

Care and management of your child with

# Sickle Cell Disease

A parent's guide



Oni L, Dick M, Smalling B, Walters J  
2nd Edition

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# FOREWORD

Perhaps you have just been told that your newborn baby or older child has sickle cell disease and have learnt that it is an inherited blood disorder. Parents like yourselves are often shocked, confused, angry and afraid, you cannot believe it, you may feel responsible or blame each other for 'giving' your child a disease. Often parents don't know what to say to relatives and friends. They are bewildered about how to care for their child and what to expect.

Such feelings are natural to all parents when their child has been found to have an inherited disorder, whether it is sickle cell disease or any other inherited condition.

We hope that this handbook will help you come to terms with some of these feelings and help you to learn more about sickle cell disease so that you will feel confident about looking after your child and will know how and what to tell relatives and friends.

You may be living in an area where there are few people with sickle cell disease and your local health workers may not have cared for many people with sickle cell disease. If that is the case, it is worth mentioning this book, or taking it with you when you consult the health workers, especially on your first visit.

You may already have been told frightening things about sickle cell disease, or may know a friend or member of your family who has the condition. Sickle cell disease is very variable and affects people in different ways, even people in the same family. There are quite a lot of myths and popular misconceptions about sickle cell disease. We hope that this handbook will dispel some of these and give you clear information.

We hope that the handbook is easy to use and understand. Some of the scientific terms may be new to you. These are briefly explained in the glossary (page 84). We intend the book to be an introduction and a start for you to get further information from your doctor, specialist nurse/counsellor, specialist social worker, paediatrician or haematologist. You may want to read it all the way through. You can also just dip in to particular sections when you want to find out about a specific aspect of sickle cell disease.

Sickle cell disease affects both males and females. Apart from a medical condition called priapism, which only affects males, all the things discussed in this book relates to both sexes. But for fluency the term 'he' is used in the first half of the book and 'she' in the second.

**Please remember that not all the things mentioned in this book will affect your child.**

## PARENTS SHARE THEIR EXPERIENCES

"I remember when I was first told that my son has sickle cell. He was eight months old and I had taken him to the hospital because he had a temperature ... They did all these tests and the doctor told me, well I cried and cried and cried, I must have cried for ever. Every time I look at him I worry, what if I can't tell when he's in pain or sick. I used to watch him all the time; I used to check him when he is sleeping. At first I didn't want any one else to look after him, even my husband ... From the time I found out I just kept waiting for the crisis ... you know the crisis? ... the sickle cell crisis? Every day I wait for the crisis. After about a year of waiting and he didn't have one I got even more worried ... maybe it is all happening secretly in his little body ... so I used to just take him to my doctor (GP) anyway, just to be sure ... he did not have his first crisis until he was three years old. In the end you learn to take each day as it comes and cope with it when it comes ..."

*Mother of a twelve-year-old, with sickle haemoglobin C disease*

"When my baby was born she looked lovely and I was very proud, but when I was told she has sickle cell anaemia I was very upset. I thought she was going to die like my brother back in Ghana and I did not want to bond with her. But with time and the support of the professionals we learn to live with her condition and we love her very much. She does not have many crises and we hope that there is going to be a cure one day."

*Mother of a ten-year-old with sickle cell anaemia*

"My baby was three months old when the haematology counsellor told me about my baby's situation. I cried for five minutes non-stop and the first question that came to my head to ask was, "What am I going to do?" ... My partner didn't believe it. He said, "The doctors are lying." I told my closest friend. She said, "This boy looks very well and healthy, there must be a mistake." She then cuddled me and we both cried ...The sickle cell clinic is a great help. Now that my baby is two years old I am coping very well because he can tell me where the pain is and if he is tired ..."

*Mother of a two-year-old with sickle cell anaemia*

"I remember the day I was told he has sickle cell as if it was yesterday. I was totally devastated and would not accept the news at first. Over the years, with the help of the sickle cell team and being a Christian, I have been able to accept my son's illness and give him my full support."

*Mother of a nine-year-old, with sickle cell anaemia*

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“ You learn not to let it take over your life ... yes, it can be hard at times but you learn to cope. You have to because otherwise if you can't cope how will she learn to cope as she is growing up? The most important thing for me is that our daughter has to have a normal childhood, yes there may be some things she can't do but there are a lot of things she does better than her mates. The school says she is very bright for her age: her reading age is two years ahead of her class, that's because I take time to work with her. Our daughter has about three crises a year and then she misses school ? but the school is very good, her teacher lets me know what they are doing in class.”

*Father of a nine-year-old with sickle cell anaemia*

“ It took me a long time to come to terms with it; in fact my husband accepted it quicker than me. When we found out, my daughter was two months old. My husband just said, “ Well, it's happened ? it's happened.” That made me angry ... we had a big row after the nurse left. I felt he did not understand how serious this thing is ... or that he did not care, but when I look back on it now that was his way of coping with the news. He just wanted to know every thing about it so that he could look after our daughter. I stayed angry for weeks, and I could tell my husband was worried too but he was trying not to show it. She has had two crises, one when she was about a year old and another one when she first started school (aged four and a half). I cope, the whole family is involved. We made sure my mum and his dad know everything about it, and other family members because they baby sit for us... My mum is great, she calms me down when I get worried and start to panic. We have been lucky; our daughter has not really had many problems. We try to do all the things that will help her to stay well. Our nurse counsellor is really good. If I am worried I just ring her up”

*Mother of a seven-year-old with sickle haemoglobin C disease*

“ When my boy was born and we were told that he has sickle cell anaemia we were devastated. Me and my husband were blaming each other. We felt so ashamed we did not want to tell our relatives. Over the years we had a lot of support from the sickle cell counsellors and the hospital doctors. We know a lot more about sickle cell now and although we will always worry about our child, we have the confidence to look after him. We pray to God that a cure will be found.”

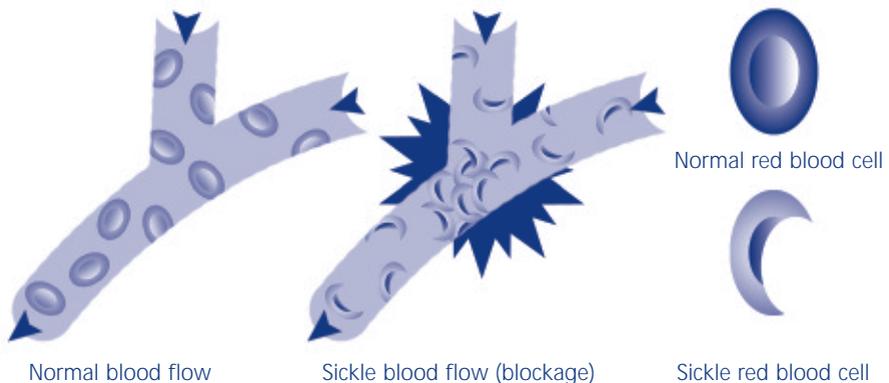
*Mother of an eight-year-old with sickle cell anaemia*

## WHAT IS SICKLE CELL DISEASE AND SICKLE CELL CRISIS?

Sickle cell disease is a term covering a number of different but similar conditions that affect haemoglobin. Haemoglobin gives blood its red colour and is responsible for carrying oxygen from the lungs to all parts of the body. The types of sickle cell disease commonly seen in the United Kingdom are sickle cell anaemia, sickle haemoglobin C disease and sickle beta thalassaemia disease. These are described further on page 6.

These conditions are called 'sickle' cell because the red blood cells, which are normally round and very flexible, become shaped like a crescent moon or farmer's sickle. Red blood cells in sickle cell disease do not last as long in the body as normal red blood cells and this leads to anaemia.

Sickled red blood cells are also not as flexible as normal red blood cells and cannot always pass through the very small blood vessels. If the sickled cells get trapped in the blood vessels, blood cannot flow through easily, this causes a blockage and leads to pain in the affected part. This is known as a sickle cell crisis, or pain crisis: the pain often comes on suddenly, and may last several hours or days.



### What causes the cells to sickle?

The red blood cells change into a sickle shape when they are in the veins because they lack oxygen. They return to their original round shape when they are in the arteries where there is more oxygen. Certain things can contribute to increase sickling, for example:

- Decreased amount of oxygen (air)

- 
- Dehydration (lack of water in the body)
  - Infection
  - Sudden changes in body temperature
  - Physical exertion
  - Stress

See page 14 for more information about how to avoid some of these.

## How did our child get sickle cell disease?

Sickle cell disease is an inherited condition. This means that your child inherited an unusual haemoglobin gene from you and your partner. (See illustration of inheritance on pages 52-58).

### **Sickle cell**

- is NOT Leukaemia or cancer or HIV
- is NOT iron deficiency
- is NOT infectious or catching
- is NOT white cells eating red cells

The normal and most common haemoglobin type is haemoglobin A, which stands for adult haemoglobin. There are over 1000 unusual haemoglobin genes but the ones that are commonly seen in the United Kingdom which affect the adult haemoglobin gene are haemoglobin S (which is sickle haemoglobin), haemoglobin C, haemoglobin D, beta thalassaemia, and alpha thalassaemia. All babies are also born with baby or fetal haemoglobin called haemoglobin F, regardless of which adult haemoglobin gene they have inherited from their parents. Haemoglobin F accounts for 90% of the newborn baby's haemoglobin at birth. By one year of age this level drops to about 1% and remain at this level right through adulthood.

An inherited condition like sickle cell disease remains with a person all his life although there is research into new forms of treatment that might change the gene. Sickle cell disease varies in its severity from one person to the next for reasons that are not clear even to research scientists. It is known that inheriting alpha thalassaemia trait (also commonly known as alpha thalassaemia carrier) or the ability to carry on making haemoglobin F (baby haemoglobin) does tend to make the sickle cell disease less severe.

There are many things that you can do to keep your child healthy and it is important to recognize early signs of illness which can then be treated promptly.

## TYPES OF SICKLE CELL DISEASE

It will usually be clear from the first blood test what sort of sickle cell disease your child has inherited. Sometimes this is not the case, particularly if the two parent's haemoglobin type is not known. In these cases it may be necessary to wait until your child is about six months to a year old before being certain as to what sort of sickle cell disease your child has inherited, although a special genetic test can also be requested to get the answer sooner. Once the result is confirmed, make sure that your nurse specialist and doctor tell you what sort your child has inherited.

**Sickle cell anaemia:** is the commonest form of sickle cell disease and occurs if your child has inherited a haemoglobin S gene from both parents to give haemoglobin SS, sometimes written, HbSS. How sickle cell anaemia will affect your child is very variable and it is not possible to predict this when your child is very young. Sickle cell anaemia tends to be the most serious of the sickle cell conditions, but this is not always the case.

**Sickle haemoglobin C disease:** occurs if your child has inherited a haemoglobin S gene from one parent and a haemoglobin C gene from the other parent to give haemoglobin SC disease, sometimes written, HbSC. In general, haemoglobin SC disease is milder than sickle cell anaemia but many of the same health problems can occur.

**Sickle beta thalassaemia disease:** occurs if your child has inherited a haemoglobin S gene from one parent and a beta thalassaemia gene from the other parent, sometimes written, HbS<sup>Thal</sup>. The beta thalassaemia gene is different from the sickle cell gene, but it can be inherited with a sickle gene to cause sickle beta thalassaemia disease. In some cases a small amount of the usual Haemoglobin A is produced, in this case the child has sickle beta plus thalassaemia, sometimes written as HbS<sup>B<sup>+</sup>Thal</sup> which tends to be a mild form of sickle cell disease. When no haemoglobin A is produced the condition is known as sickle beta zero thalassaemia, sometimes written as HbS<sup>0Thal</sup>. This behaves more like sickle cell anaemia. For more information about beta thalassaemia contact the UK Thalassaemia Society or one of the specialist Centres (addresses on page 87).

**Sickle with hereditary persistence of fetal haemoglobin:** occurs if your child has inherited a haemoglobin S gene from one parent and a persisting haemoglobin F gene from the other parent. It is sometimes written, HbS/HPFH and very rarely causes any health problems.

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**Sickle haemoglobin D disease:** is a more unusual form of sickle cell disease and occurs if your child has inherited haemoglobin S from one parent and haemoglobin D from the other parent, sometimes written, HbSD. This form can be as serious as sickle cell anaemia but could be milder.

In this book we will mainly write about sickle cell disease as if it is one condition although we recognize that the different types are not all the same in the way they affect a person's health. Some are milder than others and children with the same sort of sickle cell disease may have different experiences of sickle cell disease and medical problems. Where there are obvious differences or where there are specific health problems, this will be made clear.

## SICKLE CELL TRAIT (CARRIER) AND OTHER TRAITS

Having a trait is commonly known as being a carrier and does not affect the person in any way. It is not a form of sickle cell disease and will never turn into sickle cell disease. If a person is born with a trait they are healthy and will always have a trait.

Sickle cell trait means that a person has inherited one normal haemoglobin A gene and one sickle haemoglobin S gene from their parents, sometimes written as HbAS.

Other common haemoglobin traits seen in the UK include haemoglobin C trait, where a person has inherited one normal haemoglobin A and one unusual haemoglobin C gene from their parents (HbAC). Beta thalassaemia trait is when a person has inherited one



normal haemoglobin A and a beta gene from their parents HbA $\beta$ <sup>Thal</sup>.

The importance of knowing if one has sickle cell trait or any other unusual haemoglobin trait is because the unusual haemoglobin gene can be passed on to children through genetic inheritance. How this inheritance works is explained further on page 52.

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## Why did sickle cell occur and who does it affect?

It is thought that the sickle cell haemoglobin gene was a small change (mutation) in the haemoglobin A gene many thousands of years ago in countries where malaria was common. This is why we find haemoglobin S in people whose ancestors come from Africa, Asia, the Middle and Far East and the Mediterranean.

Sickle cell trait is found in approximately:

- 1 in 4 West Africans
- 1 in 10 African-Caribbeans
- 1 in 20 Portuguese
- 1 in 20-50 Asians
- 1 in 100 Northern Greeks

It is also seen very rarely in White English people and other Northern Europeans because of intermarriage and subsequent gene mixing between different ethnic groups. It is important to realize that inheritance of sickle cell or any other unusual haemoglobin is not to do with the colour of a person's skin; it is to do with the genes they have inherited from their parents.

In areas of the world where malaria is a danger people who have sickle cell trait (HbAS) were more likely to survive malaria than those who have the usual haemoglobin (HbAA). This advantage meant that haemoglobin S was more likely to be passed down through the generations by those with sickle cell trait. Unfortunately those who inherited two of the sickle cell genes and have sickle cell anaemia (HbSS) were not protected from malaria, and were also likely to have the medical problems which are described later in this book.

For information about malaria and the need for protection see page 39.

