Management of Children with Sickle Cell Disease	University Hospitals of Leicester NHS NHS Trust **Trust Ref: C17/2016**

1. Introduction

Concise guideline for overall management of paediatrics patients with Sickle Cell Anaemia

2. Scope

Relates to Paediatric haematology teams, Paediatric Emergency medicine, Paediatric medicine and specialties (nephrology, urology, microbiology, endocrinology, surgery, gastroenterology, anaesthesia, orthopaedics, ophthalmology, ENT)

3. Recommendations, Standards and Procedural Statements

Guidelines based on:

- National Haemoglobinopathy Peer Review Standards 2014
- NICE: sickle cell acute pain episode: management of an acute painful sickle cell episode in hospital (2012)
- NHS Sickle Cell and Thalassaemia Screening Programme - Standards for the Care of Children with SCD 2009
- Caring for people with Sickle cell and thalassaemia syndromes: RCN competencies: a framework for nursing staff (2011)
- A sickle cell crisis? A report of the National Confidential Enquiry into Patient Outcomes and Death (2008)
- Transition: improving the transition of young people with long term conditions (2006)
- TCD Scanning for Children with Sickle Cell Disease (2009)

4. Education and Training

Regular teaching provided in ED, Paediatric Specialist Trainees Regional Training days and nursing training programmes.

5. Monitoring and Audit Criteria

Key Performance Indicator	Method of Assessment	Frequency	Lead
100% adherence to NICE: Sickle cell acute painful episode - Management of an acute painful sickle guidance (2012)	Retrospective audit	Annual	Dr K Kotecha

6. Key Words

Sickle cell, pain, acute chest syndrome, stroke, transfusion

This table is used to track the development and approval and dissemination of the document and any changes made on revised / reviewed versions

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1.0 INTRODUCTION

1.1 The sickle cell disorders are a heterogeneous group of disorders affecting patients in whom haemoglobin S is the major haemoglobin. They include haemoglobin SS, SC, and S beta thalassaemia.

1.2 This clinical policy has been compiled by the haemoglobinopathy team at Leicester Hospitals NHS trust. It has been formulated following reviews in service, based on current practice and research including published standards in the management of children with sickle cell disease.

1.3 It is intended for the guidance of in and out-patient management. Cases need to be assessed individually and the management tailored appropriately. The opinion of the Lead Consultant/ Deputy Lead Consultant should be sought where necessary.

1.4 Members of the Specialist Teams (contactable through Switchboard)

- CONSULTANT PAEDIATRIC LEAD/DEPUTY
- CONSULTANT GENERAL PAEDIATRICIAN
- PAEDIATRIC HAEMOGLOBINOPATHY CLINICAL NURSE SPECIALIST
- SPECIALIST REGISTRARS: PAEDIATRIC HAEMATOLOGY & GENERAL PAEDIATRICS (COVERING HAEMATOLOGY AND ONCOLOGY)
- SHO'S: COVERING PAEDIATRIC HAEMATOLOGY AND ONCOLOGY

2 DEFINITIONS

Haemoglobinopathy	A group of inherited conditions where the pathology relates to abnormalities in haemoglobin production or function. Includes haemoglobin variants and thalassemias.
Sickle cell disease	This encompasses a number of genotypes including Hb SS, Hb SC, Hb S/beta thalassaemia and some rarer compound heterozygote states where HbS dominates and results in a clinically significant condition.
Child	Trust transition age for adult care is > age 16. For the purposes of this policy a child is a person less than 16 years old.
Adolescent	This includes teenagers and young adults age up to 18 years. Some of these will be managed in the paediatric service and some in adult service

3 ARRANGEMENTS FOR EMERGENCY ASSESSMENT, ADMISSION AND SPECIALIST ADVICE

3.1 Patients can access acute care via Childrens Admission Unit (CAU) or Paediatric Emergency Department (ED)

3.2 Emergency Patients should be booked through reception and rapidly assessed & triaged

3.3 A record of observation and clinical assessment should be made

3.4 Unwell patients will be managed in the resuscitation suite according to Trust paediatric resuscitation guidelines

3.5 Paediatric attending team will be informed of acute presentation for urgent review

3.6 All patients will be rapidly moved to a bed/cubicle for prompt treatment

3.7 Medical and Nursing staff will identify individual patient care plan in front of notes, parent held copy or ward based copy to access treatment plan and follow guidance.

3.8 For acute pain crisis, analgesia if required will be administered according to guidelines aiming for first dose within 30 minutes of presentation. (Appendix 5)

3.9 A decision to deviate from the pathway should be made only following discussion with the clinical lead/deputy or a senior paediatrician or haematologist

3.10 Prompt medical assessment will follow including recording of presenting problem and documentation of treatment plan

3.11 Required analgesia and other urgent medication including antibiotics will be prescribed on medical drug chart to avoid delay in administration of subsequent doses. If required, the second dose of analgesia will be administered BEFORE patient is moved to the acute inpatient ward

3.12 The admitting paediatric team remain responsible for the inpatient episode until the next working day. The Clinical Lead/Deputy Lead should be informed of their admission and asked for advice

3.13 Out of hours (5pm -9am and weekends) patients of concern will be discussed with the General Paediatric Consultant on call who can liase with Clinical Lead/Deputy Lead

3.14 The paediatric haemoglobinopathy team will see all patients Monday to Friday

3.15 Unwell patients will be prioritised on the ward round for review

3.16 At evenings and weekends, patients will be seen by the on-call paediatric medical teams and discussed with the Clinical Lead/Deputy Lead if required

4 GUIDELINES OF MANAGEMENT OF ACUTE COMPLICATIONS

Routine Baseline Investigations:

- FBC, reticulocyte count
- U+Es, Creatinine, LFTs, LDH
- CRP
- G+S
- Red cell phenotype (if not known)

4.1 In older children presenting with an uncomplicated vaso-occlusive sickle cell crisis, a finger prick test for FBC may be done in the first instance. Note however, all patients on hydroxycarbamide (hydroxyurea) and chelation therapy with desferrioxamine, deferipone or Exjade must have routine baseline investigations as above. Other tests may be indicated based on presentation.

- CXR (respiratory symptoms/signs)
- Urine dipstick/culture, blood, throat, nose, sputum, stool, wound, CSF culture, NPA (as dictated by symptoms)
- Mycoplasma /Chlamydia serology/urinary pneumococcal antigen (respiratory symptoms/signs)
- Parvovirus serology (IgM) / DNA (Pallor/falling Hb/low reticulocyte count)

- CT/MRI/MRA head (CNS symptoms/signs)
- Arterial/capillary blood gases (respiratory distress / metabolic compromise)
- Hb S level (Acute chest syndrome or stroke)
- Yersinia serology/stool culture (patients on desferrioxamine with diarrhoea /abdominal pain)
- Serum amylase (abdominal symptoms/signs)
- Plain abdominal film /USS abdomen (abdominal symptoms/signs, features of girdle syndrome, sequestration, cholecystitis)
- USS limb / MRI limb/ limb/ joint x-ray (suspected osteomyelitis)

Admission to hospital

The following are indications for admission to hospital:

- Persistent temperature
- Pallor, lethargy, malaise, abdominal distension
- Severe pain requiring opiate analgesia
- Symptoms and signs suggesting acute chest syndrome, acute cerebrovascular event, sequestration syndrome, aplastic crisis and fulminant priapism (lasting >3hr)

Discharge from hospital

4.2 Prior to discharge ALL cases must be discussed with the general paediatric SpRs / Consultants (senior medical team out of hours). Appropriate follow up should be arranged. A short supply of analgesia should be made available.

Follow up options

- Inform Haemoglobinopathy clinical lead/Acute clinical nurse specialist to arrange early outpatient appointment if required
- All discharges out of hours must be notified to the paediatric Haemoglobinopathy team the following day by email of attendance

5 MANAGEMENT OF COMPLICATIONS

Acute Pain Crisis

5.1 This is the most frequent complication of sickle cell disease and a common reason for presentation to hospital. Typically the child will present with limb, back or chest pain. A trial of simple analgesia may have been instituted by the family. An enquiry into this as well as potential precipitating factors should be made e.g., coryzal symptoms, dehydration, over exertion.

