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NHS Sickle Cell and Thalassaemia Screening Programme

Publications Launch Webinar

Thursday 28th January 2021

Public Health England

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Welcome to NHS Sickle Cell and Thalassaemia Screening Programme's Live Webinar Thursday 28th January 2021

Chair: Professor Dame Elizabeth N Anionwu DBE CBE FRCN FQNI PHD
Emeritus Professor of Nursing, University of West London

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Menti Questions

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Counselling, Knowledge & Skills Guidelines - Background & introduction to the new Guidelines

Dr Lola Oni OBE
London North West University Healthcare NHS Trust
Chair of working Group

BACKGROUND

QUALITITES OF A GENETIC COUNSELLOR:

1. **KNOWLEDGE OF THE GENETIC DISEASE**
2. **ABILITY TO PROVIDE & IMPART UP TO DATE INFORMATION**
3. **CAPACITY TO LISTEN AND BE EMPATHETIC**
4. **KNOWLEDGE OF MECHANISMS AVAILABLE TO SUPPPORT INDIVIDUALS, FAMILIES & COMMUNITIES**
5. **ABILITY TO TAKE ACCOUNT OF SOCIO-CULTURAL DIVERSITY AND RESPOND ACCORDING TO CLIENT'S NEED**
6. **ABILITY TO BE NON-DIRECTIVE AND NON JUDGMENTAL**
7. **ABILITY TO COPE WITH REPETITION**
8. **BE ACCOUNTABLE FOR ACTIONS AND OMISSIONS**
9. **MAINTAIN CONFIDENTIALITY**

KEY OBJECTIVE OF GENETIC COUNSELLING

To provide the family with a realistic view of the situation, the nature of the inherited disorder already manifested in a family member, the risk of occurrence or re-occurrence, what this may mean in practical terms for all concerned, and to assist the family through what is often a difficult phase of their life…

Aim & Objectives

The aim of the competency is to update and provide practice guidance for those involved in providing genetic counselling services for those with and at-risk of sickle cell & thalassaemia. Practitioners include specialist nurses, antenatal & newborn screening coordinators / midwives, genetic counsellors and other relevant health and allied care professionals involved in providing the specialist counselling service

Promote development of skills, knowledge and competence in order to meet the needs of the client group and enable formalised assessment of individuals.

And, enhance the practitioner's ability to meet the requirements of their professional code e.g. NMC

Title: SCT Counselling knowledge and skills

Core competencies:

1. **Identification** - Identify individuals and families who will benefit from testing and counselling

2. **Communication** - Understand the importance of effective communication in supporting individuals and families with, or at risk of having a baby with, sickle cell and thalassaemia

3. **Supporting personal informed choice** - Advocate for the rights of all individuals to make a personal informed choice

4. **Knowledge and awareness** - Understand the genetic basis and clinical implications of sickle cell and thalassaemia

Title: SCT Counselling knowledge and skills

5. **Use of genetic information, tests and results** - Care and support individuals and their families prior to, during and following genetic testing

6. **Maintaining SCT competence** - Maintaining and updating SCT knowledge and skills through lifelong learning

7. **Accessing information and resources** - Obtaining and using information to support credible, current communication about sickle cell and thalassaemia

8. **Ongoing support** - Providing ongoing support to individuals and families with sickle cell and thalassaemia

Additional

- **Learning Outcomes and practice indicators for assessment against each competency**
- **Formal assessment tool**
- **Mapping to National Occupational Standards**
  UK Workforce National Occupational Standards for Genetics and Genomics in Clinical Practice for non- genetics healthcare staff
- **Information on sources of further support and learning**

Thank you

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A Patients Perspective

Laurel Brumant – Sickle Cell Society

Public Health England

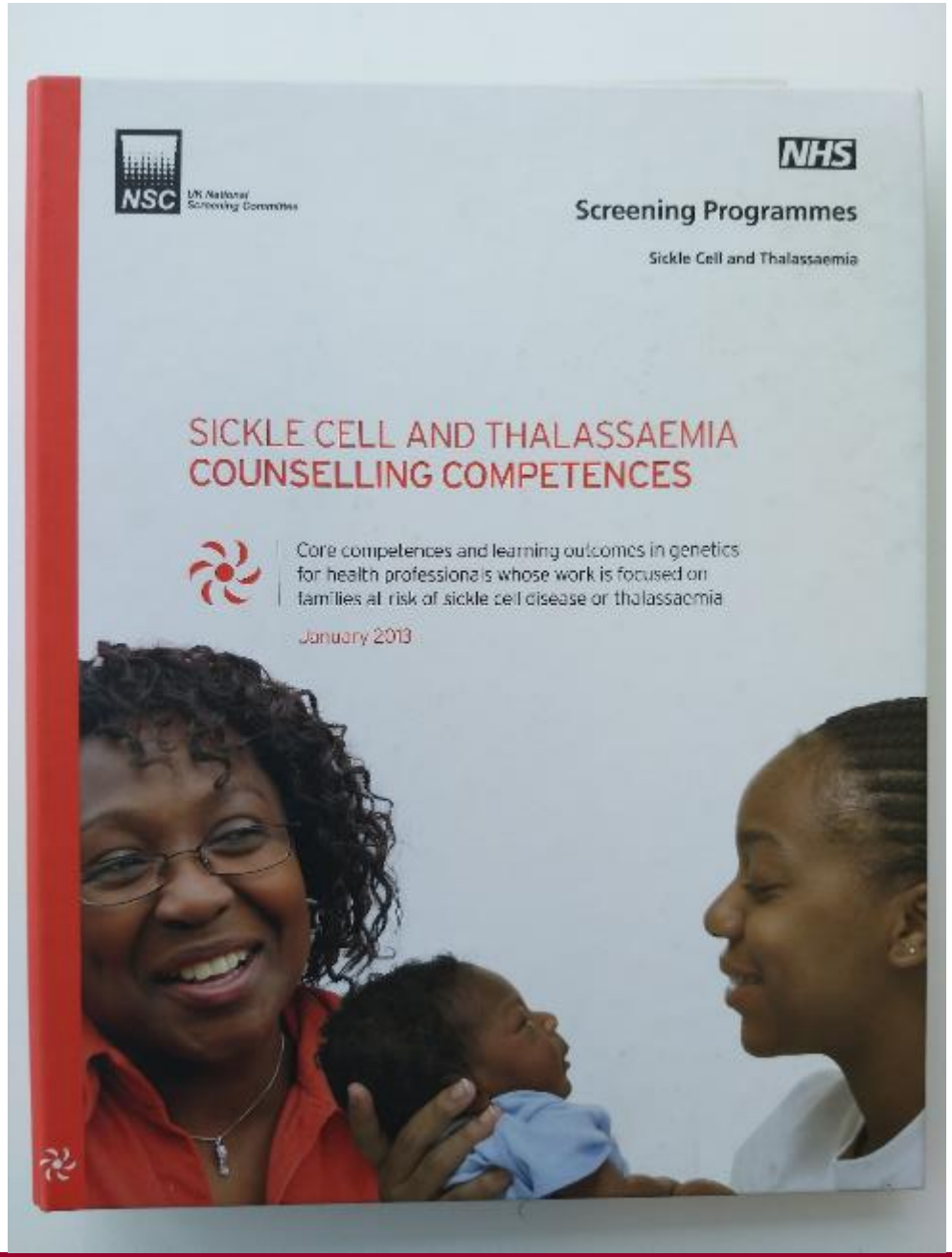
Protecting and improving the nation's health

Project delivery: Sickle cell and thalassaemia (SCT) counselling knowledge and skills

Jamili Miah – Project and Implementation Lead NBS Screening Programme

Empowering to deliver together

Organisations



Old SCT Counselling Competences

A4 folder
Over 100 pages
Too much text / repetitive
Misleading title
Nowhere to capture evidence of accomplishment
Not easily accessible online
Not well implemented

User needs

- a shorter, concise set of competences that can be achieved within very busy workloads
- an assessment record that shows trainee practitioners are working towards achieving the competences
- signposting to education and training resources for trainee practitioners, especially those working in low prevalence areas where access to an expert clinical network is not as easy

Timeline

Sep 2018

- First working group meeting
- Chair - Dr Lola Oni

March 2019

- Final working group meeting
- Piloted
- Needed to be more user friendly

Sept 2019

- Finalised
- Published
- Blog

Sickle cell and thalassaemia counselling knowledge and skills

Information and resources for health professionals who provide counselling for people at risk of having a baby with sickle cell disease or thalassaemia.

