1. Introduction and Who Guideline applies to

As survival of patients with sickle cell disease (SCD) improves, the management of chronic complications remains a challenge and requires a robust multidisciplinary team approach.

This document briefly describes management of chronic complications of:

a) end organ damage related to sickling events
b) end organ damage relating to the effects of iron overload in transfused patients
c) end organ damage relating to toxicity of chelation therapy

Chronic complications of sicke cell disease:

Organ system	Section in guideline	Description
Genitourinary	2 & 3	Chronic renal failure, haematuria, hyposthenuria, priapism, erectile dysfunction
Ophthalmology	4	Sickle eye disease
Respiratory	5	Pulmonary hypertension and chronic sickle lung
Cardiology	6	Cardiac disease and heart failure
Neurology	7	Ischaemic or haemorrhagic stroke, headaches
Musculoskeletal	8	Chronic sickle pain, avascular necrosis (AVN) of the long bones, osteoporosis / osteomalacia
Skin	9	Leg ulcers
Gastroenterological	10	Gallstones, Hepatopathy
Other	11 12	Dental Complications Complications relating to iron overload and chelation therapy

The appendix includes the annual review documents, including required schedule of tests, to identify end organ damage and monitor effectiveness of management strategies

2. Guideline Standards and Procedures

Annual Review

All Patients with Sickle Cell Disease are offered an annual review in the Hb Disorders Clinic.

1. National Haemoglobinopathy Register (NHR)

All patients are offered registration on the National Haemoglobinopathy Register. The NHR was updated in 2021, so that data inclusion is considered standard care and specific consent is not required in line with national data governance. The NHR steering committee reports directly to the NHS Clinical Reference Group for Haemoglobinopathies. Data may be used for research purposes, however, and consent for this aspect is required. The NHR is monitored and updated locally by the network data manager. Further information on the NHR can be found at https://nhr.mdsas.com/
A local database of all haemoglobinopathy patients is also maintained to aid reporting to the NHR and clinical commissioning groups. For details contact the Data Manager.

2. Renal disease:

2.1 Routine clinical review

- **All patients with SCD will be offered a urine sample screen at each outpatient appointment, (minimum frequency, annual testing), as well as a blood pressure check**.
- **Renal function testing should be performed at least annually, with prompt referral to the renal clinic indicated if there is significant deterioration in the creatinine level, or eGFR (eGFR <60 ml/min).**
- **Where urine PCR is > 50mg/mmol in normotensive patients or >30mg/mmol in hypertensive patients, treatments with ACE inhibitors should be commenced. Consider lisinopril 2.5mg daily increasing to 5mg daily if tolerated (no hypotension, renal function and potassium levelsstable).**

Urine testing is performed to detect proteinuria as an indicator of renal function, nitrates indicating urinary tract infection, haematuria and glycosuria. Symptoms of enuresis, and nocturia and priapism will be reviewed at least annually in patients with sickle cell disease.
Where urine dip test shows + protein or more, urine will be sent for ACR/PCR.

Careful control of hypertension is essential to limit renal impairment and other complications in the long term. The target blood pressure with patients with proteinuria should be 130/80 mmHg while those without proteinuria 140/90 is the target.
Chronic long-term use of non-steroidal anti-inflammatory drugs should be avoided.
Other medication (e.g. deferasirox) may also contribute to renal impairment, and appropriate dose adjustment may be needed.

The outpatient clinic provides an opportunity to reinforce the importance of continuous, regular hydration to avoid precipitation of crises

2.2 Specific renal conditions

2.2.1 Sickle cell nephropathy (affects 5-18% of patients)

Characterised by insidious onset; usually detected by regular monitoring as described above. Where significant proteinuria (PCR >50mg/mmol) is present, ACE inhibitors should be used in line with NICE guidance, as described above. Consideration for hydroxycarbamide should also be discussed. Erythropoietin in consultation with renal physicians, but higher doses may be required. Transfusion may be required and if on a regular basis, iron chelation therapy will be required or regular exchange transfusions. Where renal transplantation is likely, a perioperative plan should be available in the notes for immediate use (including transfusion plan, aiming for HbS<25%). Long term hydroxycarbamide or exchange transfusion should be considered in these patients pre and post renal transplant.