All the other unusual haemoglobin genes that have been described here probably also offer some protection against malaria.

## HOW WILL SICKLE CELL DISEASE AFFECT MY CHILD?

During the first three to six months of life, your child may not show signs of having sickle cell disease, because at birth there is a high (about 90%) level of baby haemoglobin F, sometimes written (HbF), and a very low level of sickle haemoglobin S (HbS) (about 5-10%). Over the first year of life the haemoglobin F level gets less as the child starts making more haemoglobin S. Some children continue to make higher levels of haemoglobin F even into adulthood. This can be checked by doing a special blood test. The longer your child goes on making haemoglobin F the better, because it means he will be making less haemoglobin S and is less likely to have many sickling crises. (See page 25 for an explanation of sickle cell crisis.)

### The effects of sickle cell disease

**Anaemia:** When a child is making a lot more Haemoglobin S their red blood cells will not live in the circulation as long as cells that contain the usual Haemoglobin A. The body tries to keep up by making more red blood cells but it usually cannot keep up completely and your child becomes anaemic. Your child may look pale and the palm of the hand and the lips will be paler than your own. This sort of anaemia is known as a haemolytic anaemia and is not the same as the sort of anaemia caused by lack of iron. For this reason, iron tonics or medicines should not be given unless prescribed by your child's doctor. (Also see page 14.)

Folic acid tablets or medicine may be prescribed by your doctors because the body uses folic acid when making new red blood cells. In the United Kingdom, most children get enough folic acid from their normal diet and extra folic acid tablets are usually not necessary.

**Jaundice:** When the red blood cells come to the end of their useful life they are broken down in the body. One of the substances that are produced is a yellow pigment called bilirubin. The liver clears the bilirubin from the body, but if there is a lot of bilirubin the liver may not be able to clear it all away and the yellow pigment may appear in the eyes, a condition known as jaundice. Some children may always have slightly yellowish eyes, even when they are well. Others may only become jaundiced when they are unwell, for example with coughs and colds or if they have a painful crisis.

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**Enlarged spleen:** The spleen is an organ that lies on the left side of the tummy under the rib cage. One of the first things that your doctor may notice is that your child's spleen is big and can be felt just below the rib cage. The spleen helps to clear infection from the body and also clears up old or damaged blood cells. The spleen may continue to be enlarged for some time but then reduces in size and may stop working altogether. This is because it becomes jammed with the sickled red blood cells that it is trying to clear from the body. If the spleen gets jammed with sickle cells it cannot clear the body of infection. This is why we recommend that your child takes penicillin twice daily. (See page 16 for further information on penicillin.)

Sometimes the spleen traps more blood than just the sickled red blood cells. When this happens the spleen suddenly gets very big and tender and the child becomes very pale. This is known as acute splenic sequestration. The child needs admission to hospital urgently and will probably need a blood transfusion. (See page 25 for information on spleen problems and page 34 for information on blood transfusions.)

It is important for you to learn how to feel your child's tummy to be able to know if the spleen becomes bigger than normal. Ideally this should be done daily, perhaps during bath time, but particularly if he seems unwell. The doctor or your nurse specialist will be able to show you how to feel the spleen to see if it is getting any larger.

**Pain:** One of the first signs of sickle cell disease may be painful swelling of the fingers and hands or toes and feet. This is known as dactylitis (hand foot syndrome) and may occur from about 6 months of age. If your child has been crawling or walking and then suddenly seems reluctant to do this it may be because of dactylitis. The child will need to be given regular pain killers and plenty of fluids and occasionally may need admission to hospital, (see page 22) for how to manage sickle cell crisis pain at home. The swelling usually goes down after a few days. It is unusual for children to have dactylitis after about 18 months of age. A typical painful episode or crisis after this time affects the arms or legs, but may affect the back and sometimes the abdomen (tummy).

It may be difficult when your child is young to know if they are in pain, and sometimes it is best to give a painkiller even when you are not sure. Of course, like everyone else, your child may have pain not due to a sickle cell crisis. This can be confusing but as they grow up you will learn how to tell the difference.

**Physical growth and development:** It is usual for children with sickle cell disease to be thinner and slightly shorter than children who do not have sickle cell disease; however they generally grow at a steady rate. They tend to go through puberty at an older age than usual and this means that they also go on growing for a little bit longer but eventually reach their normal adult height.

**Bed wetting (also known as nocturnal enuresis):** It is normal for children to wet the bed at night up until the age of five or six years. It may take longer for a child with sickle cell disease to become dry at night. This may be in part because the child passes very dilute urine and also because the child drinks more fluid so as to prevent dehydration. Most children will become dry on their own with support and encouragement but some need referral to a special clinic where treatment can be provided. Children who wet their beds should never be punished as it is not something that they have conscious control over. If your child is over six and still wets the bed, ask your doctor to refer him to the bed wetting clinic.

### Will my child have all these symptoms?

Not necessarily. Children, especially those who have sickle cell anaemia, HbSS or those with sickle beta zero thalassaemia (HbS $\beta^0$ Thal) are usually anaemic and may get jaundiced when they are unwell. Some children are mildly jaundiced all the time. Not all children with sickle cell disease have a big spleen and not all children get dactylitis (hand foot syndrome). Some children rarely experience pain although this is the commonest symptom of sickle cell disease.

Children who have haemoglobin SC or S $\beta^+$ Thal tend to be only very slightly anaemic and usually do not get jaundiced unless they have a crisis. An enlarged spleen in these sorts of sickle cell disease is quite a common finding but does not usually cause any serious problems.

For some other complications of sickle cell that may affect a child with sickle cell disease see page 25.

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# WHAT CAN I DO TO KEEP MY CHILD WELL?

In the first few months of life, your child will grow and develop like any other baby and will not be affected at all by sickle cell disease. This is because he will still be producing a lot of baby haemoglobin F and not so much haemoglobin S. After about six months it is possible that sickle cell problems may occur. There are basic precautions that you can take to help keep your child well and these are outlined below. Please remember that it is not always possible to prevent a sickle cell crisis, see page 22 for how to manage a sickle cell pain crisis.

## Diet and nutrition

All growing children need protein, carbohydrates, fat, vitamins and minerals. These they will get from a diet containing, fish, meat, fresh fruit and vegetables. It is recommended that we should all eat five portions of fruit and vegetables every day. Children with sickle cell disease do not need special food. They should eat the same foods as the rest of the family. If your family is vegetarian it would be best to talk to your health visitor to check that your child is getting enough protein and fat in his diet.



Sometimes children with sickle cell disease eat things which are not nutritious, such as chalk, paper or foam. This is known as pica and the cause is not known. It is usually not harmful but it is worth mentioning it to your child's doctor.

Children with sickle cell disease are more at risk from certain infections, which include food poisoning caused by salmonella infection. Chicken and eggs can be infected with salmonella. It is important to cook these and other foods thoroughly. Salmonella can lead on to a bone infection called osteomyelitis. (See page 27.)

It is important to thoroughly defrost frozen food before cooking and to make sure chilled foods from the supermarket are cooked according to the maker's instructions. Extra care needs to be taken if re-heating previously cooked food. Make sure the food is heated right through, especially if you are using a microwave oven.

Parents often worry that their child with sickle cell disease is not eating enough and is not putting on weight. This is very rarely the case. Children with sickle cell disease tend to be thin but they usually grow at a steady rate. Your child will be routinely weighed and measured at the outpatient clinic. Should there be a problem with growth this will be identified early. Your child needs to be encouraged to develop feeding skills at the

appropriate age and eat food at regular mealtimes with the rest of the family. It is not a good idea to try and force a child to eat as this may make him more reluctant to eat normally.

If fasting is part of your family's religious practice, you need to consider the special needs of your child with sickle cell disease. Although he should not be brought up any differently from his brothers and sisters, fasting for long periods of time may cause health problems.

Keeping your cultural and religious practice is part of staying healthy. Before your child reaches the age when they are expected to start fasting, it may be helpful to organize a meeting with your religious leader, nurse specialist or doctor so that you can discuss together your child's specific health and religious needs.

### What about giving extra vitamins or iron?

Your child does not need extra vitamins unless your family eats a special diet, for example, vegetarian or vegan, in which case it may be necessary to get advice from your health visitor or a dietician. If your child becomes more anaemic than usual, folic acid supplements may be prescribed by your doctor. This helps the body to make more red blood cells.

Some clinics prescribe folic acid, 5mg once a day, routinely, but a normal balanced diet will contain sufficient folic acid and daily supplements are generally not required in the UK.

Tonics containing iron or iron tablets should not be given. Your child is anaemic because the red blood cells are more fragile and do not live as long as the usual red blood cells. He does not have the sort of anaemia caused by insufficient iron in the diet. If he does need iron the doctor will tell you and will prescribe the right amount for your child's specific needs.

If you are giving your child any herbal (e.g. agbo) or complementary medicines do remember to tell your child's hospital doctor at the outpatient clinic because it may be important for them to consider this when prescribing other treatments for your child.

### Avoiding things which may trigger a sickle crisis

Sometimes your child may have a sickle cell crisis for no apparent reason. There are some situations that can trigger a crisis or make it worse and these are described below. It can be difficult knowing what triggers a crisis when your child is young but this is something that you will learn by experience. As your child grows up it will be important for him to start to learn what sorts of things or situations trigger his sickle cell crisis, so that even if you are not there he will know to avoid those things.

**Infection:** is an important trigger, but it may be difficult to avoid some of the common viral infections such as coughs and colds. Children with sickle cell disease are more prone to certain bacterial infections because their spleen does not work properly. Pneumococcal infection can be avoided by taking penicillin twice daily and having regular pneumococcal immunizations (see page 18). Salmonella infection can be avoided by re-heating food

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thoroughly and ensuring that eggs and chicken are properly cooked. All children should take advantage of the routine immunization programme which will protect them from whooping cough, haemophilus influenza, mumps, measles and German measles as well as the less common ones: polio, diphtheria and tetanus. It is also advisable to be immunized against influenza (flu) every year. If your child is travelling it is important to consider whether he needs any special medications, for example, malaria-preventing medications (see page 39).

**Inadequate fluids:** Dehydration (when the body does not have sufficient fluid) can occur if your child has a fever, is unwell or has diarrhoea or vomiting. It can also occur during hot weather or after strenuous exercise if your child does not have enough to drink. Insufficient fluid makes the blood flow more slowly and the red cells stickier so that sickling is more likely to occur.

Because his kidneys are not able to concentrate urine he will pass urine more frequently and the urine will be diluted, this may lead to extra fluid loss from the body causing dehydration. Insufficient water in the body (dehydration) can trigger a sickle cell crisis so extra fluid intake is important at all times, particularly during hot weather or if your child is unwell and has a fever (see page 21 for how to manage fever). Plain water or diluted juice should be encouraged and fizzy drinks avoided because these may cause tummy pain in some children.

When your child is well he should be encouraged to drink as much as he wants. Food also contains water, so if your child is eating and drinking normally there is no need to be concerned. If he is unwell he will need extra fluids.

The minimum amounts required are outlined below:

<b>Child's weight</b>	<b>Amount recommended</b>
Less than 10 kilograms (kg)	150mls for every kg body weight in 24 hours
10-20kg	80mls for every kg body weight in 24 hours
20kg and above	40mls for every kg body weight in 24 hours

**Example, If your child weighs 25kg: 25kg x 40mls = 1000mls**

The minimum amount of fluid he requires in 24 hours is therefore 1000mls (1 Litre). If your child is not able to drink the minimum amount recommended and he is unwell, especially if he has a fever, vomiting or diarrhoea, he will need to be seen by a doctor. This is in case he needs to be admitted to hospital for treatment to prevent dehydration.

**Extremes of cold and heat (temperature):** changing from a cold to a hot environment or vice versa can bring on sickling. It is therefore important not to overdress your child when he is indoors but to make sure he has sufficient outer layers when he goes outside. Chilling quite commonly occurs after swimming, and your child should dry off and get

dressed as quickly as possible. If the swimming pool water is too cold your child should be advised not to swim. It is important to discuss this with your child's school teachers so that they are aware of the reason for and the importance of this advice.

**Stress and anxiety:** can affect the body. A certain amount of anxiety can be helpful because it motivates us to perform, but too much can trigger a sickle cell crisis and this should be avoided. If your child is feeling stressed by school, or if his illness or something else is worrying him, it may be worth discussing this with your specialist nurse, doctor, social worker or psychologist.

## Routine medication

**Penicillin:** Making sure that you give your child penicillin twice a day is one of the most important things that you can do for your child. Children with sickle cell disease are 600 times more likely to get pneumococcal infection than other children. This is because the spleen does not work properly and in a young child the spleen is an important part of the body's defence against infection.



Pneumococcal infection may cause pneumonia or meningitis. The symptoms can develop rapidly, making your child very ill very quickly, even before you have time to get medical help, and can be fatal. The pneumococcus can be stopped from taking a hold by giving your child penicillin tablets or syrup twice daily. If your child is allergic to penicillin then erythromycin, another antibiotic, may be prescribed by your doctor.

For the penicillin to be effective it must be taken twice at around the same time every day. Make sure that supplies do not run too low. If your child is unwell and has been put on another antibiotic by your GP, check whether you can stop the penicillin and remember to start again once the other antibiotic is finished. Penicillin should be continued if your child is admitted to hospital unless other antibiotics have been prescribed as mentioned above.

Taking regular penicillin does not weaken the body in any way. Resistance of the pneumococcus to penicillin is not a problem in the UK, although it has been reported in other countries. Penicillin taken regularly has been shown to protect against pneumococcal infection.

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### The dose of penicillin is as follows:

62.5mg twice a day until one year of age;

125mg twice a day from one until five years of age;

250mg twice a day from age five years onwards.

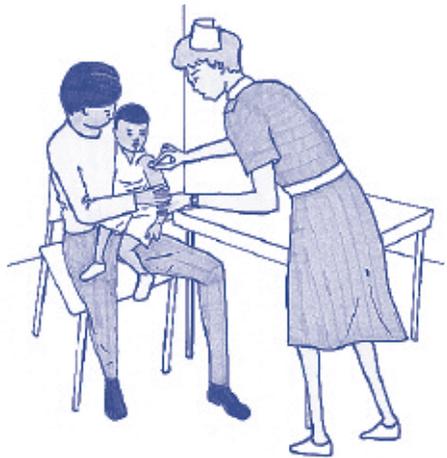
It is strongly recommended that penicillin is given throughout childhood and carried on into adulthood. It is probably better to give penicillin in tablets rather than syrup. This is because tablets can be kept at home for a longer time and because most syrup medications contain sugar, which is harmful to teeth. If syrup is taken, it is worth giving your child a drink of water after the medication so as to rinse the mouth.

