Clinical Guidance

Guidelines for Outpatient Management of Adults with Sickle Cell Disease and Thalassaemia

Summary
Guidance for clinicians reviewing patients with a diagnosis of haemoglobinopathy in the outpatients. What to do for routine monitoring and annual reviews. These comprehensive guidelines are intended for use as a reference for medical, nursing staff and all health care professionals.

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Adult sickle cell team based in:
Department of Haematology,
4th Floor, Southwark Wing,
Guy's Hospital, Great Maze Pond, SE1 7EH
Telephone: 02071887188
To arrange or cancel appointments) - 0207188 2743/2724

Medical team
Consultant         Dr J Howard       Ext 81432
Consultant         Dr R Kesse-Adu    Ext 52715
Specialist Registrar Bleep 0248
SHO                Bleep 2283

Nursing team
Advanced Nurse Practitioner
Mr Neill Westerdale Bleep 1843 / Ext 82710 / Mob 07770683947
Clinical Nurse Specialist (Teenagers and Young Adults)
Mr Luhanga Musumadi Bleep 2256 / Ext 82710 / Mob 07770678851
Clinical Nurse Specialist
Ms Judith St Hilaire Bleep 2868 / Ext 82710 / Mob 07920711266
Research Nurse
Ms Claire Woodley Ext 51152

Psychology Team
Consultant Health Psychologist Dr Nicky Thomas ext 82725
Clinical Psychologist Dr Heather Rawle ext 82718
Health Psychologist Ms Mina Abedian ext 82718

Other numbers
P.A. to Dr Howard/Dr Kesse Adu (Ms Anne Oddotte) 81432
Haematology Clinic 82743 / 82724
Haematology Day Unit 82727
Out of hours Haematology SpR on call via switch (02071887188)

Liaison physicians
Obstetrics Mr Eugene Oteng-Ntim – Ex 86876, pager 851183
Midwifery team: Thames Team – pager 822161
Orthopaedic Mr Marcus Bankes – Ex 84470
Orthopaedic CNS: Michelle Finlayson – Ex 84470, bleep 2077
Renal: Dr Cormac Breen – Ex 85709
Ophthalmology: Mr Moin Mohamed
Neurology: Dr Paul Holmes – Ex 83962
Transition: Dr Baba Inusa – Ex 84676
Respiratory: Dr Nick Hart
Chronic pain team: Dr Doug Justins
Audiology: Mr Presanna Premanchandra, Principle Audiological Scientist, Audiology dept, Guys
Tissue Viability Team: Bleep 1277/24: Pager 84312/817424: Ext 82518/Fax 86642

Sickle Community Team
South East London Sickle and Thalassaemia Centre, Wooden Spoon House,
5 Dugard Way, Off Renfrew Road, Kennington, SE11 4TH
Tel: 0203 049 5989 / 0203 049 5993 Fax: 0203 049 6069 / 85
Outpatient Management of Sickle Cell Disease

Organisation of Outpatient Care

Referrals to the haemoglobinopathy clinic should be made to Dr Jo Howard.

The clinics are held in Haematology 2 (4th Floor, Southwark Wing, Guy’s Hospital) on:
- Tuesdays 1400-1700
- Fridays 0900-1200

The clinic is attended by:
- The medical team
- Clinical Specialist Nursing team
- The Psychology team
- Community sickle team.

The aims of regular clinic review are to:
- Monitor medical and psychosocial progress
- Establish baseline observations for comparison in acute illness
- Educate patients in the management of sickle-related problems
- To encourage adherence to treatment - particularly prophylaxis and immunisation programmes

Frequency of outpatient attendances
The majority of patients will be seen every six months.
All patients should have an annual review once a year
Some patients will have shared care and will be seen at GSTT once a year, and at their local centre for their other visits.

Patients who should be seen more frequently than six monthly
- On hydroxycarbamide (two monthly)
- Thalassaemia patient on transfusion (4 monthly)
- Renal dysfunction
- Chronic pain, on long term opiates
- Other chronic health issues
What happens in the Sickle Cell Clinic?

Patient arrives and books in at the reception desk.
Patient details are checked by receptionist
Patient information leaflets are given if appropriate
Notes are given to phlebotomist

Patient is given urine pot and asked to provide sample

Patient is called by phlebotomist and blood samples are taken.
Notes are put in the ‘waiting to see doctors’ pile

Patient is called by health care assistant and observations are taken,
and the record is given to the patient.