2.2.2 Hyposthenuria

Hyposthenuria is associated with nocturia and enuresis. Although it is more common in childhood a history of enuresis should prompt further investigation (e.g. overnight O2 saturation).
Both enuresis and nocturia are associated with an increased risk of dehydration, which can precipitate a sickle crisis. Continued reinforcement of need for hydration should be given at every opportunity. Patients should aim for fluid intake of 3-4 L/ day.

2.2.3 Urinary tract infection

Patients with SCD are at increased risk of UTIs due to hyposplenism.
Asymptomatic UTI may be associated with sickle cell crisis. Renal tract sepsis should be considered in patients with SCD and fever. Urine dip test at each OPD, with MSU where screen is positive for nitrites or WBC, is advisable. Referral for Urology review is indicated in the event of recurrent urinary tract infections, pyelonephritis, haematuria, or complications of priapism

2.2.4 Renal papillary necrosis

Occurs due to medullary infarction and can affect both SCD patients and carriers. It presents with microscopic or frank haematuria. Treatment is supportive with maintenance of high urinary flow and blood transfusion if necessary.
Renal ultrasound may be helpful but definitive diagnosis may require CT urography. Colic pain may be caused by passage of clots.

2.2.5 Renal medullary carcinoma

Although extremely rare, it is a rapidly fatal malignancy, as usually already metastasised at presentation, associated with sickle gene (carriers as well as SCD). Haematuria should alert for prompt investigation with renal USS or CT/MRI. Other symptoms include flank pain, weight loss, abdominal pain and fever.

2.2.6 Acute Kidney Injury (AKI)

Acute Kidney injury can occur in the context of acute chest syndrome. It should be managed as any other case of AKI. Nephrotoxic drugs, including NSAIDs should be avoided.

3. Priapism

- **All men with SCD should be educated about priaprism and be asked about both stuttering and fulminant episodes as part of their annual review.**

3.1 Background

Priapism is defined as a persistent erection lasting more than four hours and is a medical emergency requiring hospital assessment
Priapism in sickle cell can present at a young age (from childhood) and is caused by vaso-occlusion caused by sickling in the penis. Lifetime risk 29-42%. Is common in young men with sickle disease but the exact burden is unclear as research and self-reporting are both lacking. It is undoubtedly painful and can result in penile damage and impotence which occurs as a consequence of prolonged attacks. Early discussion with boys and their parents is strongly recommended.
There are various types of priapism: ischaemic, non-ischaemic and stuttering (recurrent, intermittent).
Stuttering priapism can occur as episodes lasting less than 3 hours, often occurring in the morning and recurrent painful can occur with either on its own or with sickling elsewhere in the body. It can be relieved by exercise (e.g. running upstairs), a warm baths, but sometimes is so frequent that medication may be required to lessen or reduce the episodes.
It is important to identify those affected, usually by direct questioning in routine clinic visits as it is often a difficult subject for patients to address.

3.2 Management of stuttering priapism

Self-help measures are advised (mild exercise, hot baths, relaxation techniques)
Medication with ephedrine or etilefrine can be used
Role of sildenafil and hydroxycarbamide are uncertain but may be considered

Drugs which may be useful Drug	**Dose**	**Side effects/monitoring**
Ephedrine	15-60 mg po usually at night	High blood pressure – check 2 weekly, tachycardia
Etilefrine (short acting)	5-25mg po at night, if persistent problems	High blood pressure
Sildenafil (Viagra)	50 mg daily	Headache, diarrhoea, dyspepsia, flushing Possible increase in sickle cell crises, priapism
Tadalafil (Cialis)	5mg daily	Similar to sildenafil

4. Retinopathy

- **All patients with SCD should be informed about the risk of eye complications and asked about visual symptoms at their annual review.**
- **All patients should have baseline retinopathy screening and those with a history of retinopathy should have regular ophthalmic review.**
- **All patients on regular chelation therapy should attend for yearly ophthalmology review to exclude toxicity. Urgent ophthalmology referral is required if any acute change in vision occurs.**

Annual ophthalmology review is advised if possible for all patients with SCD, to exclude or treat sickle retinopathy. Eye casualty in the Windosr eye clinic, LRI. Outside of opening hours, patients should attend A&E for urgent review when necessary.
Retinopathy can be classified as non-proliferative (NPR) and proliferative (PR). The distinguishing element is neovascularisation. Care should be taken, as NPR can be asymptomatic in early stages. PR can be complicated by vitreous haemorrhage and retinal detachment.