5.2 The mainstay of the management of sickle cell crisis is supportive and includes:

- Pain relief
- Fluid replacement (in accordance with Trust Paediatric IV fluids guideline)
- Antibiotics

5.3 **Pain Management and PCA - please see separate pain flowchart**

Fluid Replacement

5.4 **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.

5.5 Children with sickle cell disease need individualised 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. **Hyperhydration is not routinely recommended**.

5.6 The oral / enteral route is always preferred. However, in those who are unable to tolerate this due to pain, abdominal / respiratory problems, IV fluids may be required in accordance with the paediatric IV fluid guidelines.

5.7 Intravenous therapy should be stopped once the patient is stable and pain is controlled.

5.8 Adequate oral intake should be documented.

Antibiotics

5.9 Sickle cell patients are particularly susceptible to severe overwhelming blood borne infections.

5.10 Important organisms to consider are Streptococcus Pneumoniae, E coli and Salmonella species. The use of prophylactic penicillin has decreased the incidence of pneumococcal infections.

5.11 Always look for a focus of infection when the patient is febrile and organise cultures.

5.12 Any child with a **sequestration syndrome, chest syndrome, or septic, must receive IV antibiotics:**

- >6 months: IV Ceftriaxone 80mg / kg once daily max 4g/day). Consider dose reduction after 24 hours or when clinically stable. (First dose can be cefotaxime in A&E)
- 1-6 months: IV Cefotaxime 200 mg / kg / day in 3 divided doses, after 3 days reduce to 50 mg / kg / day.

5.13 Any child with two temperatures of 38.0°C and one at 38.5°C, but who appears mild to moderately ill should receive IV antibiotics:

- > 6 months: IV Ceftriaxone 50 mg / kg/ once daily.
- 1-6 months: IV Cefotaxime 150 mg / kg / day in 3 divided doses.

5.14 If signs of chest infection as well as the above add in oral Clarithromycin (to cover mycoplasma, for dosage see BNFC). Also do mycoplasma titres, if it is positive a 10 day course is required, otherwise stop.

5.15 Patients who are clinically well and are afebrile should continue penicillin prophylaxis. If the child then enters either category 5.12 or 5.13 (see above) they should start on IV antibiotics.

5.16 If the decision is taken to treat for osteomyelitis antibiotics should be chosen to cover for Salmonella, Streptococcus and Staphylococcus spp. Always discuss cases with Microbiology.

Intravenous Therapy – note consider insertion of PICC line

5.17 Start treatment with Intravenous Ceftriaxone +/- Intravenous Flucloxacillin (for dosage see BNFC). This usually depends on culture results. Ciprofloxacin is well absorbed orally and can be used orally or intravenously for Salmonella positive cases .Intravenous therapy should be between 7 – 14 days.

5.18 Decisions over the total length of treatment will depend on the certainty of diagnosis and clinical course, and will need involvement of the orthopaedic and microbiology teams.

Oral Therapy

5.19 Again duration and type of antibiotic depends on certainty of diagnosis and culture results. Treatment of osteomyelitis including IV therapy course is usually for a total of 6 weeks.

5.20 Suggested antibiotics include :

- Ciprofloxacilin (for dosage see BNFC): Salmonella

- Flucloxacillin (for dosage see BNFC): Staphylococcus

5.21 Augmentin and Clindamycin (for dosage see BNFC):

5.22 Use in culture negative cases (common, especially if prior use of antibiotics), Gram negative bacteria, anaerobic bacteria. Augmentin provides cover for Salmonella and they both provide cover for staphylococcus.

5.23 Clindamycin has good bone penetration.

- Surgical Debridement

Under no circumstances must surgery be contemplated without prior discussion with the on call paediatric consultant (who can liase with Clinical lead/deputy)

5.24 If any concerns, discuss antibiotic usage with the microbiologist.

5.25 In suspected sepsis, hydroxycarbamide (hydroxyurea) and chelation therapy should be stopped due to the risk of cytopenia.

6 EMERGENCY MANAGEMENT- POTENTIALLY LIFE THREATENING CONDITIONS

6.1 Splenic and Hepatic Sequestration

Acute Splenic Sequestration (More common in young children).

Presentation:

- Sudden onset of tachypnoea, pallor, abdominal pain, and splenic enlargement.
- Precipitated by fever, dehydration, and hypoxia.
- Rapid sequestration of red cells can lead to a sudden fall of the haemoglobin and death from hypoxic cardiac failure with pulmonary oedema.
- May have a more insidious onset.

Investigation:

- FBC + reticulocyte count
- G+S (+red cell phenotype if not previously performed)
- Blood culture
- U&Es, creatinine, LFTs, CRP
- Store serum for virology

Management:

- Assess the need for volume expansion and site a cannula.
- Crystalloid should be used with caution as this may exacerbate heart failure.
- Organise for phenotypically matched red cell transfusion without delay (if in extremis uncrossmatched O negative).
- Broad-spectrum antibiotics to cover pneumococcus and haemophilus:- see antibiotics section.
- Patients with recurrent splenic sequestration should be considered for splenectomy after prior administration of Pneumovax, Men C and Hib Vaccines

Hepatic Sequestration

Presentation/Investigation/Management

- Less common than splenic sequestration in children but treated in the same way.

6.2 Abdominal pain due to Biliary Colic / Sepsis / Girdle syndrome

Presentation/Investigation/Management

- Biliary colic+/- biliary sepsis or girdle syndrome
- Investigations include U+Es /LFT/amylase/USS abdomen /plain abdominal film
- Girdle syndrome presents with severe abdominal pain, abdominal distension, hepatomegaly, reduced bowel sounds, this should be managed as for ACS
- All biliary complications should be discussed with the paediatric surgical team
- Manage with IV fluids, IV antibiotics (ceftriaxone +/- metronidazole), analgesia, consider ERCP/MRCP

6.3 Acute Chest Syndrome

Presentation:

- A common cause of death and may be a postoperative complication in older children. Characterized by "T-shirt" distribution of pain, signs of lung consolidation (often bilateral), high fever, tachycardia, and tachypnea.
- Coughing is a late symptom.
- Physical signs may precede X-ray changes by up to 12 hours. However chest X-ray changes can also precede signs.
- **Falling Hb without evidence of splenic or hepatic sequestration is an indication for chest X-ray. A rapid deterioration requires urgent treatment.**
- **It is sometimes difficult to tell the difference between a chest syndrome and a chest infection, early transfusion is often appropriate and frequently life saving**.

Investigations:

- As per routine baseline investigations, in addition:
- HbS% (only new patients and those on transfusion programme)
- Capillary blood gas
- Chest X-ray
- Sputum and blood cultures
- Serum/urine for atypical screen (mycoplasma AB, urine pneumococcal antigen)

Management:

- Most important is prompt recognition of acute chest syndrome.
- IV fluids as in painful crisis (watch carefully for fluid overload and reduce fluids when exchange transfusion is completed).
- Pain management, consider fentanyl PCA (avoid hypoventilation from over sedation).
- Maintain target oxygenation (>94%) and monitor with pulse oximetry.
- Regular bronchodilators by nebuliser.
- Prevent further atelectasis using incentive spirometry, CPAP and PEEP, chest physiotherapy.
- Discuss with paediatric haemoglobinopathy team

- Early CICU / anaesthetic referral.
- Ventilatory support may be required.
- Treat underlying infection (Ceftriaxone 80mg/kg plus clarithromycin, see BNFC).
- Transfusion (as discussed below).

Transfusion:

- Decisions regarding transfusion are best guided by patient's clinical condition.
- The purpose of transfusion is to:
  - enhance oxygen-carrying capacity
  - improve tissue oxygen delivery
  - reduce HbS concentration to reduce sickling
  - prevent progression to acute respiratory failure
- Transfusion commonly results in impressive improvement within hours.
- Simple transfusion is indicated for patients with:
  - mild or moderate chest syndrome, particularly with falling Hb levels
  - aim for Hb level of no more than 10g/dL.
- Exchange transfusions are used to:
  - reduce the Hb S concentration rapidly;
  - whilst avoiding the problems associated with increased fluid volume and viscosity.
- Exchange transfusion is indicated when there is evidence of:
  - clinical deterioration
  - worsening x ray changes
  - suggested hypoxia (pulse oximetry less than 90%).
  - see guidelines on exchange transfusion.

