SCHEDULE 2 – THE SERVICES

A. Service Specifications

Service Specification No:	170126S
Service	Specialist Haemoglobinopathy Services (All ages) • Specialist Haemoglobinopathy Teams
Commissioner Lead	*For local completion*
Provider Lead	*For local completion*

1. Scope

1.1 Prescribed Specialised Service

This service specification covers Specialist Haemoglobinopathy Teams (SHT) to support the provision of specialist and non-specialist haemoglobinopathy services (All ages).

This specification should be read in conjunction with service specification 1644.

1.2 Aim of the Service

The overall aim of the service is to reduce levels of morbidity and mortality and improve the experience of all haemoglobinopathy patients by reducing inequities and improving timely access to high quality expert care.

Specialist haemoglobinopathy services include all care provided by specialist haemoglobinopathy teams including inpatient care where the cause of admission is related to haemoglobinopathy and outreach when delivered as part of a provider network. This applies to provision in adults and children.

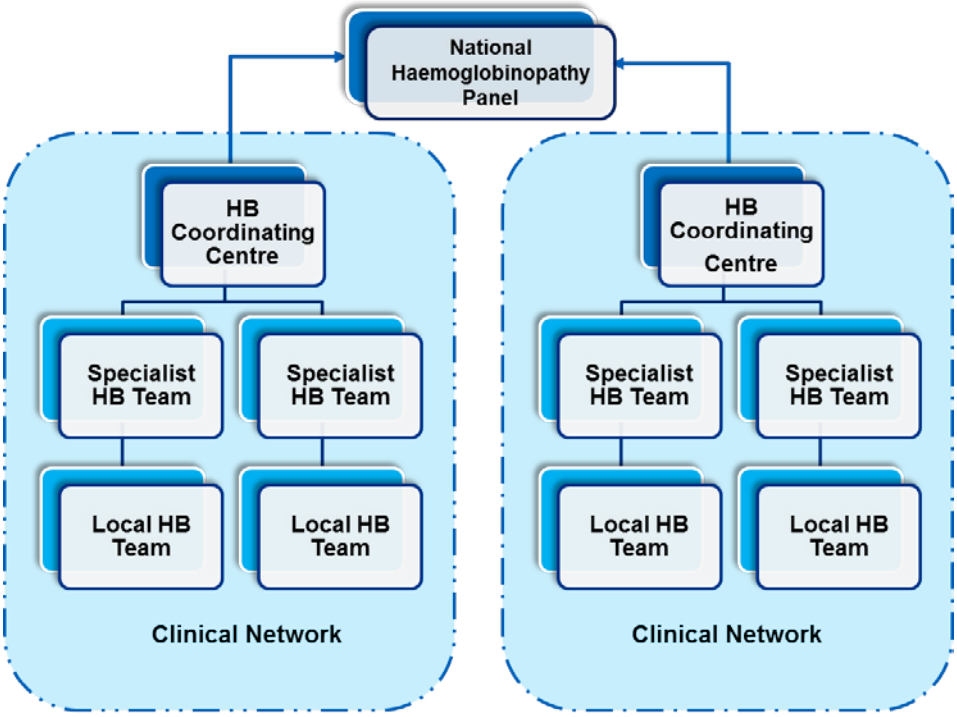
The purpose of this specification is to outline the responsibilities of Specialist Haemoglobinopathy Teams (SHTs) and the relationships that need to be in place with the local Haemoglobinopathy Coordinating Centre (HCC), the wider health economy and patients.

This specification applies to: all children and adults with Sickle Cell Disease and Thalassaemia. It also applies to children and adults with other inherited anaemias if they require intermittent or long term transfusion and/or chelation therapy. This includes Blackfan Diamond anaemia, pyruvate kinase deficiency and congenital sideroblastic anaemia amongst other rare disorders.

1.3 Specialist Haemoglobinopathy Teams

The SHTs will deliver care with local Haemoglobinopathy Teams LHTs and other local hospitals and care providers for a defined caseload of patients within a network of care. All English Trusts treating patients with a haemoglobinopathy are expected to be part of a network,

The diagram below describes the network and relationship between the teams. Depending on location and prevalence, a HCC could be responsible for providing network support for one, two or several SHTs.