5. Chronic respiratory disease

- **Oxygen saturation should be measured at least annually in the outpatient's clinic. Where pO2 is repeatedly low (<95%), hypoxia should be confirmed by obtaining arterial blood gases and if confirmed, investigated accordingly.**

In these cases, and where individuals have had recurrent chest crises, frequent admissions, or symptoms of shortness of breath, poor exercise tolerance, excessive daytime sleepiness, priapism, snoring, early morning headache, then lung function tests and overnight oxygen monitoring should be requested, clearly stating diagnosis and usual Hb on the request form. Prompt antibiotic treatment of infection and cessation of smoking should be encouraged. High resolution CT scan may be more helpful than CXR and 6-minute walk test can be used as a marker of functional ability.
Obstructive causes and sleep disorders should be considered and investigated. Obstructive sleep apnoea is usually due to tonsillar hypertrophy or other causes of deep disordered breathing. It can precipitate painful episodes and cause neurological events.

6. Cardiology

- **All patients should be assessed for cardiac symptoms and undergo a cardiac examination at each annual review**

6.1 Pulmonary hypertension and other cardiac complications (heart failure / arrhythmias)

- **Echo should be performed routinely every 3 - 5 years in asymptomatic individuals and should be repeated annually in patients with previously elevated tricuspid regurgitant jet velocity (TRV)**

Affects about 5-30% of SCD patients
Chronic haemolysis releases free haemoglobin which leads to NO deficiency causing acute and chronic pulmonary vasoconstriction.
Echocardiography should be undertaken regularly in all patients: interval depends on clinical condition, severity of disease and symptoms.

Requests for echocardiography should be cleared marked with 'sickle cell patient, to exclude pulmonary hypertension and request TRV to be measured [if present]. The following advice should then be followed:

1. If normal: repeat every 3-5 years if asymptomatic. Repeat sooner if symptoms occur
2. If TRV >290cm/sec: Refer to cardiology team for assessment and consideration of right heart catheterisation
3. If TRV 250-290 cm/sec: Check pro-BNP and consider 6 minute walk test. If concerns, discuss with cardiology.
4. If TRV raised but no intervention performed: Annual echo.

6.2 Other cardiac problems

Include left sided heart disease or diastolic dysfunction. ACE inhibitors, diuretics and beta-blockers may be used as recommended by experienced cardiologist, under joint care with the haematologist.

7. Neurological complications

- **Headache is common and the incidence of migraine is increased in people with SCD. This should be treated with standard medical management but referral to chronic headache services considered**
- **MRI/A Brain scanning should be performed if any concerns regarding neurological symptoms, especially if patients have not undergone routine TCD screening in childhood (i.e. if born outside of the UK)**
- **Annual review will include psychosocial review, offer of assessment, and referral on for further cognitive investigation by psychology team as appropriate.**

Neurological complications such as stroke or TIA should be treated as a medical emergency. Following acute management and referral to Stroke team, Leicester Royal Infirmary the Haemoglobinopathy team must provide a management plan for long term stroke prevention, including transfusion plan (consideration of exchange transfusion), antiplatelet or anticoagulation therapy and intensification of surveillance (BP, renal function) as necessary.

There is a high level of risk in sickle cell disease patients of ischaemic (24% in those with sickle cell disease over 45years) or haemorrhagic stroke including sub arachnoid haemorrhage (10.8% in adults). There is no recognised screening technique in adults and treatment should be in accordance with general principles in adults including the use of Thrombolysis and anticoagulants as necessary. Prompt exchange transfusion should be done and long term exchange transfusion or hydroxycarbamide is advisable.