Patient is called to see doctor

Further reviews as appropriate:
Patient is reviewed by acute nurse specialist
Patient is reviewed by psychologist
Patient is reviewed by community nurse
Patient is reviewed by research nurse

Patient is given follow-up appointment and books this with receptionist
New Sickle Cell Patients

New patient referrals may be received from the transition service, general practitioners, other hospitals, other departments in the hospital or following inpatient attendances.

First consultation:

Document full personal and family history to include:
- Date and place of diagnosis
- Frequency and severity of pain episodes
- Previous hospital admissions
- Any previous ITU admissions
- Any other sickle complications
- Surgical history
- Transfusion history
- Any children, siblings or parent with sickle cell disease
- Full past medical history

Discuss lifestyle issues:
- Alcohol/smoking/other drugs
- Housing
- Education/college/work

Discuss acute and chronic complications of sickle cell disease including:
- Pain management
- Fever
- Acute chest syndrome
- Stroke
- Osteomyelitis
- Leg ulcers
- Avascular necrosis
- Renal dysfunction or proteinuria
- Renal papillary necrosis
- Priapism
- Pulmonary hypertension
- Headaches
- Retinopathy
- Cholecystitis
- Splenic and hepatic sequestration
- Aplastic crisis

Discuss medications and vaccinations:
- Infection prophylaxis (penicillin V 250mg bd or alternative)
- Folic acid (5mg od)
- Hydroxycarbamide
- ACE inhibitors
- Analgesia used at home and on hospital admission
- Standard vaccinations
- Pneumovax, flu vaccine, hepatitis B

Genetic counselling:
- Discuss inheritance
- Family screening if appropriate
- Partner screening if appropriate
Discuss pregnancy, contraception and sexual health

Refer for review by multidisciplinary team:
  o Clinical nurse specialists
  o Psychology
  o Community nurse

Discuss format of clinic, day unit service and give the contact details for the sickle team and give information on how to access A+E (leaflets available)
Consent for National Haemoglobinopathy Registry and any ongoing trials.

**Full examination:**

**Document:**
  o Height and weight
  o Each patient will a record of their vital signs including: Heart rate (HR), blood pressure (BP), oxygen saturations and urinalysis checked by the clinic HCA please review these and document them in the notes
  o Perform Cardiovascular, Respiratory, Abdominal and Neurological examination

**Investigations**

**Bloods:**
  o Full blood count (FBC) and reticulocytes (retics)
  o Confirmatory HPLC testing
  o Alpha phenotype (molecular)
  o G6PD
  o Electrolytes and creatinine, LFTs, LDH, Bone profile
  o Blood group and extended red cell phenotype, antibody screen
  o Viral serology (HIV, Hep B and Hep C)
  o Ferritin and folate

**Additional tests:**
  o Protein: creatinine ratio
  o Echocardiography
  o Respiratory function tests
  o Trans-cranial dopplers

Other tests as indicated by clinical information.
Regular Sickle Patients attending Outpatients (steady state review)

At each consultation, the history should include:
- Number of painful episodes since last attendance
- Other illnesses since last attendance
- Admissions since last attendance (type of crisis, length of admission)
- Pain at home and analgesia use
- Days off work or school
- Specific sickle issues since last attendance
  - Chronic pain
  - Transfusions since last attendance
  - Leg ulcers
  - Kidney/Urinary problems
  - Liver and gastro-intestinal problems
  - Lung disease
  - Memory problems

Medications and immunisations
- Penicillin
- Folic acid
- Analgesia
- Any received since last review and date on which given

Discuss any:
- Holiday or travel plans
- Need for vaccination and infection prophylaxis pre holiday
- Any new or ongoing psychological or social needs.
- Plans for pregnancy and contraception

Refer for review by multidisciplinary team:
- Clinical nurse specialists
- Psychology
- Community nurse

National Haemoglobinopathy Registry and Clinical Trials

Examination
- Each patient will a record of their vital signs including: HR, BP, oxygen saturations and urinalysis checked by the clinic HCA please review these and document them in the notes
- Perform Cardiovascular, Respiratory, Abdominal and Neurology systems examination

Investigations
- FBC and reticulocyte count
- Electrolytes and creatinine, LFTs, LDH
- Protein creatinine ratio

Others as relevant.
Comprehensive Annual Review for Sickle Cell Patients

All patients should have a comprehensive annual review once yearly. This visit will include review by the clinical nurse specialist and psychology team.