The Specialist Haemoglobinopathy Team (SHT) will work with a specific Haemoglobinopathy Coordinating Centre (HCC) to ensure that the roles and responsibilities for its caseload of patients are clear.

The SHT will agree and monitor compliance with network care pathways and treatment protocols (elective and emergency) for its caseload of patients. The SHT will also support the provision of coordinated expert care and advice within the network.

The SHT will provide 24/7 advice for other clinical teams both within the hospital and with other local hospitals. This does not mean 24/7 access to specialists, but there must be a minimum standard of 24/7 cover. This may be either directly or as part of a shared-care arrangement with other SHTs, as the aim is to provide equitable access to specialised care.

It will be up to the SHT to decide how best to incorporate and develop on-line and existing sources of advice. NHS Blood and Transplant (NHSBT) may provide information on transfusions, although not advice on whether to transfuse or not. The NHR is being developed, which may enable it to be used as a

resource.

The SHT will support the provision of routine, non-complex care for its local population and will be responsible for ensuring that they all have an annual review. This may be in LHTs and local hospitals, depending on the needs of the individual patient. However, wherever the care is provided, it will follow a consistent approach through network, regional and national collaboration.

The SHT will demonstrate close working with local commissioners and other providers to capitalise on the expertise available outside of SHT (including secondary, primary, community and voluntary) when designing the care pathways and including discharge planning. Failure to comply with this arrangement will be reported to the responsible commissioner.

The SHT, with support from its HCC, will oversee and support the production of a training and development plan for all healthcare staff involved in the delivery of haemoglobinopathy care in its network area. The responsibility for resourcing appropriate training for healthcare staff remains with the employing organisations.

The SHT will ensure all consented patients in their network area are registered on the National Haemoglobinopathy Registry (NHR). The SHT and newborn screening laboratory will be responsible for ensuring all children identified by the neonatal screening programme are incorporated into the care system via the NHR, following the relevant guidelines from the screening programme. The SHT will make sure that individual records are complete and up to date.

1.4 National Haemoglobinopathy Panel

The National Haemoglobinopathy Panel will work alongside the national Specialised Haemoglobinopathy Clinical Reference Group (CRG), HCCs, SHTs and other key bodies in haemoglobinopathy care.

It will provide SHTs and HCCs access to national expert clinical opinion with regard to the treatment of complex patients. Referrals will be accepted via the SHT or HCC MDT arrangements, depending on suitability for the patient and the local network.

HCCs are responsible for constituting and managing the Panel. HCCs may also ask SHTs to nominate clinical representatives/ specialists.

SHTs should have access to Clinical Transfusion Specialist expertise via the National Haemoglobinopathy Panel to advise on patients with complex transfusion needs

The frequency of meetings will depend on the clinical needs of complex patients. The patient's primary consultant or their representative shall present the case for panel discussion.

The NHP will work alongside the Specialised Haemoglobinopathies Clinical Reference Group (CRG), the HCCs, the SHTs and other key bodies in haemoglobinopathies care in order to:

- Drive the delivery of a nationally consistent approach to care envisaged by the CRG and approved by commissioners
- Coordinate the actions taken at SHT and HCC levels to deliver access to specialist oversight and to reduce unwarranted variation
- Provide SHTs and HCCs access to national expert clinical opinion with regard to the treatment of complex patients
- Support the introduction of commissioned innovative therapies by acting as a national panel to consider individual patients most able to benefit and to enable patients have access to these therapies, irrespective of where they live.

The clinical responsibility for a patient remains with the treating clinician.

1.5 National Haemoglobinopathy Registry

This model will be supported by the National Haemoglobinopathy Registry. This will act as a national repository for:

- Data on patient management including annual review
- Patient information including details of local haemoglobinopathy services and contacts
- Guidelines and protocols, including special requirements for blood to reduce risk of red cell antibody formation and for the management of patients who have formed antibodies and are at the risk of delayed haemolytic transfusion reactions
- Educational materials, including the need for special requirements for transfusions
- Information on transfusion management of patients including whether on exchange (whether automated or manual) or top up transfusion and the development of adverse events in particular red cell antibody formation.

Whilst these materials may already be on other websites, inclusion on the NHR will support patients who may access services across the country.

SHTs are responsible for entering data on their own patients into the Registry and for data quality. HCCs will be responsible for auditing and benchmarking data quality in their network.