Cognitive problems may be identified by patient, clinician or psychology team.
Patients with new onset neurological problems such as headache or epilepsy will be reviewed at the regional MDT following consultation with an experienced neurologist.

8. Musculoskeletal complications

8.1 Osteoporosis / osteomalacia

- **Patients should have bone profile and Vitamin D testing at least annually and appropriate replacement advised if low.**
- **If Vitamin D <30nmol/ treat for deficiency with loading dose 300000iu (e.g. 4000iu daily for 10 weeks) then maintenance of 800iu/day. Levels of 30-50nmol/l require supplements of 800iu/day.**

Patients with Hb disorders are particularly prone to metabolic bone disease. Factors such as diet, sunlight exposure, sex hormone levels and hypoparathyroidism and expansion of the marrow cavity will all contribute.
Serum calcium levels and Vitamin D levels must be measured regularly in all patients at least annually. If below normal range, Vitamin D level must be measured and replaced. Following correction with supplementation, vitamin D levels should be repeated, but with a minimum interval of 1 year in view of slow response to treatments. High doses of Vitamin D may be necessary for patients with hypoparathyroidism.

Patients with severe deficiency may require higher doses.
Dexa Scans are appropriate if any concern about osteoporosis/ osteopenia. Advocate regular exercise and balanced diet including dairy products, fish etc. where possible. Advice against smoking and excess alcohol consumption should be given when patients are reviewed in clinic.

8.2 Arthropathy / Avascular Necrosis

Patients with SCD may be subject to avascular necrosis or degenerative joint disease. Atypical pain, limited movement of a joint must be investigated with plain X-ray.

- **If changes are apparent on X-ray, referral to orthopaedic specialist without further imaging is appropriate. If no changes are reported on X-ray, MRI may be considered to exclude early avascular necrosis**.

Management plan should be defined in consultation with relevant orthopaedic surgeon. If any intervention is advised, a perioperative plan must be provided by the haemoglobinopathy team. Early involvement of the physiotherapy and the occupational therapy team is essential.

Generally cement - less joints are preferred when joint replacement is required. If joint replacement is required then planning about pre-operative transfusion and post-operative infection prevention and thromboprophylaxis is required

8.3 Chronic Pain

- **Patients should be asked about chronic pain as part of their annual review and use of opioid analgesia should be reviewed at each clinic visit.**

Pain will be assessed at each clinic. Where there are issues of chronic pain, pain relief may be offered using WHO treatment ladder.

It is clear from work done elsewhere that multidisciplinary teams, usually including a psychologist , are very helpful in this situation. The relief of chronic pain is often not achieved by use of opiates and in most cases other strategies are required

Psychology support team in clinic will routinely enquire about pain control and discuss strategies for pain control. Where these measures fail, referral may be made to the Pain Team. Patients with chronic pain not controlled by mild opiate analgesia should also be discussed at the regional MDT.

Chronic pain, either intermittent due to painful sickle cell crises or due to a complication of sickle cell disease (e.g. avascular necrosis of hip), occurs in many sickle cell patients. This should be included in every sickle cell review undertaken. Unexpected or unusual pain should not be attributed to sickle cell disease but other possibilities should be sought.

Close liaison with other prescribers (e.g. GP) may be necessary to check doses and frequency of prescription.

Choices of analgesia

Table 2: Choices of analgesia include Medicine	**Dose range**	**Comment**
Paracetamol	1g orally qds	
NSAID	Care with renal impairment. If regular use, may need PPI cover.	
Dihydrocodeine	30mg orally every 4-6 hours In young fit people may need doses up to 60-90mg qds.	**Be aware that this drug needs to be metabolised to have full effect and ~25% of the Afro-Caribbean population don't have the gene.**
Tramadol	50-100mg orally NOT more often than 4 hourly (usually not more than 400mg required in 24 hours)	Better as a regular analgesic as superior for neuropathic type pains. Care if patient already on tri-cyclic antidepressant (added serotonin effect) and not with MAOIs.
Fentanyl patch	25-100µg/72 hours Start at lowest dose, assess at 24 hours (oral morphine sulphate equivalent is <135mg	For constant level of pain only. No use in variable pain. Needs good skin blood flow to be effective. Plasma doses of fentanyl reliant on skin blood flow.

	in 24 hours to 25micrograms patch) Also available as lozenge	Pyrexia will cause greater uptake.
Morphine salts	5-20mg orally every 4 hours	For constant level of pain only. No use in variable pain unless supplemented with other analgesics.
Gabapentin	300mg orally on day 1, increasing every 24 hours if required to max dose of 1.8g (in 3 divided doses)	For chronic neuropathic pain. No good for acute flare-ups. Reduce dose in renal impairment as renally excreted.