This follows the same procedure as for steady state review but additionally should include:

- Assessment of progress in general and review of patient’s knowledge of the condition
- Review of information provided by local unit or other professionals eg community, other specialists
- Concerns about concentration, memory or other indicators of cognitive function
- Discussion of other treatments (eg hydroxycarbamide)
- Discussion of National Haemoglobinopathy Registry and Clinical Trials

On clinical review document (pro-forma):
- Number of hospital admissions
- Number and severity of crises (days off college/work)
- Management of chronic or intermittent pain at home
- Other complications
  - Chronic pain
  - Leg ulcers
  - Headaches
  - Neurological symptoms
  - Visual symptoms
  - Abdominal pain or discomfort
  - Respiratory symptoms
  - Chest pain
  - Kidney or urinary problems
  - Priapism

Discuss;
- Pregnancy plans, partner screening, contraception, sexual health
- Assessment of psychosocial problems
- Planning for elective surgery

Review and list medications, all patients should be on:
- Penicillin (or alternate if Penicillin allergic) prophylaxis
- Folic acid
- Analgesia (PRN and or Regular use)
- Any other medication

Review vaccinations
- Flu vaccine – yearly in the community
- Hepatitis B –
  - Pneumovax (every 5 years – please administer in clinic if this has not been done in the last 5 years and put in the notes. If has been given, check pneumococcal Abs)

Clinical examination; check and document
- Each patient will a record of their vital signs including: HR, BP, oxygen saturations and urinalysis checked by the clinic HCA please review these and document them in the notes
- Perform Cardiovascular, Respiratory, Abdominal and Neurology systems examination

Investigations
- FBC and reticulocyte count
- Electrolytes and creatinine, LFTs, LDH
o Protein creatinine ratio
o Ferritin at annual review on all patients and at steady state review if on transfusion program

Request:
o Echocardiography if:
  • Exertional dyspnoea
  • New murmur
  • Loud P2
  • Low oxygen saturations (<95% on air with the other symptoms suggestive of pulmonary hypertension as listed here)

(Echo repeated yearly if abnormal otherwise alternate yearly)

o Transcranial Doppler (TCD) tests (U/A carotid and trans-cranial dopplers) on new patients, if neurological symptoms, or if previous abnormality

o MRI Brain and MRA if:
  • Headaches or new onset (or progression) of cognitive impairment
  • (consider TCD as well)

o Respiratory function tests + CT chest if:
  • Dyspnoea (exertional or rest)
  • Reduced exercise tolerance
  • Oxygen saturations <96% on air
  • Persistently abnormal chest examination

o Sleep study if: Symptoms suggestive of obstructive sleep apnoea or nocturnal hypoxia

o Check Hepatitis B antibody titre every 5 years

o Renal USS if:
  • Proteinuria with PCR > 50 on 2 occasions
  • Haematuria

  Ferriscan Liver and T2*Cardiac MRI if serum ferritin persistent >1000mcg/L

  X-ray +/- MRI of any area with chronic pain especially if progression in symptoms

  Consider other investigations as appropriate
  Please see appendix 1 for GP letter
**Comprehensive Annual Review for Sickle Cell patients on an exchange (EBT) or top up transfusion program**

Follow the same procedure as for un-transfused patients but **additionally**:

Review and document:
- The indication for transfusion, if short term or long term:
  - If short term transfusion/EBT then review need to continue
    (please discuss with consultant in clinic)
  - If appropriate discuss alternatives e.g. Hydroxycarbamide
- Ensure written consent for transfusion program is filed in notes
- The patient’s understanding of the of the reasons and aims of their EBT/transfusion program
- Document any recurrence of the original problem(s) for which patient is on the program e.g. TIAs in a patient on transfusion for secondary stroke prevention
- Intervals between EBT/ transfusions if exceeding the prescribed interval;
  - Enquire and document the reason(s)
  - If non-compliance please inform consultant in clinic
- Review and document pre and post transfusion HbS% and if achieving targets
- Any vascular access issues

**Medications:**

*All patients not on chelation with evidence of iron loading must be reviewed by consultant haematologist to discuss commencing chelation*

Patients on chelation:
- Ensure and encourage compliance
- Document date of last:
  - Ophthalmological and audiometry review (Desferrioxamine or Desferasirox)
  - If none in past year please request

**Additional Investigations**
- Blood group and antibody screen
- Ferritin and CRP
- Direct antiglobulin screen
- Endocrine profile (glucose, thyroid function, bone profile, sex hormones)
- Virology (HIV, Hepatitis B and C)
- Ferriscan of liver: **Yearly if abnormal.** Two yearly if normal and ferritin normal.
- Cardiac T2* MRI:
  - Every 2 years if T2* >20 ms
  - Yearly if T2*= 10-20 ms
  - 6 monthly if T2*<10 ms
  - 3 monthly if T2*< 10 ms and any evidence of cardiac impairment
**New patients (Thalassaemia):**

New patient referrals may be received from the transition service, general practitioners, other hospitals, other departments in the hospital or following inpatient attendances.