1.6 How the Service is Differentiated from Services Falling within the Responsibilities of Other Commissioners

Specialist Haemoglobinopathy Services include care provided by the SHT for which an expert multidisciplinary team is required and as outlined in section 2.1 and standards A-C, below.

It does not include routine inpatient, day case and outpatient care, which is funded through existing tariff arrangements.

Routine outpatient care is not funded through specialised commissioning. However, some local pricing arrangements mean SHTs are funded to undertake

some elements of outpatient care, such as annual reviews and the management of individuals with complex needs. Further work on this will be progressed as part of future tariff arrangements.

2. Care Pathway and Clinical Dependencies

2.1 Care Pathway

Haemoglobinopathies and rare inherited anaemias (RIA) are lifelong conditions and patients will access both on-going routine, as well as specialist, care throughout their lifetime. Patients' care will be delivered as close to home as possible. Whilst rare and sometimes complex, the ongoing and routine monitoring and treatment of patients with haemoglobinopathy and RIA can be managed with the use of protocols, pathways and access to specialist expertise either at the SHT or HCC/ national panel level.

The configuration of care provision will be based on networks linked to local prevalence, expertise and availability of service providers; this may include acute hospitals, community care, primary care and the voluntary sector.

Pathways for paediatric and adult thalassaemia, sickle cell and RIAs feature both routine and specialised care. The role of the SHTs is to deliver and support the delivery of both specialised and non-specialised aspects of care.

2.2 Referral

New patients enter the service either as babies (including those notified through the NHS neonatal Sickle Cell and Thalassaemia Screening Programme) or as new arrivals to England notified via GPs, community services, emergency departments and other clinical specialities. Patients may also move geographical area and require registering for local services. In all cases, the care provider should be notified in accordance with the specification for care and should in turn notify the HCC.

2.3 Care

SHTs are required to deliver and support the delivery of both specialised and non-specialised aspects of care. Patients enter paediatric and adult specialist care through the neonatal screening programme and or new arrivals into UK by referral from a local hospital or transfer from other SHTs.

Responsibility for planning and overseeing the delivery of care rests with the SHT. All SHTs have to enable access to the same level of care. For some services this will be delivered directly by the SHT. For others, the SHT might make arrangements:

- with LHTs in the network especially those which have a significant patient caseload and well developed services

- with another SHT, where it cannot provide a particular aspect of care.

For both, this will be through a sub-contracting arrangement and such providers will be reimbursed to assist in the delivery of these functions. It is the responsibility of SHTs to establish local agreements with providers in accordance with national standards and guidelines. As a minimum, all SHTs should provide:

- Paediatric and adult out-patient review and care; annual reviews; referral for specialist diagnostic investigations; discussion of disease modifying treatments; discussion of new treatments and new trials; and neurocognitive assessment and review.
- Transition care from paediatric to adult services
- Support from psychologist with specialist interest in haemoglobinopathies
- Specialist support and advice on conditions such as transfusion reactions; severe or recurrent painful vaso-occlusive episodes; sickle acute chest syndrome and aplastic crisis
- Advice on complex care surgery
- Initiation of hydroxycarbamide treatment, blood monitoring and dose escalation as appropriate
- Advice on acute organ failure
- Transfusion management including decisions regarding initiation and cessation of elective transfusion programmes
- Prescription and routine monitoring of iron chelating drugs

Providing clear pathways exist, an SHT may refer the following interventions to other SHTs within their network:

- Neurocognitive assessment and review
- MRI assessment of liver and cardiac iron
- Management of complications related to iron overload and management of endocrine and growth
- Management of complex patients and those with co-morbidities. This may entail working with other SHTs/HCC to establish expert clinics, such as renal and cardiac
- Advice and referral for stem cell transplant, novel and curative therapies
- Specialist advice for the management of pregnancy in conjunction with expert obstetric teams.

2.4 Pregnancy

All women of child bearing age should receive personalised pre-pregnancy and maternity care planning from specialised services. Therefore, women with haemoglobinopathies must be referred immediately once they are pregnant to a high risk obstetric clinic In addition the patient must be reviewed early on within their first trimester by the SHT team to monitor any disorder/disease specific aspects of

pregnancy management; this must include access to termination of pregnancy and specialist advice regarding contraception. The individualised care plan must cover the antenatal, intrapartum and postnatal periods. It must include clear instructions for shared care with secondary services, including escalation and transfer protocols and clear guidelines for planned and emergency delivery.