6.4 Pnemococcal and Haemophilus Septicaemia/Meningitis

- Consider pneumococcal / haemophilus septicaemia or meningitis in any febrile sickle cell child.
- Treat with IV antibiotics without waiting for culture results.
- See antibiotic guidelines.

6.5 Malaria

- Needs urgent anti-malarial therapy appropriate to the zone of infection.
- Inquire about travel history.
- Contact Microbiologist or Hospital for Tropical Diseases (via switchboard) for up-to-date information.
- Transfusion is often necessary as haemoglobin may fall significantly due to increased haemolysis.

6.6 Aplastic Crisis

Presentation:

- Onset of profound anaemia over 1 – 3 days with reticulocytopenia and without sequestration.

- Due to transient marrow hypoplasia induced by parvovirus.

Investigation:

- As per routine baseline investigations, in addition:
- Parvovirus DNA and antibody titres.

Management:

- If there is no reticulocyte response or the patient is cardiovascularly compromised consider transfusion.
- Immunity appears to be lifelong.

6.7 **Pyrexia associated with chelation therapy**

- Patients on chelation therapy with desferrioxamine presenting with pyrexia and/or diarrhoea /abdominal pain should be treated for Yersinia Enterolitica with oral ciprofloxacin. Stool for culture and yersinia serology should be organised. Chelation therapy must be stopped.
- Note risk of neutropenic sepsis with deferiprone (agranulocytosis) and deferasirox (cytopenia).

7 ACUTE NEUROLOGICAL COMPLICATIONS (SEE APPENDIX 3 & 4 FOR FLOWSHEETS)

7.1 These may present as: acute severe headaches, transient ischaemic attacks, seizures or arterial ischaemic stroke.

Arterial Ischaemic Stroke

7.2 10% of children with Sickle Cell Disease will have a stroke before the age of 20 and 25% of this group will have evidence of ischaemic brain injury on MRI, even if they have been clinically asymptomatic.

Presentation:

7.3 There is often a significant delay between stroke onset and diagnosis in children usually because of failure to recognise the significance of the acute clinical presentation. The commonest clinical presentation of childhood arterial ischaemic stroke (AIS) is acute hemiparesis. About 20%of AIS is referable to the posterior circulation and here clinical signs may include ataxia, vertigo and vomiting. Seizures occur in 20% of children .

7.4 Acute neurological signs may not be clear cut in a child with AIS due to sickle cell disease , who present more commonly with ‘soft signs’ and there should be a low threshold to suspect the diagnosis in this group of children.

Any new neurological signs in children with sickle cell disease should be evaluated as potentially being a stroke

Investigations:

1. Brain MRI is recommended for the investigation of children with clinical stroke.
2. Brain MRI should be undertaken as soon as possible after presentation. If brain MRI will not be available within 48hrs, CT is an acceptable initial alternative.
3. Brain Imaging should be undertaken urgently in children with clinical stroke who have a depressed level of consciousness at presentation or whose clinical status is deteriorating.
4. Consider transthoracic cardiac echocardiogram within 48hours after presentation in all children with arterial ischaemic stroke.
5. Consider investigating for another underlying prothrombotic tendency.

Management: ABC

- Blood pressure should be maintained at an adequate level to optimise cerebral perfusion.
- Adapted from RCP guidelines:

1. All children with stroke should have regular assessment of conscious level and vital signs.
2. Urgent exchange transfusion should be undertaken to reduce %HbS to < 30% and raise haemoglobin to 10.0g/dl (avoid >11g/dl).
3. If the patient has had a neurological event in the context of severe anaemia (e.g. splenic sequestration or aplastic crisis), or if exchange transfusion is going to be delayed for more than 4 hours, urgent top-up blood transfusion should be undertaken.

Multidisciplinary assessment: Adapted from RCP guidelines

- As soon as possible after admission, all children following stroke should have an evaluation of:
  - swallowing safety
  - feeding and nutrition
  - communication
  - pain
  - moving and handling requirements
  - position requirements
  - risk of pressure ulcer
- All children affected by stroke should have a multidisciplinary assessment within 72hours of admission to hospital.

Secondary Prevention

1. Regular blood transfusion (every 3 to 6 weeks) should be undertaken to maintain the HbS% < 30% and the Hb 10 -11.0g/dl.
2. Transfusion may be stopped after 2 years in patients who experience stroke in the context of a precipitating illness, e.g., aplastic crisis and whose repeat vascular imaging and Trans Cranial Doppler velocities are normal at this time.
3. After 3 years a less intensive regime maintaining HbS < 50% may be sufficient for stroke prevention.
4. Those who cannot receive blood transfusions because of allo-immunisation, auto-antibody formation, lack of vascular access or non-compliance with transfusion or chelation may be considered for treatment with hydroxyurea (hydroxycarbamide).
5. Addition of aspirin (for dosage see BNFC), neuro-revascularisation procedures and bone marrow transplantation should be considered.

8 MANAGEMENT OF PRIAPISM

8.1 A sustained, painful and unwanted erection of the penis. The mean age at which priapism occurs is 12 years and by the age of 20, as many as 89% of males with SCD will have experienced one or more episodes of priapism.

8.2 Typically, priapism affects the corpora cavernosae, very rarely the corpus spongiosum may be affected. Penile ischaemia and acidosis begin to occur about 6 hours into a sustained priapic episode.

Triggers

- fever

- dehydration
- cold exposure
- full bladder
- REM sleep
- alcohol
- sexual arousal.

Stuttering priapism

- Recurrent
- Pain of variable intensity
- Erection lasting < 3 hours
- Penis may not be fully erect
- Low risk of cavernosal fibrosis and impotence
- Risk of subsequent fulminant attack

Fulminant priapism

- Severe pain
- Duration >3 hours
- Penis fully erect
- High risk of cavernosal fibrosis and impotence
- Urgent intervention indicated

General principles of management of priapism

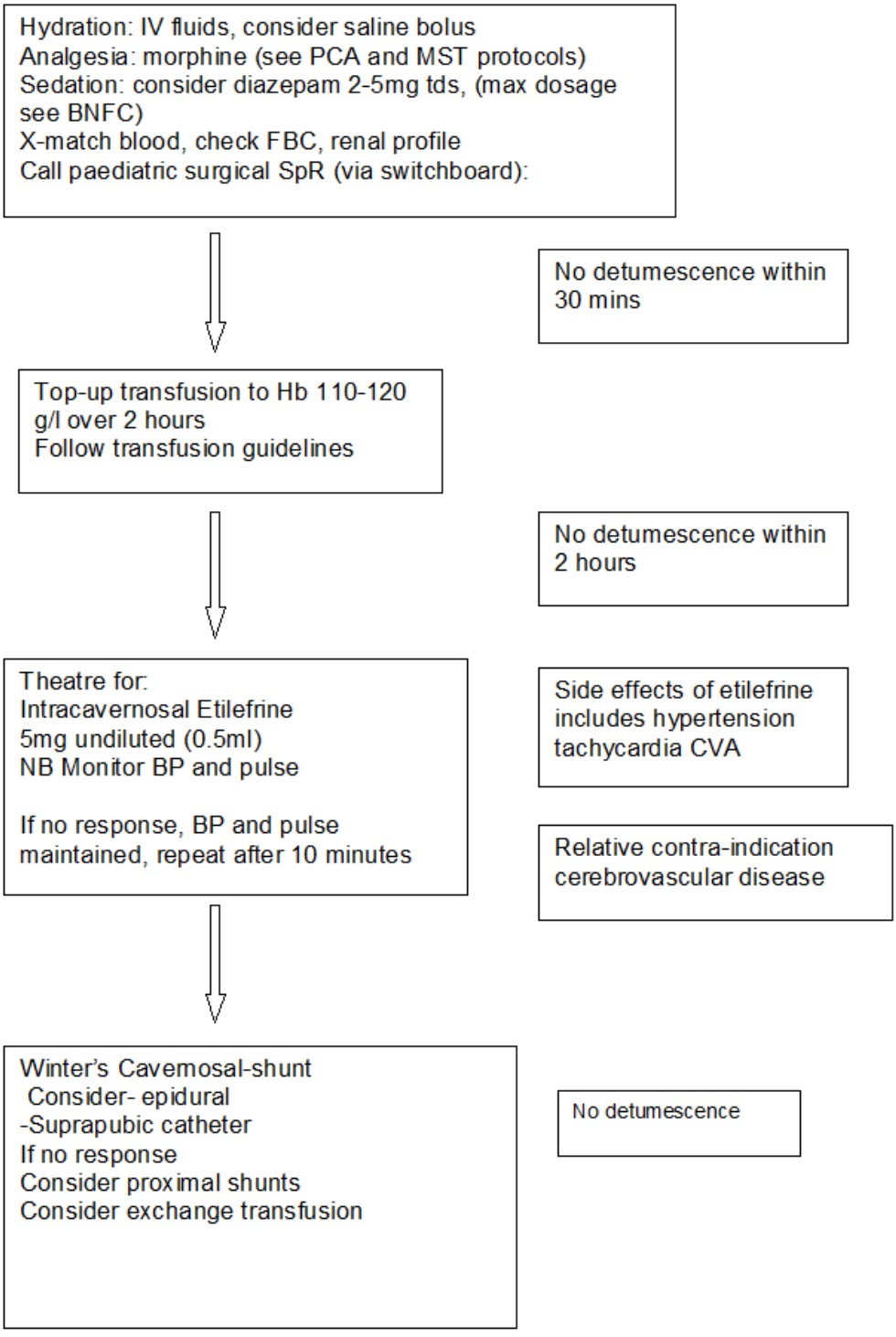
- Attempt to urinate
- Try warm bath
- Try Vigorous exercise
- Hydration
- Analgesia-usually parenteral opiates
- Sedation may be required in severe cases