**First consultation:**

Document full personal and family history to include:

- Date and place of diagnosis
- Transfusion history including indication, frequency and venous access
  - If transfused ascertain cumulative volume transfused over preceding 12 months
- Any symptoms of anaemia (especially in un-transfused intermedia patients)
- Chelation history (when commenced, drug(s), doses, side effects, compliance)
- Cardiac history -
  - Palpitations/dyspnoea etc
- Endocrine history -
  - Diabetes, menstruation, erections, any hormonal therapy e.g. HRT etc
  - Any known blood borne viral infections e.g. Hep C
  - If previous splenectomy
  - Any previous hospital/ITU admissions and what for
  - Vaccination history
  - Any children, siblings or parent with thalassaemia
  - Full past medical + social + drug history
- Date last reviewed by:
  - Cardiologist
  - Endocrinologist
  - Last audiometry assessment (if on chelation)
  - Ophthalmology review
- Alcohol/smoking/other drugs
- Education/college/work

Discuss acute and chronic complications of thalassaemia, iron overload and iron chelation;

- When and why transfusion is initiated, frequency and target haemoglobin
- Symptoms of the endocrinopathies (diabetes, hypogonadism etc)
- Symptoms of cardiac disease
- Pulmonary hypertension
- Aim of iron chelation (and encourage compliance in those on therapy)

Discuss medications and vaccinations

- Bisphosphonate/Vitamin D and calcium supplements
- Hydroxycarbamide
- All patients should be vaccinated against Hepatitis B
- All splenectomised patients should be vaccinated as per the vaccination protocol and on antibiotic prophylaxis

Genetic counselling

- Discuss inheritance and partner +/- family screening if appropriate

Discuss pregnancy and contraception

Refer for review by multidisciplinary team (as for the new sickle patient)

- Clinical nurse specialists
- Psychology
- Community nurse
Discuss format of clinic, day unit service and give the contact details for the medical/nursing team and give information on how to access A+E (leaflets available)

Consent for National Haemoglobinopathy Registry and any ongoing trials.

**Full examination:**

**Document:**
- Height and weight
- Blood pressure and heart rate
- Oxygen saturations
- Urinalysis
- Random blood glucose
- Perform Cardiovascular, Respiratory, Abdominal and Neurological examination

**Investigations**

**Bloods:**
- Full blood count and reticulocytes
- Confirmatory HPLC testing and molecular testing
- Alpha phenotype (molecular)
- G6PD
- Electrolytes and creatinine, LFTs, LDH, Bone profile, vitamin D
- Endocrine: thyroid profile, LH/FSH
- Blood group and extended red cell phenotype, antibody screen
- Viral serology (HIV, Hep B and Hep C)
- Ferritin and folate

**Additional tests:**
- Protein : creatinine ratio
- Oral glucose tolerance test
- Echocardiography
- Respiratory function tests
- Dexa scan
- US abdomen
- All new patients who have not had a cardiac or liver iron assessment in the previous 12 months must have a Ferriscan Liver and Cardiac T2*MRI
- Refer all new patients for cardiology, endocrinology and ophthalmology review

Other tests as indicated by clinical review.
Regular Thalassaemia Patients attending Outpatients (steady state review)

At each consultation, the history should include:
- Any subjective problems – (enquire specifically about vision and hearing if on chelation)
- Energy levels
- Number of transfusions (if any) since last review
- Other illnesses including infections since last attendance
- Specific cardiac and endocrine history
- Medications and immunisations
  - List medications and note any dose changes

Discuss any
- Holiday or travel plans (arrangements for transfusion while away if appropriate)
  - Need for vaccination and infection prophylaxis pre holiday
- Any new or ongoing psychological or social needs.
- Plans for pregnancy and contraception

Refer for review by multidisciplinary team
  - Clinical nurse specialists
  - Psychology
  - Community nurse

Discuss any ongoing trials and National haemoglobinopathy registry

Full examination:

Document:
- Blood pressure
- Oxygen saturations
- Urinary dipstick
- Perform Cardiovascular, Respiratory, Abdominal and Neurological examination
- Document spleen size if palpable