Whilst your child is still young you can give tablets by crushing them with a spoon and mixing the powder with a little unsweetened fruit juice to make it pleasant.

**Folic acid:** The body needs folic acid to make new red blood cells. Generally, in the United Kingdom peoples' diet is rich in folic acid, and children with sickle cell disease do not need to take supplements routinely. Foods that contain folic acid are green leafy vegetables. Many hospitals still give folic acid to children with sickle cell disease, 5mg once a day. If this is recommended by your doctor it will do no harm.

### Childhood immunizations

Your child should get the same immunizations that other children get. These immunizations include diphtheria, whooping cough, tetanus (DPT), polio, haemophilus influenza (Hib), meningitis C, measles, mumps and rubella (German measles) (MMR). These are fully explained in the parent-held record (or baby book) and if in doubt about the timing check the book and ask your health visitor. There is no reason why a child with sickle cell should not have any of the routine childhood immunizations. In fact it is perhaps even more important that your child should be fully protected because children with sickle cell disease are more prone to infections, and infections can trigger a sickle cell crisis.



## Other immunizations

**Prevenar (conjugate pneumococcal vaccine)** This vaccine gives protection against pneumococcal infection and so is very important. From 2007 this vaccine will be included in the primary course of immunizations recommended for all children in the UK irrespective of whether they have sickle cell or not and will be given at 2, 4 and 13 months.

***Pneumovax*** This gives protection against a greater number of bacterial types that cause pneumococcal infection than Prevenar. The vaccine is given at around two years of age and then every five years thereafter. This is given routinely to all children with SCD and your GP or clinic doctor will prescribe it.

**Prevenar and Pneumovax give protection against pneumococcal infection but it is important that your child continues to take penicillin as well.**

**Meningivac** This gives protection against meningococcus types A and C which cause meningitis. Even if your child has had the Men C, which protects against meningococcus type C, he should also have this one before travelling to some parts of the world, e.g. Africa, as it will protect against meningitis A.

***Hepatitis B*** Your child will probably have a blood test in the outpatient department to check whether he has been in contact with hepatitis. Hepatitis B is occasionally passed from mother to baby in the womb. If this is the case the baby is offered a course of three immunizations to clear the infection. Many hospitals will recommend that your baby has a course of immunizations to protect him from hepatitis B starting at one year old in case he should require a blood transfusion in future. Very occasionally hepatitis B can be passed on through a blood transfusion but the blood transfusion services in the UK are extremely careful to screen all blood donors for hepatitis B (see page 35). If your child were to need regular blood transfusions for any reason and had not received an earlier course of immunization, then his doctor may advise a course should be given.

**Influenza (Flu)** it is advisable to ask your GP for a flu vaccination for your child every year.



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# VISITING THE HOSPITAL OUTPATIENT CLINIC

Once the diagnosis of sickle cell disease has been made, your child will be referred to a general children's clinic or one that specializes in sickle cell disease. This clinic may be known as the Paediatric Haematology or Sickle Cell Clinic and will be staffed by some or all of the following people:

- Consultant paediatrician (child health specialist doctor)
- Consultant haematologist (specialist doctor in blood disorders)
- Consultant paediatric haematologist (a specialist in both child health and blood disorders)
- Sickle cell nurse specialist /counsellor
- Specialist psychologist
- Specialist social worker/social support officer
- Specialist children's nurse
- Clinic receptionist /clerk
- Phlebotomist (person taking blood)
- Other specialist doctors e.g. heart doctor, eye doctor, brain scanning doctor

Many families find that visits to the outpatient clinic are useful, especially in the early years when there is a lot to learn about the condition. As your child gets older, frequent visits to the clinic may not be necessary. Discuss with your doctor how often your child needs to be seen. For the school age child appointments can be offered during the school holidays so that your child does not miss too much schooling. It is a good idea to keep in contact with the clinic even if your child is keeping well and let them know if you are unable to keep your appointment or if you move house. Always remember to arrange another appointment if you cancel or miss an appointment.

## Why does my child need to go to clinic?

The purpose of the clinic is for the doctors and nurses to check your child's health and development, for you to get information about sickle cell, learn how it may affect your child, and meet other parents and families of children with sickle cell disease. Your child will usually be weighed, measured and have a physical examination. Whilst your child is young it is a good idea to take your baby book to the sickle cell clinic. The doctor and nurse specialist will fill it in, so that you will have a record of all contacts and treatment in the hospital.

## Getting Information and support

The clinic also offers an opportunity for discussion about issues such as how to explain the condition to school teachers and what to do when travelling to another country. As your child gets older, he may find it helpful to talk with the doctors, nurses, psychologist and social worker about the illness and how he is coping.

## Having Blood tests and other investigations

As well as the medical check-up there are a number of tests that will be carried out on your child but these may not be done every time your child comes to the clinic.

Whilst he is well a blood test and a urine test once a year may be sufficient. (See page 64 for an explanation about blood tests.) In addition a special test to check the blood flow through the brain should be carried out. This is known as a transcranial doppler scan (TCD) and is important to check if your child might be at risk of having a stroke (see page 28).



## Contact between the hospital clinic and your GP

The doctor at the clinic will keep your GP fully informed about your child's condition and the amount of penicillin that he needs to take. You should go to your GP if you need to renew your child's prescription. If your child is unwell it is always best to see your GP in the first instance. If there are specific concerns that you wish to discuss with the hospital team then it is usually possible to ask for an earlier appointment than the one given to you for a routine visit. If your child is unwell and you cannot manage the illness at home (see page 29 for information about medical emergencies), you should either call your GP urgently or take your child directly to the hospital Accident & Emergency department.

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# MANAGING ILLNESS AT HOME

## Fever

A fever or raised temperature may be an early sign that your child is unwell. It is a good idea to keep a thermometer at home so that you can measure the temperature. Your child will probably look sweaty and feel hot to the touch if he has a raised temperature.

Your child has a fever if his temperature stays above:

100°F or 38°C - Taken in the mouth (if your child is above 8 years old)

99°F or 37.5°C - Taken in the armpit (if he is younger)

It is not a good idea to take the temperature in the rectum (bottom) because this can cause damage to the muscles in the rectum.

## How to use a thermometer

There are two main types of thermometer available glass and digital thermometer.

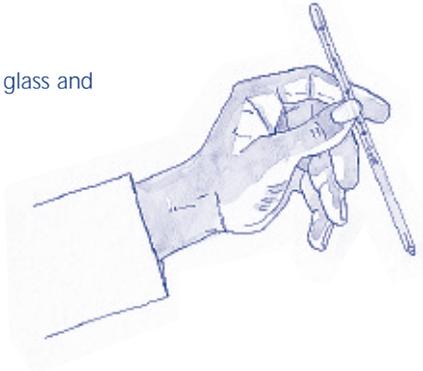
**Glass thermometers** - have a bulb of protected mercury at the end. They have an arrow pointing to 98.6°F (Fahrenheit) / 37.0°C (Centigrade), showing the normal oral temperature.

These thermometers can be used in the mouth or the armpit.

If the temperature is taken in the mouth the thermometer should be kept in the mouth under the tongue for at least 3 minutes. This method can be used when your child is above 8 years old, make sure that the mouth is firmly closed when the thermometer is in place. If taking the temperature under the armpit hold the arm firmly to the side for at least 4 to 5 minutes. Before taking your child's temperature with a glass thermometer, shake the thermometer down towards the silver end, to get the silver or coloured bar to below 95°F / 35.1°C.

To read the thermometer:

- Turn the thermometer until you can see the silver or coloured bar.
- Line up the end of the coloured bar with the degree bar.
- Read the mark. Each mark on the thermometer usually stands for two tenths (2/10ths) of a degree.



- After reading the temperature, wash the thermometer with soap and cold water and put it back into its case to protect it from breaking.

If you are not sure how to use a glass thermometer properly, ask your pharmacist, nurse or doctor to show you.

**Digital thermometers** - can be used in the mouth or armpit. Many parents like using them because they are fast and easy to read. The digital thermometer will show you the exact temperature in numbers. After use wash the tip with warm water and soap and put back in its case.

Do not give your child anything hot or cold to eat for half an hour before taking an oral temperature. Food and drink can change the reading by warming or cooling the child's mouth.

**Forehead thermometer** - is often preferred by parents for use with babies and young children (can be used from 3 months to 12 years). This form of thermometer is a tape, which you hold at both ends and press against your child's dry forehead for at least 15 seconds. It displays the child's temperature in a few seconds. The green color signifies normal temperature and red signifies that the child has a raised temperature. The tape is usually reusable.

### What to do if your child has a raised temperature

- Give Paracetamol or other prescribed pain medication (see page 23 for dosage)
- Give plenty of fluids to drink
- Remove most of his clothes
- Sponge him with warm (tepid) water

Don't chill your child too quickly. The room should be of normal temperature, approximately 21°C. If you sponge him the bath water should be lukewarm, not cold. Take his temperature about every half hour to check that it is going down.

If the temperature does not come down and stays above 38.5°C, you should ask your GP to visit, or take your child to the surgery or to hospital.

### Managing sickle cell pain at home

Mild sickle cell pain can be managed at home. In young babies and toddlers it may be difficult to know whether they are in pain or not. It is likely that you will notice that your child is not behaving as normal. He may be fretful and miserable, persistently crying, or only crying when moved. As your child gets older you will find that he gets better at being able to tell you where the pain is. Sometimes, as in dactylitis (hand foot syndrome), you may see swelling of the part which hurts or it may feel warm to your touch.

## Giving Pain killers (Analgesics)

It is a good idea to keep a supply of paracetamol (Calpol, Disprol) medication at home. If your child is in pain give the Paracetamol regularly every 4 hours but do not exceed the amount recommended on the bottle.

### The usual dose of Paracetamol / Calpol/ Disprol is:

3 Months to 1 year old	60mg to120mg	every 6 hours
1 Year to 5 years old	120mg to 250mg	every 6 hours
6 Years to 12 years old	250mg to 500mg	every 6 hours
12 Years and above	500mg to 1gm	every 6 hours

Can be given every four hours but **DO NOT** give more than four doses in 24 hours.

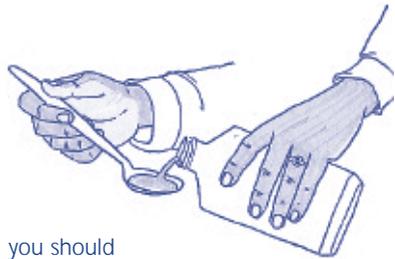
Your doctor may also prescribe Ibuprofen (Junifen) which helps relieve inflammation. This can be given with the paracetamol.

### The usual dose of Ibuprofen is:

1 to 2 years	50mg	every 6 to 8 hours
3 to 7 years	100mg	every 6 to 8 hours
8 to 12 years	200mg	every 6 to 8 hours
Above 12 years	400mg	every 6 to 8 hours

Give all medications according to the doctor's instructions and follow the instructions on the bottle very carefully.

The doctor may prescribe other pain killers for your child to use at home and as your child gets older he will know which one works best for him.



### Aspirin should not be given to a child under the age of 16 years.

If your child is getting no relief from the painkillers, you should call your GP or take your child to the Accident & Emergency department.

As well as giving regular pain relief it is a good idea to also try some of the other treatments described below.

## Extra fluids

Children with sickle cell disease should always be encouraged to drink plenty of fluids even when they are well. When they are unwell extra fluids can help thin out the blood and unclog the sickle cells in the small blood vessels. Dehydration (not enough water in the body) is known to be one of the causes of sickle cell crisis and pain.

## Warm baths

Let your child soak in a warm bath for a while. Check that it is not too hot and do not let it get cold because this can trigger another crisis. When a person is in pain sometimes gentle exercise in the warm water feels good and relieves anxiety.

## Using warm moist towels or heat pads

Soak a face towel in warm water and wring it out then use it to gently massage the painful area. This can be very soothing and will often relieve pain. Do not let the towel get cold as this will make the pain worse. Heat pads (bought from the chemist) can be put on the painful area. They are either electric or non electric.

**Electric pads** – will have a temperature dial which needs to be set at the required heat temperature, but always check the manufacturer's instructions.

**Non electric pads** – may need to be heated in a bowl of warm water, again check the manufacturer's instructions.

You can apply the warm towels or heat pads as often as you wish if they help.

## Massage

Touch can be very comforting. Use warm baby oil or lotion and gently massage painful areas to relax tense muscles and increase blood flow.

## Quiet play and distraction

Complete bed rest may not be needed. Sometimes cutting back on physical activity can be helpful. Find things your child can do quietly indoors for a while, e.g. read a book, watch a video or put together a jigsaw puzzle, colouring. Anything that can help distract your child's attention from the pain will be helpful (see information on Self-Help on page 95).

## When to seek medical and nursing help

If you have tried all the above remedies and your child still has a temperature or his pain is not relieved by the medications you have given him it is best to seek medical help. Call your GP, who will be able to advise you about what to do next and may visit your child at home. During working hours, you can call your nurse specialist/counsellor for advice.

There are some medical emergencies when your child will need prompt medical attention. These are explained under medical emergencies on page 29. In these situations you will need to get your child to the hospital straight away. Some hospitals have an arrangement that you can take your child straight to the children's ward. If your hospital does not have this arrangement, take your child to the Accident and Emergency department.

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## SOME MEDICAL PROBLEMS

Please remember that sickle cell disease is very variable and your child may never get any of the following problems or he may have some at different times in his life. Sometimes it is possible to manage a medical problem at home with the help of your GP. Sometimes it will be necessary for your child to go to hospital. Your child may need medical attention if any of the following occur.

### Painful episode (sickle cell crisis)

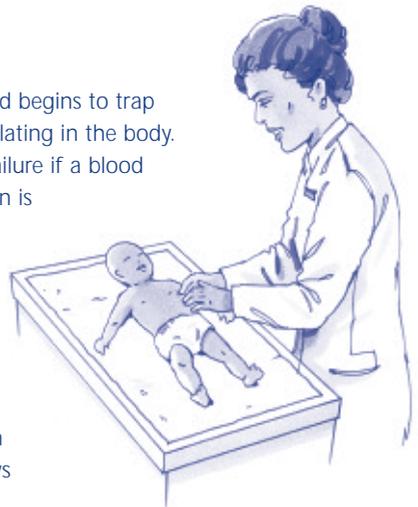
This is the most commonly seen medical problem and is caused by sickled red blood cells becoming caught up and blocking small blood vessels. It can occur in any part of the body - muscles and bones, tummy or chest - and will cause pain in that part. Pain may be mild but sometimes it can be very severe and will be very frightening for your child, see page 22 for information on how to manage pain at home. Painful crises can sometimes be triggered by your child getting cold or hurting himself or by a viral infection, for example flu, but many times there is no obvious cause.

