Standard Operational Procedure for Haemoglobinopathies Screening and Referral

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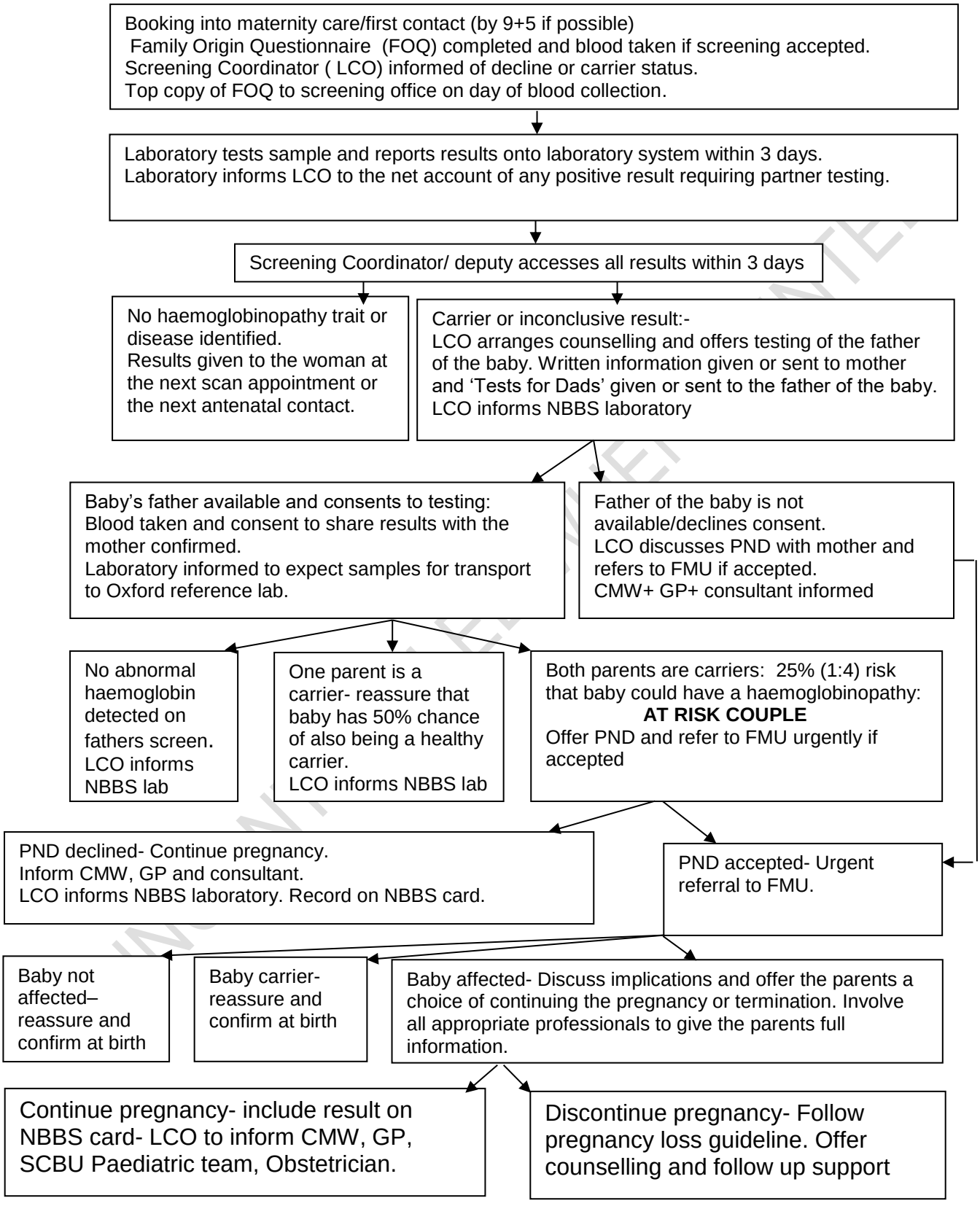
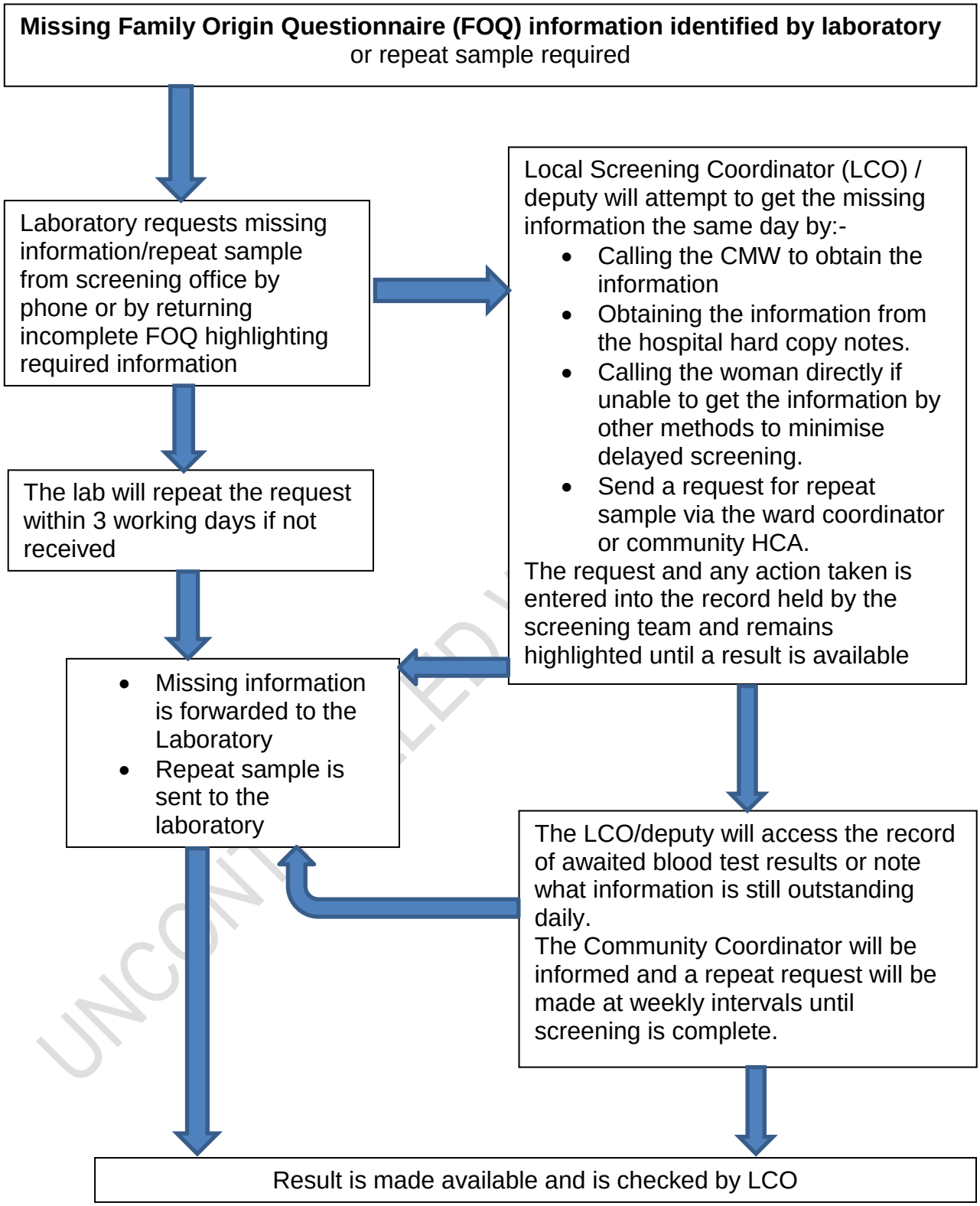
Appendix A

Linked Antenatal and Newborn Haemoglobinopathy Pathway



Appendix B



Appendix C

Isle of Wight NHS Trust
Tracker for all pregnant women who are carriers of an haemoglobinopathy

Patient Addressograph

EDD…….