Onward referral if pain relief problematic in community

Applicable to	Those with chronic pain related to haemoglobinopathy
Check	Use of analgesia appropriate
If still problems	Consider referral to chronic pain team in the community for further advice, use of alternative therapies
Referral in hospital service	Ask GP to refer to Chronic Pain team

9. Chronic leg ulcers

- **Annual review should include questioning about leg ulceration and inspection of the lower extremities for active or healed ulcers.**

Affects predominantly males with HbSS disease. The mechanism of ulcer formation is poorly understood but they usually form around the medial or lateral malleoli. Early intervention is required to prevent progression to chronic ulceration which is difficult to treat. Elevation, modern wound dressings and treatment of infections is essential. In difficult cases, review by the plastic team (skin grafting) might be needed.Zinc levels should be measured in patients with leg ulcers and supplements offered to those with deficiency.

10. Gastrointestinal and hepatic complications

- **Liver function tests should be monitoried at least annually.**

Gallstones - present in at least 30%, if symptomatic suggest laparoscopic cholecystectomy (ursodeoxycholic acid may be used) . Splenic and hepatic sequestration occurs - largely supportive. Mesenteric syndrome is rare characterised by upper abdominal pain and presenting as acute bowel pseudo-obstruction, again managed conservatively but challenge is to distinguish from acute abdominal. Acute intrahepatic sickling (right upper quadrant pain hepatic enlargement and tenderness, coagulopathy, very raised bilirubin with moderate liver enzyme rise. Early exchange transfusion is the recommended management. Chronic sickle hepatopathy can vary from mild liver enzyme elevation to cirrhosis and should be managed in conjunction with a centre experienced in liver problems in sickle patients (hydroxycarbamide not indicated)

11. Dental Complications

Dental complications can occur and may be directly related to a diagnosis of sickle cell disease. Community dentist treatment should always be considered first line but if specific concerns arise, patients should be referred to Oral and Maxillofacial surgery (OMFS) for specialist review if concerns. Any emergencies require liaison with the OMFS on call team.

12. Complications of transfusion overload and chelation therapy

Patients who are on transfusion programmes (top-up or exchange transfusion) are at risk of iron overload. Ferritin and LFTs should be checked every 3 months. Iron chelation should be considered in all patients on a regular transfusion regimen who have received at least 20 top-up transfusion episodes or have a liver iron concentration of >7mg/g dry weight. Monitoring with T2* MRI is required to assess cardiac and liver iron overload.

Usual monitoring is with Ferriscan technique which measures liver iron. Cardiac iron overload is much less common with sickle cell disease but if rising liver burden or clinical concern a cardiac T2 * can be done at Glenfield hospital, Leicester via ICE request.

Specifc information about dosing of iron chelation and appropriate management can be found in the UHL Thalasaemia guideline

12.1 Endocrinopathy

Low sex hormone levels may be a feature of iron overload and should be corrected as part of the regular endocrine review. Endocrine failure is a common complication of poor chelation, and screening will be carried out at regular intervals and always as part of the annual review of patients on transfusion programmes.

- Thyroid function: TSH every three months
- Reproduction: menstrual and sexual history annually as appropriate
- FSH, LH annually for females; Testosterone at least annually for males
- GTT annually. If this is not carried out, then fructosamine levels and random blood glucose levels when attending for transfusions.
- Calcium and Vitamin D – consider hypoparathyroidism in refractory cases: may require higher Vitamin D doses

12.2 Audiometry

Must be offered annually to all patients receiving chelation therapy to exclude toxicity (high tone deafness). Possible complications of chelation therapy should be monitored regularly. Regular monitoring of the therapeutic index may help to avoid toxicity. Further information is available on the iron chelation guideline.