Acute management of fulminant priapism (See diagram below)

General points

8.3 Many patients are not aware that priapism is a complication of sickle disorders and may be reluctant to discuss it. Stuttering priapism is under-diagnosed, symptoms should be specifically asked for at outpatient clinic visits.

8.4 It is vital to attend for treatment as early as possible. Delay may increase the risk of cavernosal fibrosis and impotence. Discuss WXH patients with Lead Clinician in hours or paediatric & surgical team out of hours.


Hydration: IV fluids, consider saline bolus
Analgesia: morphine (see PCA and MST protocols)
Sedation: consider diazepam 2-5mg tds, (max dosage see BNFC)
X-match blood, check FBC, renal profile
Call paediatric surgical SpR (via switchboard):
No detumescence within 30 mins
Top-up transfusion to Hb 110-120 g/l over 2 hours
Follow transfusion guidelines
No detumescence within 2 hours
Theatre for:
Intracavernosal Etilefrine
5mg undiluted (0.5ml)
NB Monitor BP and pulse
If no response, BP and pulse maintained, repeat after 10 minutes
Side effects of etilefrine includes hypertension tachycardia CVA
Relative contra-indication cerebrovascular disease
Winter's Cavernosal-shunt
Consider- epidural
-Suprapubic catheter
If no response
Consider proximal shunts
Consider exchange transfusion
No detumescence

Management of Stuttering Priapism

Indication: Stuttering Priaprism – prophylaxis and treatment

Diagnosed if: recurrent pain of variable intensity, erection lasts <3hours. Penis may not be fully erect, low risk of cavernosal fibrosis/impotence, risk of subsequent fulminant attack.

General point: It is vital to treat as early as possible to reduce the risk of cavernosal fibrosis and impotence.

Availability: Named patient medicine, hospital pharmacy only

- Preparations kept in Pharmacy
- Etilefrine injection 10mg in 1ml
- Etilefrine 25mg capsules
- Etilefrine 5mg tablets

8.5 **Dose: INJECTION**

Paediatrics age <16 years – Acute Fulminant Priapism, lasting >4hrs 5mg (0.5ml) undiluted injected by intra-cavernosal route.

Adults/>16yrs: 5-10mg undiluted, by intra-cavernosal injection under local anaesthetic (done by Urology team).

8.6 **Dose: ORAL**

Paediatrics - Treatment **and** prophylaxis

- 0.5mg/kg at night OR 0.25mg/kg bd (depending on when onset of symptoms is more common e.g. night only or throughout the day and night).
- Continue for a total 4 weeks and then stop/wean depending on recurrences.
- Monitor blood pressure (follow up at 1 week and then monthly).

Monitoring:

8.7 During therapy, patients should be seen weekly on acute assessment unit or outpatient clinic (or in local doctors surgery if prior agreement has been obtained) for assessment of response and of side effects.

8.8 Blood pressure should be checked and treatment stopped if blood pressure above 90th centile for age (See PEWS CHART) or experiencing increased headaches or any symptoms suggestive of TIA.

Other treatment options

8.9 For prophylaxis against stuttering and fulminant episodes consider:

- Flutamide 250 mg tds
- Casodex 150 mg OD
- Zoladex 3.6 mg s/c monthly

Note: that these treatments are expected to cause impotence for the duration of therapy. For more information on Etilefrine see appendix 5. All patients should be referred for urology input.

9 PROTEINURIA / HAEMATURIA

Definition

- Early morning albumin / creatinine ratio > 3.0 mg/mmol or > 1+ proteinuria in a dilute urine specimen.
- Nephrotic syndrome: proteinuria ≥ 3+ on dipstick (UA/UC >200mg/mmol), oedema, plasma albumin <25g/l, ± hyperlipidaemia.
- Haemastix positive does not necessarily mean there will be proteinuria.

Aetiology

- Transient, orthostatic, glomerulonephritis, nephrotic syndrome, tubulo-interstitial disease.

Sickle Cell Nephropathy

- Possibly due to mesangial phagocytosis of sickle cells, an immune complex mediated process, glomerular hypertrophy or glomerular injury by hyper filtration. Kidneys show focal segmental glomerulosclerosis. May progress to full blown nephrotic syndrome and end stage renal failure.

Investigations:

Urine

- urine albumin/creatinine ratio on first morning urine and pm ambulatory sample
- urine microscopy and culture
- urinalysis for blood and glucose

Blood

- FBC, renal profile

Radiology

- Renal tract ultrasound

Once orthostatic proteinuria has been excluded

- ESR , EMU osmolality
- Autoimmune/ vasculitic screen
- DMSA scan

Management:

- STOP NSAIDS, review all medication. Use codeine phosphate instead for mild/moderate pain, monitor BP

When to refer to Nephrology and indications for biopsy

- Duration > 6 months
- Excretion > 1 gm / 24 hours
- Family history of renal disease
- Macroscopic haematuria
- Sustained hypertension
- Renal failure

- Hypocomplementaemia
- Age less than 1 year or greater than 10 years
- Hypertension, proteinuria, increasing severe anaemia, and haematuria predict renal failure in Sickle cell patients.

10 BONE AND JOINT PROBLEMS

Osteomyelitis causative organisms:

- *Salmonella species*
- *Staph aureus, Haemophilus influenzae*
- *E-Coli and other Gram -ve bacteria (such as Klebsiella spp.)*
- *Enterobacter spp*
- *Mycobacterium spp*

Diagnosis of osteomyelitis:

10.1 This is difficult in sickle cell disease. Signs and symptoms could be similar to acute pain crisis. Clinical distinction can be difficult especially with the increased use of antibiotics in painful vaso-occlusive crises.

Clinical Features:

10.2 These usually include: pain, swelling, tenderness. Usually the child is systemically unwell. The commonest sites are the femur, tibia, humerus. Remember that fever may be modest. Presentation could be acute or occur over a period of a few weeks. Suspect osteomyelitis if pain is unusual and does not resolve as expected.

Investigations:

- FBC, CRP , aerobic /anaerobic blood cultures
- Stool samples (Salmonella)
- Throat swabs for M,C&S
- NPA for virology if viral infection is suspected
- Bone, pus or tissue samples should be sent to Microbiology for M, C&S and FB, and for 16S rDNA molecular studies.

Radiology:

10.3 **X-ray:** Early changes include osteopenia and periostitis , periosteal reactions. Similar changes are also seen in acute pain crisis. Early X-rays are then of limited value. X-ray changes do not appear until about 10 days after infection.