Investigations

Bloods:
- Full blood count and reticulocytes
- Electrolytes and creatinine, LFTs, LDH, bone profile, random glucose
- Blood group and antibody screen
- Ferritin and folate

Other tests as relevant
Comprehensive annual review for thalassaemia patients

Document
- Any new problems
  - Objective problems: Leg ulcers, recent fractures
  - Subjective: energy levels
- Transfusion history over the prior 12 months (if applicable)
  - Ascertain cumulative volume transfused
  - Review indication (short or long term)
  - Review venous access and document any problems
- Chelation therapy, drug, dose and compliance
- Any cardiac symptoms
  - Palpitations/dyspnoea etc
- Endocrine history
  - Diabetes, menstruation, erections, any hormonal therapy e.g. HRT etc
- Any hospital admissions and reasons
- Any planned surgery/admissions
- Date of last and next review by:
  - Cardiologist
  - Endocrinologist
  - If on chelation: Last Ophthalmology & Audiometry assessment
- Alcohol/smoking/other drugs
- Education/college/work any issues associated with diagnosis

Review and document medications and vaccinations dates
- Bisphosphonate/Vitamin D and calcium supplements
- Hydroxy carbamide
- All patients should be vaccinated against Hepatitis B
  - All splenectomised patients should be vaccinated as per the vaccination protocol and on antibiotic prophylaxis

Genetic counselling
- Discuss inheritance and partner +/- family screening if appropriate
- Discuss pregnancy and contraception

Refer for review by multidisciplinary team (as for the new thalassaemia patient)
- Clinical nurse specialists
- Psychology
- Community nurse

Consent for National Haemoglobinopathy Registry and any ongoing trials.

Full examination:

Document:
- Height and weight
- Blood pressure and heart rate
- Oxygen saturations
- Urinalysis
- Random blood glucose
- Perform Cardiovascular, Respiratory, Abdominal and Neurological examination (and if on desferrioxamine review infusion sites)
**Investigations**

**Bloods:**
- Full blood count and reticulocytes
- Electrolytes and creatinine, LFTs, LDH, Bone profile, Vitamin D level
- LH, FSH, Thyroid profile, PTH
- Blood group and antibody screen
- Viral serology (HIV, Hep B and Hep C) in regularly transfused patients
- Ferritin and folate

**Additional tests:**
- Protein:creatinine ratio
- Oral glucose tolerance test
- Echocardiography
- Respiratory function tests
- Dexa scan
- US abdomen
- Ferriscan of liver: Yearly if abnormal. Every two years if normal (and serum ferritin also normal).
- Cardiac T2* MRI:
  - Every 2 years if T2* >20 ms
  - Yearly if T2*= 10-20 ms
  - 6 monthly if T2*<10 ms
  - 3 monthly if T2*< 10 ms and any evidence of cardiac impairment

Other tests as indicated by clinical information.
Special consideration

Patients on Hydroxycarbamide:
On commencing Hydroxycarbamide patients need to attend for a blood count 2 weeks after commencement and be reviewed in the OPD 4 weeks later
All patients established on Hydroxycarbamide need to be reviewed 8 weekly in the OPD

Patients on Long term transfusions/Erythrocytopheresis
OPD review at least 6 monthly, need for continued transfusion documented or reasons for discontinuing.
All patients MUST have iron status, Vaccination status documented and monitoring forms completed annually. Iron chelation commenced by consultant haematologist when Ferritin reaches 1000ng/L

Specialist clinics

Renal clinic: Joint clinic with Dr C Breen
Held every 1-2 months in Haematology 2 clinic
For referral - discuss with Dr Howard/Dr Kesse-Adu and book in via clinic clerks
Referral guidelines – see appendix 2

Orthopaedic clinic: Joint clinic with Mr M Bankes
Held every 1-2 months in Haematology 2 clinic
For referral - discuss with Dr Howard/Dr Kesse-Adu and book in via clinic clerks
Quick referral guidelines- see appendix 2

Neurology clinic: Joint clinic with Dr P Holmes
Held every 2-3 months in Haematology 2 clinic
For referral – discuss with Dr Howard/Dr Kesse-Adu and book in via clinic clerks
Quick referral guidelines – see appendix 2

Obstetric clinic: Joint clinic with Mr Oteng-Ntim/Dr Robinson
Held last Thursday of the month in Antenatal clinic STH
Referral guidelines – see appendix 2

Pulmonary hypertension clinic: Refer to joint pulmonary hypertension clinic at King’s College Hospital run by Prof Thien and Dr Coglan or locally to Dr Cathy Head for her joint clinic with Dr G Coghlan