Carrier of (Specify HB variant)…….....

Date of Sample to screen for haemoglobinopathy……Gestation……./40

Date of Carrier/Affected result: ……Signature (and print name)……

Date patient informed of result: telephone: ……Signature (and print name)……

Date patient informed of result in writing (copy placed in file): ……Signature (and print name)……

Baby's Father Testing offered: YES: ☐ NO: ☐ Tests for Dad's PIL Given: YES: ☐ NO: ☐

Baby's Father Testing accepted: YES ☐ NO: ☐

Date of Baby's Father Testing: …… Baby's Father Result: …….

Baby's Father not available, PND offered - YES: ☐ NO ☐ PND Accepted YES: ☐ NO: ☐

IF PND Accepted date of referral to FMU……Signature (and print name)……

Normal Baby's Father result : date patient informed of result:

By telephone: ……..Signature (and print name)……

In writing: ……..…Signature (and print name)……

Abnormal Baby's Father Result: **AT RISK COUPLE**

Date patient informed and urgent referral to FMU offered: ……..…Signature ……

Date of Appt at FMU ……..

Notification of an 'At Risk' Couple for completed ☐ Sent to SCBU ☐

Appendix D

Antenatal and Newborn Screening Programmes

ANTENATAL AND NEWBORN SCREENING
'AT RISK PREGNANCY' ALERT FORM
to be completed for all HIGH RISK couples

Maternal details

Surname		**First Name**
DOB	**NHS Number**	**EDD**
Haemoglobinopathy Result		**Place of test**
		Date tested **Gestation**

Paternal details

Surname	**First Name**
DOB	**NHS Number**
Haemoglobinopathy Result	**Place of test**
	Date Tested

Print name:
Contact telephone number:

Date:
Trust / Maternity unit:

Please send completed form to
Toby Greenfield Senior Biomedical Scientist, at pho-tr.PortsmouthScreening.nhs.net
* in the event of a baby being identified as carrier status, maternal screening or parental status can help inform the subsequent information sent from the Newborn Laboratory to GP and Health Visitor

Appendix E

SCBU Notification of an 'AT RISK' couple identified via antenatal testing.

Addressograph

Obstetric Consultant……..

Mothers Haemoglobinopathy status……

……..

Fathers Haemoglobinopathy status……

……..

EDD……..

Gestation when at risk status identified……

Prenatal Diagnosis (PND) accepted Y/N…….

Date and type of PND…….

Outcome of PND…….

Singleton/Multiple pregnancy ……

***Send this form to SCBU**

Appendix F

Haemoglobinopathy 'At Risk' Couple- Newborn follow- up

Addressograph (Mother)

EDD……

Mothers Haemoglobinopathy Satus…….
……

Fathers Haemoglobinopathy Status……
……

Prenatal Diagnosis……

Addressograph (Baby)

Gestation at birth……...

Date of Newborn Bloodspot test……

Newborn Bloodspot result……..

Repeat required Y/N reason……

Repeat Date and result……

If haemoglobinopathy confirmed- detail of referral……

……..

……..

***Return to Screening Office when complete.**

1. Purpose/Background:

Haemoglobinopathies, such as Sickle Cell Disorder and Thalassaemia, are inherited genetic blood conditions of which there are over a thousand variants with effects ranging from mild to very severe.

Infants are at risk of inheriting these disorders only if both parents are carriers and/or suffer from one of the disorders. All women are offered screening in pregnancy to assess the risk of the baby inheriting a haemglobinopathy to give information to the mother regarding her own status and to enable discussion of choices should the baby be identified as being affected.

Although some haemoglobinopathies are common in some parts of the world such as Africa, Asia, the Caribbean, the Middle East and the Mediterranean it is important to note that no haemoglobinpathy is exclusive to any single ethnic group. Therefore it is possible for anyone, regardless of family history or ethnic background, to be a carrier of an abnormal gene.