10.4 **Ultrasound:** Rapid, non- invasive and easy to target areas of maximum pain. Changes are non-specific and findings are similar to those seen in acute pain crisis.

10.5 **MRI:** Useful in monitoring treatment.

10.6 No single imaging technique can reliably distinguish acute infection from infarction.

Treatment:

10.7 See under antibiotics section

10.8 Under no circumstances must surgery be contemplated without prior discussion with the on call consultant. Most patients will require blood transfusion prior to general anaesthetic – see policy for general anaesthesia.

Avascular Necrosis of the Shoulders and Hips

10.9 This complication should be suspected particularly in older children with persistent pain affecting the hip, shoulder, knee, leg or groin. Pain may be worse on movement though also occurs at rest. Often there is restriction of movement at the hip and shoulder joint.

Diagnosis:

10.10 An x-ray should be considered in those patients with persistent / prolonged or recurrent pain.

10.11 MRI may show early changes.

Management:

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10.12 Analgesia, adequate rest, avoidance of prolonged weight bearing.

10.13 Refer for physiotherapy, consider hydrotherapy.

10.14 Programme of gradual non weight bearing exercise – particularly swimming, cycling.

10.15 All cases should be discussed and referred for orthopaedic assessment.

10.16 Consider review of Hydroxyurea (hydroxycarbamide) therapy if thought to be precipitated/exacerbated by this.

10.17 Consider transfusion programme in those requiring surgery, debilitated by pain or restricted movement as may prevent progression of damage. Review transfusion programme at 6 monthly intervals.

11 EYE PROBLEMS

11.1 Patients should be made aware of sickle cell related eye complications. Routine ophthalmological screening is not indicated, however community opticians review should be encouraged. Patients / parents should be advised to report changes in visual acuity, altered / distorted vision, presence of floaters as a matter of urgency and referral made to Paediatric ophthalmology clinic for assessment.

11.2 All patients on iron chelation therapy should have annual screening in the ophthalmology clinic for chelation associated cataract / retinopathy formation, see chelation guidelines.

12 TRANSFUSION IN SICKLE CELL DISORDERS

12.1 See haemoglobinopathy transfusion guidance also.

12.2 Anaemia alone in an otherwise well child is not an indication for transfusion unless Hb falls to 5 g/dl or lower, in which case discuss with the haematologist with details of previous results. Check reticulocyte counts. Use Kell compatible, Rhesus compatible blood matched for antibody status.

Simple or 'top-up' transfusion:

12.3 Indicated for acute anaemia e.g. aplastic, sequestration crisis or acute bleeding. To calculate volume of packed cells required use guide below:

- If pre-transfusion Hb 9.5-10.0g/dL transfuse 10ml/kg
- If pre-transfusion Hb 9.0-9.4g/dL transfuse 12ml/kg
- If pre-transfusion Hb <9.0g/dL transfuse 15ml/kg

Note:

- Do not transfuse to above 11g/dl,
- The volume transfused should be capped at 3 units for children > 50kg
- Document indication for transfusion clearly in notes

Chronic transfusion programme in day unit setting:

12.4 **Definition:** Repeated transfusions to keep Hb S < 30% over a period of time.

12.5 Transfuse at 3 to 4 week intervals to suppress erythropoiesis and keep Hb S <30%. Aim for Hb no more than 11g/dl .

12.6 Patients vary in the frequency and amount of blood required to suppress Hb S production. In children with SC disease it is usually necessary to start with an exchange transfusion. In other children with HbSS (particularly those with a high initial haemoglobin level) exchange may be necessary at times.

Prior to commencing transfusion programme potential complications of transfusion must be discussed and documented.

- Transfusional iron overload.
- Transfusion transmitted infection – aim for Hepatitis B /C + HIV screen prior to starting.
- Transfusion reactions.
- Ensure hepatitis B vaccination status satisfactory.

Monitor:

- Hb, Hb S%, Red cell alloantibodies monthly
- Serum ferritin every 3 months
- Start iron chelation when Serum ferritin is 1000ug/L or after 10 transfusions (See Chelation protocol, appendix 5)
- Annual viral screen (HIV, hepatitis B & C) + check immunity to Hep B vaccination –anti-HBs Ab (done in January every year)

13 POLICY FOR GENERAL ANAESTHESIA

13.1 All children with SS and SC disease should be discussed with the Paediatric Haemoglobinopathy team and Anaesthesia when they are booked for surgery so that a coordinated plan can be made for their care.

13.2 It is the responsibility of the surgical team to ensure a bed is organised and the patient admitted.

13.3 These patients should be scheduled early on the operating list to ensure they are not likely to be cancelled, and to avoid prolonged fasting time.

13.4 Care should be taken to avoid factors which may precipitate the development of a crisis. These include hypoxia, dehydration, acidosis, cold and pain. The majority of crises in the perioperative period occur postoperatively.

13.5 All patients should receive overnight IV hydration the evening prior to surgery requiring general anaesthetic.

13.6 All transfusions will be organised by the paediatric haemoglobinopathy team.

13.7 A valid G & S sample must be organised for all patients. A HbS % is not necessary pre surgery unless exchange transfusion is required (see below).

Transfusion Recommendation for Elective Surgery:

13.8 A three tiered approach is recommended:

1. **Low risk surgery** short procedures with minimal risk of perioperative complications, e.g. grommets or GA for scans in children who have no other risk factors:
   - **Hb should be ≥ 7g/dl.**

2. **Intermediate risk surgery:** tonsillectomy, splenectomy, laparoscopic cholecystectomy, hip/knee replacements. History of obstructive sleep apnoea. Children with a history of recurrent chest problems or other chronic health problems:
   - **Simple transfusion to an Hb of 11g/dl, regardless of HbS levels.**

3. **High-risk surgery:** thoracic, major upper abdominal surgery or neurosurgery and children with a history of severe sickle related problems e.g. previous CVA. Consider in eye surgery, surgery involving tourniquets:
   - **Transfuse or exchange transfuse to reduce the HbS level to <30%.**
   - **Total Hb should not be > 11g/dl.**

13.9 Children with sickle cell disease may be difficult to exchange transfuse and will require early consultation. Exchange transfusion is difficult to perform, particularly in small children and should not be carried out unless absolutely necessary.

13.10 Children and their parents should be involved in the decision to transfuse whenever possible.

13.11 Children requiring emergency surgery should be treated similarly if time allows. If this is not possible, blood should be cross- matched and standing by in case of perioperative problems requiring emergency exchange transfusion.

13.12 All patients must be discussed with the Haematology team at presentation.

14 COMMON SURGICAL PROCEDURES

Adenotonsillectomy

14.1 This is normally indicated for children with tonsillar / adenoidal hypertrophy and confirmed obstructive sleep apoea (OSA) demonstrated on sleep study.

14.2 The input of the ENT team and respiratory teams will be required when planning for this procedure.

14.3 Children with severe OSA and/or additional risk factors (eg cerebrovascular disease, chronic lung disease etc) should also be discussed with CICU for management prior to the procedure.

Laparoscopic cholecystectomy

14.4 This is indicated for children with recurrent biliary colic,/ cholecystitis.

14.5 A thromboprophylaxis risk assessment should be carried out and treatment instituted as appropriate, (see trust thromboprophylaxis policy).

Splenectomy

14.6 This is normally indicated for children with recurrent splenic sequestration (>2 episodes requiring transfusion therapy or 1 life threatening episode) or chronic hypersplenism. Cholecystectomy could be considered at the same time for those children with recurrent biliary colic / cholecystitis.

14.7 A thromboprophylaxis risk assessment should be carried out and treatment instituted as appropriate, (see trust thromboprophylaxis policy).

14.8 If not previously vaccinated, 1 month prior to scheduled splenectomy the patient should receive pneumococcal, HiB conjugate, Men C conjugate. 5 yearly pneumococcal vaccination and penicillin prophylaxis should be recommended.

14.9 Thrombocytosis is common post splenectomy. For sustained platelet counts >1000x10^9 low dose aspirin may be considered.

15 HYDROXYUREA (HYDROXYCARBAMIDE)

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. There is accumulating experience of its use in children, and a significant benefit in survival has been demonstrated a significant benefit in survival in those given hydroxyurea.