Published 28 September 2020
From: Public Health England

Documents

SCT counselling knowledge and skills overview

Ref: PHE publications gateway number GW-1587
HTML

SCT counselling knowledge and skills guide

Ref: PHE publications gateway number GW-1587
HTML

SCT counselling knowledge and skills assessment record

Ref: PHE publications gateway number GW-1587
MS Word Document, 140KB

Additional resources

Ref: PHE publications gateway number GW-1587
HTML

Job description example

Ref: PHE publications gateway number GW-1587
HTML

SCT example counselling form

Ref: PHE publications gateway number GW-1587
PDF, 894KB, 4 pages

This file may not be suitable for users of assistive technology.

▶ Request an accessible format.

Mapping to national occupational standards

Ref: PHE publications gateway number GW-1587
ODT, 97.8KB

This file is in an OpenDocument format

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Documents

SCT counselling knowledge and skills overview

Ref: PHE publications gateway number GW-1587
HTML

SCT counselling knowledge and skills overview

Published 28 September 2020

Contents

1. Knowledge and skills required
2. Definitions
3. Renewal and review of learning

Print this page

This guidance describes the provision of high quality, competent and sustainable counselling services for people potentially at risk of having a baby with a clinically significant haemoglobinopathy.

Haemoglobinopathies include haemoglobin variants such as sickle cell and the thalassaemias such as beta thalassaemia. These conditions occur as a result of a genetic alteration (mutation) in the haemoglobin gene. More than 1,000 mutations have been identified that result in haemoglobinopathies.

Each individual has two copies (alleles) of the haemoglobin gene, one from each parent Most haemoglobin mutations are clinically insignificant in the carrier state. That is wher an individual inherits one copy of the usual haemoglobin gene and one unusual (altered) haemoglobin gene. For example, an individual who inherits normal haemoglobin A and sickle haemoglobin S is a sickle cell carrier (HbAS).

Other common carrier states include:

- haemoglobin C carrier (HbAC)
- haemoglobin D carrier (HbAD)
- haemoglobin E Carrier (HbAE)
- beta thalassaemia carrier (HbAβThalassaemia)

The above are all healthy carrier states and individual carriers do not experience any clinical manifestation or symptoms.

However, there is a genetic relevance to being a carrier. When the altered haemoglobin gene is passed on from a carrier parent to a child in combination with another altered haemoglobin gene from the other parent, the combination may result in a mild or severe

2. Definitions

There are 8 **core competences** relating to the knowledge, skill or attitude required for a professional to perform their role.

The **learning outcomes** are the desired end result of education or training aimed at enabling the professional to become competent.

The **practice indicators** provide a measure of how the competence would be demonstrated in a practice setting.

To enable practitioners to develop the necessary competences, links to additional learning resources are provided.

We have also created a pro forma job description for a haemoglobinopathy specialist as an example of how the competences might be incorporated into a job description and appraisal.

3. Renewal and review of learning

Practitioners should discuss and demonstrate the practical indicators with their assessor. This can be in a clinical setting or during a formal appraisal. The assessor should define the time frame by which they expect a practitioner to have met all 8 core competences. Ideally this should be within the first year of being appointed to the relevant post.

Once all 8 core competences have been met, training should be reviewed at least every 3 years.

Assessors are those with knowledge, skill and experience in sickle cell and thalassaemia genetic counselling and who have completed the Genetic Risk Assessment and Counselling or equivalent course.

Practitioners may have more than one assessor when completing the assessment document in an agreed timeframe.

[SCT counselling knowledge and skills guide]

Ref: PHE publications gateway number GW-1587
HTML

Guidance
SCT counselling knowledge and skills guide
Published 28 September 2020

Contents
1. Identification
2. Communication
3. Supporting personal informed choice
4. Knowledge and awareness
5. Use of genetic information, tests and results
6. Maintaining SCT competence
7. Accessing information and resources
8. Ongoing support

Print this page

1. Identificat

1.1 Core compet
will benefit fron

Learning outcome: und
predisposition to gene
Practice indicators:

1. Obtain family history
2. Draw a multigenerati
3. Document potentiall
a family history. To i

Learning outcome: b
offspring, at risk of SC
Practice indicators:

1. Explain the relevant i
2. Interpret a multigene
3. Describe the factors
on family history. To i
transplant and surrog
4. Explain why some inc
history of that condit

Learning outcome: be
their family
Practice indicators:

1. Outline the impact ar
2. Describe how an indi
condition.
3. Explain how to gain assistance from genetics and other relevant specialists and peer support services.

2. Communication

2.1 Core competence: understand the importance of effective communication in supporting individuals and families with, or at risk of having a baby with SCT

Learning outcome: understand the influence of ethnicity, culture, religion and ethical perspectives

Practice indicators:

1. Describe the potential effects of culture, ethnicity, religion and values on an individual's use of genetic information and services.
2. Explain the importance of sensitive communication when exploring cultural, religious and ethical perspectives.
3. Demonstrate the appropriate use of SCT information to meet an individual's needs taking into consideration their cultural, ethnic, religious and ethical perspectives.

SCT counselling knowledge and skills assessment record

Ref: PHE publications gateway number GW-1587
MS Word Document, 140KB

Title		SCT counselling knowledge and skills – **1. Identification**				
Core competence		**Identify individuals and families who will benefit from testing and counselling.**				
Learning outcomes	**Assessment outcome/comments**	**Initials and date**	**Assessment outcome/comments**	**Initials and date**	**Final assessment outcome/comments**	**Initials and date**
1. Understand the importance of family history in assessing predisposition to genetic conditions						
2. Be able to make genetic risk assessments for individuals, or their offspring, at risk of sickle cell and thalassaemia						
3. Be aware of the potential impact of sickle cell and thalassaemia on an individual and their family						
4. Be able to make appropriate referrals to genetic services and other agencies that are available at local/regional levels						

7

Title		SCT counselling knowledge and skills – **2. Communication**				
Core competence		**Understand the importance of effective communication in supporting individuals and families with, or at risk of having a baby with, sickle cell and thalassaemia.**				
Learning outcomes	**Assessment outcome/comments**	**Initials and date**	**Assessment outcome/comments**	**Initials and date**	**Final assessment outcome/comments**	**Initials and date**
1. Understand the influence of ethnicity, culture, religion and ethical perspectives						
2. Be able to communicate effectively with individuals, families and colleagues`						

8

Title		SCT counselling knowledge and skills – **3. Supporting personal informed choice**				
Core competence		**Advocate for the rights of all individuals to make a personal informed choice**				
Learning outcomes	**Assessment outcome/comments**	**Initials and date**	**Assessment outcome/comments**	**Initials and date**	**Final assessment outcome/comments**	**Initials and date**
1. Recognise the importance of providing sickle cell and thalassaemia information and support fairly and accurately without coercion or personal bias.						
2. Respect personal informed choice						
3. Be aware of potential misuse of sickle cell and thalassaemia information						

Title		SCT counselling knowledge and skills – **4. Knowledge and awareness**				
Core competence		**Understand the genetic basis and clinical implications of sickle cell and thalassaemia**				
Learning outcomes	**Assessment outcome/comments**	**Initials and date**	**Assessment outcome/comments**	**Initials and date**	**Final assessment outcome/comments**	**Initials and date**
1. Understand the genetic basis of sickle cell and thalassaemia						
2. Recognise the clinical implications of sickle cell and thalasssaemia						
3. Educate health and allied practitioners about sickle cell and thalasssaemia						

Guidance

Ac

Publis

6. Maintaining SCT counselling knowledge and skills

Introduction to the Counselling Skills used in Genomic Medicine

Category: Taught courses Tags: Communicating genomics, Genomic testing

This course provides an introduction to the knowledge, communication and counselling skills as well as the appropriate attitudes and behaviours necessary to support patients and their families whose care will be influenced by genomic investigations.

This module forms part of the HEE Genomics Education Programme's Master's in Genomic Medicine framework.

Description | Funding rules | Timetable | FAQs | Make an enquiry

During this course, you will learn how to communicate and provide appropriate support to patients and their families. There will opportunities to develop these essential counselling skills through the use of role play in theoretical and practical sessions. The course also explores the importance of recording a family history and ways to communicate pathogenic and/or uncertain results.

