered is warmed in the blood warmer.

Do not start until compatible blood is available on the ward

1. Set up a bag of normal saline and run 500mls over 15 to 30 minutes to ensure the patient is adequately pre-hydrated (reduce rate/and or volume if concern over fluid overload or cardiovascular compromise).
2. Ensure the blood is warmed prior to infusing.
3. To venesect: remove 450-500mls of blood over 15-30minutes.
4. Ensure local transfusion policies are adhered to and documentation completed.
5. Calculate the amount to be exchanged, dependant on the starting haemoglobin, as follows:

- Hb >8.0g/dl 5-8 units
- Hb 6-7.9g/dl 4-6 units
- Hb <6g/dl up to 4 units

Exchange Procedure if starting Hb >8.0g/dl

Venesect 1_{st} unit	***whilst***	Replacing with 500mls normal saline stat
Venesect 2_{nd}unit	***then***	Transfuse 1_{st} unit over 30-40 minutes
Venesect 3_{rd} unit	***then***	Transfuse 2_{nd} unit over 1 hour
Venesect 4_{th} unit	***then***	Transfuse 3_{rd} unit over 2 hours

Re-check FBC, HbS and electrolytes at this stage

If repeat Hb<9g/dl	Transfuse 4_{th} unit and consider 5_{th} unit (3hrs each)
If repeat Hb>9g/dl	Restart from "venesect 1_{st} unit"

*N.B. By removing **two** units of blood before transfusing the 1_{st} unit, this method results in more efficient lowering of the HbS %. However if the patient is cardiovascularly unstable, or becomes hypotensive during the venesection, the replacement transfusion should be started sooner, i.e. after venesection of the 1^{st} unit.*

If starting Hb 6-7.9g/dl

Venesect 1_{st} unit	***whilst***	Transfusing 1_{st} unit
Venesect 2_{nd}unit	***then***	Transfuse 2_{nd} unit over 1hr and 3_{rd} and 4_{th} units over 3hrs

Further exchange may be required (see "Hb 8g/dl") if insufficient clinical improvement/impact on HbS level

If starting Hb<6g/dl

Top up transfusion to 8-10g/dl (rate depending on clinical condition and baseline Hb) initially, discuss with Consultant Haematologist.

Formal exchange may be required (see "Hb 8g/dl") if insufficient clinical improvement/impact on HbS%

5. Post procedure

Remove Kimal needle but leave cannula in, flush with normal saline.
Monitor vital signs at 15 and 30 minutes post procedure
Take bloods 30 minutes post procedure for:

- Full blood count
- HbS percentage
- Electrolytes, calcium and magnesium

Avoid final Hb of >11g/dl (risk of hyper viscosity) or <7g/dl

Watch for development of **hyperkalaemia** and **hypocalcaemia** during the exchange.

Ensure all transfusion documentation is completed correctly and returned to blood transfusion as per local policy.

Acknowledgements:
Thank you to the Haematology Department, Manchester Royal Infirmary, for allowing reference and adaptation of their guidelines for the management of Adults with Sickle Cell Disease.