Clinical haematology

# Pregnancy and sickle cell disease (SCD)

## Information for patients, relatives and carers

### Introduction

This leaflet aims to answer your questions about pregnancy, sickle cell disease (SCD) and its effects on the baby. If you have further questions please ask your midwife, obstetrician, haematologist, clinical nurse specialist (CNS) or community nurse specialist.

### Planning a pregnancy

It is important to tell your haematologist/clinical nurse specialist (CNS) or GP early if you are planning a pregnancy or are in the early stages of pregnancy. This will give you the best chance of a healthy and successful outcome. At this stage we will:

- carry out a medical review in the haematology clinic to plan the best care for you during pregnancy
- refer you to joint haematology and obstetric clinic at Queen Charlotte's & Chelsea Hospital

Your pregnancy with be managed jointly with the obstetric team at Queen Charlotte's & Chelsea Hospital for the full length of your pregnancy and for six weeks after the birth.

The effects of SCD in pregnancy vary

**Genetic counselling**

You are advised to have genetic counselling to assess whether you are at risk of having a child with SCD. This involves a simple blood test to analyse if your partner carries a gene for an unusual type of haemoglobin. If your partner is a carrier of a sickle gene there is a 1 in 2 (50 per cent) chance that your baby could inherit SCD, or 1 in 2 (50 per cent) chance your baby would carry the gene themselves.

**Prenatal (before birth) diagnostics test**

We can offer a prenatal diagnostics test for your fetus during pregnancy. This can be done by:

- chorionic villous sampling (CVS), which involves removing and testing a small sample of cells from the placenta
- amniocentesis, which involves removing and testing a small sample of cells from amniotic fluid, which surrounds the fetus in the womb (uterus).

Termination of pregnancy can be discussed if the fetus is affected by SCD, if this is known in the first trimester.

**Postnatal (after birth) diagnostics test**

A diagnostic test can also be done after pregnancy. This is called newborn blood spot screening.

## Clinic attendance

The combined specialist clinic is on alternate Monday mornings between 09.30 and 12.00 noon.

You will see a consultant obstetrician, consultant haematologist and specialist midwife. You're encouraged to attend the clinic regularly for monitoring, scans, and investigations. This will help us to provide you and your baby with the best care.

Visits tend to be:

- every 4 weeks until you are 28 weeks' pregnant
- then every 2 weeks until 36 weeks
- then weekly until delivery

Visits can be arranged more frequently if necessary, and a specific check-up is arranged for 6 weeks after the birth.

At your first appointment we will carry out a general review of your health and arrange further tests as needed.

The clinic is in the Centre of Fetal Care, on the 2nd floor of Queen Charlotte's & Chelsea Hospital. Telephone: 020 3313 3998

## How pregnancy could affect your SCD and related complications

SCD in pregnancy affects women differently depending on your health status before becoming pregnant.

The most common issues are:

- you have more painful crisis during your pregnancy, especially towards the end. This usually gets better after delivery)
- you may become more anaemic as your pregnancy progresses. Only take iron supplements if your haematologist advises
- there is an increased risk of the placenta not functioning well, resulting in reduced growth of the baby (intrauterine growth restriction). This will be monitored by regular scans during your pregnancy
- your baby will need close monitoring at every stage including during labour

- you may be at increased risk of high blood pressure. This is monitored closely at the antenatal (during pregnancy) visits.
- you will need frequent ultrasound scans because you have a high-risk pregnancy. The usual schedule is:
    - early dating scan between 11 to 14 weeks
    - anomaly scan with uterine artery Doppler at 20 weeks
    - monthly scans for growth and fluid volume from 28 weeks

## Medicines advice

Before you get pregnant (conception) or in early pregnancy you will be advised to stop the following medicines due to the risks to the baby:

- hydroxycarbamide – stop at least 3 months before conception. (This does not affect fertility in women)
- enalapril
- esferasirox (Exjade®) or desferrioxamine (Desferral®) or other iron chelation agents – stop at least 3 months before conception or immediately after your pregnancy is confirmed
- non-steroidal anti-inflammatory drugs (NSAIDS); ibuprofen, naproxen or Nurofen®
- pregaballin and gabapentin
- ramadol

Women with SCD are at increased risk of blood clots during pregnancy, particularly if they are less mobile. So, we may advise you to take a preventative daily dose of a clotting agent, like tinzaparin. You will need to take this during your pregnancy and for up to 6 weeks after birth.