If the pain is severe and the pain killer you are using at home has not worked it may be best for your child to go to hospital where he can be given stronger pain medications.

### Sudden enlargement of the spleen (acute splenic sequestration)

In this condition the spleen suddenly gets very big and begins to trap lots of blood. This reduces the amount of blood circulating in the body. The blood count drops rapidly and can cause heart failure if a blood transfusion is not given immediately. This complication is more commonly seen in children who are under five years of age. Once the spleen has behaved in this way it is possible for the same thing to happen again and the doctor may advise having the spleen removed. This operation is called a splenectomy.

It is important for you to learn how to feel the spleen for any enlargement. If your child appears pale, shows signs of being unwell, and his spleen becomes much enlarged he needs to see a doctor straight away. Ask your doctor or nurse specialist to show you how to feel for the spleen.



## Aplastic crisis

This condition is due to a virus infection (parvovirus) which stops the body making new red blood cells for a short time. This causes the blood count to fall but not as rapidly as in acute splenic sequestration, discussed earlier. A blood transfusion is usually necessary. The body makes antibodies against the parvovirus so once your child has recovered he will not be infected by the parvovirus again.

## Meningitis

This is an infection of the surface of the brain. Meningitis can be caused by different infections. In sickle cell disease, the pneumococcus is one of the commonest causes of meningitis. This is why it is important for your child to take penicillin daily (see page 16.) To confirm the diagnosis blood tests and a lumbar puncture (a small amount of fluid is taken from a space surrounding the spine) are done. If an infection is present antibiotics are usually given intravenously (through a drip) to clear it.

## Chest infection/acute chest syndrome

Children with sickle cell anaemia are more prone to infections and may get pneumonia, an infection of the lung. Sometimes sickling of the red cells can occur in the lung when there is no infection, but the medical signs are the same. For this reason, the term 'acute chest syndrome' is used for both conditions. There may be cough, fever, chest pain and the breathing rate may be faster than normal. Pain may also be felt over the back and abdomen. If your child needs admission because of an acute chest syndrome, a chest X-ray will be taken and your child will probably be started on antibiotics. The level of oxygen in your child's blood will be measured, using a machine called an oxygen saturation monitor. It may be necessary in some cases to give oxygen through a face mask and to give your child a blood transfusion. (See page 34 for information on blood transfusion.)

## Avascular necrosis of the femoral head (painful hip)

This is due to sickling in the topmost part of the thighbone. Sometimes it is seen on an X-ray just by chance but at other times it can cause a painful hip and a limp. This sort of crisis can usually be managed at home but you should let your doctor know about it. Pain in the hip may last for several months and it may be necessary to avoid bearing weight on the affected leg.

Avascular necrosis can also occur in other parts of the body, for example the shoulder joint or elbow.

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## Osteomyelitis (infection in the bone)

Osteomyelitis can be difficult to distinguish from a painful crisis but it does not occur very often. In both conditions, there may be swelling and tenderness of an arm or leg and the skin feels hot. In the early stages, X-rays are often normal, and your doctor may ask the orthopaedic (bone specialist) doctor to explore the swelling to look for infection. If osteomyelitis is confirmed, intravenous antibiotics will be given for at least 6 weeks.

## Haematuria (blood in the urine)

Blood may be noticed in the urine due to sickling of red blood cells within the kidney. It is usually painless and may last a few weeks. There is no particular treatment and the bleeding will stop on its own. There are other causes of bleeding in the urine that are not to do with sickle cell disease so tell your doctor if you notice blood in your child's urine. The doctor will probably ask for kidney scans and urine tests.

## Gallstones

When red blood cells are broken down some of it goes towards making bilirubin, a yellow pigment which is responsible for the yellow coloration of the whites of the eyes in many children with sickle cell anaemia. The excess bilirubin may also form gall stones in the gall bladder, a sac which produces bile and lies behind the liver.

Most children with sickle cell disease over the age of ten years have gallstones and these can be shown by doing an ultrasound of the abdomen (tummy). Sometimes the stones get caught in the gallbladder tube and cause tummy pain, particularly on the right side. If gallstones start to cause problems it is advisable to have the gallbladder removed, because the stones can sometimes cause a serious infection known as cholecystitis. Not having a gall bladder does not cause any health problem and your child will still be able to make enough bile.

## Priapism (painful erection of the penis)

Priapism is caused when sickled red blood cells block the blood vessels of the penis, causing a hard and painful erection. This can occur at any age and is more common at night or early morning after a long period of being under warm bed clothing. Going to the toilet to pass urine and empty the bladder often helps to relieve priapism. Your child may need pain killers. A warm bath or shower and gentle walking to improve blood circulation may help. Sometimes the penis becomes soft again on its own without any treatment. If this happens on a few occasions you should let your child's doctor know. If the painful erection persists for longer than two hours your child should be seen in the hospital

because an operation may be needed to flush out the red cells that are causing the obstruction. Do not apply ice packs as this may make the situation worse.

## Stroke

A small proportion of children suffer from a stroke (damage to a part of the brain) resulting in weakness down one side. If weakness occurs, particularly if there is no associated pain, your child should be seen straight away. He may need special imaging tests of his brain and a blood transfusion (see page 29 for information about medical emergencies). Sometimes the weakness does not last very long and goes away by itself within a few hours or days but it is still very important that your child is seen in hospital as a full blown stroke may occur. Without blood transfusion it is possible that the stroke be more extensive and cause more permanent damage. It is usual for the initial signs of weakness to get better after the child has been transfused. Unfortunately there is quite a high risk that another stroke may occur and to prevent this happening monthly blood transfusions are recommended.

It is now possible to carry out an ultrasound test (Transcranial Doppler Scan) to see if your child may be at risk of having a stroke (see page 66). It is advisable that this is done on a yearly basis from about the age of 2 years.

## Eye problems

Different problems can affect all parts of the eye and can sometimes affect vision. Some hospitals carry out routine eye screening in children with sickle cell disease. If your child complains of blurring of vision or pain in the eye, you should consult your doctor immediately.

## Headaches

Headaches are quite common in sickle cell disease, probably due to an increase in blood flow to the brain. Headaches do not tend to last very long and can usually be treated with pain killers, for example, paracetamol. You should seek medical advice if the headaches are very frequent or if they persist for more than two hours at a time. Your child should be seen straight away if as well as a headache he has a temperature above 38.5°C, a rash, or is vomiting or has a stiff neck. (See page 29 for medical emergencies.)

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# MEDICAL EMERGENCIES

Situations when your child needs to be seen by a doctor straight away:

<b><i>Fever</i></b>	Temperature 101°F or 39°C or higher
<b><i>Headache</i></b>	Severe headache, dizziness or stiff neck
<b><i>Breathing difficulty</i></b>	Pain or trouble breathing
<b><i>Abdominal pain</i></b>	Severe pain or swelling of the tummy
<b><i>Colour</i></b>	Very pale palms of the hand or lips
<b><i>Spleen</i></b>	Sudden enlargement
<b><i>Penis</i></b>	Painful erection lasting for more than two hours
<b><i>Change in behaviour</i></b>	Appears confused or drowsy or unable to speak
<b><i>Fits, convulsions</i></b>	Body spasms and loss of consciousness
<b><i>Weakness</i></b>	This may affect one side of the body or both and may include inability to walk

If you have difficulty reaching your GP, take your child to the Accident and Emergency department. In an emergency call for an ambulance.

Tell the medical and nursing staff that your child has sickle cell disease as soon as you arrive in the hospital.

## WHAT TO EXPECT IF YOUR CHILD IS ADMITTED TO HOSPITAL

Your child may need to come in to hospital because he is unwell or because he is to have an operation or a particular investigation or treatment that cannot be done as an outpatient. Being admitted to hospital can be quite frightening for a child and a worrying and daunting experience for the parents and family. If possible try to make sure that someone your child knows and trusts stays with him whilst he is in hospital. This is particularly important when your child is young.

### Getting to know the children's ward

It may be helpful to get to know the children's ward before your child needs admission to hospital.

- When attending clinic, the staff can arrange for you and your child to visit the children's ward. This is also a good idea for brothers, sisters, grandparents and other Carers.
- Most children's units have booklets or leaflets telling you about the different staff and the facilities available.
- You may be introduced to a play specialist or nursery nurse who can give you ideas on how to prepare your child for a hospital stay.



Try to plan ahead about what you will do if your child needs to come into hospital, especially in an emergency:

- a) Who will take your child to hospital?
- b) Who will stay with your child in hospital? There are usually facilities within most children's units for parents to stay with their children. For a young child, your familiar voice and touch can be very comforting.
- c) Who will visit your child and how often?
- d) Who will look after your other children when you are at the hospital?
- e) What arrangements have you made with your employer if your child becomes ill?
- f) How will you deal with the cost of visiting the hospital, or loss of income if you are not able to go to work?

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Your nurse specialist, ward staff or the hospital social worker are all available if you want help with any of these plans.

## Common reasons for hospital admission

Your child may be admitted to hospital for:

- Medical investigations
- Pain relief
- Other medications
- Intravenous fluids
- Blood transfusion
- An operation

## Medical investigations

Most investigations, for example, X-rays or ultrasound scans can be done on an outpatient visit. Some investigations, for example, MRI scan, may require having an anaesthetic and your child may need to come into hospital as a day case. (See page 66.)

## Pain relief medication

You will probably be able to manage painful crises at home using paracetamol and ibuprofen or any other painkiller that your doctor has prescribed. If these medicines do not control the pain, your child will need a stronger painkiller and it is likely that he will need to be admitted to hospital. A stronger painkiller, such as morphine, can be given by mouth but if your child is unwell and not drinking it can be given in a drip into a vein (intravenous), or under the skin (subcutaneous).

You should let the doctors and nurses know which painkillers he has already taken and at what time. Your child may know the best painkillers for him and which ones helped in previous painful crises.

Some parents are concerned about using very strong painkillers, particularly ones like Morphine and Pethidine because they can be associated with drug dependency (this is where the body becomes too used to the medication and cannot do without it). Pain experts suggest that dependency is extremely unlikely to occur if the painkiller is used properly in the early stages of the crisis and the effect is closely monitored. What is important is that enough pain medication should be given in the early stages to help your child cope better with the pain.

Many hospitals use a series of drawings such as faces or bodies to help children indicate where the pain is and whether it is mild or severe. When children are very young they may not be able to say how bad their pain is and the parents have to help the doctors and nurses by telling them how severe they think their child's pain is. Older children may prefer to use numerical scales to score the severity of their pain from 1 to 5 or 1 to 10, with 1 indicating 'the least pain' and 5 or 10 indicating 'the worst pain ever', depending on the type of scale used.

When a child is old enough he can control (within safe limits) the amount of painkiller he gets through a special pump. This method is known as patient controlled analgesia (PCA), the drug is given through a drip in the vein. Generally from about 6 to 8 years old most children can start taking part in managing their own pain relief. Children will often turn the pump into a game, using the button on the PCA machine to 'zap' the pain away. This will help your child to feel that he has some control over the pain and will make him feel less frightened and powerless.

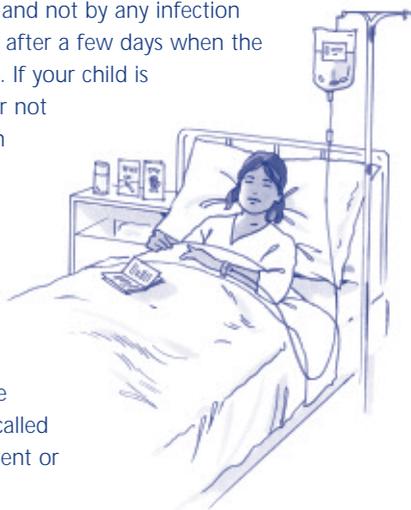
Many hospitals have a pain control team to help people with all types of pain. The team is usually made up of experts, such as anaesthetists, psychologists and specialist nurses, who will advise on how to best manage your child's painful crisis. The team may advise on other ways to help your child cope with the pain, such as relaxation (see page 95 for Self Help Manual details), and this can be used with the medications. The Play specialist can help you practise some of the relaxation methods with your child.

## Need for other medications

Antibiotics are commonly given, particularly if your child has a high temperature. The temperature may be caused by the painful crisis and not by any infection and if this is the case the antibiotics may be stopped after a few days when the results of all the tests show that there is no infection. If your child is feeling very unwell, and especially if he is vomiting or not able to take fluids orally, the antibiotics may be given directly into the vein rather than by mouth.

## Intravenous fluids

Children with sickle cell disease need to drink more fluid than other children. If your child is not able to drink the minimum amount recommended (see page 15) then he may need to be given fluids through a vein (intravenous, commonly called a drip) to ensure that he gets sufficient fluids to prevent or correct dehydration.



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## Going home after a hospital admission

When your child is ready to go home is a good time to discuss any questions or concerns you may have about caring for your child. You may be given medicines to take home. Remember you will need to continue penicillin medications as well.

If your child goes back to school before finishing a course of antibiotics, it may be necessary for him to take the medication to school and you will need to inform the school nurse and or your child's class teacher. If giving the medication in school is a problem discuss this with your specialist nurse, clinic doctor, GP or contact one of the sickle cell Centres or voluntary organizations listed on pages 87-94.



Sometimes after being in hospital, a child may not return to his normal behaviour pattern when he gets home and may show some of the following behaviour:

- Trouble sleeping at night
- Wetting the bed again after being dry at nights
- Wanting more attention than before
- Being more unruly than before
- Asking for a feeding bottle or dummy though he no longer uses one
- Not wanting to be parted from you
- Refusing to go to nursery/school

These situations occur because your child is reacting to having spent time in hospital, away from you and the rest of the family. Usually these problems are mild and do not last long. If they extend beyond a few days or become difficult to cope with talk to your nurse specialist, doctor, play specialist or sickle cell clinic psychologist.

## Blood transfusion

A blood transfusion may be necessary for a number of different reasons which include:

- Worsening anaemia: for example, in acute splenic sequestration (see page 25)
- To improve oxygen content in the blood: for example, if your child develops sickling in the Lungs (known as an acute chest syndrome)
- To reduce sickle haemoglobin: for example, following a stroke or before having an operation.

### Types of blood transfusion

1. Top-up or simple transfusion. A small amount of donated blood is given in addition to the child's own blood.
2. Exchange or partial exchange transfusion. A small amount of the child's blood is removed and donated blood is given to replace this. This method may be used, for example, to treat a child following a stroke, so as to quickly reduce the level of sickle haemoglobin and prevent further sickling.
3. Long-term blood transfusion. This involves giving a transfusion on a regular basis to keep the sickle haemoglobin level low enough so that the occurrence of further sickle cell complications can be reduced. Regular transfusions do eventually lead to iron overload.