2.5 Interdependence with other Services

Within the defined network area, interdependencies with other services include:

- HCCs, other SHTs, LHTs and local care services
- National Haemoglobinopathy Panel
- Acute providers
- Primary care
- Community healthcare services
- Antenatal and new-born regional screening programmes
- Genetic counsellors and specialists
- Local authority and public health
- Education providers
- Social care
- Voluntary sector
- Sickle Cell and Thalassaemia Screening Programme

3. Population Covered and Population Needs

3.1 Population covered by this specification

This specification will apply to: all children and adults with Sickle Cell Disease and Thalassaemia and other inherited anaemias requiring transfusion and chelation therapy such as Blackfan Diamond anaemia, pyruvate kinase deficiency and congenital sideroblastic anaemia amongst other rare disorders.

The service outlined in this specification is for patients ordinarily resident in England*; or otherwise the commissioning responsibility of the NHS in England (as defined in Who Pays?. Establishing the responsible commissioner and other Department of Health guidance relating to patients entitled to NHS care or exempt from charges). **Note**: for the purposes of commissioning health services, this excludes patients who, whilst resident in England, are registered with a GP Practice in Wales, but includes patients resident in Wales who are registered with a GP Practice in England.

3.2 Understanding Local Population Needs

There are about 1,500 patients with thalassaemia and 15,000 with SCD in England and circa 1,000 patients with rare inherited anaemias. A large number are under 19 years of age. Around 250 babies are born in England each year with SCD, compared with 20-30 babies with thalassaemia.

Nearly all SCD affected children born in England, and the majority with thalassaemia, will be identified by the NHS Sickle Cell and Thalassaemia Screening

programme. Other new patients may present through immigration or late diagnosis. The patient population is unevenly distributed through the country, making equity of access a priority.

Life expectancy for both conditions is progressively improving and is now likely to be in excess of 50 years. Maximising quality of life is an important factor in the organisation of care and in treatment decisions.

Patients with rare inherited anaemias are scattered throughout England and are often diagnosed late and managed by local haematologists and paediatricians. The numbers of patients in this population are unknown but expected to be relatively small.

Services will need to develop to meet the needs of their local populations. This may mean using outreach and technology to share expertise.

The increasing life expectancy of individuals with these conditions means that the overall prevalence will increase and services will need to develop to meet the needs of older patients with the additional comorbidities encountered with age.

Over time, the advent of new treatments such as gene therapy has the potential to change the prevalence of haemoglobinopathy disorders.

4. Outcomes and Applicable Quality Standards

4.1 Evidence Base

This specification has been developed on the basis of clinical consensus, taking into account other examples of networks and multi-disciplinary teams.

- The standards, guidelines and quality requirements referred to include:
- The National Haemoglobinopathy Project: A guide to Effectively Commissioning High Quality Sickle cell and Thalassaemia Services (2011), East Midlands Specialised Commissioning Group.
- Quality Standards. Health Services for people with Haemoglobin Disorders v3.1. 7. Dec 2017 http://www.wmqrs.nhs.uk/review-programmes/view/haemoglobin-disorders-2014-16-reviews-adults-and-children
- Royal College of Nursing – Caring for people with sickle cell disease and thalassaemia syndromes – a framework for nursing staff (2011).
- Sickle Cell Disease in Childhood – standards and guidelines for clinical care – second edition (2010). First edition 2006.
- Trans-cranial Doppler Scanning for Children with Sickle Cell Disease – standards and guidance (2009).
- Specialised Services National Definitions Set (SSNDS) 3rd edition – specialised haemoglobinopathy services (all ages) – Definition No. 38 (2009).
- NHS Sickle Cell and Thalassaemia Screening Programme.
- – Handbook for New-born Laboratories January 2017, Handbook for antenatal laboratories Nov 2017
- Standards for the Clinical Care of Children and Adults with Thalassaemia in the

UK (2016) third edition. (2008) – second edition. First edition, 2005. http://ukts.org/standards/Standards-2016final.pdf

- Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK (2018) second edition. First edition 2008.
- Standards for the Linked Antenatal and New-born Screening Programme Second Edition (2011), NHS Sickle Cell and Thalassaemia Screening Programme.
- Sickle cell disease: managing acute painful episodes in hospital, NICE (2012).
- The National Confidential Enquiry into Patient Outcome and Death (NCEPOD) report, A Sickle Crisis? (2008).
- Understanding the Contribution of sickle cell and thalassaemia specialist nurses: a summary report (2012), NHS Sickle Cell and Thalassaemia Screening Programme.
- Transition: Getting it Right for Young People, improving the transition of young people with long term conditions (2006), Department of Health. Gateway reference 5914.
- Spectra Optia for automatic red blood cell exchange in patients with sickle cell disease, NICE Medical technologies guidance [MTG28], March 2016

NHS Outcomes Framework Domains

Domain 1	**Preventing people from dying prematurely**	**X**
Domain 2	**Enhancing quality of life for people with long-term conditions**	**X**
Domain 3	**Helping people to recover from episodes of ill-health or following injury**	**X**
Domain 4	**Ensuring people have a positive experience of care**	**X**
Domain 5	**Treating and caring for people in safe environment and protecting them from avoidable harm**	**X**

4.2 Quality Indicators

Number	Indicator	Data Source	Outcome Framework Domain	CQC Key question
Clinical Outcomes				
101	Trans Cranial Doppler (TCD) monitoring	Provider via SSQD	2, 3	Safe, effective, caring
102	Timeliness of pain relief in sickle cell disease	Provider via SSQD	2, 3	Safe, effective, caring, responsive
103	Neonatal screening	Provider via SSQD	2, 3	Safe, effective, caring, responsive
104	New born treatment	Provider via SSQD	2, 3	Safe, effective, caring, responsive
105	Proportion of patients that receive an annual review	Provider via SSQD	2, 3	Safe, effective, caring
106	Assessment of adequacy of chelation - adult	Provider via SSQD	2, 3	Safe, effective, caring
107	Assessment of adequacy of chelation - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
108	Assessment of adequacy of chelation - adult	Provider via SSQD	2, 3	Safe, effective, caring
109	Assessment of adequacy of chelation - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
110	Assessment of adequacy of chelation - adult	Provider via SSQD	2, 3	Safe, effective, caring
111	Assessment of adequacy of chelation - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
112	Assessment of adequacy of chelation, sickle only - adults	Provider via SSQD	2, 3	Safe, effective, caring
113	Assessment of adequacy of chelation, sickle only - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
114	Assessment of adequacy of chelation, Thalassaemia only - adult	Provider via SSQD	2, 3	Safe, effective, caring
115	Assessment of adequacy of chelation, Thalassaemia only - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
116	Assessment of adequacy of chelation, Thalassaemia only - adults	Provider via SSQD	2, 3	Safe, effective, caring
117	Assessment of adequacy of chelation, Thalassaemia only - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
118	Length of stay emergency admissions - adults	Provider via SSQD	2, 3	Safe, effective, caring
119	Length of stay emergency admissions - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring

120	Length of stay emergency admissions - adults	Provider via SSQD	2, 3	Safe, effective, caring
121	Length of stay emergency admissions - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
122	Readmissions - sickle cell - adults	Provider via SSQD	2, 3	Safe, effective, caring
123	Readmissions - sickle cell - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
124	Readmissions - thalassaemia - adults	Provider via SSQD	2, 3	Safe, effective, caring
125	Readmissions - thalassaemia - paediatrics	Provider via SSQD	2, 3	Safe, effective, caring
126	Proportion of patients admitted as an emergency with a Haemoglobinopathy condition who die within 30 days of admission.	Provider via SSQD	2, 3	Safe, effective, caring
Patient Experience				
201	The SHT participates in PREM activity and undertakes a patient experience exercise at least annually, reviewing the results which are reported annually to the HCC for review and comment.	Self declaration	4	safe, effective, caring, responsive
202	There is agreed patient information available.	Self declaration	4	safe, effective, caring.
Structure and Process				
301	There is a multi-disciplinary team in place as per the service specification.	Self declaration	1, 2, 3	safe, effective, caring.
302	The MDT meet at least monthly and include core members listed in 301 above.	Self declaration	1, 2, 3	Well-led, safe, effective, caring.
303	The SHT has a process in place for transcranial Doppler (TCD) scanning .	Self declaration	1, 2, 3, 5	safe, effective, caring.
304	There must be transition pathways in place as defined within the service specification.	Self declaration	1, 2, 3, 4, 5	safe, effective, caring.
305	The SHT agree the HCC patient pathways as per the specification.	Self declaration	1, 2, 3, 4, 5	safe, effective, caring.
306	The SHT agree the HCC clinical guidelines as per the specification	Self declaration	1, 2, 3, 5	safe, effective, caring.
307	The SHT participates in local, HCC and national audits as required.	Self declaration	2, 4	Safe, effective
308	The SHT submits data to the National Haemoglobinopathy registry.	Self declaration	2, 4	Safe, effective

Detailed definitions of indicators, setting out how they will be measured, are included in schedule 6.

