

# Information for you

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## Sickle cell disease and pregnancy

### About this information

This information is for you if you want to know more about pregnancy and sickle cell disease (SCD). You may find it helpful if you have SCD and are pregnant, or are thinking about having a baby. It may be helpful if you are a partner, relative or friend of someone who has SCD.

### What is SCD?

SCD is a condition that affects the red blood cells and the haemoglobin they contain. Haemoglobin is the protein that carries oxygen around the body.

SCD makes red blood cells more fragile so they break down more easily than usual. Instead of the red blood cells being round, they become sickle shaped (like a crescent) and are then known as sickle cells.

In SCD the amount of haemoglobin and the number of normal red blood cells in your body is less, causing anaemia. The sickle cells can cause blockages in small blood vessels. If this happens in your bones, it can cause severe pain and is called a 'crisis'. Crises are more common during pregnancy. Damage from sickle cells can also occur in the kidneys, lungs or eyes.

SCD can lead to high blood pressure, kidney problems, leg ulcers and damage to joints.

### What causes SCD and how common is it?

SCD is caused by a fault in the gene that makes haemoglobin. It is an inherited condition, which means it is passed down through families:

- if you have two of these genes (one from each parent), you will have SCD
- if you have one gene (from one parent), you will be a carrier of the condition; this is known as sickle cell trait.

SCD is the most common inherited condition in the world and mostly affects people whose family origins are in the Middle East, Sub-Saharan Africa, parts of India and parts of the Mediterranean. You cannot catch SCD.



Between 12 000 and 15 000 people in the UK have SCD and over 300 babies are born with the condition in this country every year. Parents of all babies born in the UK are given the opportunity to have their baby tested for SCD in the first week of life (the heel prick test).

## **Will my pregnancy be affected if I have sickle cell trait?**

No, carriers do not usually experience symptoms, but you can pass the condition on to your children if your partner is also a carrier, or has SCD (see below).

## **I have SCD. What should I think about before becoming pregnant?**

It is important that you let your SCD team know that you are planning to have a baby. They will be able to help you to be in the best possible health before you become pregnant. Until that time, they can advise you on which contraception is best for you.

If you have SCD, you should consider finding out whether your partner is also affected *before* getting pregnant. If your partner does not have SCD and is not a carrier, your baby will not have SCD.

If your partner has SCD or is a carrier, specialist counselling is available. This will help you both decide whether to have tests when you become pregnant to find out whether your baby has the condition. This can be a difficult decision for many couples and further information is available from the NHS Sickle Cell and Thalassaemia Screening Programme at: <http://sct.screening.nhs.uk> and <http://sct.screening.nhs.uk/professional-leaflets>.

Whether you are planning a pregnancy or not, you should see your SCD team at least once a year. The checks may include:

- a detailed scan of your heart (echocardiogram)
- blood pressure, urine and blood tests
- a special eye test (retinal screening) to detect problems at the back of the eyes.

## **Will I need to change my normal treatment before I become pregnant?**

If you are taking hydroxycarbamide (hydroxyurea), you should stop taking it and continue using contraception for 3 months before you become pregnant. Your SCD team will also review any other medicines you are taking.

## **What extra treatment might I need?**

People with SCD are at extra risk of infection, so you may be advised to take a daily dose of antibiotics (usually penicillin). Your vaccinations for hepatitis B, flu and pneumonia should be up to date.

You will be advised to take high-dose folic acid (5 mg) every day.

## **What are the risks to me and my baby in pregnancy?**

Most women with SCD will have a straightforward pregnancy and not have serious problems. However, painful crises can be more common during pregnancy. Cold weather, dehydration and doing too much physical activity can bring them on. You may need to take it easy. If you start to feel tired or have mild pain, you should rest.

SCD can also cause serious problems such as sudden anaemia and lung problems. If you have morning sickness (which can lead to dehydration) or have any other concerns, contact your maternity unit as soon as possible.

Pregnant women have a higher risk of developing blood clots in the legs (venous thrombosis) compared with women who are not pregnant. SCD makes you even more likely to develop a venous thrombosis. You can find out more about this from the RCOG patient information leaflet *Reducing the risk of venous thrombosis in pregnancy and after birth: information for you*, which is available at: [www.rcog.org.uk/en/patients/patient-leaflets/reducing-the-risk-of-venous-thrombosis-in-pregnancy-and-after-birth](http://www.rcog.org.uk/en/patients/patient-leaflets/reducing-the-risk-of-venous-thrombosis-in-pregnancy-and-after-birth).

There is also a higher chance that you may get pre-eclampsia (a condition of high blood pressure and protein in the urine) in later pregnancy. You can find out more about this from the RCOG patient information leaflet *Pre-eclampsia: information for you*, which is available at: [www.rcog.org.uk/en/patients/patient-leaflets/pre-eclampsia](http://www.rcog.org.uk/en/patients/patient-leaflets/pre-eclampsia).

SCD may also affect the growth of your baby because it can affect how your placenta works. You can find out more about this from the RCOG patient information leaflet *Having a small baby: information for you*, which is available at: [www.rcog.org.uk/en/patients/patient-leaflets/having-a-small-baby](http://www.rcog.org.uk/en/patients/patient-leaflets/having-a-small-baby).

You are more likely to go into labour early. If you don't, you are likely to be advised to have your labour started off (induced) at some point before your due date, to reduce the risks to you and your baby. You are also more likely to need a caesarean section. Your obstetrician and midwife will talk to you about your options.

## What extra care will I receive when I am pregnant?

You should be looked after by an obstetrician, a midwife and a haematologist (a blood specialist) with expertise in SCD. If you have not had the recommended tests in the previous year, they should be carried out. Your vaccinations for hepatitis B, flu and pneumonia should be updated if necessary. These vaccinations are safe in pregnancy.

You should be seen at the antenatal clinic at least every 4 weeks until your 24th week, and then every 1–2 weeks until you have had your baby. At each visit you will have your blood pressure checked and your urine tested. As well as the routine scans, you should have extra scans to check that your baby is growing normally.

Blood transfusions are not routinely given during pregnancy, but may be needed. If so, this will be discussed with you.

Your risk for thrombosis (blood clots in your legs or lungs) should be assessed in early pregnancy. If you have any other risk factors that make you more likely to get a blood clot, for example being overweight, you may be advised to have daily heparin injections throughout your pregnancy. This is safe to take while you are pregnant and should be continued for 6 weeks after your baby is born to reduce the risk of blood clots.

You should ask the team looking after you for contact details of whom you should call (usually your maternity unit) if you develop problems such as a sickle crisis, so that you can be seen promptly if you have difficulties in between clinic appointments.

## What medicines should I take in pregnancy?

If you are taking hydroxycarbamide (hydroxyurea), this should be stopped as soon as you know you are pregnant. Stopping it should not affect your health.

You should take:

- high-dose folic acid (5 mg) daily throughout your pregnancy
- your daily antibiotic
- low-dose aspirin (75 mg) daily from early pregnancy to reduce the risk of pre-eclampsia.

You can take painkillers such as paracetamol and codeine. Like all pregnant women, you should not take painkillers such as ibuprofen before 12 weeks and after 28 weeks of pregnancy without talking to your doctor as they could cause problems for your baby.

## **What if I have a crisis during pregnancy?**

If you become unwell, contact your maternity unit as soon as possible so that you can be seen urgently by medical staff and given treatment.

You will be given strong painkillers, oxygen to breathe, and fluids through a drip in your arm if you are dehydrated. You will also be checked for other causes of your symptoms and you may be given antibiotics. You should also be given heparin injections to reduce the risk of blood clots. You will be monitored closely – often in a high-dependency area of the hospital. Your baby's wellbeing will be checked.

## **What happens in labour?**

You should have your baby in a hospital that is able to manage SCD complications. You will be kept warm to reduce the risk of developing a crisis in labour. You may be given fluids through a drip to prevent dehydration and you may need oxygen. Blood that is suitable for you will be available in case you need a transfusion. Your baby's heartbeat will be closely monitored in labour.

You should be able to have a vaginal birth if there are no complications.

## **What about pain relief?**

You should see an anaesthetist before you go into labour to discuss pain relief. All the usual methods should be suitable for you except pethidine as it could cause complications.

## **What happens after my baby is born?**

You should be kept warm and well hydrated and you may be given extra oxygen to prevent a crisis. You will be encouraged to get up and about to help stop blood clots forming in your legs. You should be offered special stockings and daily heparin injections (to help thin the blood) for at least a week to reduce the risks further. You may be advised to continue heparin for 6 weeks if you have any other risk factors, for example caesarean section.

Breastfeeding is recommended, and you will be given the support you need.

Parents of all babies in the UK are given the opportunity to have their baby tested for SCD (together with other health checks) from a heel prick blood spot sample on about day 5 after birth.

## **What about contraception?**

Progesterone-only pills, injections (Depo-Provera), implants (Nexplanon), the Mirena coil and barrier methods (sheaths and caps) are safe and effective. The combined estrogen/progesterone oral contraceptive ('the pill') and copper coil can be used but only if the above methods are unsuitable for you. You can talk to your GP or family planning specialist.

## **Key points**

- SCD is the most common inherited condition in the world.
- Most women with SCD will have a straightforward pregnancy and a healthy baby.
- If you are planning a pregnancy, let your SCD team know so that they can review your medicines and vaccinations and make sure your checks are up to date.

- You and your partner can meet a specialist or counsellor to discuss the chance of SCD being passed to your baby and the tests available to you.
- A specialist team will look after you and your baby very closely during pregnancy.
- Having your baby at some point before your due date is usually advised.
- You will be given support to breastfeed.

## Further Information

NHS Sickle Cell and Thalassaemia Screening Programme: <http://sct.screening.nhs.uk> and <http://sct.screening.nhs.uk/professional-leaflets>

NHS Choices – Sickle cell anaemia: [www.nhs.uk/conditions/Sickle-cell-anaemia/Pages/Introduction.aspx](http://www.nhs.uk/conditions/Sickle-cell-anaemia/Pages/Introduction.aspx)

Sickle Cell Society: [www.sicklecellsociety.org](http://www.sicklecellsociety.org)

## Making a choice

### Shared Decision Making

If you are asked to make a choice, you may have lots of questions that you want to ask. You may also want to talk over your options with your family or friends. It can help to write a list of the questions you want answered and take it to your appointment.



### Ask 3 Questions

**To begin with, try to make sure you get the answers to three key questions if you are asked to make a choice about your healthcare.**

1. What are my options?
2. What are the pros and cons of each option for me?
3. How do I get support to help me make a decision that is right for me?

These resources have been adapted with kind permission from the MAGIC Programme, supported by the Health Foundation

\* Ask 3 Questions is based on Shepherd HL, et al. Three questions that patients can ask to improve the quality of information physicians give about treatment options: A cross-over trial. Patient Education and Counselling, 2011;84: 379-85



<http://www.advancingqualityalliance.nhs.uk/SDM/>

## Sources and acknowledgements

This information has been developed by the RCOG Patient Information Committee. It is based on the RCOG Green-top guideline *Management of Sickle Cell Disease in Pregnancy* (July 2011). The guideline contains a full list of the sources of evidence we have used. You can find it online at: [www.rcog.org.uk/womens-health/clinical-guidance/sickle-cell-disease-pregnancy-management-green-top-61](http://www.rcog.org.uk/womens-health/clinical-guidance/sickle-cell-disease-pregnancy-management-green-top-61).

The RCOG produces guidelines as an educational aid to good clinical practice. They present recognised methods and techniques of clinical practice, based on published evidence, for consideration by obstetricians and gynaecologists and other relevant health professionals. This means that RCOG guidelines are unlike protocols or guidelines issued by employers, as they are not intended to be prescriptive directions defining a single course of management.

This information has been reviewed before publication by women attending clinics in London, Birmingham and Manchester.

A glossary of all medical terms is available on the RCOG website at: [www.rcog.org.uk/womens-health/patient-information/medical-terms-explained](http://www.rcog.org.uk/womens-health/patient-information/medical-terms-explained).