15.1. Hydroxycarbamide is an S phase-specific cytotoxic agent, which has been used for many years to treat myeloproliferative conditions. It has also been used to treat children with secondary polycythaemia. Its side effects include bone marrow suppression, gastro-intestinal disturbances, and increased skin and nail pigmentation. It is potentially tetatogenic and there is concern about the risk of malignancy.

15.2. The mechanism of action is not certain. Three effects may be important:

- Increase in fetal haemoglobin content within the red cell, inhibiting sickle haemoglobin polymerisation.
- Decreased adhesion molecule expression on the surface of the red blood cell, reducing red cell-endothelial adhesion.
- Reduction of white cell and platelet count. This may also impair the sickle cell/endothelial interaction and reduce inflammatory process in the microvasculature.

Indications for use

- Offer to all patients with homozygous sickle cell disease, sickle beta thalassaemia, haemoglobin SC disorder as has shown significant benefit in survival

PREVIOUS INDICATIONS REMAIN IF PATIENTS NOT ON HYDROXYUREA

- Recurrent painful crises, significantly interfering with lifestyle. In practice, this would be more than three hospital admissions with painful vaso-occlusive crises per year
- Recurrent chest crises, two or more a year
- Avascular necrosis
- Consider also in those children with high risk transcranial Doppler velocities where transfusions are not possible.

Exclusions

- Anaemia with Hb <6g/dl at baseline
- Platelets <100x10 /dl, Neutrophil count <1x10 /dl
- Decompensated cardiac, renal, liver or pulmonary disease

Procedure Prior to Starting Therapy

15.3. The benefits and hazards of using Hydroxycarbamide should be considered for each individual patient, and discussed with the patient and parents on 2 separate occasions. Give detailed explanation of treatment, including nature of possible side effects, and a copy of the Patient Information Leaflet. Where appropriate, discussion about the risks of becoming pregnant or fathering a child while on hydroxycarbamide should occur

15.4. Sperm banking should be considered in older male adolescents

15.5. Ensure that the patient is willing to attend regularly to monitor blood counts

Baseline investigations

- Height and weight centile. Tanner stage
- FBC, reticulocytes
- Renal, liver, bone profile

Dosage

- Start at 15mg/kg, as a single dose
- Dispense no more than 2 weeks of tablets at a time
- Dose can be increased by 5mg/kg every 8 weeks
- Ideally aim for dose that controls symptoms while maintaining neutrophils 1.5 x 109/L and Platelets> 150x109 /L
- Continue regular penicillin V and folic acid

Monitoring of Therapy

Weekly: FBC for first 4 weeks, then 2 weekly for next two months. Then monthly when dose stable. Monitor 2 weekly if dosage changed.

Monthly: FBC (when on stable dose), HbF, reticulocyte count, biochemistry, toxicity recording, compliance monitoring.

Three-monthly: clinical assessment: frequency of crises, other adverse sickle events, height and weight centile, Tanner stage, toxicity recording, compliance monitoring

Dosage Modification

- If neutrophils <1.5x109 /L, platelets <100 x109 /L, Hb>2g/dl below baseline - stop medication
- Recheck FBC weekly
- When counts back to normal, restart hydroxycarbamide 5mg/kg below previous dose and continue to check FBC weekly until count remains stable
- **May need to be stopped during an acute admission when sepsis suspected**

Duration of Therapy

15.6. Assess clinical response, HbF% and adverse events at 3 and 6 months.

15.7. If HbF increment >2.5 fold, continue therapy.

15.8. If increment, <2.5 and/or clinical response not apparent after 6 months, discuss discontinuation of therapy with the patient and parents

16 OUTPATIENT MANAGEMENT

The Aims of the Clinic are to:

- Monitor progress of the children: medical, educational and psychosocial.
- Establish baseline observations for comparison in acute illness.
- Educate parents and children in the management of sickle related disease.

New Patients

- Usually referred following neonatal screening, but also patients presenting from overseas.

Registration:

16.1 **FBC, RETICULOCYTE COUNT**

16.2 Haemoglobinopathy screen (sickle solubility test, Hb electrophoresis, Hb A2 and F estimation By HPLC), G6PD screen.

16.3 Blood group, red cell phenotype

16.4 Take full family history including names, ages and address(es) and plans for future children.

16.5 Parents and all siblings should have a full blood count, haemoglobinopathy screen (if not already performed)

16.6 Explain to parents the probable diagnosis and its implications, including genetic counselling.

16.7 Issue haemoglobinopathy card and demonstrate splenic palpation

16.8 At each visit the sickle cell clinic proforma should be followed

16.9 Measure height and weight document on growth chart, pulse oximetry, examine the child including for palpable splenomegaly.

16.10 Faltering growth should be managed with the nutrition team and nutritional intervention considered.

16.11 Document any sickle-related or other illness since last visit, immunisation status, school progress and attendance and holiday plans. Ask about bed-wetting, priapism (boys). Sleep study should be organized for symptoms of upper airways obstruction.

16.12 Enquire about learning/ behavioural concerns, discuss at psychosocial MDT. Healthy lifestyle advice should be given.

16.13 Ensure regular supply of penicillin V, folic acid provided by GP. Discuss use of analgesics and ensure GP prescribes supply.

Arrange follow up:

- 3 monthly until age 12 months
- 4 – 6 monthly ages 1-3
- 6 monthly thereafter, unless clinical indication for earlier review
- 12 monthly for children with HbSC disease if clinically stable

Screening investigations

16.14 Transcranial doppler screening (TCD) should be organised for all children with homozygous sickle cell disease or HbS/thalassaemia over the age of 2 at least annually (more frequently depending on results – see TCD screening protocol).

16.15 Screening for pulmonary hypertension should occur 5 yearly from age 15 with echocardiogram (request TR jet velocity). An echocardiogram might be required earlier than this for investigation of hypertension, unexpected heart murmur, disproportionate cardiomegaly or on the advice of the cardiologist. Screening for chronic sickle lung disease should be organised 5 yearly from age 15. Those patients with recurrent acute chest syndrome or asthma should be referred for respiratory input.

16.16 Screening for sickle nephropathy should be organised annually with urinalysis from age 15 and urine protein creatinine ratio (UPCR) organised for those with significant proteinuria. See table below for other screening investigations.

Communication with patient / MDT

16.17 Copies of correspondence should be sent to patient, GP and community services. This will have contact details of the lead consultants. Patient information sheet detailing services provided and list of contacts should be made available to new patients. Patient information should be made available covering discussion in clinic e.g. TCD.

Annual review

16.18 This should be done by a member of the Haemoglobinopathy Team

16.19 It should include review and documentation of acute episodes and complications over the previous year, current medication, transfusion parameters, iron monitoring and iron chelation therapy for those on regular transfusion

16.20 Checking to ensure that routine screening investigations and vaccinations are up to date.

16.21 Discussion of disease-related issues relevant to the patient and family

16.22 If appropriate, a discussion of treatment options including hydroxyurea, transfusion and bone marrow transplantation.

16.23 Routine investigations at annual review are shown in the accompanying table.

16.24 The relevant information is prompted by use of the out-patient clinic proforma contained on ICE

Policy for DNA

16.25 Patients who DNA on 3 consecutive occasions or young infants on the first appointment should be followed up by the community team and a letter sent to GP and patient. Referral of children who move to another region should be organised by the specialist centre and community teams ensure appropriate community input.

17 PREVENTION OF INFECTION IN CHILDREN

17.1 Antibiotic prophylaxis for prevention of pneumococcal disease is mandatory

17.2 **ALL** children with Sickle Cell Disease (including HbSS, HbSC, HbS/Beta thalassaemia) are prescribed Penicillin V by the age of 3 months. This should be taken lifelong.

Dosage:

- Birth to 1 year
- 3 months to 5 years 125 mg Penicillin V suspension b.d.

- 5 years and onwards 250 mg Penicillin V suspension/tablets b.d.

17.3 Try to get the children taking tablets as soon as possible (crushed and mixed with some fruit juice). Pharmacies may be prepared to prescribe the dry suspension, or to receive a batch of request prescriptions from the surgery to avoid collecting a repeat prescription for the suspension every week.