People may inherit more than one abnormal haemoglobin gene with varying degrees of significance.

Where a person inherits only one abnormal gene they are referred to as having a 'trait' or being a 'carrier'.

Some variants are easily detected in the laboratory whilst others are biochemically silent.

2. Scope:

This guideline applies to all staff working in the Isle of Wight NHS Trust Maternity services.

3. Responsibilities

It is the responsibility of all Midwifery Nursing and medical staff to:

- Access read understand and apply this SOP
- Attend any mandatory training pertaining to the SOP

It is the responsibility of the department to:

- Ensure the SOP is reviewed as required in line with trust and national recommendations
- Ensure the SOP is accessible to all relevant staff

4. Procedure:

4.1 Community Midwife (CMW):

Is responsible for:

- Ensuring all pregnant women who have accessed maternity care, either through their GP or directly, are booked if possible before 10wks of pregnancy.
- At the booking appointment the booklet 'Screening for you and your baby' will be given, screening choices will be explained and offered. If English is not the woman's first language the community midwife should download and print translations of 'Screening for you and your baby' for the woman. Should a suitable translation not be available the telephone translation service should be used. Easy read simple versions are also available.
- All women will be offered screening for Sickle Cell and Thalassaemia at booking or first contact. If first contact is when the woman is in labour screening will be offered at the earliest opportunity following delivery. (NSC SC/T Standard 1).
- Known carriers, or carrier couples, should be directly referred to the local Screening Coordinator (LCO) who will ensure immediate counselling and referral for prenatal diagnosis (PND) if accepted.
- The Family Origin Questionnaire (FOQ) is a tool used in low prevalence areas of England, which includes the Isle of Wight, to identify the risk to the baby of inheriting a haemoglobin variant. It should be completed with information from the woman regarding her own family origin and that of the father of the baby. It is important to ask for family origins as far back as possible through the generations and tick all that apply (NSC SC/T Standard 3).
- Where a donor egg has been used the family origin of the donor is required on the FOQ and the blood of the father of the baby should be taken.
- If both donor egg and donor sperm have been used, and the donors are not available for testing, it is not possible to offer screening and the couple should be referred to the Local Screening Coordinator (LCO) for counselling.
- It should also be noted on the form where family origin is unknown (adoption).

- If the mother has had a bone marrow transplant she should be tested and testing of the biological father should also be offered immediately.
- If screening is declined the reasons for declining screening should be explored and documented. The LCO should be informed of any woman who declines to be tested to ensure she has been given all the relevant information to make her choice.
- The form also requires entry of the EDD and gestation at the time of the blood sample. The ideal is to have a result by 10 weeks gestation (NSC SC/T Standard 2).
- Women should be informed that Thalassaemia may be indicated by a blood test rather than diagnosed. Further tests may be requested.
- Documentation of the screening discussion, leaflets given, any test declined, all screening accepted and all samples taken must be made in the maternal notes.
- Where consent is given blood samples (One pink top and one purple top bottle) will be obtained and sent to the laboratory the same day with the completed bottom copy of the FOQ form.
- The top copy of the FOQ form must be brought to the screening office on the day the blood is taken.
- The CMW will access results for all tests accepted and ensure they have been documented in the hand held maternity notes at the next contact.
- The CMW should contact the LCO to check she is already aware of any abnormal blood results and that appropriate action has been taken.

4.2 Haematology lead for laboratory services:

Is responsible for ensuring:

- That all laboratory staff are aware of the National screening committee recommendations and that staff will receive adequate training and competency assessment for all aspects of the screening process.
- That all staff are aware of multidisciplinary and multiagency failsafe pathways to safeguard the efficient screening of all women who accept it.