4.3 Commissioned providers are required to participate in annual quality assurance and collect and submit data to support the assessment of compliance with the service specification as set out in Schedule 4A-B

5. Applicable Service Standards

5.1 Applicable Obligatory National Standards

https://www.nice.org.uk/guidance/qs58
https://www.nice.org.uk/guidance/mtg28

5.2 Other Applicable National Standards to be met by Commissioned Providers

5.3 Other Applicable Local Standards

Section A: Core standards (mandatory), these are the standards that the SHT must meet directly i.e. they must have the clinical expertise and facilities within their organisation.

A1 Clinical Leadership

- Medical Leadership – the SHT will have a named medical lead at consultant level. This must be a haematologist/paediatric haematologist or a paediatrician with expertise in haemoglobinopathies. Dependent on configuration of acute care there may be two medical leads to cover paediatric and adult care.
- The SHT will have a named medical deputy at consultant level responsible for haemoglobinopathy care. There may be two deputies i.e. one each for paediatric and adult care.
- Nursing leadership – the SHT will identify a lead nurse. There may be two deputies i.e. one each for paediatric and adult care. The lead nurse will support all nurses across the SHT and linked hospitals
- The SHT, working with the HCC, will be responsible for data, audit and outcome monitoring for all the patients under its care.

A2 Prevention and management of neurological complications of SCD through transcranial Doppler (TCD) scanning in childhood; specialised neuro- radiology, neurology and neuropsychology services.

The SHT must be able to demonstrate responsibility for:

- The coordination of access to TCD screening for all eligible children.
- The expert clinical management of those children and adults identified at risk of stroke and other neurological impairment to minimise the risk.
- The multidisciplinary team management of complex neurological abnormalities.
- Compliance with training and quality assurance schemes established to support continuous quality improvement.

A3 Expert Multidisciplinary Care for Complex Patients

The SHT is responsible for the management of complex patients using a multidisciplinary team approach. Indicators of complexity include but are not limited to:

- Multi-system disease including organ damage.
- Mono system disease for example hepatic and renal disease.
- Abnormal neurology (see standard A2).
- Psychological and psychosocial problems.

- Pregnancy (see standard A9).
- Surgery (see standard A8).
- Orthopaedic issues.
- Endocrine complications.
- Cardiac complications especially related to iron overload.
- Infection prevention and control requirements.

The multidisciplinary team should include the following professionals:

- Medical lead, nursing representation (acute and community) and psychology.
- The multidisciplinary team may require input from physiotherapy, neurology, cardiology, radiographer and sonographer.
- All patients to be reviewed at least annually by the SHT or under the supervision of the SHT.
- Multidisciplinary team teams will review and oversee the overall progress of all patients with clinical complexities to optimise overall care. Note: for children this will include growth, development and academic achievement.

A4 Initiation, Modification and Cessation of Long-Term Transfusion Regimes and Preventative Therapy in SCD&T

This standard is associated with standard A5

- The initiation, modification and cessation of long-term blood transfusion regimes should be under the responsibility of the SHT.
- Regular administration and monitoring of transfusions should be carried out locally wherever possible and sessions should be designed to fit around the requirements of patients wherever possible (e.g. evening clinics and out of hours transfusion).
- Access to automated red cell exchange transfusion for sickle cell patients needing long term transfusion therapy should be available

A5 Initiation, Modification and Cessation of Long-Term Iron Chelation. Monitoring of Complications of Chelation

This standard is associated with standard A5

- The initiation and amendment of long-term iron chelation regime is the responsibility of the SHT.
- The regular administration of iron chelation regime can be carried out locally wherever possible.
- The SHT will have access to cardiac and liver magnetic resonance scanning (this does not necessarily need to be on site).
- The SHT will have access to neuro-psychological, psychosocial and social worker support for patients that struggle with adherence.