AT A GLANCE

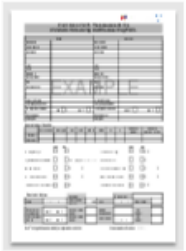
- Up to 6 weeks
- Accredited
- Part Time

introduces the knowledge, communication and counselling skills as well as the appropriate attitudes and behaviours necessary to support patients and their families whose care will be influenced by genomic investigations.

1). See rs) –

directed individuals' sections (e.g. [redacted]).

See also list of Health Education England resources in section 6 below.



SCT example counselling form

Ref: PHE publications gateway number GW-1587
PDF, 894KB, 4 pages

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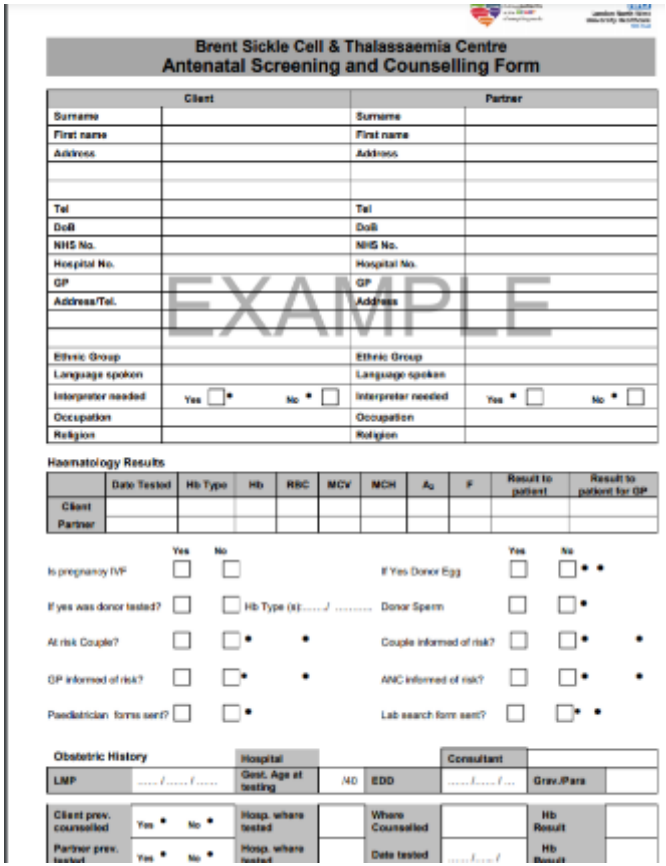
▶ Request an accessible format.



Brent Sickle Cell & Thalassaemia Centre
Antenatal Screening and Counselling Form

Client		Partner	
Surname		Surname	
First name		First name	
Address		Address	
Tel		Tel	
DoB		DoB	
NHS No.		NHS No.	
Hospital No.		Hospital No.	
GP		GP	
Address/Tel.		Address	
Ethnic Group		Ethnic Group	
Language spoken		Language spoken	
Interpreter needed	Yes ☐ No ☐	Interpreter needed	Yes ☐ No ☐
Occupation		Occupation	
Religion		Religion	

EXAMPLE

Haematology Results

	Date Tested	Hb Type	Hb	RBC	MCV	MCH	A_2	F	Result to patient	Result to patient for GP
Client										
Partner										

	Yes	No		Yes	No
Is pregnancy IVF	☐	☐	If Yes Donor Egg	☐	☐
If yes was donor tested?	☐	☐ Hb Type (s):...../.....	Donor Sperm	☐	☐
At risk Couple?	☐	☐	Couple informed of risk?	☐	☐
GP informed of risk?	☐	☐	ANC informed of risk?	☐	☐
Paediatrician forms sent?	☐	☐	Lab search form sent?	☐	☐

Obstetric History		Hospital			Consultant	
LMP	...../...../.....	Gest. Age at testing	/40	EDD	...../...../...	Grav./Para
Client prev. counselled	Yes ☐ No ☐	Hosp. where tested		Where Counselled		Hb Result
Partner prev. tested	Yes ☐ No ☐	Hosp. where tested		Date tested	...../...../	Hb Result

Details of Children

Name	DoB	Hospital	M/F	Hb Type	Comments

Counselling Details
Date of appointments: [1] ...../...../..... [2] ...../...../..... Gest. age at counselling: ...../40

Attended with partner ☐ Attended alone ☐ Partner attended alone ☐ • Did not attend ☐ •

Partner Screening: Blood sample taken ☐ • Laboratory forms given / Sent ☐ • date sent .....

Reason for not attending (if known): .....

EXAMPLE

Information discussed	Yes	No	N/A	Client Initial
1. Difference between blood group and Hb type	☐	☐ •	☐	• .....
2. What is a red blood cell and its function	☐	☐ •	☐	.....
3. Types of haemoglobin (normal and abnormal)	☐	☐ •	☐	.....
4. Population affected and proportion	☐	☐ •	☐	.....
5. Clinical effect of trait/disease	☐	☐ •	☐	.....
6. Genetic and health implications for nuclear and extended family	☐	☐ •	☐	.....
7. Testing offered to other family members	☐	☐ •	☐	.....
8. Client understanding checked	☐	☐ •	☐	.....

Prenatal Diagnosis

Discussed?	☐	☐ •	☐ .....
Offered?	☐	☐ •	☐ .....
Accepted?	☐	☐ •	☐ .....

If PND not accepted, reason given.....
If PND accepted, name of Dr: ..... Centre referred to: ..... Result of PND: .....

At risk- couple letter to parents to inform Centre of birth: Yes ☐ No ☐ If no state reason: .....

Termination of pregnancy? Yes ☐ No ☐ Gestation when PND Declined: .....

Post ToP contact • • •
Outcome of contact: .....

Neonatal Outcome
Registered Name: ..... DoB: ...../...../..... Sex: M ☐ • F ☐ Lab No: .....
NHS No: ..... Neonate Result Hb type: ..... Date Parents informed of baby's result: .....
PND Centre informed of Neonatal result (if relevant): Yes ☐ No ☐

Mapping to national occupational standards

Ref: PHE publications gateway number GW-1587
ODT, 97.8KB

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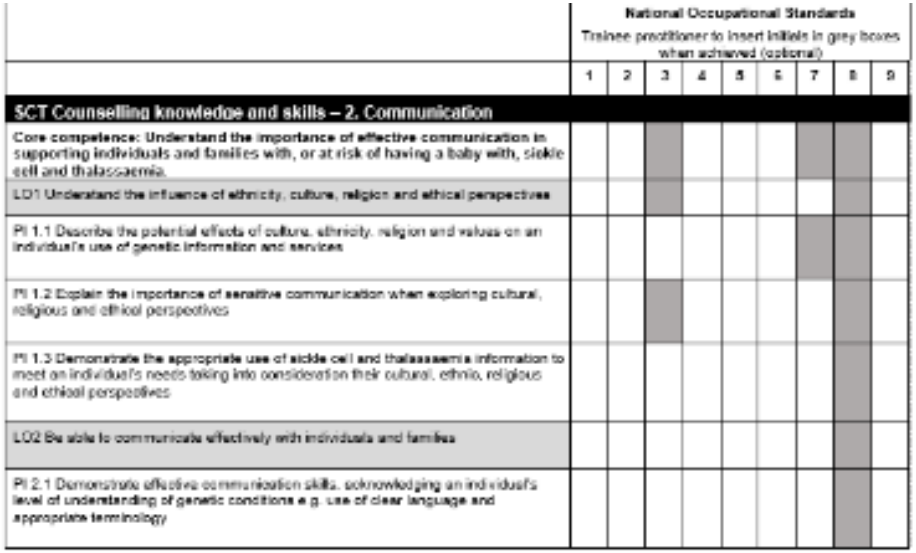
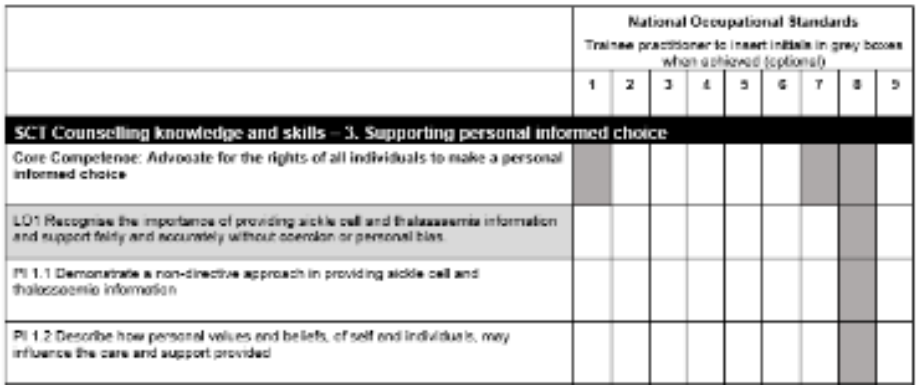
Skills for Health NOS for Genetics and Genomics