- The maintaining of internal failsafe pathways for follow up of samples requested
- That information is requested from the LCO when FOQ forms are incomplete (NSC SC/T standard 3) to ensure minimal delay to screening.
- That repeat samples are requested via the LCO when inadequate samples are received.
- The LCO is informed of any carrier or condition status of the woman within the recommended timeframe and requesting testing of the baby's father where necessary.
- That national and local guidelines are followed where a positive result is identified.
- That accurate data is collected to ensure compliance with key performance indicators (KPI)

4.3 Local Screening Coordinator (LCO):

Is the designated lead for antenatal and new born screening programmes and is responsible for ensuring:

- That screening for sickle cell and thalassaemia is based on National Screening Committee recommendations.
- That all staff are aware of current pathways and recommendations through development and delivery of education and training programmes to enable staff to give accurate information to the women that they are then able to make informed choices.
- That multidisciplinary and multiagency pathways and communications are in place.
- That guidelines and failsafe's across the antenatal and new born screening programmes are developed and updated.
- That there is regular liaison with local screening steering groups, regional screening meetings and national screening forums to ensure up to date information is shared appropriately.
- Production of audit data as required and requested.

- Reporting and acting on any incident where SC/T standards do not comply with UK NSC standards.
- That a result is available for every woman who accepts SC/T screening within 3 days of the laboratory receiving the sample (NSC SC/T standard 4).
- That when informed of an incomplete FOQ by the haematology lead the missing information is obtained urgently to minimise any delay to screening. (Appendix B)
- That there is a clear result or decline recorded for every woman who books for pregnancy care on the Isle of Wight.
- That every woman who declines screening has the chance to discuss her choice and that she receives at least one reoffer of screening at 20weeks.

4.4 Positive results

- That any positive result is acted on immediately. Any woman with a carrier status is counselled by a specifically trained member of staff and given information regarding her status and testing of the baby's father. The woman will be contacted by phone or, if unable to contact by phone, a community midwife will be asked to visit to offer an appointment with the LCO/trained staff. If the CMW is unable to speak to the woman a letter will be sent with an invitation to contact the LCO.
- The SC/T Pathway (Appendix A) will be followed and the 'Tracker for pregnant women who are carriers of an haemoglobinopathy' (Appendix C) will be commenced and attached to the maternity notes hard copy
- That counselling and written information 'Tests for Dads' (downloaded from NSC website) is provided where testing of the father of the baby has been advised and he is available. Testing of the baby's father will be arranged if he accepts screening. Explicit consent to share paternal results with the mother will be sought.
- The LCO will liaise closely with the IOW laboratory lead for Haemoglobinopathy to ensure samples are sent urgently to the Oxford reference laboratory.

- If the father of the baby is not available or declines to be tested the LCO will discuss and offer prenatal diagnosis (PND) (NSC SC/T Standard 5)

4.5 Normal/ 'not at risk' result-

- That reassurance by phone is provided to the couple and also that a letter confirming results is sent where the tests of the father show normal haemoglobin. The CMW will be informed and asked to make a note on the NBBS sample card of the carrier status.
- That, as this is a linked antenatal and newborn screening pathway, the Portsmouth NBS laboratory is informed by email, using the electronic template in the screening folder, of any woman with a carrier status and of the fathers result where available. (Appendix D)

4.6 Couple 'at risk' result-

- That all 'at risk' couples are contacted, counselled immediately, and offered urgent PND. (NSC SC/T Standard 5)

  -PND is declined- the LCO will ensure that the choice is documented in the hand held and hospital notes and will inform the GP, the obstetric consultant, CMW and the NBS screening laboratory (Appendix D).

  A paediatric alert and 'At Risk' couple (Appendix E) will be sent to SCBU and placed in the notes.

  An orange paediatric alert sticker will be placed on the maternity hard copy.

  -PND accepted- the LCO will refer the couple to Southampton Fetal Medicine Unit (FMU) for either CVS or Amniocentesis and will have a result within 5 days of the PND procedure (NSC SC/T standard 6 and 7)

  PND results confirm:

  - that the baby is a carrier or not affected- the couple will be contacted and informed immediately and the result will be documented in the notes. The CMW will also be informed and asked to record the result in the hand held notes.