17.4 For those children who are genuinely allergic to Penicillin, Erythromycin is prescribed instead.

Dosage:

- < 2 years 125 mg Erythromycin o.d.
- 2-8 years 250 mg Erythromycin o.d.
- > 8 years 250-500 mg Erythromycin o.d.

17.5 Prescriptions should be given by GP.

Pneumococcal immunisation:

Conjugate Pneumococcal vaccine = Prevenar (PCV)

17.6 This is part of the universal immunisation programme and will be offered routinely at two, four and thirteen months of age.

Pneumovax

17.7 All children with Sickle Cell Disease should receive Pneumovax at 2 and 5 years and 5 yearly thereafter (to be given by GP services) Those children > age 5 not vaccinated should receive 1 dose of prevenar followed by pneumovax 1 month later.

17.8 Children < age 5 not vaccinated: follow BNFC prevenar catch-up guidelines.

Routine childhood immunisation programme:

17.9 Follow standard NHS guidance

Prevention of Haemophilus Influenza type b:

17.10 Older children (over 1 year) who have not previously been immunised should be offered a single dose of Hib vaccine (to be given by GP services)

Prevention of Meningococcal disease:

17.11 If missed, give 2 doses 1 month apart between ages 4 months and 2 years.

17.12 Other children and adults who have not previously been immunised should receive a single dose of Meningococcal C conjugate vaccine.

17.13 If traveling to countries where Meningitis A is endemic they should be offered quadrivalent meningitis A, C, Y, W135 vaccine.

17.14 All given by GP services

Influenza vaccine:

17.15 Should be given every year in October/November. To be given by GP.

Malaria prevention:

17.16 Children with Sickle Cell Disease are at increased risk of severe malaria if travelling to endemic areas.

17.17 Many children in Leicester visit West Africa. Parasite resistance to standard prophylaxis is common and specific advice is especially important. GP to supervise prophylaxis.

17.18 Regimes vary depending on destination and resistance patterns (consult Hospital for Tropical diseases or Travel clinic).

17.19 Chemoprophylaxis should be started before travel and be continued after return. G6PD status should be checked before starting.

Hepatitis B vaccination:

17.20 All non immune children with Sickle Cell Disease should be vaccinated against Hepatitis B. Ideally this should start at 12 months of age. Those children not born in the UK should be vaccinated at the earliest opportunity.

17.21 Vaccination to be three injections at 0, 1 and 6 months (to be given by GP service)

17.22 Vaccinations are recorded in the hospital records and the Child Health Records Booklet.

18 BONE MARROW TRANSPLANTATION

18.1 Bone marrow transplantation should be discussed as a curative intervention for all patients.

18.2 Tissue typing of patients, parents and siblings should be carried out if clinical indication for transplantation.

18.3 Bone marrow transplantation could be considered for patients on sickle modifying intervention

- for primary or secondary stroke prevention
- recurrent acute chest syndrome
- avascular necrosis
- recurrent severe acute pain / chronic pain
- considered for parental preference

18.4 A thorough discussion and explanation of BMT procedure, benefits and risks should be offered to parents of Children with a clinical indication for BMT and histocompatibility testing of siblings organised on Ward 27 Oncology Daycare

18.5 If there is willingness to proceed, the child and family should be formally referred to Birmingham Childrens Hospital, giving summary of medical condition, relevant investigations, and a copy of the HLA typing results.