1. GTC1.2014 Identify where genetics and genomics are rele
   practice (https://tools.skillsforhealth.org.uk/competence/sh
2. GTC2.2014 Identify individuals with, or at risk of, genetic c
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
3. GTC3.2014 Gather multi-generational family history inform
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
4. GTC4.2014 Use multi-generational family history informati
   pedigree (https://tools.skillsforhealth.org.uk/competence/sh
5. GTC5.2014 Recognise a mode of inheritance in a family
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
6. GTC6.2014 Assess the genetic risk associated with a conc
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
7. GTC7.2014 Organise a test that uses genetic technologies
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
8. GTC8.2014 Communicate genetic and genomic informatio
   families and other healthcare staff
   (https://tools.skillsforhealth.org.uk/competence/show/html/i
9. GTC9 Use genomic information in clinical decisions-makin
   (https://tools.skillsforhealth.org.uk/competence/show/html/i



	National Occupational Standards Trainee practitioner to insert initials in grey boxes when achieved (optional)								
	1	2	3	4	5	6	7	8	9
SCT Counselling knowledge and skills – 2. Communication									
Core competence: Understand the importance of effective communication in supporting individuals and families with, or at risk of having a baby with, sickle cell and thalassaemia.									
LO1 Understand the influence of ethnicity, culture, religion and ethical perspectives									
PI 1.1 Describe the potential effects of culture, ethnicity, religion and values on an individual's use of genetic information and services									
PI 1.2 Explain the importance of sensitive communication when exploring cultural, religious and ethical perspectives									
PI 1.3 Demonstrate the appropriate use of sickle cell and thalassaemia information to meet an individual's needs taking into consideration their cultural, ethnic, religious and ethical perspectives									
LO2 Be able to communicate effectively with individuals and families									
PI 2.1 Demonstrate effective communication skills, acknowledging an individual's level of understanding of genetic conditions e.g. use of clear language and appropriate terminology									



	National Occupational Standards Trainee practitioner to insert initials in grey boxes when achieved (optional)								
	1	2	3	4	5	6	7	8	9
SCT Counselling knowledge and skills – 3. Supporting personal informed choice									
Core Competence: Advocate for the rights of all individuals to make a personal informed choice									
LO1 Recognise the importance of providing sickle cell and thalassaemia information and support fairly and accurately without coercion or personal bias									
PI 1.1 Demonstrate a non-directive approach in providing sickle cell and thalassaemia information									
PI 1.2 Describe how personal values and beliefs, of self and individuals, may influence the care and support provided									

Feedback

Resources are reviewed every 3 years

Email your feedback/suggestions to PHE.screeninghelpdesk@nhs.net

Thank you

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Menti Questions

Jamili & Lola

Public Health England

Protecting and improving the nation's health

NHS Sickle Cell and Thalassaemia Screening Programme's Live Webinar, 28th January 2021

Comfort Break

Public Health England

Protecting and improving the nation's health

NHS Sickle Cell & Thalassaemia Screening Programme

Publications go digital

Cynthia Gill – SCT Programme Advisor
Siobhan Ryan – SCT Project Lead

Digital by default

All the programme resources are now digital

- Transition to digital is supported by user research which show that most women benefit from, and expect digital information
- We are supporting services in England to move to using digital versions of the antenatal and newborn (ANNB) screening and carrier leaflets
- Digital versions of the leaflets:
  - Are easy to edit, so they always contain the most up to date information
  - Are in a format that can be used with assistive technologies
  - Work well on smaller screens such as phones
  - Can be saved to a smart phone home screens like an app

Accessible Information

Leaflets are required to meet Government accessibility standards

The new format we are using:

- Makes the leaflets easy to find with search engines
- Can be accessed on a variety of devices
- Can be printed out by HCPs to be given to those who are not online

The language:

- Remains in “Plain English” – allowing the information to be easily understood
- Jargon and abbreviations are avoided
- Technical terms are fully explained

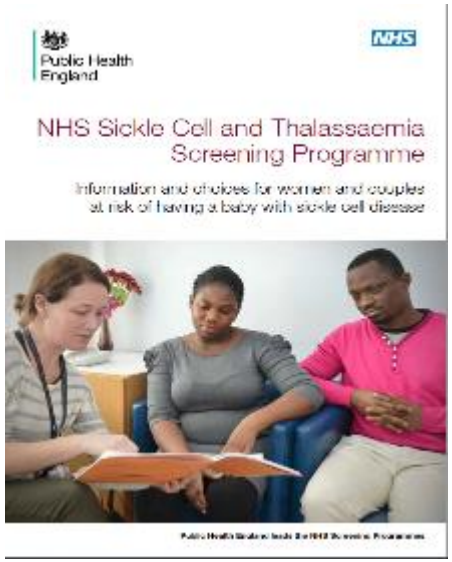
The plan

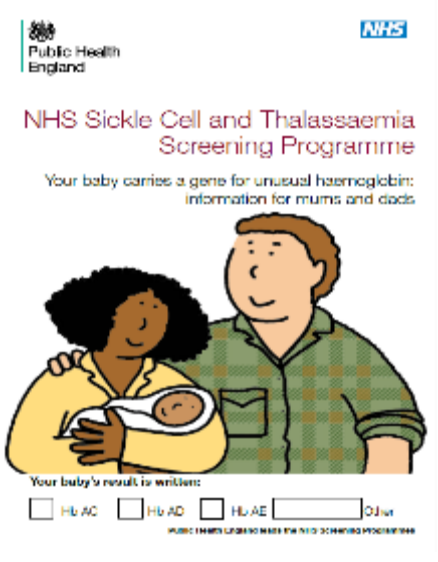
Printed screening and carrier leaflets are being phased out, with a move to information which is digital by default

We have created a digital format for
- 8 antenatal leaflets for adult carriers
- 2 antenatal leaflets for women and couples at risk of having a baby with a haemoglobinopathy
- 2 newborn leaflets for parents of babies who carry a gene for unusual haemoglobin