### Transfusion before an operation

Children with sickle cell disease may need surgical operations for problems unrelated to their sickle cell disease: for example, to remove the appendix.

There are sickle cell-related problems that sometimes lead to an operation. Because they are prone to infections children with sickle cell disease many get enlarged tonsils and adenoids which may cause problems with noisy breathing and snoring and the tonsils and adenoids may need to be removed surgically.

Splenectomy (removing the spleen) is normally recommended after one or two episodes of acute splenic sequestration (see page 25) or cholecystectomy (removing the gall bladder) may be necessary if gallstones develop (see page 27) and are troublesome.



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Before an operation, the anaesthetist and surgeon must be informed that your child has sickle cell disease. Some operations or investigations, for example circumcision or tooth extraction or MRI scan, can be done safely without giving a blood transfusion. Extra oxygen may be all that is required. In other cases a blood transfusion may be advisable, particularly if your child has already had certain complications of sickle cell disease such as acute chest syndrome. The blood transfusion will be given a few days before the operation, usually on a day visit, in order to increase the blood haemoglobin.

### **Blood safety**

Every precaution is taken to ensure that the blood that your child is given is the correct match for him and that there is no risk of infection. This is done by:

- Grouping and cross-matching - A small sample of blood is taken to find out your child's blood group. This is then cross-matched against the blood that is going to be given. This cross matching reduces the possibility of antibodies (see page 84) being made which would reject the blood that is being transfused.
- Reducing the risk of infection - All donated blood in the United Kingdom is tested for HIV, hepatitis and other infections. Blood that does not show evidence of infection is used for transfusion.
- Reducing the risk of allergic reaction - blood transfusion can sometimes cause a rash, itching and a high temperature because of an allergic reaction. These side effects are reduced by giving blood without the white blood cells and this should be normal practice for children with sickle cell disease.

### **Religion and blood transfusion**

Blood is sometimes used in an emergency and may be the only means of saving a person's life, for example, following a splenic sequestration or aplastic crisis (see pages 25-26). Some peoples' religious beliefs are opposed to blood transfusions. If having blood transfusion is against your religious or social beliefs it is useful to discuss this with your doctor, nurse specialist or religious leader, preferably during one of your routine clinic visits, rather than waiting until an emergency occurs and the doctor suggests giving your child blood.

As with all other treatments the hospital staff will talk to you about any suggested treatment and will seek your verbal or written consent before they give treatment to your child, except in an emergency when it may be necessary to give treatment without consent so as to save a person's life.

## Managing iron overload

When blood transfusions are given on a regular basis over a long period of time, more iron than is needed builds up in the body. Excess iron can damage the body's organs, such as the liver or heart, and so it is important that the iron is not allowed to build up to a high level.

### - Desferrioxamine

At present the best way to remove excess iron is by injecting a medication (iron chelator) called desferrioxamine (desferal for short) under the skin or into a vein. If your child needs desferrioxamine, you will be shown how to give it at home. A small needle is inserted under the skin and the medication is given slowly over eight hours during the night using a small pump. Ideally it should be given at least five nights a week. Some hospitals give the desferrioxamine whilst the child is in hospital and receiving their regular blood transfusion. This helps to reduce the iron level even further.

### - Deferiprone

Deferiprone, unlike desferrioxamine can be taken by mouth. It is not as effective as desferrioxamine but it may be used for children who have great difficulty with desferrioxamine injections for any number of reasons. For children who are not using their desferrioxamine properly or regularly, as prescribed by their doctor, every effort is made to help them improve their use of the treatment, by giving them frequent counselling and talking to them about alternative ways of using their desferrioxamine so that they will feel more committed to using it regularly. If these efforts fail, or if the child is unable to take desferrioxamine for other medical reasons, deferiprone may be considered as an alternative treatment.

### - Exjade

Like deferiprone this can also be given by mouth and has been shown in research trials to be safe and effective. It is now licensed for use in the UK.

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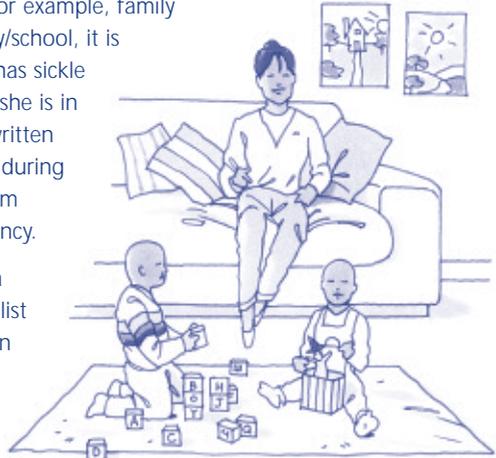
# YOUR CHILD AWAY FROM HOME

## Carers, child minders and nurseries

If your child is being cared for by others, for example, family members, child minder, baby sitter, nursery/school, it is important that they know that your child has sickle cell disease, in case she becomes ill whilst she is in their care. It may be useful to give them written instructions on how to manage your child during a sickle cell crisis and remember to tell them where you can be contacted in an emergency.

You can obtain leaflets, booklets and extra copies of this book from one of the specialist Centres or voluntary organizations listed on pages 87-92 of this book or from the NHS Sickle Cell and Thalassaemia Screening Programme. These will help your child minder or Carer get an understanding of sickle cell disease and the specific

needs of your child when you are not there. It may be useful to ask your nurse specialist or health visitor to help you explain and advise the child minder or Carer on how to prevent illness and what actions to take in an emergency.



## Your child in school

Whilst she is in school, as long as your child is well, there should be no restrictions on any school activity. Provided the school knows that your child has sickle cell disease, teachers will be able to take sensible precautions to make sure she is not exposed to things which may trigger a sickle cell crisis, for example they can:

- a) Make sure she is kept warm and away from draughty windows
- b) Allow her to take part in physical activities but recognize that she may get tired more quickly than other children. Every child is different and the amount of exercise that she is able to cope with will need to be judged in school.
- c) Prevent chilling after physical exercise and not allowing her to swim if the water or pool environment is cold. As soon as she has finished swimming it is important for her to have a warm shower, get dry and dressed immediately, to prevent chilling of the body. You need to pay particular attention to wet hair because a lot of heat is lost through the scalp.

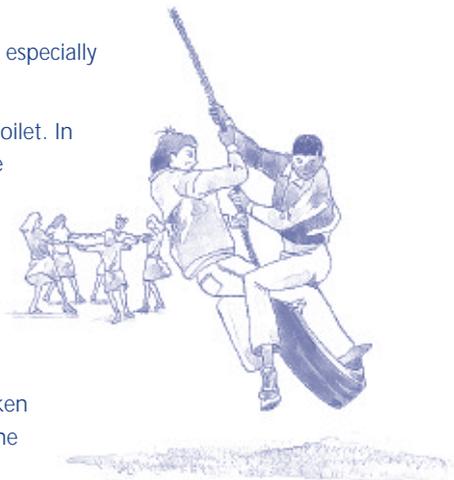
- d) Make sure she is able to drink enough in school, especially during the summer months.
- e) Be sympathetic when she requests to go to the toilet. In sickle cell disease the kidneys do not concentrate the urine very well, which means that your child may need to go to the toilet more often than most children.

When she starts school it is worth making a specific appointment to meet with her school nurse. Discuss your child's sickle cell disease, how it affects her, what precautions need to be taken in the school environment to keep her well, how she behaves when she is in pain and what teachers need to look out for, and actions to take when she is unwell. Some children try to cope with the pain or hide the fact that they are feeling unwell especially when they are among their peers in school.

The school nurse, in cooperation with your specialist nurse or health visitor, will be able to help you inform and advise teachers and other school members, whilst making sure that your child is not stigmatized or singled out from her classmates. If your child experiences bullying or being teased because of her sickle cell disease this should be discussed with the school and it may be helpful to seek the advice of your school nurse, health visitor or specialist nurse.

### Educational progress

Sickle cell disease may affect your child's educational progress. This may be because she is frequently unwell and has to miss school. Children may suffer from stroke and this may affect their learning ability and their behaviour. Routine TCD scans (see page 66) will help show if your child is at risk of stroke but it is also important to alert the doctors if your child, having been progressing normally starts to find school work hard. This may be because she has suffered a 'silent' stroke which can be diagnosed on an MRI scan (see page 66). A clinical psychologist will be able to assess your child and find in what areas she may need extra help with her school work.



Some hospitals provide lessons for school children whilst they are in hospital if this is not available your child's teacher should be able to provide some class work for her to do provided she is well enough.

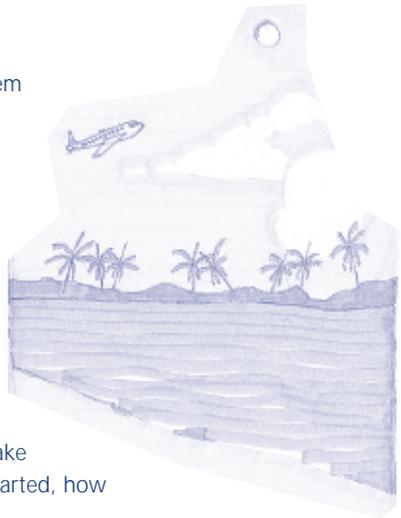
All schools are able to provide some extra teaching, but if she is experiencing considerable difficulties in keeping up with the other children in class, it may be necessary to get the local education authority to agree to extra teaching support. To do this a formal assessment is carried out and a 'statement of educational need' is produced. This is a legal contract between the parents and the education authority. It defines what extra teaching support is going to be provided in school.

If you have concerns about your child's progress in school it is best to talk to your child's class teacher. Every school has a special educational needs coordinator (SENCo), who can give you advice and information about how the school can help your child.

## Travelling and going abroad

Going on holiday or travelling is not usually a problem for children with sickle cell disease. Certain precautions need to be considered, depending on whether the journey is within the UK or abroad. Air travel should not cause any complications and extra oxygen is not required as all modern aircraft are pressurized to maintain steady oxygen levels.

As soon as you know the date you are travelling, talk to your pharmacist, GP or clinic doctor tell them which country you are going to and seek their advice. They can tell you which vaccines, immunizations or special drugs your child need to take before going away, how soon the drug should be started, how much she should take and for how long.



## Malaria prevention and medication

When travelling to a malarial zone children with sickle cell disease must be protected against the possibility of getting malaria. Malaria can be serious in all children but may be fatal in sickle cell disease due to the spleen not working properly. Wearing socks and long sleeve garments in the evenings when mosquitoes are known to be most active can help prevent bites and using insect repellent sprays and creams on exposed skin areas especially when going out in the evening is also useful.

Anti-malarial medications are recommended. Generally these are started at least two to three weeks before you are due to travel, so that your child will be protected against malaria on arrival at your destination. Dosage will vary depending on the type of medication and the age of your child, and when to start taking the drug will depend on the date you are due to travel. Follow medication instructions carefully. It is recommended that the anti-malarial medication be continued for at least two to four weeks after returning from your journey.

Some forms of malaria are resistant to the usual malaria medications, for example, chloroquine. **Therefore it is important to tell the pharmacist exactly which country you are travelling to** so that the right medication can be recommended. Before giving anti-malaria medications it is important to know if your child has a deficiency of an enzyme called G6PD (see page 65). This will normally have been checked in the sickle cell clinic. Ask your doctor or nurse specialist about the result of this test and for more information about G6PD.

People with sickle cell trait growing up and living in a malaria area develop some natural resistance to malaria, but if they live away from the malaria area for any length of time this natural resistance is quickly lost. All members of the family, including those with sickle cell trait, will need to take anti-malaria medications when visiting a malaria area.

## Vaccinations

When travelling abroad it may be necessary for your child to have an anti-meningitis immunization (see page 18). For some countries, vaccination against hepatitis and yellow fever is also recommended. Talk to your GP or hospital doctor well in advance in order to leave enough time for your child to obtain the relevant immunizations and/or vaccines before you travel.

## Other Medications whilst abroad

Your child will need to continue her penicillin medication, and if she normally takes folic acid, this should also be continued. Depending on the level of health care available in the country you are going to; consider whether you need to take a supply of the pain medication she normally uses, for example, Paracetamol and Ibuprofen. It is worth discussing this with your GP or hospital doctor.

## Travel insurance

Remember, not all countries have a free health service. For peace of mind when taking your child abroad it is worth having travel insurance, even if you are going 'back home'. If you have lived in the UK for a long time, things may have changed from when you were

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there last. Check that you book your travel and insurance with a reputable company. When you book inform the airline that your child has sickle cell disease.

## Medical reports and other documents to take with you

Obtain a letter from your hospital doctor and take this with you just in case your child becomes unwell whilst you are abroad. The letter should say:

- The type of sickle cell disease your child has
- Up-to-date information about her usual blood levels
- Any operations or complications she has had
- Medication which she takes regularly
- Any special treatment that your child is having, for example, regular blood transfusion.
- The type of analgesic (painkiller) which helps her best

If travelling with strong painkillers (especially opiates such as morphine) and syringes and needles remember to ask your doctor to write about this in your letter. You would not want to be accused of carrying drugs illegally.

## Care whilst travelling

It will be necessary to give extra fluids so as to prevent your child from becoming dehydrated. Should she complain of pain, especially in the chest, please inform the air hostess/steward as it may be necessary to give her oxygen. Airplanes can be quite cool, even whilst travelling in a hot country. Take some warm clothing in case your child feels cold. Depending on the country and time of year you travel some hot countries can be quite cold in the evenings. And remember some tropical countries have cool periods called hammattan. The warm clothing may be useful when you arrive at your destination.

If the journey is long you should encourage your child to walk about a little. Sitting for long periods of time is not advisable because it slows down blood circulation, especially in the lower part of the body, and this can trigger a sickle cell crisis.

## Preventing an illness whilst abroad

Whilst on holiday your child will need to keep taking her routine daily medications. Depending on which country you are travelling to, it is worth asking your haematologist to find out the name of a specialist doctor or sickle cell centre in the area you are travelling to.

If you are unsure about the cleanliness of the water supply in the country you are going to it may be worth taking water sterilization tablets with you. They are available in most

chemists otherwise use bottled water, provided you are sure the local bottled water is safe for drinking. Remember, because it is bottled water does not mean it is safe, especially in a country where there is limited monitoring of safety standards. Boiling the water is a possible solution, even for bottled water if you are not sure about its purity. If you need to use sterilization tablets, be sure to follow very carefully the instructions for use.

If your child becomes unwell with diarrhoea and or vomiting there is a simple recipe recommended by the World Health Organization to prevent dehydration. Mix the following for your child to sip/drink:

- Four-finger scoopful of sugar (about 1oz / 30gms)
- plus a thumb and finger pinch of salt (about 1/2 teaspoon)
- plus a full cup of water (about 6 - 8ozs / 150 - 250mls)

Alternatively, you can buy sachets of an oral re-hydration powder mix called Dioralyte to take with you. This is available from your local chemist.