A6 Acute Management of Severe and Life Threatening Complications of SCD and Thalassemia

The SHT will develop guidelines to implement the NICE guidance on the management of acute painful episodes. The SHT will be able to clinically manage or have referral pathways for the following range of complications for SCD:

- Fulminant sepsis.
- Acute sickle lung syndrome.

- Acute splenic or hepatic sequestration.
- Ischaemic and haemorrhage stroke.
- Subarachnoid haemorrhage.
- Acute renal failure.
- Multi-organ failure.
- Billiary obstruction.
- Priapism.
- Post-transfusion hyperhaemolysis and severe delayed haemolytic transfusion reactions
- Acute ophthalmological complications (for example complications of sickle retinopathy/central retinal artery occlusion).
- Osteonecrosis of major joints (for example hip, shoulder).

The SHT will be able to manage or have agreed referral pathways the following complications for thalassaemia and rare inherited anaemias:

- Heart failure and cardiac arrhythmias.
- Infection prevention and control.
- Post-splenectomy sepsis.
- Acute endocrine disturbances (for example hypocalcaemic tetany).
- Acute hepatic decompensation.

The SHT will offer formal liaison support to any acute provider within its network area.

A7 Long-Term Specific Therapy for Severe Complicated SCD and Thalassaemia (Complex Long-Term Conditions Management)
This standard links to standard A3 relating to annual reviews and multidisciplinary team management of complex patients.
The SHT will be able to clinically manage (or have referral pathways within the network as agreed by the HCC) a range of progressive and often irreversible complications in both outpatient and in-patient settings. In SCD, these include:

- Stroke.
- Chronic sickle lung syndrome.
- Pulmonary hypertension.
- Chronic renal impairment.
- Avascular necrosis of the hips, spine and shoulders.
- Retinopathy.
- Chronic leg ulceration.
- Chronic pain.

In thalassaemia major and intermedia, these complications include:

- Endocrine dysfunction (growth hormone deficiency), hypogonadotrophic, hypogonadism, hypothyroidism, hypoparathyroidism, diabetes, (which may require insulin treatment).
- Cardiac dysfunction.
- Chronic liver disease (cirrhosis portal hypertension, hepatic failure, hepatocellular carcinoma, often associated with transfusion- transmitted

hepatitis B or C).
- Bone problems (avascular necrosis, osteoporotic fractures of the hips and spine, disc disease).
- Gallstones.
- Leg ulceration.
- Iron overload.
- Pulmonary hypertension.
- Thrombosis.
- Retinal damage.
- Pseudoxanthoma
- Chronic pain.

The SHT must be able to initiate, modify and cease long-term medication regimes. For instance, to prevent or mitigate sickle painful episodes. The monitoring of such drug regimens is not a specialised function but any modification based on the outcomes of that monitoring remains specialised.

The SHT must be able to provide psycho-social/psycho-neurological support to complex patients struggling to manage their condition.

A8 Peri-Operative Management of Sickle Cell and Thalassaemia Patients Requiring Surgery

In principle all elective surgery, and where possible all emergency surgery, should be carried out in at the SHT. For practical purposes this may not be possible or desirable and it will be for the Local Area Team and the SHT to agree surgical pathways.

The SHT will demonstrate close liaison between haematologists, paediatricians, surgeons and anaesthetists. Surgeons and anaesthetists will have experience in the effective peri-operative management of SCD&T patients.

Where a local acute provider is required to deliver an emergency operation, it should liaise with the SHT.

The SHT is required to have pathways in place to manage emergency scenarios.

A9 Management of Pregnant Women with SCD and Thalassaemia

Complex pregnancy refers to any pregnant woman that has SCD or thalassaemia. A high risk carrier couple identified by the Antenatal Screening Programme do not require specialised care during the pregnancy unless a specific complicating factor has been identified.

All high risk pregnancies must be managed through a multidisciplinary team approach between obstetricians and haematologists. Pathways should be agreed by the SHT and HCC.