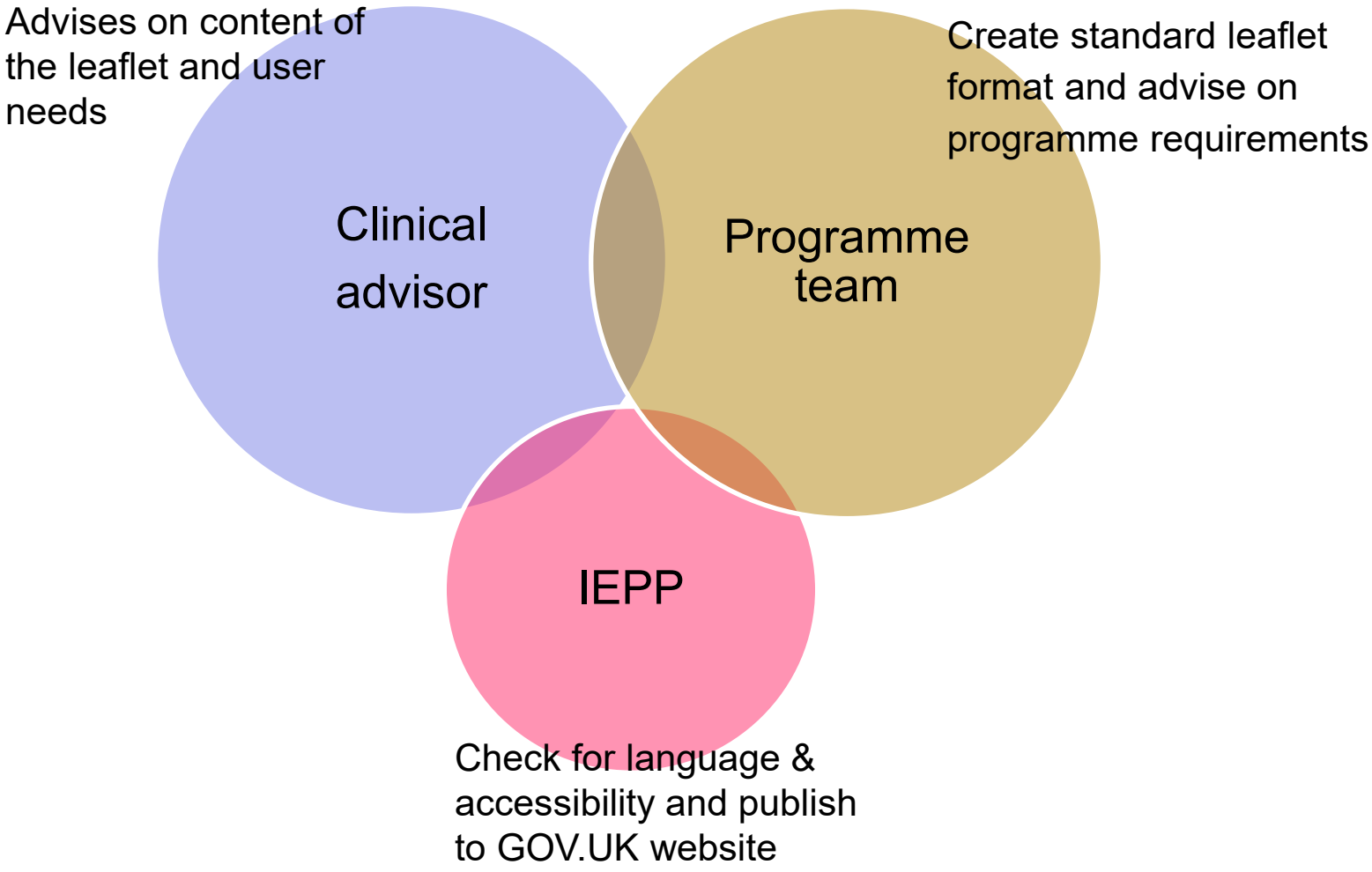
And we blogged to inform professionals in October 2020

https://phescreening.blog.gov.uk/2020/10/14/sickle-cell-and-thalassaemia-carrier-leaflets-go-digital/





The process


Advises on content of the leaflet and user needs
Clinical advisor
Create standard leaflet format and advise on programme requirements
Programme team
IEPP
Check for language & accessibility and publish to GOV.UK website

Aim of antenatal carrier leaflets

To inform the pregnant woman of what her carrier status means to her and her baby

Has information specific to her carrier status

To invite baby's biological father for screening

Clear unambiguous language

Links to other PHE publications

If your baby's biological father has an unusual haemoglobin gene it is important to identify the type of gene and the chance of your baby inheriting a serious haemoglobin condition. For this reason, we will also invite the biological father for screening. He will only know he carries a gene for unusual haemoglobin if he has a blood test to check his haemoglobin type.

If the test shows your baby's biological father is a carrier of an unusual haemoglobin gene you will be offered specialist counselling and, if necessary, diagnostic testing.

Please let your healthcare professional know if you:

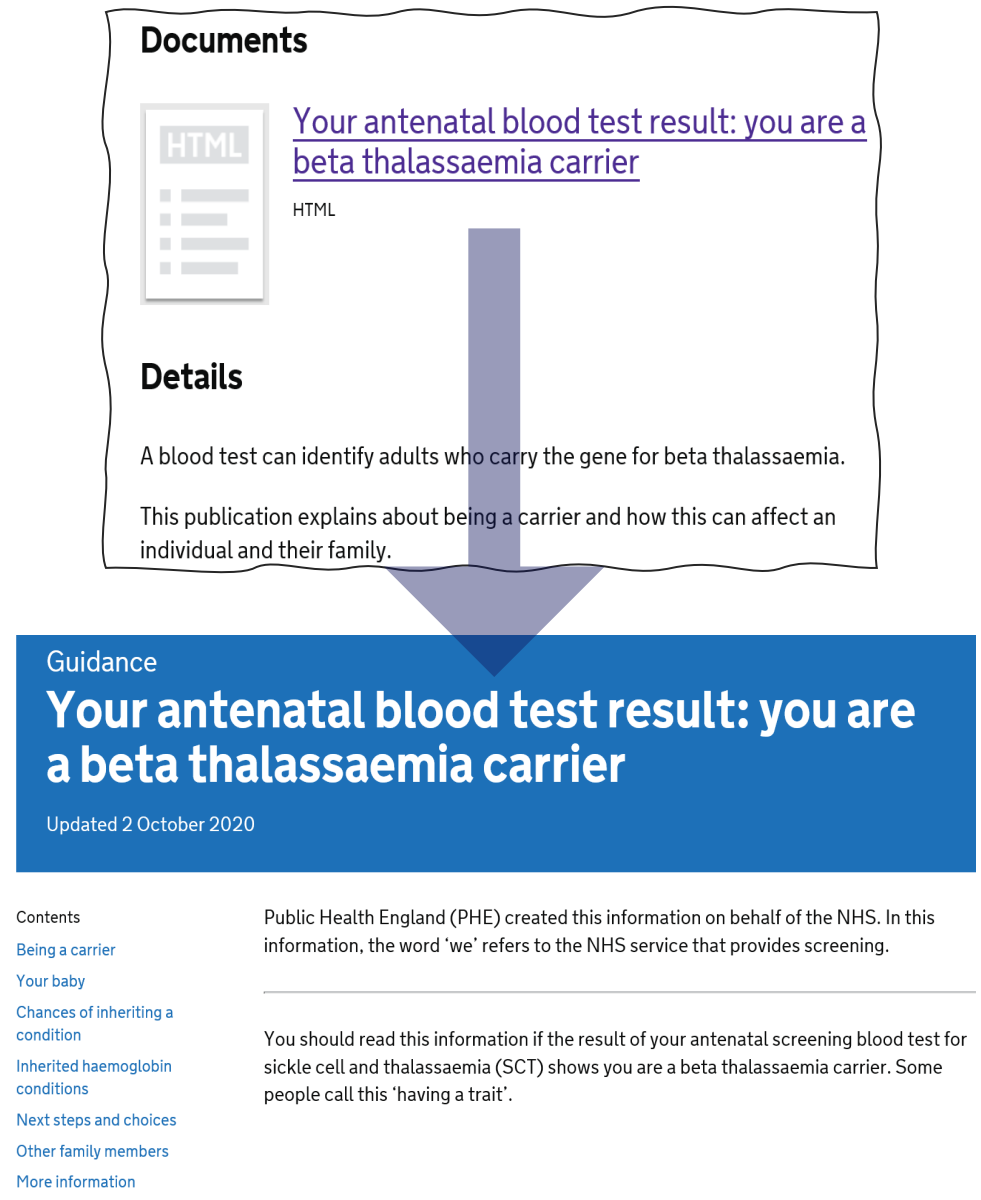
- became pregnant as a result of fertility treatment with donor sperm or a donor egg
- have had a bone marrow or stem cell transplant
- are pregnant as a surrogate

As a beta thalassaemia carrier your red blood cells are smaller than usual and your haemoglobin level is lower than normal. This is different to iron deficiency anaemia. Always ask your healthcare professional to check your iron levels before taking iron supplements.