### Checklist before you travel

1. Letter from the doctor (haematologist or paediatrician)
2. Special immunization/vaccination required
3. Family doctor (GP) for anti-malarial medication and other routine medication, (e.g. penicillin V, folic acid, painkillers etc.)
4. The name and address of the sickle cell centre or a reputable doctor in the area you are visiting
5. Extra fluids for the journey
6. Travel insurance if going abroad. For advice about travel planning it is highly recommended that you talk to your health visitor, school nurse, specialist nurse / counsellor or contact one of the voluntary organizations listed on pages 91-92.
7. Thermometer (you may need to check if your child has a fever)
8. Water sterilization tablets
9. Oral re-hydration sachets

These simple precautions will help to keep your child well whilst you are abroad.

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# FEELINGS AND FAMILY RELATIONSHIPS

## How you may feel when told that your child has sickle cell disease

Parents go through different emotional phases when they are told that their newborn child has sickle cell disease.

If you did not know that you and your partner have the sickle cell gene or any of the other unusual haemoglobin genes before having your baby the chances are that the diagnosis of sickle cell disease will have come as a shock to you and your family. You may also find it difficult to accept especially if it is unexpected. You may feel upset, angry or guilty that you have unknowingly given your child this condition.

You may disbelieve the diagnosis; feel confused, anxious, depressed, or even frightened because you do not know how this condition is going to affect your child, and the rest of your family. You may feel helpless because you are unable to take away your child's illness. If you are religious you may feel like blaming God or feel that God does not care about you any more, you may ask "How could God allow this condition to affect my child?" Some parents go through a behaviour change, resenting everything and everyone around them and hating this 'disease' that appears to have come uninvited into their family.

Some or all of these feelings are common and natural when parents are told that their child has a long-term illness such as sickle cell disease.

Sometimes these feelings will go away quickly, and you feel you have come to terms with the illness only to find yourself having these feelings again at a later time. For example, this may happen when your child has her first sickle cell crisis or illness. Do not be alarmed if this happens. Sometimes your feelings and emotions are beyond your control, you just feel them and have to accept them when they occur. People have different ways of coping with their feelings, but the important first step is to recognize how you feel and go through the experience without feeling guilty that you feel the way you do.

## Public attitude to sickle cell disease

The impact of sickle cell disease on your child and family can also be determined by what other people think about it. Many people have little or no knowledge of sickle cell disease, and there are a lot of myths, taboos and misconceptions. Some people think that it is a condition that affects only black people.

Some people hold strong cultural or religious beliefs: for example, some believe that a child has been born with this disease because God is punishing the child's parents.

Unfortunately, these myths and cultural beliefs from the public, friends, or even family members may lead to a negative attitude towards children with sickle cell disease or to the stigmatization of the child and their family. As a parent this may make you feel sad, rejected and ashamed that you gave your child this condition for which she is being stigmatized.

These attitudes and experiences within your extended family, community or society create an opportunity for you to teach others about sickle cell and how it affects your child and family.

## Who can help?

It may help to talk about these feelings initially with a friend, a member of your family, your religious leader, nurse specialist / counsellor, health visitor, doctor, other health worker, social worker or anyone you trust will understand how you are feeling. They will be able to support you as you go through these feelings and experiences. Sometimes talking to other parents who have been through a similar experience, or talking to adults with sickle cell disease will help to reduce your fears, anxieties, and concerns about sickle cell disease.



However, if you find that it is still difficult to cope you may find it helpful to ask to see a psychologist. A psychologist is a specially trained health care professional who helps people deal with personal issues and or emotional problems affecting their lives. A psychologist is not a psychiatrist, they do not deal with mental illness and will not prescribe any medication, but will talk to you in depth about your feelings and help you determine what to do about a problem. Seeing a psychologist does not mean you or your child is 'crazy' or that there is something wrong with you or your family. It means that you are taking an active step to prevent some of the consequences of stress, which comes with having a family member with any chronic illness. Talk to your doctor or nurse specialist if you wish to see a psychologist.

As you learn more about your child's condition it will hopefully become less frightening or worrying. You will get to know what things affect your child as an individual. You will learn how to manage, how to plan your child's care and how to get the best help available. Most important, your child will be developing her own individual personality to become that little person you get to know and love.

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As you get to know what you can do to help your child and family live as normal a life as possible, you will realize that sickle cell disease need not be as fearful or hurtful as you first imagined. The unknown is often more frightening or distressing than reality.

There are a lot of myths about sickle cell disease. If you hear anything that worries or concerns you, talk to your nurse specialist or doctor at the clinic. You may find that these stories are just 'old wives' tales or half-truths.

Social research has shown that many families with a child with sickle cell disease tend to be much more supportive of each other, develop strong ties, cope very well emotionally and build effective mechanisms for dealing with the illness. This is one of the positive aspects of having a child with a chronic illness like sickle cell disease.

## Your young child with sickle cell disease

Children with sickle cell disease are often very strong psychologically and emotionally and able to cope with their illness. Your child's ability to cope with her illness depends on a number of factors: the severity of your child's illness, individual personality, your attitude to the illness, and your ability to handle your child's feelings. Your child may feel guilty because of her illness and that she may be causing a lot of worry for the family, both emotional and possibly financial. Giving your child space to grow and become independent is an important part of helping her to cope with her sickle cell disease.

Many children come to understand their illness around age six to seven years, when they may realize that their condition is permanent. This may create fear and anxiety for your child and she will need all your support to come to terms with this knowledge about herself.

Your child may start to blame you or show signs of resentment towards you for giving her this disease, or she may be jealous of her siblings who do not have the disease. She may have difficulty relating to other children at school, especially if her teachers and classmates have little or no knowledge of sickle cell disease, or she may try to be very brave about it all.

The way you personally deal with pain and symptoms may conflict with the images that your child sees on television and outside of the home. Children need people to believe them when they say they are in pain or feel unwell, and for adults to help them learn to cope with pain or get relief and get better. Developing a relationship where your child feels secure in telling you and others when she is in pain or feeling unwell is an important start to your child developing a positive attitude to her illness. She learns to trust her body's messages and herself to interpret those messages accurately and, more importantly, she learns to trust you and those caring for her.

Giving your child positive images of herself, showing her that you love her even though you may not love her illness is an important part of your child being able to develop a positive attitude which will prepare her for living with her illness even when you are not there.

It is tempting to over-protect a child with a chronic illness, but this can do more harm than good for the child as they face society outside of the home. It is important to raise your child with sickle cell disease in the same way as you would your other children. Every child needs care, love, support, encouragement, and where necessary discipline. They still need to learn the difference between right and wrong, what is acceptable social behaviour and what is not, even if they have sickle cell disease.

## Your teenager (adolescent) with sickle cell disease

Most children and their parents find the teenage years a difficult time. Sickle cell disease may make this time even more difficult. Your child may only now start to understand sickle cell disease and what it means for her future, she may find this a bit daunting or even frightening, as she starts to think about choosing a career, building intimate relationships and parenthood. On the other hand she may be more emotionally mature than her friends.



Sometimes the frequency of painful crises increases at this time because of hormonal changes going on in the body and changes in social lifestyle, and adolescents may find themselves spending more time in hospital, which of course is very disruptive to their lives. They may find it hard to keep up with school, college or university work or to take part in sports and social activities. Should your child have frequent episodes of painful crises and hospitalizations, she may envisage a life which is constantly associated with pain and hospitals. These thoughts may trigger feelings of depression and she may need some emotional support and encouragement during this stage.

Your child may want to join in adolescent fashions, which in some cases may not be good for her health, for example, wearing skimpy clothes when it is cold, smoking, drinking alcohol, staying out late and getting too tired physically.

Adolescence can be a distressing time for your child, and she will need a lot more encouragement and support. It is sometimes helpful to talk to someone who knows about sickle cell disease and how it can affect her life, an older person with sickle cell disease perhaps.

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## Coping with Sickle cell disease

As your child gets older she needs to learn to cope with her illness effectively and to recognize that she can play an active part in keeping herself well, reducing the chances of getting ill and coping with crisis if and when they occur. A self-help manual aimed at children with SCD has been developed by a specialist psychologist. It may help your child to learn about her condition and the various techniques that she can use to deal with painful episodes as well as ways she can manage other aspects of living with sickle cell disease (see page 95 for details).

## Brothers and sisters

Brothers and sisters may feel ignored, rejected, jealous, and even angry at the amount of attention you are giving to your child with sickle cell disease. Sometimes siblings may feel guilty that they do not have the illness and their brother or sister has it and is suffering so much pain when they are not. It is also important for siblings to understand that your child was born with sickle cell disease, so they will not have the fear of catching it.

Other members of the extended family, especially grandparents, may be anxious as they start to understand how the family connection and genetics work, and they may feel guilty about the part they have played in passing on this genetic condition through the family line.

## How do you handle brothers' and sisters' feelings?

You will need to help siblings understand that their feelings are natural and that you are there for them as much as for the child with sickle cell disease. Let them know you love them and want to continue giving them as much of your time as you can. Give them time to talk about their feelings and encourage them to take part in caring for their sibling when appropriate. Try to share your time equally with all your children, recognizing that each child has different needs at different times.

Share how you feel let your family know that you all have needs, including you and your partner. This will help to build trust, encourage mutual support and family togetherness, especially when your child with sickle cell disease is unwell.

## Dealing with grandparents and other family members

Encourage grandparents and other family members to help support you and your child with sickle cell disease. Teach them about sickle cell disease so that when required they are able to help care for your child without you or they worrying that they would not know what to do during a painful crisis or an emergency. If you have difficulties answering their questions talk to your nurse specialist / counsellor, health visitor or doctor. Ask them to help you explain about sickle cell disease to your children and other family members if necessary.

## PRACTICAL ISSUES FOR COPING

Having a child with a chronic condition often creates extra demands, and challenges the family's capacity to cope. This is especially so if the child has an unpredictable condition like sickle cell disease. Demands on a parent's time, energy and financial resources may make the situation more stressful.

### Forward planning

When your child is first diagnosed, money and career is probably the last thing you will see as important parts of your life. But they are, and it is worth talking about these as soon as you feel able. Many parents find that forward planning is the key to making the situation less stressful. They are better able to adjust to the economic changes and they gain better security.

Each family's situation is unique. You will need to look at your family's specific circumstances, for example:

- Number of children in your family and their age
- Are you a lone parent or couple?
- Do you have the support of an extended family and friends?
- What are the family's income, expenditure and basic financial needs?
- Do you have security in your living circumstances e.g. do you own your home, have a mortgage, or is your home rented?
- How long will it take to pay off your mortgage?
- Do you have an insurance policy to help protect the family if anything happens to the bread winner?



Only you will be able to answer these questions realistically. Take time to plan and look at the various options available to you. It will enable you to feel confident and better able to manage your circumstances.

But remember, don't panic or take any major decisions too quickly wait until you feel able to do so. Life is not a race. It is to be lived one day at a time.

### The working parent

Having a child with sickle cell disease may make it more difficult for one or both parents to go out to work and maintain a career, especially if there is little or no extended family support. This will have an effect on the family's income.

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## Questions which parents often ask about employment issues:

**Can I go out to work? What hours can I work? Can I get home quickly if my child becomes unwell suddenly?**

This will depend on your personal circumstances, for example, whether you are a one or two parent family, number and ages of your children, whether you have extended family or friends for practical support. Secondly, the skills that you have to offer employers, the distance between your place of employment and your home will need to be considered. You may be able to negotiate with your employer flexible working hours, part-time work or job sharing.

**What and how much do I need to tell my employer, especially if I need to take more time off work than usual and how will time off work affect my rights as an employee?**

This depends on the type of work you do and how flexible your working life is. Telling your employer about your personal circumstances is your choice and you will need to weigh up the advantages and disadvantages. If you feel that your employer will be sympathetic if and when you need their support and cooperation when your child is unwell then it is definitely an advantage to tell them about your situation. Some employers are family-friendly and will allow time off work for caring for a sick child or relative.

Being truthful and honest with your employer from the outset is usually useful; your employer is likely to be more receptive and accommodating when you need time off.

Talk to your social worker and local citizen's advice bureau (CAB) for further information about your statutory entitlements to 'Carer's Leave' and other entitlements whilst working. You can also check this with your union or professional association.

**What type of job can I do that will be flexible enough for me to cope with my child's unpredictable illness and will my employer understand that I may have to have time off or leave during normal working hours if my child is unwell?**

This will depend on the type of work that you do and whether it is flexible enough for you to be able to negotiate your working hours. For example, could you do some of the work from home? Could you come into work at weekends or working in the evenings when your partner or family helpers are available to help with child care? You will need to be realistic about the level of flexibility your employer can allow.

A job centre or careers adviser will help you look at the skills and qualifications that you have already and help you consider whether you should continue in the same job or career. It may be possible to do the same job but working at home or part-time.

The Training and Enterprise Council (TEC) offer schemes and grants for skills and jobs training. Your job centre will be able to provide information about this, especially if you are thinking of a job or career change.

### Who will be competent enough and willing to look after my child with sickle cell disease when I am at work?

You will need to explore your personal family circumstances and consider whether you have sufficient family support to be able to go to work full or part-time.

Health care/social care professionals will help you look at some of the range of options for suitable day care if you wish to go back to work. Check if your employer has a day care scheme which caters for children with special needs. The health visitor/nurse specialist or social worker will also advise on how to access respite care for your child if and when it is needed.



If you need to use outside Carers such as nannies, child minders and nurseries it will be necessary to educate the persons who will be caring for your child. Seek the support of your health visitor and or specialist nurse.

### Do I need to take out a health or life insurance? What are the benefits or limitations? What do they cost, can I afford it and is it worth having?

This will depend on your personal financial circumstances and your personal values and aspirations. Talk to an independent financial adviser who will help you explore your personal situation and reach a decision that suits your and your family's needs. But make sure you talk to a reputable broker.

You may need to get practical information and advice to help you plan ahead. Talk to your social worker, citizens advice bureau (CAB), or look in library directories or search for information on the internet, for a list of useful websites see page 95. These will help you locate where to access and get the range of information you need for your forward planning:

Financial advisers are available in banks, building societies and debt counselling Centres. They can help you plan long-term. For example, they will look at whether there are any tax concessions you are entitled to and are not claiming; whether you have any assets which can bring additional income; whether you should consider an insurance policy to protect the family's income.

If you have a social worker attached to your local specialist centre or hospital discuss your employment and financial advice needs with them or visit the local CAB.

And there are a number of useful practical guidebooks from reputable bookshops which may help in your attempt to plan and organize your family life so that having a child with sickle cell does not create undue stresses for you and the rest of the family.