You will probably be prescribed low-dose aspirin (75mg once a day) from the time of your initial scan until approximately 36 weeks. This is to prevent pre-eclampsia, which is a condition that can cause serious problems for mother and baby.
We advise you to continue with:

- penicillin (250mg) or your alternative antibiotic twice daily
- folic acid (5mg) once daily

If you're anaemic (lower than your baseline) and if advised by your haematologists, only then can you take iron supplements.

Check you are up to date with your recommended vaccines:

- influenza (annually)

- pneumovax (5 yearly)
- HIB/Single Men.C
- MenACWY
- MenB
- Hepatitis B booster course.

## How to look after yourself while pregnant

Carry on using self-management to prevent crisis. We find that following this prevention plan helps:

- Drink plenty of fluids (3 litres in 24 hours) to avoid becoming dehydrated.
- Avoid alcohol.
- Eat a healthy, well-balanced diet including fibre to prevent constipation.
- If you get indigestion and it affects your appetite, eat smaller meals more frequently.
- Plan enough rest if you become more tired as the pregnancy progresses
- Slow down or reduce your activity in later pregnancy if you are struggling to work, study or cope with home life.
- Ensure you keep warm.
- Spot early signs of infection:
    - fever
    - productive cough with yellow or green sputum
    - pain passing urine
    - difficulty breathing
    - shivers

  Seek medical help as soon as possible.

## What happens if you have a crisis while pregnant

**You should go to hospital immediately if your sickle cell-related pain cannot be managed with painkillers or you develop chest pain or signs of infection.**

If you're under 20 weeks, the haematology team will manage you via the renal and haematology triage unit (RHTU) or day pain service. If you're over 20 weeks, the obstetric team will manage you on the delivery suite on the third floor of Queen Charlotte's & Chelsea Hospital.

If in pain, we will start an injectable opiate according to your individual protocol and intravenous paracetamol. Opiates are safe for you and your baby when they are needed for moderate or severe pain for short periods. It is not advised to take opiates daily during your pregnancy as this could be harmful for you and your baby.

We will give you fluids either orally or intravenously to ensure you are well hydrated.

If you have low oxygen levels, you will be given oxygen therapy. If your oxygen levels are normal or low, you will be advised to use incentive spirometry. This helps to improve lung function using a medical device called an incentive spirometer which is designed to help you take long deep breaths.

Any signs of infection will be treated with antibiotics either orally or intravenously.

Blood tests will be taken if you are unwell to check for anaemia, signs of infection and liver and kidney function.

## Blood transfusion and exchange transfusions

You may need a blood transfusion if you become anaemic below your baseline or are in a sickle cell crisis with complications.

Anaemia may be more likely to develop when you are in pain as the red blood cells are destroyed in a SCD crisis. There are two treatment options:

- simple top-up transfusion – to correct the anaemia or prevent further crisis
- exchange transfusion – to treat acute crisis or prevent crisis or complications

If you are on regular top-up or exchange transfusions this will continue as normal. The transfusions will not interfere with your pregnancy or the health of your baby.

If you have religious objections or concerns about blood transfusions it is essential that you let the obstetric and haematology team know as soon as possible so they can manage your care appropriately.

## How your labour and birth plan will be managed

The obstetrician and midwives will discuss and agree an individualised birth plan to take into account:

- your medical condition
- the condition of your baby
- your wishes for labour

Most women with SCD will carry their baby to term (37 weeks onwards). If you have had a caesarean previously, SCD should not stop you from attempting vaginal delivery.

If you've had hip replacements, it's important to consider that in your delivery plan. We normally would still aim for a normal delivery.

**Induction**

If you experience more frequent crises, are unwell, or the baby is not growing as expected it may be advised to induce labour early. Induction is more common among pregnant women with sickling disorders than without due to the possibility of complications with the baby.

Once you are in established labour it is normal to recommend continuous monitoring of the baby until birth.

An anaesthetist will talk to you about pain management. During labour you can have:

- epidural or regional block
- patient controlled analgesia (PCA)
- intravenous paracetamol

If labour does not progress following induction a caesarean section may be necessary and, in certain situations, a caesarean section may be planned without induction.

**Going home**

If you have had a vaginal delivery and there are no complications, you may be discharged within a few days of delivery.

However, if you have had a caesarean section or experienced complications, depending on your clinical situation, you may be advised to stay longer in hospital to recover.

When you're discharged, your midwife and health visitor from your local area will visit you at home for monitoring and advice.

## Caring for yourself and your baby

If your baby is well and there are no complications, they can stay with you in Queen Charlotte's & Chelsea Hospital and be discharged with you. It is not possible for you and your baby to be on Fraser Gamble ward or Weston ward post-delivery due to safeguarding and clinical issues.

If your baby was born earlier than expected or smaller than expected, he or she may be admitted to the special care baby unit (SCBU) or neonatal intensive care unit (NICU).

**Breastfeeding**
You will be encouraged to breastfeed your baby if not advised otherwise by clinical staff. If you have any concerns about breastfeeding, please discuss these with your midwife, health visitor or obstetrician.

The medicines given after birth are safe for breastfeeding with only very few exceptions. The doctors, midwives and pharmacists can answer any questions about breastfeeding and medicines.

**Blood spot test**

Your baby will be checked after birth by a 'heel prick' or 'blood spot' test. This will confirm if they may they have sickle cell disease or be a carrier.

If your baby does have SCD, he or she will be referred to the specialist paediatric haematology clinic at St Mary's Hospital.

You and your baby will also be followed up by the community nurse specialist for further support and advice in your local area.

## Support available

Many women are naturally anxious or stressed about how they are going to cope with pregnancy and a baby, particularly if their SCD condition has been severe, or if family or social support is limited.

If you are worried for any reason, please speak to your medical team, midwife, CNS and specialist social worker early on for advice about social support available to you.

If you have psychological concerns about coping during and after pregnancy, we can refer you to our clinical psychologist for counselling.

Contraceptive advice will be given at your postnatal clinic appointment, and this will include the following leaflet: Contraception advice for sickle cell disease patients.

## Benefits and entitlements during and after pregnancy

For advice on your benefits and entitlements visit: https://www.gov.uk/maternity-pay-leave

## How do I make a comment about my visit?

We aim to provide the best possible service and staff will be happy to answer any of the questions you may have. If you have any **suggestions** or **comments** about your visit, please either speak to a member of staff or contact the patient advice and liaison service (**PALS**) on **020 3312 7777** (10.00 – 16.00, Monday to Friday). You can also email PALS at imperial.pals@nhs.net The PALS team will listen to your concerns, suggestions or queries and is often able to help solve problems on your behalf.

Alternatively, you may wish to complain by contacting our complaints department:

Complaints department, fourth floor, Salton House, St Mary's Hospital, Praed Street
London W2 1NY

Email: ICHC-tr.Complaints@nhs.net

Telephone: **020 3312 1337 / 1349**

## Alternative formats

This leaflet can be provided on request in large print or easy read, as a sound recording, in Braille or in alternative languages. Please email the communications team: imperial.communications@nhs.net

## Wi-fi

Wi-fi is available at our Trust. For more information visit our website: www.imperial.nhs.uk

Clinical haematology
Published: December 2023
Review date: December 2026
Reference no: 2938