The result (1)

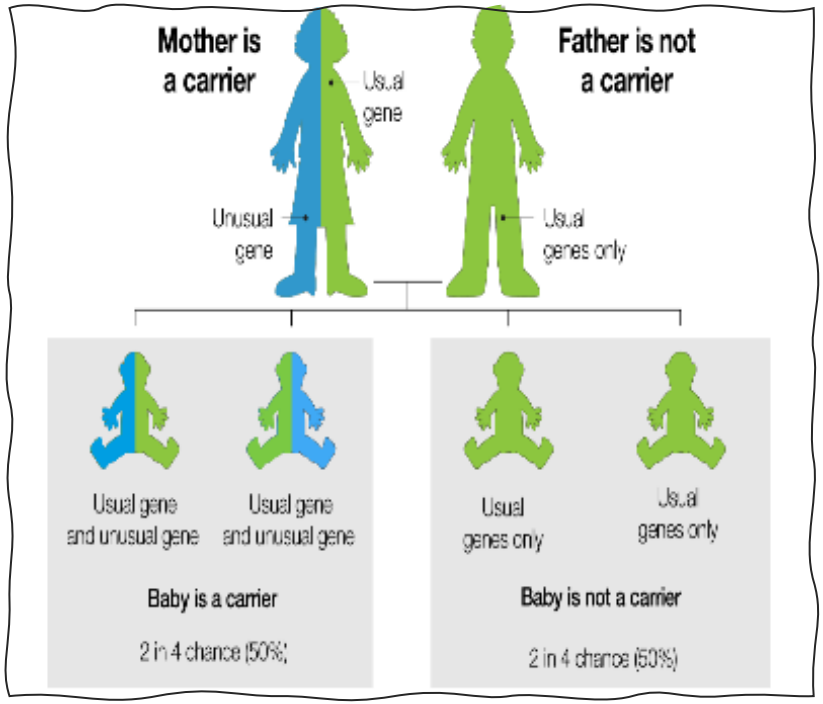
Clear headings
Standard layout

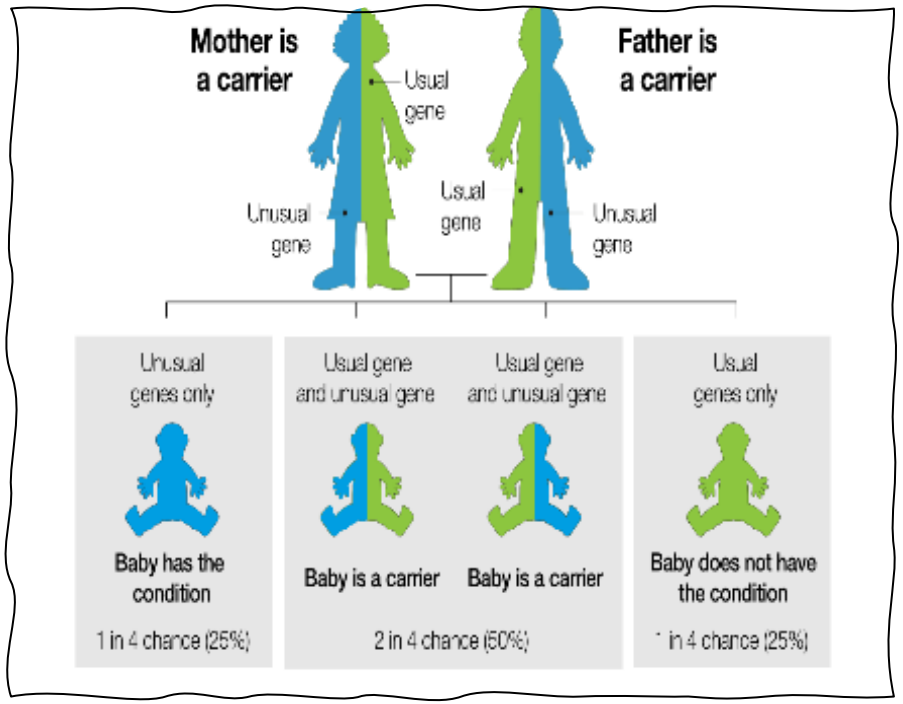
Links in the text make navigation quick and easy



The result (2)

Graphics for healthcare professionals to use to explain inheritance to women or couples and discuss next steps and choices





To consider

- Information is targeted to antenatal woman who is a carrier. What about information for carrier fathers?

- Accessible carrier information for individuals who do not read well?

- Accessible carrier information for individuals whose first language is not English?

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Laboratory Handbooks

Dr Yvonne Daniel

Handbooks

- Update cycle – 2 years
  - Current versions published 2017
- Antenatal handbook
  - Approved
  - Waiting for final review
- Newborn handbook
  - Pending final edits and approvals
- Programme handbook
  - Pending final edits and approvals
- Published as HTML

Contents

Handbooks

- Detailed guidance for the laboratories:
  - Methods
  - Instruments
  - Protocols
  - Testing strategies
  - Interpretation,
  - Reporting
  - Risk assessments

Review criteria

- Incidents
- Queries
  - Programme Helpline
  - Support Service
- Changes in practice
- New guidance
- Feedback from work shops/training events
- Advisory groups
  - UK NEQAS

What's new: Newborn

- MSMS – guidance updated
  - More users ⟶ more cases ⟶ more evidence
  - Limitations of techniques
  - More explicit in appropriate second test techniques

- Linkage with UKAS

What's new: Antenatal

- Language more consistent across handbooks
- Removed some schematics due to increasing complexity
- Emphasised co-inheritance of conditions

- Linkage with UKAS

NHSE – Genomics Laboratory Hubs

- 7 Genomics Centres
- 4 Haematology
- Implementation on going
- Ultimately no direct charge to requestor
- PHE engagement:
  - Reporting
  - Testing strategies for DNA transfused babies
  - Ensure that it meets the needs of the screening programme

Thank you

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Menti Questions

Siobhan, Cynthia and Yvonne

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Two New Publications and the Consultation with Sickle Cell Service Users

Iyamide Thomas, NHS Engagement Lead, Sickle Cell Society

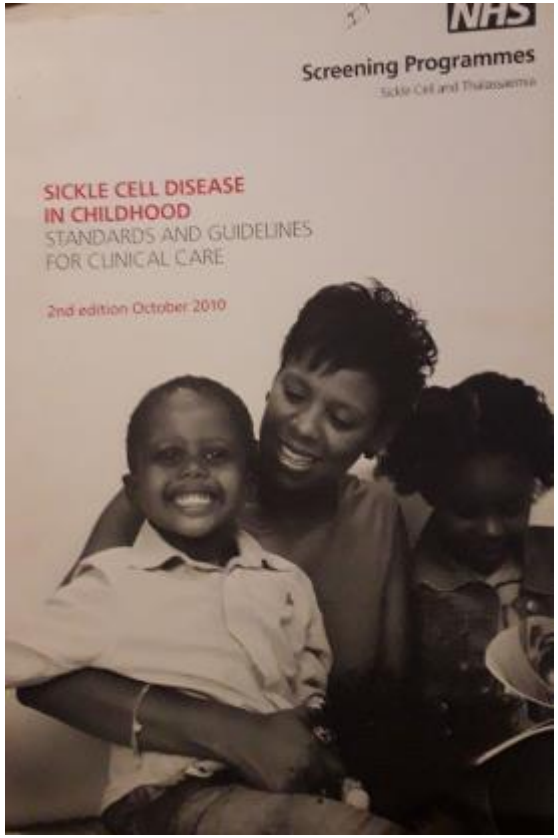
Background & Objectives

- In 2018 NHS SCT Programme Commissioned Sickle Cell Society and UKTS to work collaboratively with them in 'Engagement, Outreach and Programme Development' Project. Work Specification included:

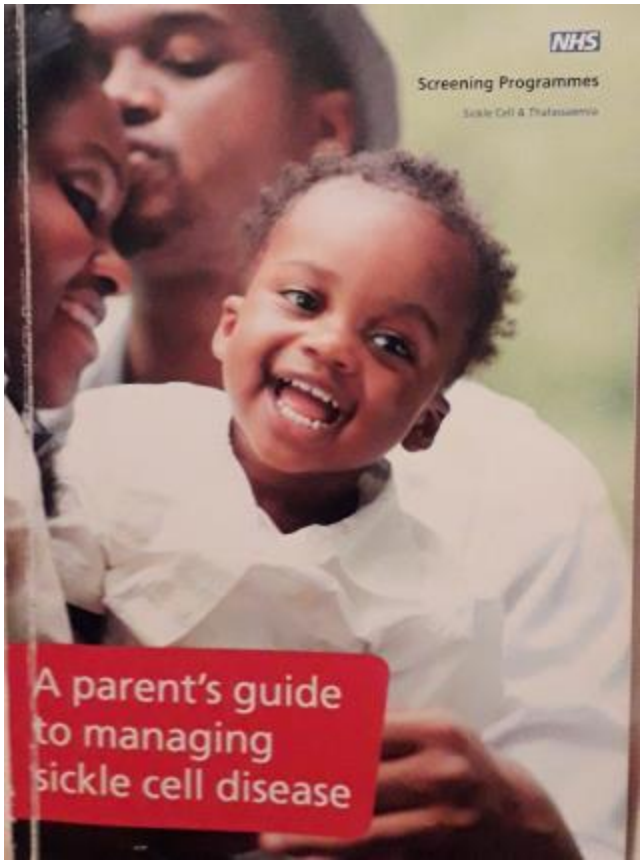
- **Review and update publication *'Sickle cell disease in childhood: standards and guidelines for clinical care' (2010 Edition) –'Paediatric Standards'***
- Project Advisory Group decided that this publication should remain focused on the clinical management of sickle cell so added:
- **Update of *'Parent's Guide to Managing Sickle Cell Disease' to include the wider determinants of health relevant to living with sickle cell (2012 Edition) – 'Parents Handbook'***

Updating Two New Publications

‘Paediatric Standards’



‘Parents Handbook’



Consultation Outputs

Consult stakeholders (i.e. sickle cell service users) to solicit opinion on any changes they might want to see and whether the Parents Handbook (PH) should be reproduced in print or electronic format.