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## SOCIAL CARE AND WELFARE RIGHTS

Although it is well known that many people with disabilities and their Carers are unable to work, few are aware of their entitlement to benefits and how to claim these and may be living in poverty.

Information and leaflets regarding benefit entitlement and their rates can be obtained, free of charge from your local job centre plus office, social security office or job centre. Some leaflets can also be found in post offices. Leaflets on housing benefit and council tax benefit are available from your local council.

You can get more information from the Department for Work and Pension, job centre plus and the Pension Service's websites. Their addresses are: [www.dwp.gov.uk](http://www.dwp.gov.uk), [www.jobcentreplus.gov.uk](http://www.jobcentreplus.gov.uk), [www.thepensionservice.gov.uk](http://www.thepensionservice.gov.uk)

The Benefit Enquiry Line is a free confidential telephone service set up for people with disabilities, their Carers and representatives.

Phone: 0800 88 22 00 Text phone: 0800 24 33 55

Disability Living Allowance (DLA) Helpline gives advice on existing DLA claims and send out application packs to new claimants.

Phone: 0845 712 3456 Text phone: 0845 722 4433

The CAB will be able to advise you on a range of earnings-related and other benefits which you may be entitled to, whether you are working or not. This may include housing, welfare and state benefits. They will advise you on whether you are entitled to family income support, invalid care allowance, disability living allowance, family fund and a range of other benefits means and non means-tested benefits.

Many local sickle cell & thalassaemia centres also offer welfare advice and or are able to direct you to local areas for support.

Support to purchase essential items such as a washing machine, a fridge, or clothing can be applied for through your local community support worker, social worker or specialist nurse or counsellor. Eligibility for meeting the criteria when applying for welfare grants are usually based on your income and the severity of the individual's disability.

## PREGNANCY AND FUTURE BIRTHS

Since you already have one child with sickle cell disease, there is a possibility of your having another child with sickle cell disease if you are with the same partner, or if you have a child with a different partner who also has sickle cell or other unusual haemoglobin. Knowing this means that planning to have another child may be an emotional and worrying time for you. It may be useful to talk to your nurse specialist/counsellor, health visitor or doctor when you are planning to have another child, they will tell you about the services and options available to you.

### What are the chances of having another child with sickle cell disease?

If you have a new partner it is important to get your new partner tested.

To work out which haemoglobin type your child could inherit you will need to know which haemoglobin type you and your partner have.

### How do I find out which type of haemoglobin my partner and I have?

If one or both of you have not been tested for sickle cell you will need to have a special blood test called haemoglobin electrophoresis and a full blood count. This test can be done by your GP or you can visit one of the sickle cell/thalassaemia Centres listed on page 87-91.

### What can our child inherit?

Remember the chances are the same for each and every pregnancy.

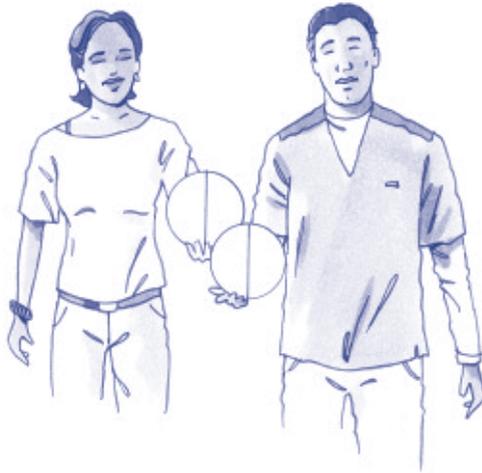
Here are a few examples:



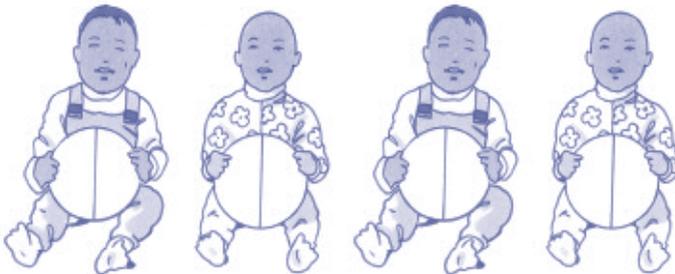
### Example 1

If both parents have the usual and most common haemoglobin combination AA (HbAA), EACH and EVERY time they are expecting a child:

There is a 4 in 4 chance (100%) that their child will inherit the usual and most common haemoglobin combination (HbAA.) There is no possibility of their children inheriting any unusual haemoglobin.



Usual  
(HbAA)



Usual  
(HbAA)

Usual  
(HbAA)

Usual  
(HbAA)

Usual  
(HbAA)

### Example 2

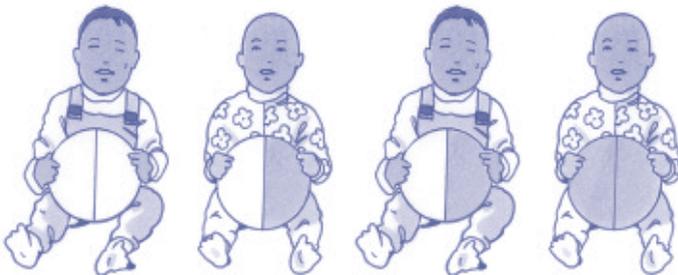
If both parents have sickle cell trait, haemoglobin AS (HbAS), EACH and EVERY time they are expecting a child:

There is a 1 in 4 chance (25%) that their child could inherit the usual haemoglobin (HbAA), a 2 in 4 chance (50%) that their child could inherit sickle cell trait (HbAS) and a 1 in 4 chance (25%) that their child could inherit sickle cell anaemia (HbSS.)



Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)



Usual  
(HbAA)

Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)

Sickle cell anaemia  
(HbSS)

### Example 3

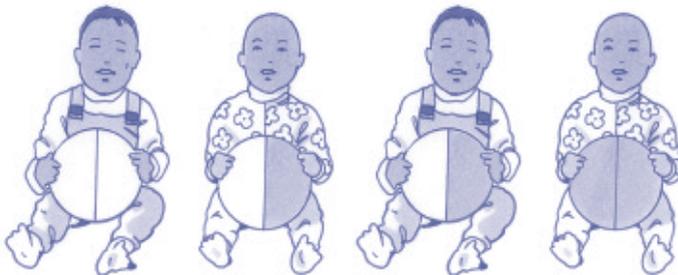
If one parent has sickle cell trait (HbAS) and the other parent has haemoglobin C trait (HbAC), EACH and EVERY time they are expecting a child:

There is a 1 in 4 chance (25%) that their child could inherit the usual haemoglobin (HbAA), a 1 in 4 chance (25%) that their child could inherit Haemoglobin C trait (HbAC), a 1 in 4 chance (25%) that their child could inherit sickle cell trait (HbAS) and a 1 in 4 chance (25%) their child could inherit sickle haemoglobin C disease (HbSC.)



Sickle cell trait  
(HbAS)

Haemoglobin C trait  
(HbAC)



Usual  
(HbAA)

Haemoglobin  
C trait (HbAC)

Sickle cell trait  
(HbAS)

Sickle Haemoglobin  
C disease (HbSC)

### Example 4

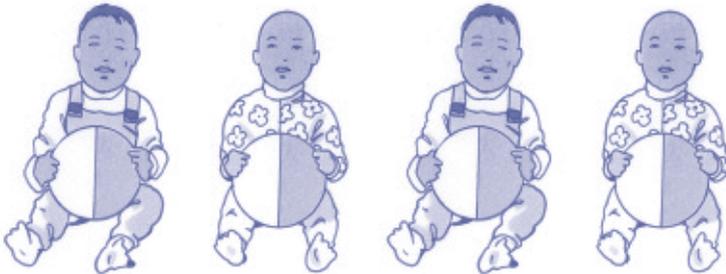
If one parent has the usual haemoglobin (HbAA) and the other parent has sickle cell anaemia (HbSS), EACH and EVERY time they are expecting a child:

There is a 4 in 4 chance (100%) their child will inherit sickle cell trait (HbAS.)



Usual  
(HbAA)

Sickle cell anaemia  
(HbSS)



Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)

### Example 5

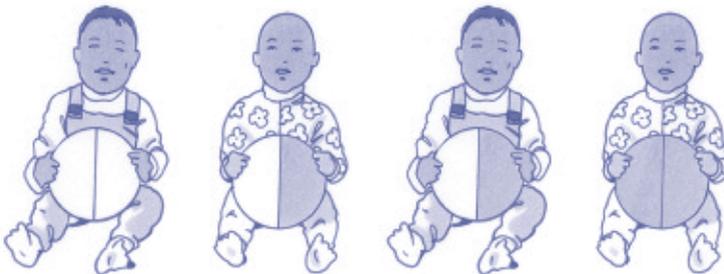
If one parent has sickle cell trait (HbAS) and the other parent has beta thalassaemia trait (HbA $\beta$ <sup>Thal</sup>), EACH and EVERY time they are expecting a child.

There is a 1 in 4 chance (25%) that their child could inherit the usual haemoglobin (HbAA), a 1 in 4 chance (25%) that their child could inherit sickle cell trait (HbAS), a 1 in 4 chance (25%) that their child could inherit beta thalassaemia trait (HbA $\beta$ <sup>Thal</sup>) and a 1 in 4 chance (25%) that their child could inherit sickle beta thalassaemia disease (HbS $\beta$ <sup>Thal</sup>.)



Sickle cell trait  
(HbAS)

Beta thalassaemia  
trait (HbA $\beta$ <sup>Thal</sup>)



Usual  
(HbAA)

Sickle cell trait  
(HbAS)

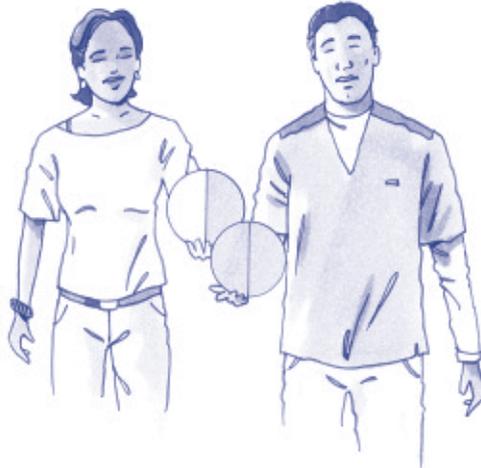
Beta  
thalassaemia  
trait (HbA $\beta$ <sup>Thal</sup>)

Sickle beta  
thalassaemia  
(HbS $\beta$ <sup>Thal</sup>)

### Example 6

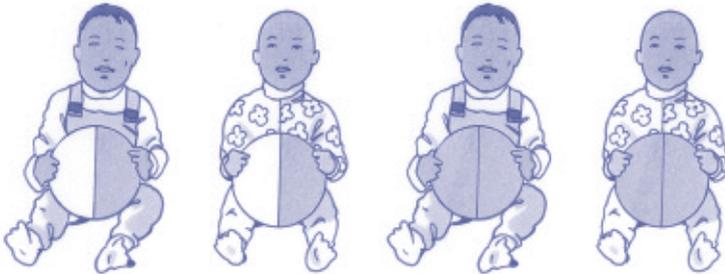
If one parent has sickle cell trait (HbAS) and the other parent has sickle cell anaemia (HbSS), EACH and EVERY time they are expecting a child:

There is a 2 in 4 chance (50%) that their child could inherit sickle cell trait (HbAS) and a 2 in 4 chance (50%) that their child could inherit sickle cell anaemia (HbSS.)



Sickle cell trait  
(HbAS)

Sickle cell  
anaemia (HbSS)



Sickle cell trait  
(HbAS)

Sickle cell trait  
(HbAS)

Sickle cell  
anaemia (HbSS)

Sickle cell  
anaemia (HbSS)

**REMEMBER IN ALL THESE EXAMPLES, THE FOUR POSSIBLE CHANCES ARE THE SAME FOR EACH AND EVERY PREGNANCY**

## Can an unborn baby be tested in the womb?

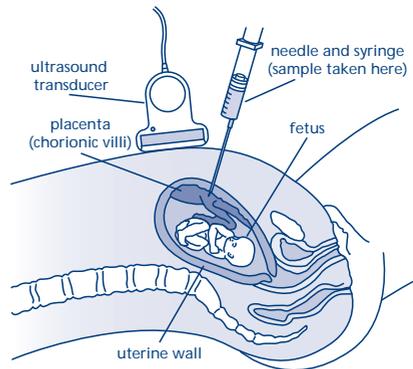
There are a number of ways of testing to find out if the unborn baby has a genetic condition. These tests are called pre-natal diagnosis (PND). They will tell you which haemoglobin type your baby has inherited from you and your partner. It is worth thinking about these options before you become pregnant so that you will have had a chance to find out as much as possible about the tests beforehand.

The type of test you are offered will depend on how many weeks pregnant you are. There are three types: Chorionic villus sample (CVS) Amniocentesis and Fetal blood sampling (FBS.) Most women will attend a Regional fetal medicine centre or hospital as an outpatient and local anaesthetic may or may not be used.

### Prenatal diagnosis

#### a) Chorionic villus sample (CVS)

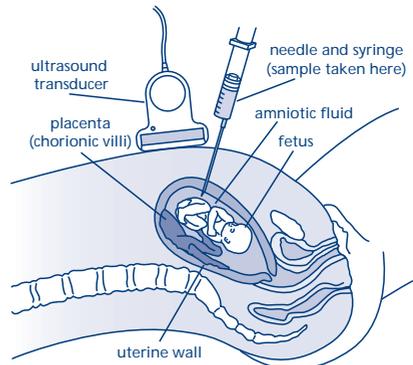
This test can be done after 10 weeks of pregnancy. A small piece of the placenta (the afterbirth) is taken and sent for testing. The result is usually available within one week.



#### b) Amniocentesis

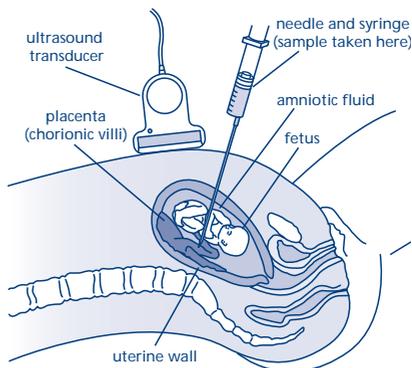
This test can be done from around 14 weeks of pregnancy. A small amount of the liquid around the baby, called amniotic fluid, is taken and sent for testing. The result is usually available within one week.

The sample for Chorionic villus sample and Amniocentesis is not taken from any part of the baby's body, but there is an increased risk of miscarriage of about 1% as a result of having these diagnostic tests. The risk calculation does not take into account the miscarriage rate that can occur in some pregnancies even where no test is done.