  -that the baby is affected- the couple will contacted and informed immediately and offered an appointment to discuss the options which are:-

1) To continue the pregnancy

LCO will inform the CMW, GP, Paediatric Consultant, Obstetric Consultant and NBS laboratory (Appendix D).

A paediatric alert and an 'At Risk' couple- Newborn follow up form (Appendix E) will be sent to SCBU and a copy will be placed in the notes.

An orange paediatric alert sticker will be placed on the maternity notes hard copy.

The decision will be documented on the hand held and hospital notes.

2) Not to continue the pregnancy

The couple will be counselled by the LCO (or deputy) regarding methods of termination of pregnancy.

5 Implementation/training/awareness

- This is a review of a current document and it formalises current practice.
- Once ratified it will be available in all clinical areas within the Maternity Unit and on the intranet.
- All new, reviewed and ratified documents are notified to staff via the monthly maternity newsletter
- Mandatory maternity updates are provided and all maternity staff are required to attend on an annual basis.The NSC e-learning screening modules are mandatory for all staff . Quarterly newsletters update staff on any changes.
- The LCO will be available to give tailored information to new staff or students.

6. Auditable Standards

What aspects of compliance with the document will be monitored	What will be reviewed to evidence this	How and how often will this be done	Detail sample size (if applicable)	Who will coordinate findings	Which group or report will receive findings
Staff Training	Completion of screening e-learning on E-LfH	Quarterly	Training records for all relevant staff groups	The LCO	Audits will be reported to the HOM, at the

					Screening steering Group and a in the quarterly screening newsletter
Timely screening at booking	Data collected by the screening team	Annual	Data from a 6 month timeframe	The LCO	Audits will be reported to the HOM, at the Screening steering Group and a in the quarterly screening newsletter
Evidence of discussion of screening results	Data from randomly selected postnatal maternal notes	Annual	30-60 sets of maternal notes	The LCO	Audits will be reported to the HOM, at the Screening steering Group and a in the quarterly screening newsletter

Where failings are identified the LCO will take appropriate action to improve the service.

The LCO ensures timely submission of all required quarterly Key Performance Indicator (KPI) reports and a detailed annual report all of which are discussed at local level in the Screening Steering Group and at regional level at the Hampshire and Isle of Wight (HIOW) meetings.

7. Related Documents:

Guidelines:

http://www.nice.org.uk

www.rcog.org.uk/guidelines

www.bpas.org

SOP for the management of stillbirth, late intrauterine fetal death

SOP for the management of Termination for Abnormality

SOP for referral to the Fetal Medicine Unit

Patient Information:

'Screening for you and your baby'

8. References:

- NHS Sickle Cell and Thalassaemia Screening programme standards (2017)
- NHS Public Health functions agreement 2017-18 Service specification no.18
- NHS Sickle Cell and Thalassaemia Screening Programme Handbook for Laboratories (2017)
- NHS Sickle Cell and Thalassaemia Screening Programme Handbook for antenatal laboratories (2017)
- PHE Guidance: Understanding haemoglobinopathies July 2018
- PHE Guidance: Sickle cell and Thalassaemia overview July 2018
- PHE Sickle Cell and thalassaemia supporting information March 2019

9 DISCLAIMER

It is the responsibility of staff to check the Trust intranet to ensure that the most recent version/issue of this document is being referenced.

DOCUMENT HISTORY					
Date of Issue	**Version No.**	**Next Review Date**	**Date Approved**	**Director Responsible for Change**	**Nature of Change**
April 2012	1.0	23rd April 2015	23rd April 2012		New document. Ratified at Maternity CSG
August 2017	2.0	15th August 2020	15th August 2017	Clinical Director for SWCH	Reformatted for easy use. Based on national guidelines. Ratified at Maternity CSG
June 2020	SOPv1	June 2023	25th June 2023	MCSG	Converted to SOP reviewed and ratified