- Design and pilot user questionnaire at support group (PH only)
- Finalise questionnaires for hard copy and online consultation
- Disseminate hard copy to parents in Brent and Milton Keynes SCaT for on site completion (PH only)
- Conduct service user focus group in central London
- Design & Disseminate advertising graphics for online questionnaires
- Analyse questionnaires and produce report for Editorial Teams of both publications to incorporate user feedback as necessary

Consulting Parents Face-to-Face

Parents' Support Group

Parents Focus Group in Euston

Galvanising Online User Input

HAVE YOUR SAY

Give your feedback on the New Draft

Paediatric Standards

Find our more here:
bit.ly/paediatricstandards

SICKLE CELL SOCIETY
Celebrating 40 YEARS 1979 - 2019

Galvanising Online User Input

HAVE YOUR SAY

Give your feedback on

A Parent's Guide to Managing Sickle Cell Disease

Find our more here:

bit.ly/parentsguidefeedback

Celebrating 40 YEARS 1979 - 2019

Some Summary Results for Parents Handbook

- 51 completed questionnaires returned – half done online
- **20% respondents knew nothing about SCD before the PH**
- **97% stated they found the 2012 edition useful for reasons such as**:

"Gives different signs and symptoms of the disease and what to look out for"

Parents who did not find the book useful said:

"*Sometimes have difficulty reading the English or medical jargon"*

- **90% felt a printed copy of the PH should be given to all new parents**
- **67% said child's symptoms less manageable without book**

What format of the PH will you find most useful?

65% (Both internet and hard copy) 33% (Hard copy) 2% (internet copy)

What new information would you like in the Handbook?

- "Parents' Experiences", "Breaking news of SCD to child", "Direct references to legal /statutory support for parents to access when making bids to employers for flexible working" " Extra curricular activities like ballet" "More information on potential need for counselling psychological support, particularly in adolescence and onwards"

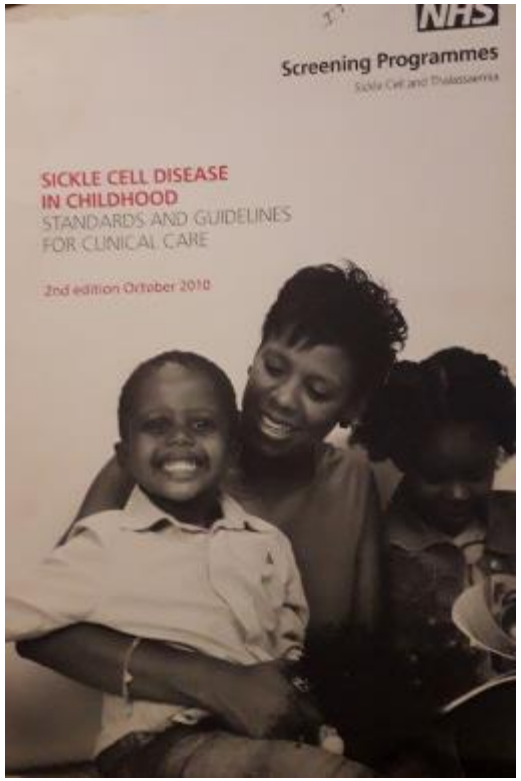
Parents' Handbook – What was Said

"I personally have learnt so much from this book. It has been like our second doctor in my house"

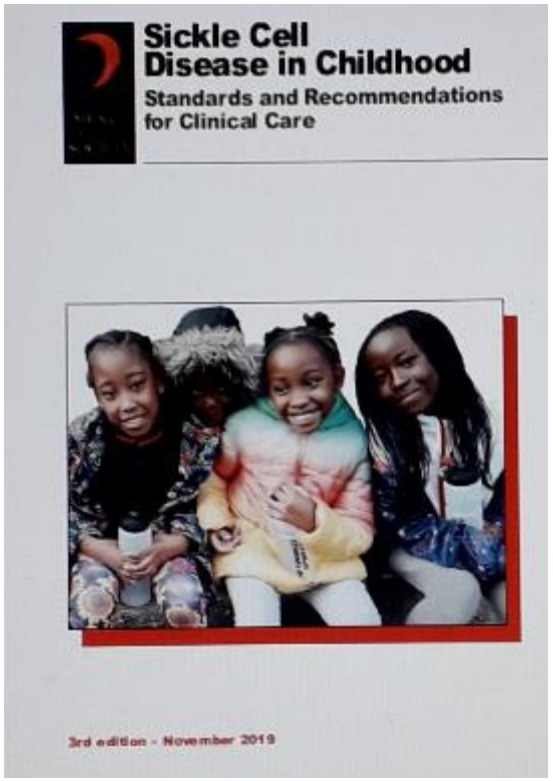
"The book is like the sickle cell bible, very useful"

Paediatric Standards – Updated!

Second Edition - 2010

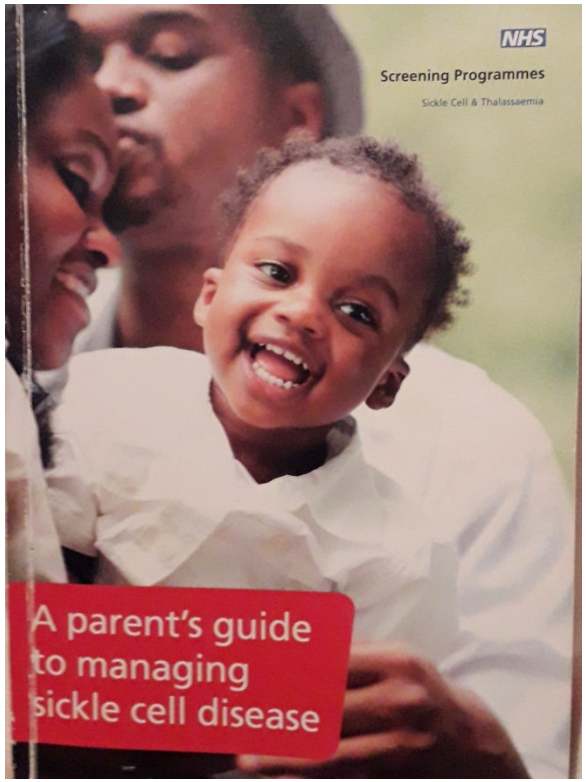


Third Edition - 2019



Parents Handbook – Updated!

Third Edition - 2012



Fourth Edition - 2021

Two New Publications and the Consultation with Sickle Cell and Service Users

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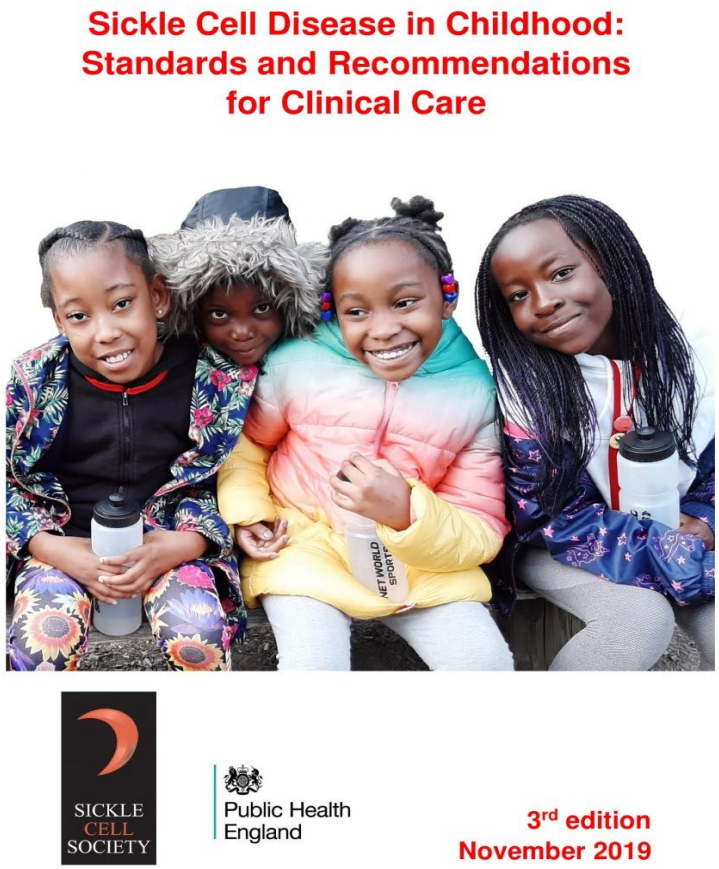
Sickle Cell Disease in Childhood: Standards and Recommendations for Clinical Care.

