Clinical haematology

Pregnancy and sickle cell disease

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/CNS or GP early on if you are planning a pregnancy, or in the early stages of pregnancy to maximise your chances 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 and we will refer you to Queen Charlotte's & Chelsea Hospital. Your pregnancy with be managed jointly with the obstetric team at Queen Charlotte's & Chelsea Hospital for the duration of your pregnancy and for six weeks after the birth.

The effects of SCD in pregnancy are variable. All women with SCD during pregnancy within the North West London Network are advised to be referred to the joint haematology and obstetric clinic at Queen Charlotte's & Chelsea Hospital.

During this period you will be 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.

We can offer a prenatal diagnostics test for your baby during pregnancy, either by way of chorionic villous sampling or amniocentesis (amniotic fluid test). A diagnostic test can also be done after pregnancy and is referred to as a newborn blood spot screening.

Termination of pregnancy can be discussed in the event of the foetus being affected by SCD, if known in the first trimester.

Clinic attendance

The combined specialist clinic is held on alternate Monday mornings between 09.30 and 12.00 noon and you will see a consultant obstetrician, consultant haematologist and specialist midwife. We encourage you to attend clinic regularly for monitoring, scans and investigations as this will help us to provide you and your baby with the best care. Visits tend to be every 4 weeks until 28 weeks gestation, and then every 2 weeks until 36 weeks, and 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 located on the 2nd floor of Queen Charlotte's & Chelsea Hospital in the centre of fetal care (telephone 020 3313 3998).

How will pregnancy affect my SCD and are there related complications?

SCD in pregnancy affects women differently depending on your health status before becoming pregnant. The most common issues that may occur are:

- increased number of painful crisis during your pregnancy especially towards the end, (usually resolves after delivery)
- as your pregnancy progresses you may become more anaemic (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 the pregnancy
- close monitoring of the baby is recommended at every stage including during labour
- there may be an increased risk of high blood pressure. This is monitored closely at the antenatal visits. Ultrasound scanning will be done at frequent intervals due to your high risk pregnancy as follows:
    - early dating scan between 11-14 weeks
    - anomaly scan with uterine artery Doppler at 20 weeks
    - monthly scans for growth and fluid volume from 28 weeks

Medicines advice

Before 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
- Desferasirox (Exjade®) or Desferrioxamine (Desferral®) or other iron chelation agents - stop at least 3 months before conception or immediately after confirmation of pregnancy
- non-steroidal anti-inflammatory drugs (NSAIDS); Ibuprofen, Naproxen or Nurofen®
- Pregaballin and Gabapentin
- Tramadol

During your initial review we may advise you to take a preventative daily dose of a clotting agent throughout your pregnancy and for up to 6 weeks post-birth such as Tinzaparin. Women with SCD are at increased risk of blood clots during pregnancy, particularly if they are less mobile.

Aspirin (75mg) once a day is normally prescribed from the time of your initial scan until approximately 36 weeks to prevent pre-eclampsia.

We advise you to continue with Penicillin (250mg) twice daily or your alternative antibiotic and 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 if you are up to date with your recommended vaccines: Influenza (annually), Pneumovax (5 yearly), HIB/Single Men.C, MenACWY, MenB and Hepatitis B booster course.

How do I look after myself whilst pregnant?

Continue as before your pregnancy to prevent crisis by self-management. The following prevention plan can help you:

- 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 experience indigestion and it affects your appetite, eat smaller meals more frequently
- plan adequate rest if you become more tired as the pregnancy progresses
- you may need to 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
- identify early signs of infection; fever, productive cough with yellow or green sputum, pain passing urine, difficulty breathing and shivers. Seek medical help as soon as possible

What happens if I have a crisis whilst 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 on a daily basis 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 through the use of 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

Blood transfusions are sometimes given if you become anaemic below your baseline, or 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 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 will my labour and birth plan 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, and 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 have had hip replacements it is important to discuss a suitable delivery plan for you, though normally we would still aim for a normal delivery.

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 amongst 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. In terms of pain management, during labour you can have an epidural/regional block or patient controlled analgesia (PCA) and intravenous paracetamol as required. This will be discussed with you by an anaesthetist.

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.

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

Your baby will be checked after birth by a heal prick blood test to confirm the genetics i.e. if they may 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.

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: 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 3313 0088** (Charing Cross, Hammersmith and Queen Charlotte's & Chelsea hospitals), or **020 3312 7777** (St Mary's and Western Eye hospitals). 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

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