A10 Clinical Governance and Audit

The SHT will adopt a clinical governance and leadership function. This will include:

- Reporting all adverse events to commissioners and the National Haemoglobinopathy Registry (NHR)
- Reporting adverse transfusion events to SABRE/SHOT
- Undertaking an agreed number of clinical/quality audits as agreed with the HCC
- Participating in any peer review process
- Ensuring compliance with network clinical guidelines and protocols
- Submission of data to support local and national benchmarking (e.g. NHS England quality dashboards)

A11 Patient and Carer Engagement

The SHT will ensure public and patient engagement (PPE) through:

- User or user group representation at meetings
- User involvement in service planning and development

The SHT will promote user feedback and engagement with all healthcare providers.

A12 Education and Research

- The SHT will be able to provide practical training to all relevant clinical staff including junior doctors and nurses and other allied health professionals
- Training for nurses should meet a recognised competency framework for nursing haemoglobinopathy patients.
- All counsellors or healthcare professionals who counsel couples at risk of an affected pregnancy should have undertaken the relevant training.

SHT must demonstrate a research portfolio – possibly linked to clinical and cost effectiveness of certain aspects of care.

In order to deliver this service staff must be supported and trained to build capability for the consistent delivery of high quality, evidence based care for patients. This will be underpinned by robust clinical governance and an educational and training plan to embed learning and professionalism.

A13 Timely Access to Critical Care (Adult)

Unless a Children's Trust, the SHT must have an adult Intensive Therapy Unit (ITU) on site.

A14 Transition

The SHT should develop, provide and oversee a protocol for adolescents transitioning between paediatric and adult services with adequate facilities and staff trained to be sensitive to the special needs of this group of patients and which meets national guidance

A14 Laboratory services

- United Kingdom Accreditation Service (UKAS) / Clinical Pathology Accreditation (CPA) accredited laboratory services with satisfactory performance in the National External Quality Assessment Service (NEQAS) haemoglobinopathy scheme and Medicine and Healthcare Products Regulatory Agency (MHRA) compliance for transfusion should be available at all times (other than genetics services which can be provided in working hours only). SHT must be able to access the range of laboratory tests and transfusion support to manage elective and emergency patients.

- Transfusion laboratories must be aware of the special requirements of haemoglobinopathy patients and ensure that national guidance has been incorporated these into their transfusion guidelines and standard operating procedures.

SECTION B: Collaborative standards (mandatory) – these are standards that the SHT may deliver in collaboration with other SHTs to ensure clinical and cost effectiveness. In addition, some elements will be super-specialised and will be limited to a very small number of providers nationally.

B1 Timely Access to Critical Care (Paediatric)

If the SHT does not have a Paediatric intensive Care (PICU) on site, they must demonstrate formal arrangements with either other SHTs or other acute Trusts with PICU.

B2 Access to a Comprehensive range of Clinical Specialists Experienced in Treating Haemoglobinopathy Patients

The SHT must have demonstrable arrangements in place that recognise the challenges that patients face in travelling long distances, access to the following specialists:

- Experienced nurse specialising in the conditions
- Acute and chronic pain team
- Consultant cardiologist
- Consultant respiratory physician
- Consultant teams with experience in managing pulmonary hypertension
- Consultant nephrologist and access to renal replacement therapy and transplant
- Consultant hepatologist
- Consultant urologist with expertise in managing priapism, erectile dysfunction
- Consultant neurologist and acute stroke service
- Consultant ophthalmologist
- Consultant endocrinologist
- Contraception and sexual health services

- Genetic counselling and fertility services
- Consultant obstetrician
- Consultant general surgeon
- Tissue viability service/leg ulcer clinic
- Psychologist and other mental-health services

B3 Access to Bone Marrow and Stem Cell Transplantation

Both of these interventions are deemed super-specialised and will be available at only a few centres nationally. The SHT will have formal processes in place to consider patients for such clinical interventions and to refer to the National Panel.

6. Designated Providers (if applicable)

To be completed following provider selection exercise.

7. Abbreviation and Acronyms Used in this Document

HCC: Haemoglobinopathy Coordinating Centre
SHT: Specialist Haemoglobinopathy Team
LHT: Local Haemoglobinopathy Team
NHP: National Haemoglobinopathy Panel
HEE: Health Education England
SCD: Sickle Cell Disease
MDT: Multi-Disciplinary Team
CRG: Clinical Reference Group
TCD: Trans-Cranial Doppler
MRI: Magnetic Resonance Imaging
RIA: Rare Inherited Anaemia