Chronic pain clinic: referrals to be sent to Dr Doug Justins
Respiratory symptoms: referrals to be sent to Dr Nick Hart
Persistant priapism or erectile dysfunction - Refer to Mr Paul Hegarty
Gynaecological problems refer to Mr Rajesh Varma
Referral for sperm banking before Hydroxycarbamide to assisted conception unit (use referral form)
Referral for pre-implantation genetic diagnosis – to assisted conception unit (use referral form)
Did Not Attend (DNA) policy:

New and follow up patients:

- First DNA – repeat appointment sent (usually 2 months, review urgency in referral letter/clinic notes)
- Second DNA – repeat appointment sent (2-3 months). Plus standard letter to GP and patient.
- Third DNA – (Please check patient not admitted) if no explanation then patient discharged. Letter to patient and GP

Hydroxycarbamide patients (or high risk patients with high clinical concern)

- First DNA – 2 week appointment
- Second DNA – patient is phoned by medical or nursing staff to rearrange appointment
- Third DNA – patient is phoned and letter sent to GP and patient
Appendix 1

Annual review letter to GP

Dear Dr…,

Re: Patient

Mr/Ms ….. was seen in the multidisciplinary sickle annual review clinic on DATE

List active problems:
Diagnosis and genotype: SC/SS/Sb+..)
Any complications: Osteomyelitis/priapism/Stroke etc

Document in past 12 months:
Number of acute painful episodes both requiring hospital admission and not
Any chronic pain and site
Any blood transfusion in past 1 2 months
Any other hospital admission and surgical procedures
Days off work due to sickle cell disease

Other problems

Medication list

Vaccinations
List those received in past 12 months and document when others received

Examination findings

Investigations
FBC
Echo: date and result
Lung function: date and result
Transcranial Doppler: date and result

Plan

Yours sincerely
Appendix 2

Specialist clinic quick referral guidance
(Please discuss all potential referrals with a sickle consultant)

Joint Renal clinic (Guidance on management of early proteinuria, hypertension and haematuria in sickle patients on intranet): referral to joint clinic if:
- BP not controlled on 2 agents
- Persistent proteinuria despite ACEI
- A non sickle cause for renal disease
- Worsening creatinine/eGFR
- For consideration for erythropoietin therapy

Joint Orthopaedic clinic
Refer patients with:
- Chronic hip pain: X-Ray evidence of avascular necrosis
- Chronic hip pain: normal X-Ray, but MRI evidence of avascular necrosis
- Chronic hip pain: no evidence of avascular necrosis
- Chronic shoulder pain: X-Ray or MRI to be done before referral
- Other chronic joint pain: Imaging to be performed before referral
- Follow up after hip surgery
- Recurrence of pain after hip surgery
- Follow up of patients with osteomyelitis.

Joint Neurology
Refer patients with:
- Chronic intermittent headache: perform MRI+MRA and TCD before referral
- Previous history of stroke: MRI within last year
- Patients on long term transfusion for primary or secondary stroke prevention – to be reviewed every 2 years with recent imaging.
- Symptoms of transient ischaemic events: perform TCD, carotid artery Doppler, MRA+MRI brain
- Fits or seizures

Joint Obstetrics
Check partner status and arrange testing for haemoglobinopathy if not known
Check medications and stop all teratogenic medications: e.g. Hydroxycarbamide, Bisphosphonates, Exjade etc
Consider starting aspirin 75mg daily
Referral form must be faxed to Kylie Gould (Tower team midwife) with copy to Dr Robinson (copy clinic letter to both Dr Robinson and Dr Oteng-Ntim)

Respiratory (Specialist joint clinic in set up) refer to respiratory physicians if
- Saturations<93% on room air
- Dyspnoea at rest on or on minimal exertion
- Excessive daytime somnolence (with sleep study booked)
- Nocturnal cough
- Features of hypercapnia e.g. morning headaches

Endocrinology: refer to endocrinology if:
Thalassaemia
- impaired glucose tolerance
- hypothyroidism
- hypoparathyroidism
- impaired fertility or abnormal sex hormones
Sickle
- as above (impaired sex hormones/testosterone most common cause for referral

Chronic pain: refer to chronic pain if;
- pain on a daily basis, requiring regular analgesia
- frequent unexplained admissions to hospital
- evidence of bony infarction/chronic bony damage without surgical solution
- CONSIDER INPUT PROGRAMME - chronic pain management programme.