3. Education and Training

Ongoing training required for haematology trainees involved in seeing patients in outpatient clinic – to be included in regional training programme.

4. Monitoring Compliance

What will be measured to monitor compliance	How will compliance be monitored	Monitoring Lead	Frequency	Reporting arrangements
Data as required by NHS England haemoglobinopathy dashboard	NHS England Haemoglobinopathy Dashboard	Amy Webster	Quarterly	SSQD

5. Supporting References (maximum of 3)

Sickle Cell Society Standards for the clinical care of adults with sickle cell disease in the UK, February 2018

6. Key Words

Sickle cell disease, Chronic complications

	CONTACT AND REVIEW DETAILS
Guideline Lead (Name and Title) **Dr Amy Webster, Consultant Haematologist**	**Executive Lead**
Details of Changes made during review: **Reference to dental complications added** **Wording around iron chelation guidance updated**	

Appendix 2: Specialist clinician advice

Renal:	Dr Jorge Jesus-Silva Consultant Nephrologist	Via email
Urology:	Mr Duncan Summerton, Consultant Urologist	LGH 0116 258 8260
Bone disease:	Dr Faizanur Rahman, Consultant Chemical Pathologist Dr Prashanth Patel, Consultant Chemical Pathologist	LRI 0116 258 6560 0116 258 6550
Orthopaedics:	Hip: Mr Andrew Brown/Mr Steffan Hutchings	LGH 0116 258 4217
Ophthalmology:	Mr James Deane, Consultant Ophthalmologist	LRI 0116 258 6864
Cardiac:	Dr Aidan Bolger, Consultant Cardiologist	GGH 0116 250 2930
Cardiac MRI:	Dr Aparna Deshpande, Consultant Radiologist	GGH
Respiratory: (including sleep disorders)	Dr Alys Scadding, Consultant in Respiratory Medicine	GGH Leicester 0116 250 2640
Endocrinology:	Dr Miles Levy, Consultant Physician and Endocrinologist Dr Ragini Bhake, Consultant Physician and Endocrinologist	LRI 0116 258 5866 0116 258 5157
Stroke team:	Dr Amit Mistri, Consultant in Stroke Medicine	Via email
Chronic Pain Team:	Dr Mahesh Kodivalasa, Consultant Anaesthetist (Lead clinician, Pain Service)	LGH 0116 258 4661

Patient Details (or affix addressograph) Name: Hospital number: DOB:	Name of reviewer: Date of review: Place of review: UHL / NGH / KGH / NUH / RDH / Other
	Diagnosis: NHR registered? Yes / No Card Issued? Yes / No **Transition Patient?** Yes / No **Centre change in previous 12 months?** Yes / No
Emergency care plan Does patient have an up to date care plan? At home In hospital	Yes / No
Past Medical History: **To include known sickle complications**	
Chronic health review: **Consider physical, psychological, financial and emotional implications**	
General health review: Any new or ongoing concerns? Chronic pain: To include site, severity, regular analgesia use Systems review: To include: Neurology Renal Respiratory Sleep disordered breathing Orthopaedic Eyes	 Yes / No Details: Date of last ophthalmology review: Outcome
Acute Issues: **Refer to MDT if severe crisis/event, >3 admissions in 12 months or consideration of disease modifying therapy**	
Has the patient undergone any surgery in the previous 12 months?	Yes / No If yes, details:

Number of acute presentations: • **ED attendances** • **Unplanned inpatient admissions** **Total number of bed days in hospital:** **Number of painful crises managed at home:** Further information regarding crises (including triggers):	

Serious Adverse Event: Please highlight as appropriate and refer to HCC MDT for discussion

Cardiac Failure		Ischaemic Stroke		Haemorrhagic Stroke	
DHTR: Associated with new antibody		DHTR: Associated with known antibody		DHTR: Not thought to be antibody related/ Hyperhaemolysis	
Intrauterine Death		Pneumococcal Infection			

Relevant clinical details:

Other new co-morbidities: *Can refer to HCC MDT to discuss, but not mandated.*

Sexual health and Psychosocial review:

In the past 12 months, has the patient conceived/fathered a child? ***If Yes: What was the outcome:***				Yes	No
Live Birth?	Yes			No	
Additional information (if relevant):	*PreTerm (<36/40 delivery)*	Yes	No	Spontaneous Miscarriage	Therapeutic abortion
	Mode of delivery	Caesarean Section	Vaginal Delivery		
Discuss fertility/contraception/pre-pregnancy counselling/pregnancy complications					

Psychosocial review (highlight input required if applicable)	Referred to Psychology (not seen)	Psychology review ongoing or completed	Other Mental health input

Notes:

Transfusion review: **<u>Refer to MDT if hyperhaemolysis or other complex transfusion issue</u>**

Has the patient ever been transfused?	Yes / No If yes: Details
Does the patient have known red cell antibodies?	Yes / No If yes: Details

Hospital number: Date of review:

Has the patient received red cell transfusions during the last 12 months? Method of transfusion If on long term transfusion, review and document regarding: • Indication • Frequency and units transfused • Vascular access • Efficacy (pre -HbS% levels)	Yes / No If yes: Indication: Top up transfusion: Long Term □ Urgent/one off □ Exchange transfusion: Long Term □ Urgent/one off □

Vaccination and Medication review:

Vaccination Review: Tick if administered and provide date (if known):

Influenza	□	Menitorix (Meningitis C + HiB)	□
Pneumovax (PPV 23)	□	Prevenar (PCV13)	□
Hep B	□	COVID	□
Meningitis B	□	Other (please specify)	□
Men ACWY	□		

Hydroxycarbamide:

Eligible	Yes	No
Offered	Yes	No
Offered and declined	Yes	No
Currently Taken	Yes	No

Novel Therapies and Clinical Trials:

Eligible	Yes	No
Offered	Yes	No
Offered and declined	Yes	No
Currently Taken	Yes	No

If on Hydroxycarbamide: (MTD = Maximum Tolerated Dose) Indication: Current dose: When was last dose increase: What is the MTD? HbF%:	If Yes: Which therapy/trial: Response to therapy: Yes / No Further information:

Allergies?: Yes / No

Details:

Penicillin V	
Folic Acid	
Vitamin D	
ACE inhibitor	

Other regular medication:

Observations and Examination findings:

BP		Cardiovascular: Respiratory: Spleen size: Liver size: Leg ulcers? Other (please specify)
Oxygen Sats		
HR		
Weight		
Height		

Hospital number: Date of review:

Routine Investigations: **Refer to MDT if significant proteinuria, eGFR <60ml/min, persistently abnormal LFTs or pulmonary hypertension on echo**

Mandatory bloods: Repeat annually unless otherwise stated		
Test	*Date*	*Result*
LFTs		
U&E		
Haemoglobin		
Vitamin D		
Bone Profile		
Ferritin		

Echocardiogram: *Repeat every 3-5 years unless previously abnormal or new symptoms*	
Date:	*Result:* (Document TRV if reported)

Urine testing: *Annual PCR required if dipstick protein or patient on chelation*	
Date:	*Result:*

Tests required for regularly transfused patients Not applicable □		
Test	*Date*	*Result*
Virology		
TFTs		
FSH/LH		
Testosterone		
GTT		

Specialist Investigation **Refer to MDT if abnormal MRI/MRA, sleep study requiring intervention or worsening iron overload**

All patients (if indicated):		
Test	*Date*	*Result*
MRI/MRA		
Pulmonary Function		
Sleep Study		
Other (please specify)		

Selected Regularly Transfused patients:			
Test	*Indicated:*	*Date*	*Result*
Ferriscan	Yes / No		
Cardiac T2*	Yes / No		

Management Plan

Investigations required?	Yes / No Details:
Specialist referral indicated?	Yes / No Details:
Other actions required/discussed?	Yes / No Details:
Interested in research studies?	Yes / No
MDT referral required? *(Note if NHP referral required or completed)*	Yes / No
Follow up appointment	

Hospital number: Date of review: