

Pregnancy and sickle cell disease

The sickle cell and obstetric (pregnancy and childbirth) teams at Guy's and St Thomas' look after women with sickle cell disease (SCD) before, during and after pregnancy. Many women with SCD have questions about the effect pregnancy will have on their SCD, whether the SCD will affect the baby, and what will happen during pregnancy. This leaflet attempts to answer some of these questions. Please ask the SCD team if you would like to discuss further.

Why it is important to plan and discuss my future pregnancy with my haematologist?

There are a number of health issues related to sickle cell disease (SCD), which may need investigation or treatment before pregnancy. So it is important to see your haematologist when you are planning a pregnancy so we can work with you to increase your chances of a healthy and successful pregnancy.

We will carry out a health review to give us the information we need to maximise your physical well-being before pregnancy and plan how best to look after your health during pregnancy. We will review any medications you may need to start or stop taking before or during your pregnancy, and we will also give you a clear plan of who to contact when you know you are pregnant.

We will arrange for you to have a heart scan (echocardiogram or 'echo'), an eye test, and blood tests to review your general health and review medications. Common SCD medications that you will need to stop taking before you conceive include:

- **hydroxycarbamide** (hydroxyurea, Hydrea® or Siklos®) – a treatment for severe SCD
- **enalapril** – a medicine that protects your kidneys
- **deferasirox** (Exjade®) or **desferrioxamine** (Desferal®) or other iron chelation agents (medicines that reduce the amount of iron in your blood)
- **ibuprofen** and **diclofenac** or related painkillers, often known as non-steroidal anti-inflammatory drugs (NSAIDs).

The haematologist or clinical nurse specialist may also offer to refer you for specialist genetic counselling before pregnancy. The genetic counsellors can assess whether you and your partner are at risk of having a child with SCD or a related inherited blood disorder.

For couples who do not want to have a child with SCD but would not want to terminate an affected pregnancy, pre-implantation genetic diagnosis (PIGD) is an option. This involves removing mature embryos (eggs) from the woman's ovary. The eggs are then fertilised in a

laboratory, with sperm from the partner or a donor. The developing embryo is tested for the condition. If the embryo does not have the condition, it is placed in the woman's womb so she can become pregnant. This process is often referred to as 'in vitro fertilisation (IVF)' or having a 'test tube baby'. This is still a very new method for couples at risk of having a child with sickle cell disease or other genetic conditions. If you are interested in this option, please discuss it with your GP, genetic nurse counsellor or a specialist nurse.

How will pregnancy affect my sickle cell condition?

SCD in pregnancy affects each woman differently. You may experience more frequent SCD painful crises, especially towards the end of your pregnancy. This increased frequency usually resolves once your baby is born. You may become more anaemic (low levels of red blood cells), particularly as the pregnancy progresses.

Life-threatening SCD complications in pregnancy are rare in the UK.

What should I do when I become pregnant?

Although it is preferable for us to see you before you get pregnant, the most important thing is to contact us immediately when you realise you are pregnant.

You can contact the sickle team on 020 7188 2710 or 07770 683947.

We will arrange to see you as soon as possible at the clinic. This is an important appointment to reassess your health and medication, arrange further appointments and scans, give advice for the first few weeks of your pregnancy and schedule your first visit to the combined pregnancy/SCD clinic. In the combined clinic you will be seen by a consultant obstetrician, consultant haematologist, advanced nurse practitioner and community nurse specialists, who have expert knowledge in managing SCD during pregnancy. This clinic is usually the last Thursday of the month, and we will see you at this clinic every month during your pregnancy.

You will also be referred to the genetic counsellors if you have not all ready seen them. They can offer to take a blood test from your partner and let you both know whether your partner carries a sickle gene. The genetic counsellors can then assess whether you and your partner are at risk of having a child with SCD and discuss tests to detect whether your unborn baby has inherited SCD or a related inherited blood disorder. In addition, the genetic counsellors can explain how a child might be affected by SCD once it is born and explain the care and support that is available

We will advise you to continue with certain medications such as penicillin 250mg twice daily and folic acid 5mg once daily. Most pregnant women with SCD should also start taking aspirin 75mg daily. This reduces the chance of developing pre-eclampsia (high blood pressure, swelling and protein in the urine during pregnancy). We assess all pregnant women to determine their individual risk of a blood clot. If your risk is considered higher than average we will offer you treatment with heparin (a medicine to reduce your risk of blood clots) throughout your pregnancy. If you have a previous history of miscarriages, the team may suggest you have treatment with heparin in addition to aspirin throughout your pregnancy.

If you have SCD pain it is important to take painkillers. However, you should not take any NSAIDs, as these can be potentially harmful to your baby. There are many medicines in this group but the most common ones include ibuprofen, diclofenac and naproxen. If your recommended painkillers are not working to control your pain at home, you must attend hospital (see 'What happens if I have a sickle cell crisis while I am pregnant?').

The Tower team midwives will also see you on a regular basis. This specialist team looks after women with a variety of medical conditions. They will advise you on how to look after yourself and your baby during your pregnancy. They will check the health of the baby and help you plan for the birth and organise antenatal classes.

They will organise their first appointment for when you are 9-13 weeks pregnant. When you attend this visit you will be given a set of maternity notes (orange book). It is important to ensure that you bring the maternity notes with you each time you visit the hospital or see anyone about your pregnancy, as the team will need to record all relevant information in this book.

During your visits to the clinics and midwives you will be offered regular blood and urine tests to monitor your health. You can also have regular scans of your baby to see that it is growing and developing normally in the womb.

What pregnancy-related complications can women with SCD have?

Most women with SCD experience very few problems in pregnancy. However, sickle cell disease can make some pregnancy-related complications more common. These include:

- reduced growth of the baby
- miscarriage
- high blood pressure, swelling of the ankles and pre-eclampsia
- preterm (premature) labour and birth
- **blood clots in the legs, causing pain, redness or swelling in the calves, known as deep vein thrombosis or DVT**
- **blood clots in the lungs known as pulmonary embolisms (PEs), causing shortness of breath and chest pain**
- sickle cell related eye complications, including bleeding in the eyes or loss of vision
- increased likelihood of caesarean section (although many women with SCD have an uncomplicated labour and birth).

If you experience any symptoms of blood clots, please go to your nearest A&E department as these may be very dangerous.

How do I look after myself while I am pregnant?

Most women with SCD seem to manage their condition and pregnancy very effectively. Here are some basic tips to look after yourself while pregnant:

Diet and fluids

- Remember to drink plenty of fluids (three litres every 24 hours) to avoid becoming dehydrated, which may trigger a sickle crisis. If you experience morning sickness and/or vomiting and become dehydrated it is important to seek medical advice early.
- Eat a healthy, well-balanced diet.
- Eat plenty of fibre; constipation may be an issue as your pregnancy progresses.
- Indigestion is a common problem in pregnancy. It is better to eat smaller meals more frequently if indigestion is affecting your appetite.

Tiredness and fatigue

- You may feel more tired as the pregnancy progresses, especially if you become more anaemic. You may need to plan for adequate rest.

- You may need to slow down or reduce your activity, particularly as your pregnancy progresses. If you are struggling with work, study or home life it may be worth discussing your concerns with your doctor, midwife or nurse specialist.
- In most cases iron supplements are not recommended in pregnancy for women with SCD, as the cause of the anaemia is related to your underlying SCD condition and not iron deficiency.
- Taking your folic acid regularly will reduce the risk of anaemia as this helps your blood make red blood cells. Folic acid is essential for any healthy pregnancy.

Keeping warm

Ensure you keep warm enough, particularly in winter by wearing warm clothing and ensuring your home and work environment are a comfortable temperature. Remember, being cold is a major cause of sickle cell crises.

Infection

- Pregnancy and SCD increase the risk of infection. Taking your penicillin as prescribed. Please ensure you have your whooping cough vaccination and annual flu vaccines, as this helps to reduce the risk of infection.
- If you have signs of infection such as a raised temperature, cough that produces phlegm, or pain on passing urine, ensure you are immediately checked by a healthcare professional. Common infections in pregnancy include:
 - **urine infections**, which may cause stinging on passing urine; passing small, frequent amounts of urine; or strong-smelling urine
 - **chest infections**, which can cause you to have difficulty breathing, a cough and producing yellow or green sputum.
- SCD crises tend to happen when you have an infection, so it is important that you see a doctor to check if any treatment is required.

Attending clinics

We encourage you to attend the regular clinics and related scans or investigations. This will help us to provide you and your baby with the best care possible during your pregnancy.

Travel

If you plan to travel abroad while pregnant please let the team know when you are making plans. We will then be able to offer advice and consider anti-malarial medication if necessary.

What happens if I have a sickle cell crisis while I am pregnant?

You should attend hospital promptly if your SCD-related pain cannot be managed by your recommended painkillers or if you have chest pain or difficulty breathing.

- Your painkillers will be reviewed and others added if necessary, which may include weak opiates such as dihydrocodeine, or stronger opiates such as morphine. Opiates are safe for you and your baby and may be needed for moderate or severe pain, which is usually only for a short time. Controlling your SCD pain will help you and your baby's health.
- We will give you fluids either orally or by a drip into your vein (intravenous, IV) to ensure you are well hydrated. This will prevent or treat a sickle cell crisis.
- We may give you oxygen, which can help improve the symptoms of your SCD crisis and reduce the risk of complications.
- We will treat any signs of infection with oral or IV antibiotics.

Blood tests are normally undertaken daily if you are unwell. These blood tests tell the doctors about your haemoglobin levels, signs of infection and how vital organs like your liver and kidneys are working.

Where should I go?

If you have a painful crisis and you are pregnant you should **not** use the SCD pain service in the haematology day unit at Guy's Hospital.

If you have an SCD crisis when you are less than 18 weeks pregnant you will need to attend A&E at St Thomas' Hospital (or a local A&E department if you are unable to travel to St Thomas'). If you are admitted to St Thomas', in most cases you will be taken to a medical ward while in crisis and reviewed by the sickle team daily until you are discharged (once your symptoms have resolved).

If you have complications such as abdominal pain or vaginal bleeding and you are less than 18 weeks pregnant, you should attend the Early Pregnancy / Acute Gynaecology Unit (EPAGU) on the 8th floor, North Wing, St Thomas' Hospital.

- The EPAGU is open Monday-Friday 8.30am-6.30pm, and weekends 9.30am-3.15pm.
- Patients attending outside opening hours should go to A&E.

If you have an SCD crisis or you have pregnancy-related concerns and you are more than 18 weeks pregnant, go to the Antenatal Day Unit (ADU) on the 7th floor, North Wing, St Thomas' Hospital. In the ADU you will be treated for your SCD crisis as normal and the health of your baby will be monitored. If for any reason you need to be admitted to the Antenatal Ward or Birth Centre, staff on the ADU will organise your admission.

- The Antenatal Day Unit opening hours are 8am-8pm, Monday to Friday, and 9am-4pm, weekends and bank holidays.
- Women attending outside these hours must attend the Hospital Birth Centre on the 7th floor, North Wing, St Thomas' Hospital.

Admission into hospital

- If you are admitted, you will be reviewed each day by the obstetric team and sickle cell / haematology teams, and they will jointly manage your care with the midwives.
- You will normally be given blood-thinning injections once a day in hospital. This is because women with SCD are at risk of blood clots and this risk increases when you are in hospital and less mobile.

Blood transfusions and exchange blood transfusions

Blood transfusions are sometimes given if your red blood cell levels become very low and you become anaemic. Anaemia may be more likely to develop when you are in pain as the red blood cells are destroyed in an SCD crisis.

The procedure will be explained to you in full, but if you would like more information on having a blood transfusion, please ask your nurse or midwife for a copy of the leaflet **Having a simple (top up) blood transfusion: Information for adult patients with sickle cell disease**.

Occasionally the haematologist may recommend an exchange blood transfusion. Again, this will be explained to you in full, but please ask for a copy of the leaflet **Having an exchange blood**

transfusion: Information for adult patients with sickle cell disease if you would like further information.

If you are on regular blood transfusions or exchange blood transfusions this will continue as normal. This treatment 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 teams looking after you know as soon as possible.

How will my labour and baby's birth be managed?

The obstetrician and the midwives will discuss and agree an individualised care plan that will take into account your wishes for labour and the birth of your baby. If caesarean section is recommended, the obstetrician will ensure that the reasons for this are explained and that you can ask questions and give your fully informed consent.

When you first go into labour you should call the Tower Team midwives on 07717 700276. If unavailable, contact the Hospital Birth Centre on 020 7188 6867 / 2975.

Many women with SCD seem to cope very well with the pain related to labour. This may be because women with SCD are used to coping and dealing with the pain that is linked to SCD crisis.

Analgesia (pain relief) commonly used in labour includes nitrous oxide and oxygen (commonly called gas and air) and, if required, an epidural, which is an injection into your back that blocks out pain from the waist down. Blood-thinning injections are also stopped around the time of your labour and birth, to decrease the risk of bleeding.

The haematology SCD team will also be informed that you have been admitted in labour and will review you throughout your admission.

Most women with SCD will carry their baby to term and will give birth normally. However, if you are developing more frequent crises, if you are unwell due to your SCD, or if the baby is not growing as well as expected, the obstetrician will consider inducing an earlier labour (usually at around 38 weeks). Induction of labour is more common among women with SCD. Once you are in established labour it is normal to recommend continuous electronic monitoring of the baby until birth although you will still be encouraged to adopt upright positions and to move around.

If labour does not establish or progress well following induction, you may need a caesarean section. In some situations this may be planned before labour. The aim is to keep you well throughout your pregnancy and enable you to have a healthy baby.

Discharge

How soon you go home after you have your baby will depend on how your baby was born and how well you are. If you have a vaginal birth and there are no complications you may be discharged within one or two days. However, if you have had a caesarean section or there are complications, you may need to spend longer in hospital. Because women with SCD are thought to be at greater risk of blood clots, you may need to carry on with blood-thinning injections at home for 1-6 weeks after the birth. Before you are discharged, the midwife will teach you or a family member how to inject blood-thinning injections.

After discharge from hospital you will be sent an appointment to be seen in the SCD clinic at Guy's Hospital. If you are not sent an appointment letter within two months, please phone the

clinic on 020 7188 2743 or contact your sickle cell nurse. If you have care elsewhere for your SCD please ensure you are followed up in your usual SCD clinic

The Tower team midwives will normally see you at your home within 10 days of the birth if you live in the area. If you live outside of the area they will arrange for your local midwifery team to contact you.

Caring for yourself and your baby

If your baby is well and there are no complications, the baby will stay with you while you are in hospital and go home at the same time as you. Some babies born before their due date, or small-for-date babies, may be admitted to the Special Care Baby Unit (SCBU), or the Neonatal Intensive Care Unit (NICU) at St Thomas' Hospital. You will be encouraged to care for your baby and spend time with them while in SCBU or NICU. You will also be encouraged to breastfeed your baby. If you have any concerns about breastfeeding, please discuss this with your midwife or doctor.

The medicines you will take after you've had your baby are generally safe for breastfeeding. The doctors, midwives and pharmacists can help you with any questions you have about breastfeeding and medicines.

If you are concerned that your baby has an increased risk of inheriting SCD or a related inherited blood condition, please tell the medical and nursing teams. They will arrange for your partner and baby to be tested. If your baby does have SCD or a related inherited blood condition, they will be referred to the specialist paediatric (children's) SCD clinic at the Evelina Children's Hospital on the St Thomas' Hospital site.

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 in cases where there is little family or social support. If you are worried for any reason please discuss this with your doctor, midwife or nurse specialist, as they can advise you about the support services available to you. If you have concerns about coping during and after pregnancy, you can contact the sickle cell psychology team directly and they will be happy to offer you an appointment. You will find their telephone numbers at the back of this leaflet.

Contact us – sickle cell team

Consultant Haematologists Dr Jo Howard and Dr Rachel Kesse-Adu
t: 020 7188 82741 (Secretary)

Specialty Registrar (sickle cell)
bleep: 0248 (via the main switchboard)

Junior Doctor (sickle cell)
bleep: 2283 (via the main switchboard)

Advanced Nurse Practitioner Mr Neill Westerdale
t: 020 7188 2710
077706 83947 (mobile)
bleep: 1843 (via the main switchboard)

Clinical Nurse Specialist (adolescent) Mr Luhanga Musumadi
t: 020 7188 2710
07770 678851 (mobile)
bleep: 2256 (via the main switchboard)

Clinical Nurse Specialist Mrs Judith St Hilaire
t: 020 7188 2710
07920 711266 (mobile)
bleep: 2868 (via the main switchboard)

Psychologists
Consultant Health Psychologist Dr Nicky Thomas
t: 020 7188 2725

Clinical Health Psychologist Dr Heather Rawle
t: 020 7188 2718

Health Psychologist Mina Abedian
t: 020 7188 2718

South East London Sickle Cell & Thalassaemia Centre
Wooden Spoon House, 5 Dugard Way
London SE11 4TH

t: 020 3049 5993

Contact us – obstetrics

Consultant Obstetrician Mr Eugene Oteng Ntim
t: 0207188 6874 (secretary)

Tower Team midwives
t: 07717700276

Hospital Birth Centre
t: 020 7188 6867 / 2975

Antenatal Day Unit
t: 020 7188 1722 / 23

Antenatal Ward
t: 020 7188 0677

Postnatal Ward
t: 020 7188 2233

Antenatal Appointments
t: 020 7188 8001

Early Pregnancy and Acute Gynaecology Unit
t: 020 7188 0864

Further sources of information

Pharmacy medicines helpline

For information about any medicines that you have been prescribed at Guy's and St Thomas' hospitals, you can speak to the staff caring for you or call our helpline.

t: 020 7188 8748 9am to 5pm, Monday to Friday

Your comments and concerns

For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.

t: 020 7188 8801 (PALS) **e:** pals@gstt.nhs.uk

t: 020 7188 3514 (complaints) **e:** complaints2@gstt.nhs.uk

Knowledge & Information Centre (KIC)

For more information about health conditions, support groups and local services, or to search the internet and send emails, please visit the KIC on the Ground Floor, North Wing, St Thomas' Hospital.

t: 020 7188 3416

Language Support Services

If you need an interpreter or information about your care in a different language or format, please get in touch:

t: 020 7188 8815 **e:** languagesupport@gstt.nhs.uk

NHS 111

Offers medical help and advice from fully trained advisers supported by experienced nurses and paramedics. Available over the phone 24 hours a day.

t: 111

NHS Choices

Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.

w: www.nhs.uk