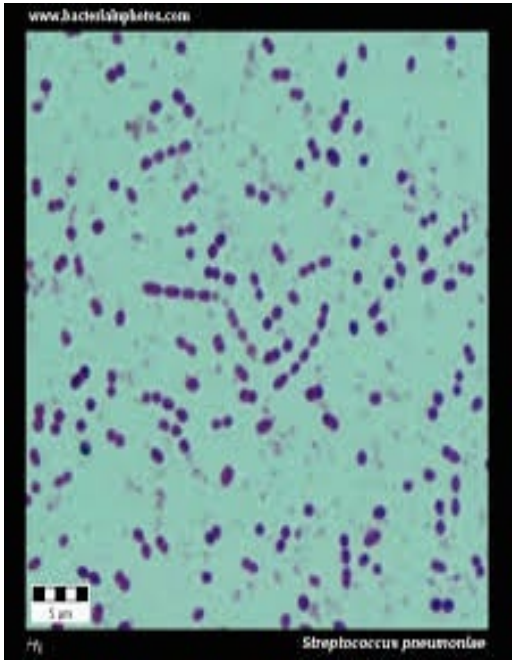
Dr Moira Dick and Professor David Rees on behalf of PHE and the Sickle Cell Society



https://www.sicklecellsociety.org/paediatricstandards/

Dr Moira Dick and Professor David Rees on behalf of PHE and the Sickle Cell Society

Sickle cell disease as it was then



- a rare childhood disease in 1970s- 90s
- considered an acute hospital issue
- childhood mortality 10% due to infection, stroke, acute splenic sequestration, worsening anaemia, acute chest syndrome
- approximately 2000 adults and children in the UK with main populations in London and Birmingham (survey 1979) Davis LR et al BMJ 1981 282
- antenatal and newborn screening was inconsistent across country
- Infants not always followed up after screening Assessment of care of children with sickle cell disease: implications for neonatal screening programmes. R I Milne BMJ 1990 300

Implications due to lack of newborn screening and quality of follow up

- infant deaths from pneumococcal sepsis or acute splenic sequestration occurred before diagnosis made
- understanding of cerebral vasculopathy limited as no MRI/TCD
- most paediatricians had little or no training or experience in sickle cell disease
- was it a paediatric or a haematological condition?
- resources and services varied enormously across the country
- widespread experience of stigma
- racist attitudes prevalent

MRI magnetic resonance imaging
TCD transcranial Doppler scanning

First edition paediatric standards 2005

- developed in 2005 on behalf of BSH to support roll out of universal newborn screening and aimed at those clinicians working in areas of low prevalence
- dealt with organisation of care as much as clinical recommendations
- audit standards were included – timeliness into care and prescription of penicillin, timeliness of Pneumovax, follow up and failsafe arrangements, coverage of TCDs
- evidence only in four areas- penicillin prophylaxis, TCD screening , hydroxyurea and incentive spirometry
- mainly based on good practice from UK and USA

BSH : British Society of Haematology

2nd edition 2010

Sickle cell disease in childhood Standards and Guidelines for clinical care

- added a standard on data collection and National Haemoglobin Registry (NHR)
- included clinical peer review quality requirements and specialist services definitions

Latest edition 2019
Standards and clinical recommendations for care of children with SCD

- recommendations on cerebrovascular disease, preoperative transfusion and hydroxycarbamide therapy updated as evidence now available in these areas
- standards strengthened in line with Public Health England guidance and Metric Definition Sets that inform the Specialised Services Quality Dashboard commissioned by NHS England
- new standards on coverage of children prescribed hydroxyurea, coverage of children on NHR and completion of annual review
- a new information technology system linked with NHR for referring infants from newborn screening into treatment introduced.

https://www.gov.uk/government/publications/sickle-cell-and-thalassaemia-screening-newborn-outcomes-system/clinical-user-guide-newborn-outcomes-system

Grade A recommendations

Requires at least one clinical trial

- the TWiTCH study looked at the safety of switching children with abnormal TCDs from regular transfusions to hydroxycarbamide and found that hydroxycarbamide was equivalent to transfusion, with no increase in TCD velocities or cerebrovascular events (A)

- children with HbSS and HbS/β^0 thalassaemia undergoing low- and moderate-risk surgery should have a preoperative transfusion to increase the Hb level to 100 g/L (A)

DeBaun MR, Gordon M, McKinstry RC, et al. Controlled trial of transfusions for silent cerebral infarcts in sickle cell anemia. N Engl J Med 2014 Aug 21;371: 669–710

WareRE, DavisBR, SchultzWH, et al. Hydroxycarbamide versus chronic transfusion for maintenance of transcranial doppler flow velocities in children with sickle cell anaemia—TCD With Transfusions Changing to Hydroxyurea (TWiTCH): a multicentre, open-label, phase 3, non-inferiority trial. Lancet 2016; 387: 661–70.

Howard J, Malfroy M, Llewelyn C et al. The Transfusion Alternatives Preoperatively in Sickle Cell Disease (TAPS) study: a randomised, controlled, multicentre clinical trial. Lancet 2013; 381: 930–8.

Hydroxycarbamide (hydroxyurea)

- hydroxycarbamide should be offered to all children with HbSS/Sβ^0 thalassaemia aged 9–42 months regardless of the clinical severity of their illness. (A) See Standard 6.
- hydroxycarbamide should be offered to all children older than 42 months who have recurrent episodes of acute pain or who have had two or more episodes of acute sickle chest syndrome (A)
- hydroxycarbamide should be offered to all children older than 42 months whose lives are significantly affected by symptoms of SCD, including those with frequent episodes of pain that disrupt normal activities (A)
- hydroxycarbamide should be offered to all children older than 42 months who are at high risk of progressive organ damage caused by SCD, including those with hypoxemia, significant albuminuria, conditional TCD velocities, or significant anaemia (steady state Hb<70 g/L) (B)

Qureshi A, Kaya B, Pancham S, et al. Guidelines for the use of hydroxycarbamide in children and adults with sickle cell disease: A British Society for Haematology Guideline. Br J Haematol 2018;181:460–75.

Standards- alignment and data collection

- Standard 1 (SCT-S08): Sickle cell and thalassaemia screening – reporting newborn screen-positive results to parents Newborn clinical outcome system (NBO)
- Standard 2 (SCT-09 HAEM 4A): Sickle cell and thalassaemia screening – timely follow up, diagnosis and treatment of newborn infants with a positive screening result NBO
- Standard 3: (HAEM 4B) Timeliness of penicillin prophylaxis NBO
- Standard 4: Coverage of pneumococcal immunisation at 2 years ? NHR
- Standard 5 (HAEM02) Coverage of transcranial Doppler (TCD) scanning ? NHR
- Standard 6: Coverage of hydroxycarbamide (hydroxyurea) therapy ? NHR
- Standard 7: Coverage of children identified through the screening programmed subsequently registered on the national haemoglobinopathy registry NBO/NHR
- Standard 8: (HAEM08) Coverage of children who have had an annual review NHR

https://www.gov.uk/government/publications/sickle-cell-and-thalassaemia-screening-newborn-outcomes-system/clinical-user-guide-newborn-outcomes-system

Acknowledgements – and thanks to everyone who took part in the consultation

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- Iyamide Thomas, Sickle cell Society
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and additional specialist input from:

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Menti Questions

Iyamide & Moira

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Thank you and closing remarks

Chair: Professor Dame Elizabeth N Anionwu DBE CBE FRCN FQNI PHD
Emeritus Professor of Nursing, University of West London