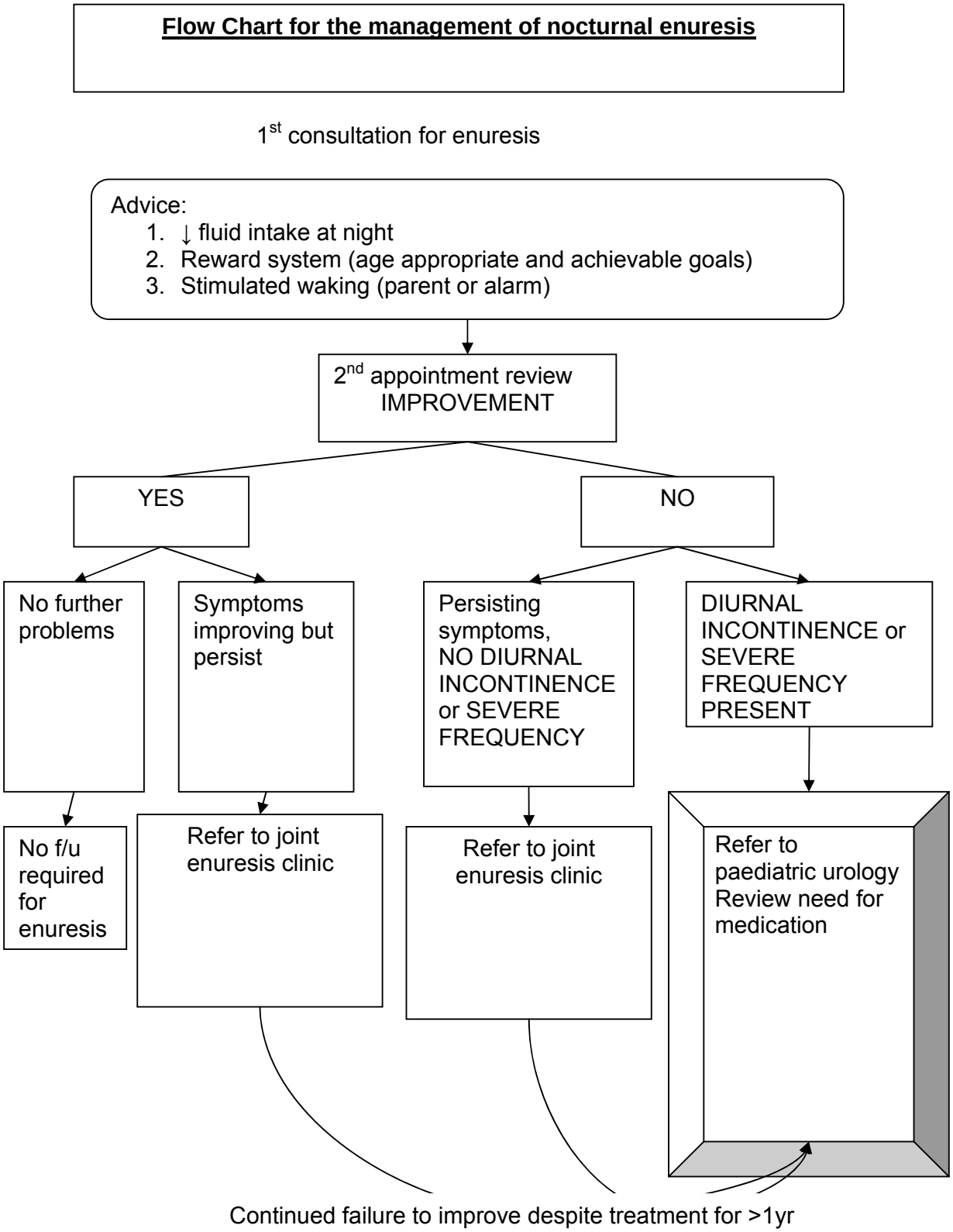
Adolescent Transition

18.6 Please see trust and haemoglobinopathy transition policy.

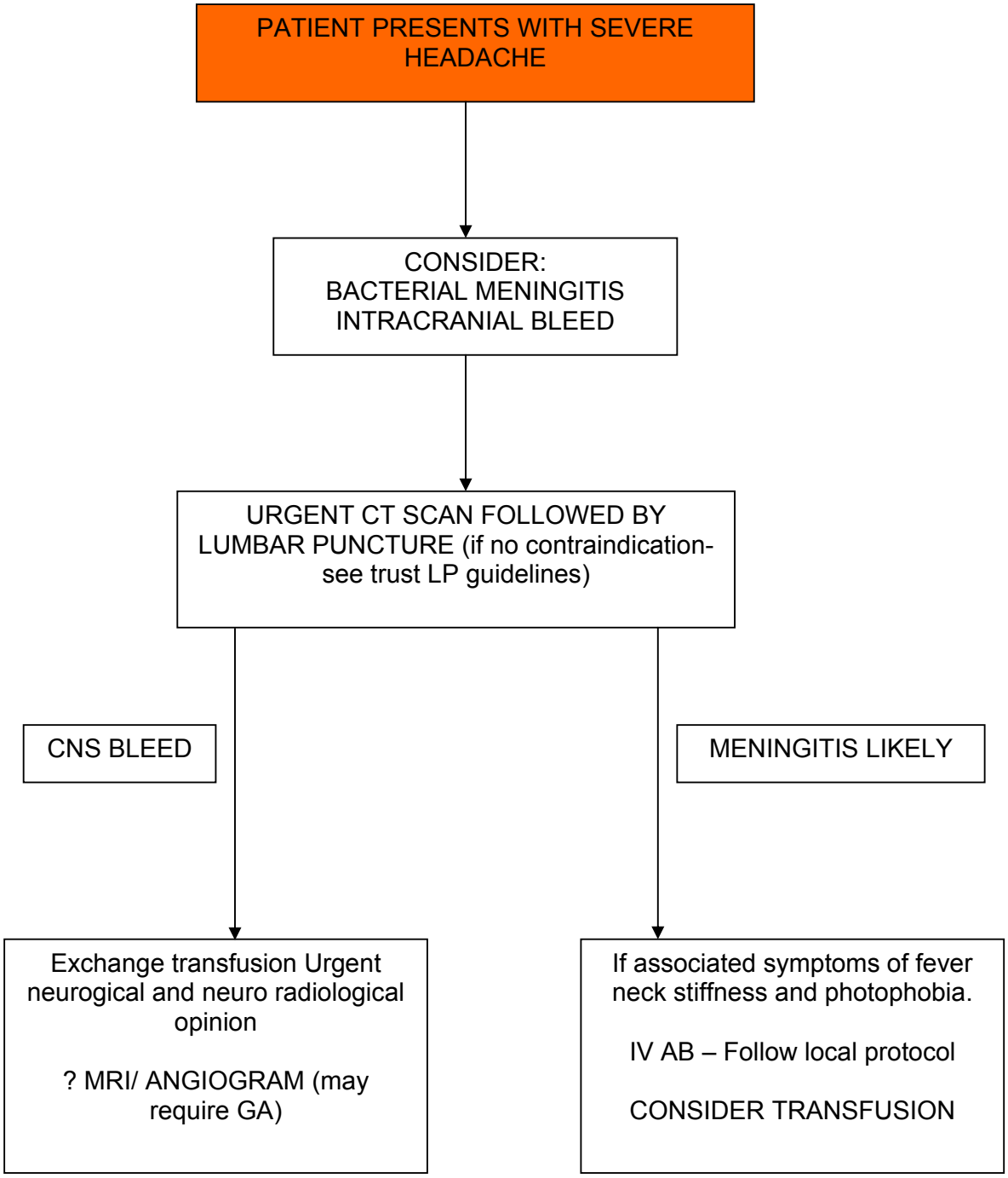
Appendix 1: Annual Investigations Table

INVESTIGATION OR INTERVENTION	IST APPT	1YR	2YRS	3YRS	4YRS	5YRS	6YRS	7-11YR, ANNUAL	12YRS	13YRS	14YRS	15YR	16-19 YRS
FBC/ Retics/ Biochem/ LDH	●	●	●	●	●	●	●	●	●	●	●	●	●
HbF level		●											
G6PD level	●												
Blood group and full red cell phenotype	●												
HepBSAg, HepC Ab		●											
Transcranial dopplers			●	●	●	●	●	●	●	●	●	●	●
Urine Prot/Creat ratio												●	
Pulmonary function tests/ ECHO / R heart pressure/ TR jet velocity												●	
Immunisation													
Prevenar	●	●											
Pneumovax			●			●		● Age 10				●	
Hepatitis B		● full course											

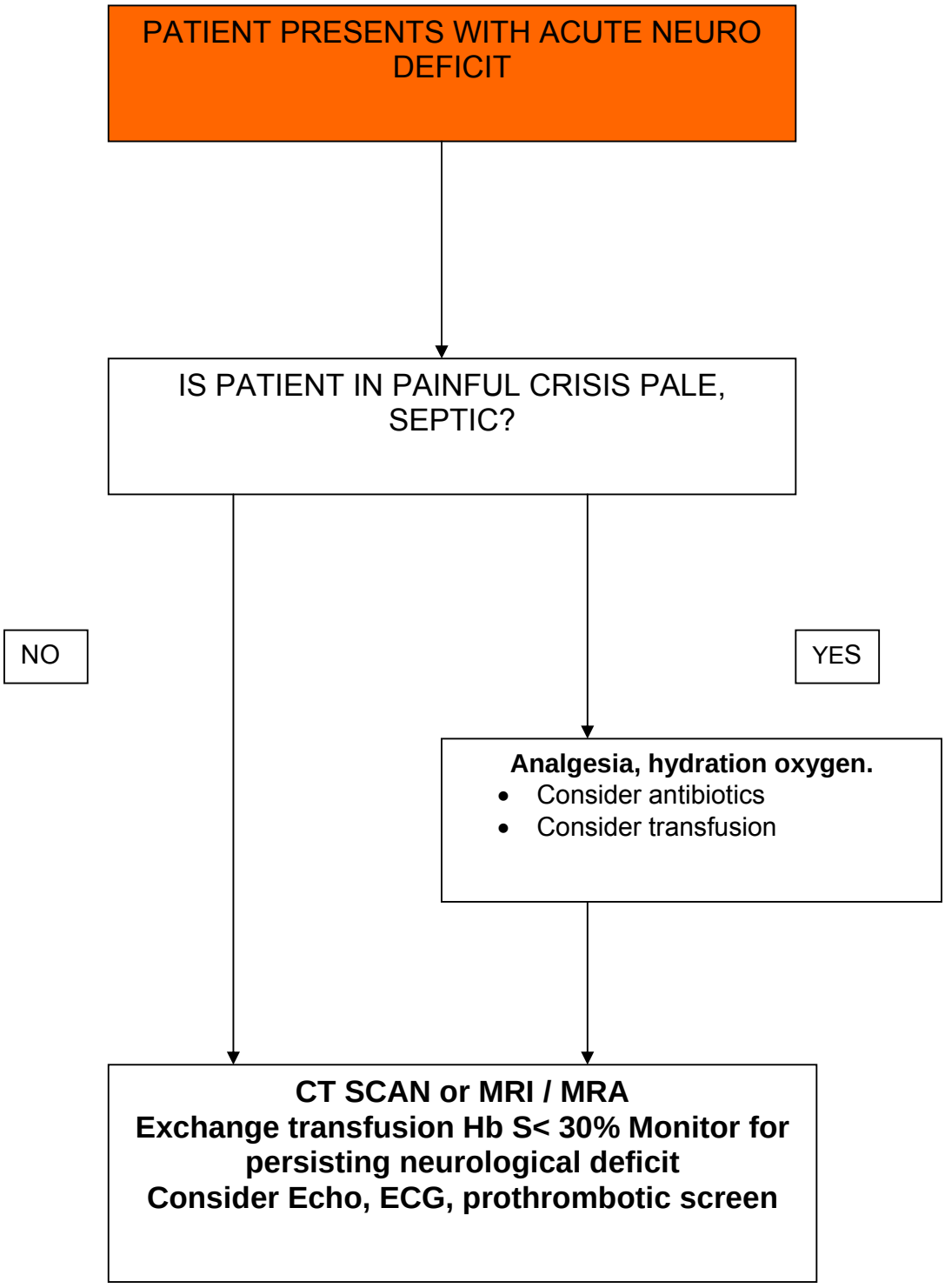
Appendix 2: Management of Nocturnal enuresis



Appendix 3: Flow chart of patients presenting with severe headache



Appendix 4: Acute Neuro Deficit



Appendix 5: Supporting Documents (see separate guidelines)

- **Paediatric Haemoglobinopathy transfusion**
- **Guidelines for monitoring and treatment of iron overload**
- **TCD Scanning policy**
- **Children Acute Sickle Pain Pathway**

DEVELOPMENT AND APPROVAL RECORD FOR THIS DOCUMENT		
Author / Lead Officer:	Dr Krishna Kotecha	**Job Title:** Consultant
Reviewed by:	Childrens Clinical Practice Group	
Approved by:	**Childrens Clinical Practice Group**	**Date Approved: March 2016**

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