### c) Fetal blood sample (FBS)

This test can be done around 16 weeks of pregnancy, but can be done up to delivery of the baby. A small amount of blood is taken from the cord and sent for testing. This is rarely offered as it has a higher risk of miscarriage than the other two diagnostic tests mentioned earlier. The result is usually available in two to four days.



If you wish to consider having any of these tests or you just want to find out more about them, talk to your GP, nurse specialist, or someone at any of the specialist Centres listed on page 87 - 94 when planning to have another baby. The decision to have a pre-natal test rests with you and your partner. The health care workers are there to give you information which will help you whilst trying to make a decision. They will not force you to make a decision either way.

### What if the result shows that the unborn baby has sickle cell disease?

You may have thought about what you would do if the result shows that the unborn baby has sickle cell disease. It could be that you decided to have the test so that you can prepare for the arrival of the child. It is possible that you wanted to avoid having a child or another child with sickle cell disease and were planning on terminating an affected pregnancy.

In either case, it may be useful to discuss your feelings with your doctor or nurse specialist. They are there to support you and your partner, no matter what decision you make about the pregnancy. If you decide to terminate the pregnancy, this will be arranged for you and you will be given as much support as you need to come to terms with what can be an emotionally difficult time for you, your partner and other family members.

There is an organization called Antenatal Results and Choices (ARC) who offer additional support to parents who are making choices about an at-risk pregnancy (see page 94 for their details).

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## Pre-implantation genetic diagnosis

Pre-implantation genetic diagnosis (PIGD) is another possible option for couples who do not want to have a child with a genetic disease like sickle cell anaemia (HbSS) yet feel for whatever reason, unable to consider termination of an affected pregnancy. The process involves removing mature eggs from the woman's ovary. The eggs are then fertilized in the laboratory with sperms obtained from her partner or donor if she wishes to use this alternative.

The developing embryo is then tested for sickle cell anaemia specifically but not for other types of sickle cell disease or other genetic conditions. If the embryo does not have sickle cell anaemia, the fertilized egg is placed in the woman's womb for a pregnancy to become established and continue. This process is often referred to as 'in vitro fertilization' (IVF) or 'test tube baby'. The 'take home a baby' rate is 20% in other words, 1 in 5 attempts will result in a baby being born.

Although IVF has been done for over thirty years PIGD is a fairly new development for couples at risk of having a child with sickle cell disease and few hospitals offer this facility. Although the service is provided in NHS hospitals only 50% of couples are able to get NHS funding. Others have to pay for the service which costs around £5,000 (2006 prices) per cycle, including all the drugs required for the treatment.

If you are interested in PIGD discuss it with your doctor, specialist nurse or contact one of the specialist Centres or voluntary organizations listed on pages 87-94 when planning to have another baby.

## DEVELOPMENTS IN THE MANAGEMENT OF SICKLE CELL DISEASE

Sickle cell disease affects people in different ways. Some people are mildly affected whilst others are severely affected, even within the same family. The reasons for this are not always clear but several things have been linked with the severity of sickle cell disease. For example, the level of baby haemoglobin F (HbF) which some people continue to make into adulthood is important. Normally the level of haemoglobin F falls to about 1% by the end of the first year of life and stays at this level right through adulthood. Children with sickle cell disease may go on producing higher than usual levels of haemoglobin F for longer and levels above 7% appear to be related to less sickling crisis and fewer complications of sickle cell disease.



### Hydroxyurea

Currently the most promising method for reducing sickling of the red blood cells is to encourage the body to carry on making fetal haemoglobin (HbF) right through into adulthood. The advantage of fetal haemoglobin is that it does not sickle and its presence prevents red blood cells from sickling.

Several drugs have been shown to increase production of fetal haemoglobin; of these Hydroxyurea is the most promising and the one that is currently being prescribed. This is a drug that has been used for many years to treat diseases in which the bone marrow is overacting and producing too many red blood cells. There is good evidence now that Hydroxyurea reduces sickling crisis in most patients.

Three hundred patients with sickle cell disease in the USA were given this drug over a three-year period. The number of sickling crises and hospital admissions were reduced by half. Fewer patients suffered from severe complications, for example, sickle lung, a complication seen in adults. Fewer patients on this treatment needed blood transfusions.

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However, Hydroxyurea may not be right for all patients with SCD. It has the potential to reduce bone marrow activity, which increases the risk of infection. It should not be used for patients who are likely to become pregnant or those who have difficulty following instructions given for the treatment.

Careful follow-up is needed, with regular visits to the clinic to check the blood levels. It is important to understand that Hydroxyurea is not a cure for sickle cell disease. It is an effective treatment for preventing or reducing sickling crisis and its effect will only last as long as the person is taking the drug.

## **Bone marrow transplantation (BMT)**

This is the only cure for sickle cell disease at the moment. BMT involves taking normal bone marrow from a 'matched' relative or donor and giving it to the person with sickle cell disease. This is after they have had treatment with strong medicines to wipe out their own bone marrow which is producing the sickle red blood cells.

Whilst this treatment has been done successfully on many patients with sickle cell disease, there are several problems. It is best done when the child is very young; using a matched brother or sister, possibly before any complications have arisen and the level of severity of their sickle cell disease is not known. As the severity of sickle cell disease can be so variable it is difficult to justify a high risk treatment like BMT in the majority of people with sickle cell disease. 9 in 10 people will survive the transplant, but 1 in 10 will not. There may be several unpleasant long-term side effects. It is often difficult to find a relative with a matching bone marrow and the treatment does not always work even among those who survive the treatment.

## **Gene therapy**

In future it is likely that we will be able to offer gene therapy, by replacing the defective gene with a normal gene, but research in this area is still in its infancy.

To find out more about any of these developments talk to your nurse specialist or counsellor, doctor or contact one of the sickle cell specialist Centres on pages 87-92.

## MEDICAL TESTS AND WHAT THEY MEAN

### Blood tests

In children blood is usually taken from the back of the hand or the inner part of the elbow. The doctor or nurse will explain the tests to you and why they are needed. To make having blood tests less painful the nurse may put a patch of local anaesthetic cream (emla) on your child's skin to make the area numb. The numbness will wear off after a few hours.

### Haemoglobin electrophoresis

This test is done to find out the type of haemoglobin a person has inherited from their parents: for example, sickle cell trait (HbAS), haemoglobin C trait (HbAC), sickle cell anaemia (HbSS), sickle haemoglobin C disease (HbSC) and sickle beta thalassaemia (HbS<sup>βThal</sup>.) This test will also show if there is any baby haemoglobin F (Hb F) present. Sometimes it is done to find out how many sickle red blood cells are present in the blood. This is called quantitative haemoglobin electrophoresis. If your child is having regular blood transfusions it helps the doctor work out how much blood transfusion your child needs.

### Fetal haemoglobin test

Some children with sickle cell anaemia remain very well with very few problems. This may be because they have inherited the advantage of continuing to produce a lot more haemoglobin F (baby haemoglobin) than usual. Some children have levels of haemoglobin F in their blood as high as 25-30% (the usual amount is below 1% in people over one year of age), because they have inherited a gene that makes their body continue to produce high levels of haemoglobin F in adulthood. Other children do not have this gene but continue to make a lot of haemoglobin F anyway, even after the first year of life.

### Full blood count (FBC)

This test examines the size, number and shape of the blood cells, how healthy they are and whether they are doing their job properly. Children who do not have sickle cell disease usually have a haemoglobin level of between 10 and 14. Children with sickle cell disease generally have lower levels and this depends on the type of sickle cell disease. For example, children with sickle cell anaemia and sickle beta zero thalassaemia (HbS<sup>β0Thal</sup>) range between 6 and 8, whilst children with sickle haemoglobin C disease (HbSC) and sickle beta plus thalassaemia (HbS<sup>β+Thal</sup>) range between 9 and 11.

It is useful to keep a record of what your child's usual haemoglobin level is. If your child's haemoglobin level falls it is usually a sign that they are becoming unwell and may need treatment to prevent complications.

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This test also measures other parts of the blood: for example, the number of white cells, which help fight infection, and the number of platelets which help the blood to clot. The number of platelets may fall if the spleen gets very enlarged.

### **Alpha thalassaemia trait test**

Normal adult haemoglobin A (HbA) contains two beta chains and two alpha chains. Sometimes one or more of the genes controlling alpha chain production is absent and this results in alpha thalassaemia trait. If your child has inherited alpha thalassaemia trait as well as sickle cell anaemia it may in fact mean that the sickle cell anaemia will be less severe. This is because the concentration of haemoglobin in each red blood cell is less and therefore the red blood cell is less prone to sickling.

### **Reticulocyte count**

This test checks the number of young red blood cells present in the blood. It will show if the bone marrow is working properly. The bone marrow is the factory where new red blood cells are made before they are released into the blood stream. Sometimes as a result of infection the bone marrow may stop making these. This will cause the haemoglobin level to fall (see aplastic crisis page 26).

### **Blood chemistry**

A number of tests are done to check the level of various substances in the blood, substances which are vital for maintaining health, growth and development. For example, this test is done to check how well the kidneys and liver are working.

### **Glucose 6 Phosphate Dehydrogenase (G6PD) Deficiency test**

G6PD is an enzyme which protects the red blood cells against chemicals. Absence of the G6PD gene is found most commonly in people who come from the areas of the world where the sickle gene is found. If the enzyme is not present and certain foodstuffs or medicines are taken, the red blood cell may be destroyed, leading to a worsening of anaemia. Some medications, including those used in the treatment of malaria, can cause this problem, so it is important for this test to be carried out before giving malaria medications. It is possible to inherit both the sickle cell gene and the G6PD deficiency gene.

### **Urine tests**

The urine may need to be checked if there is blood or protein in the urine, if there is a possibility of infection, or to look at how well the kidneys are working.

## X-Rays and scans

Your child may need to have X-rays and scans taken for different reasons.

**Chest X-ray:** usually to look for infection.

**Bone X-ray:** sometimes to look for infection or the results of sickling (avascular necrosis of the femoral head (see page 26) or to monitor growth.

**Ultrasound scan of the abdomen:** to determine the size of the spleen and kidneys and to look for gallstones.

**Echocardiogram:** like an ultrasound scan, used to look at the size and functioning of the heart.

**CT and MRI scan:** this is a picture taken to look for tissue damage, For example, it may be used to help distinguish a painful crisis from Osteomyelitis (infection of the bone) or to look at the brain to see if there has been a silent stroke or the blood vessels are more narrow than usual.

**Doppler blood flow studies:** tests to see how well blood is flowing in any part of the body. Transcranial Doppler (TCD) in particular looks at how fast the blood is flowing through blood vessels in the brain. If the blood seems to be flowing very fast this may be due to narrowing of that blood vessel and may indicate that there is an increased risk of having a stroke (see page 28).

Other scans or special tests may be requested and these will be explained to you by your doctor.

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# HEALTH AND SOCIAL CARE PROVIDERS

## Anaesthetist

If your child needs an anaesthetic for any operation, including certain dental treatments, it is important for the anaesthetist to know that your child has sickle cell disease. Apart from putting people to sleep for operations, anaesthetists also give advice about managing pain, including sickle crisis pain.

## Audiologist

Audiologists are specialists that check hearing; they are usually based in community Centres or hospitals. One of the rare complications of giving desferrioxamine (see page 36) to get rid of excess iron is hearing loss. Therefore children having this treatment need to have their hearing checked regularly by an audiologist.

## Children's nurse (paediatric nurse)

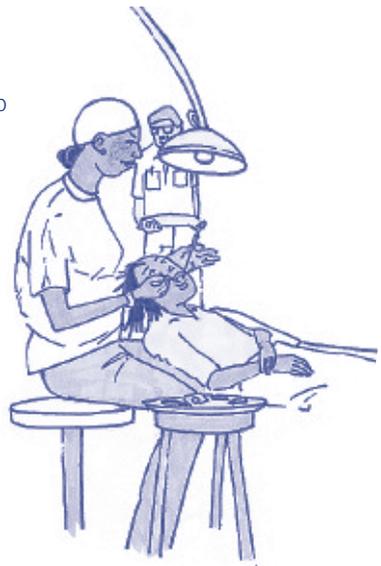
Paediatric nurses are specially trained to look after children (from premature birth up to the age of 18) with acute and chronic illness. They are involved in assessing the patient's nursing needs considering their medical, emotional and family circumstances, then planning and delivering nursing care in hospitals or other settings in conjunction with other health professionals.

## Children's centre workers

Every area will now have a children's centre a one-stop shop for all children to access local health and social services and support. These Centres are staffed by children's support workers.

## Dentist

Children with sickle cell disease may have bone problems which affect the jaw bone and subsequently their teeth. Therefore they may need special dental care and advice.



## General practitioner (GP)

Your child's GP needs to know that your child has sickle cell disease, because s/he is responsible for your child's medical care in the community and refers to specialists when necessary. The hospital doctor usually informs your GP about your child's treatment and care during hospital admissions and outpatient clinic visits.

It is also necessary for you to inform your GP if your child has been admitted to or treated in the hospital recently. When your child commences penicillin V, the initial dose will be prescribed by the hospital doctor. As this medication is given twice per day every day, it will be necessary to obtain a repeat prescription from your GP. Do remember that if your child is ill, it is always advisable to see your GP first, instead of taking your child to the hospital, unless it is an emergency.

If you move to another area, it is important to get your child re-registered with a local GP in that area. Once you have done this your child's GP health records from birth will be transferred automatically to your new GP, so that continuity of care can be maintained.

## Haematologist

Haematologists are specialist doctors who look after people with blood disorders. They, with the paediatrician and your GP usually manage your child's sickle cell disease.

## Health visitor (HV)

Your health visitor (HV) is a family visitor and offers health advice to help your family stay healthy. All children under the age of five years are seen by a HV regularly or periodically depending on the service provided in your area. They give advice on family health matters, including diet, childhood illness, immunizations, how to help your child grow healthily and how to cope with parenthood. They, with your GP or local child health clinic doctor, will check that your child is growing and developing normally.

Although many sickle cell nurse specialists are health visitors they do not replace your family health visitor. Your family health visitor advise you mainly about your family's general health and well being but most can and do advice on sickle cell and other health issues affecting any member of the family. Some areas of their work do overlap with the specialist nurse / counsellor. The hospital clinic doctor and nurse specialist do keep your health visitor informed about your child's progress, through your child's baby book or through the patient held records available in some areas. Therefore it is important that you take the book or records with you to clinic, and when your child has any other medical appointments.

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## Hospital youth worker

Increasingly hospitals are employing a person who works mainly with young people between 11 and 20 years of age. The aim is to have someone to promote their personal and social development with a range of informal educational activities which combine enjoyment, challenge, and learning. The four key parts of this role are to offer personal support, promote the young people's viewpoints, provide a diverse range of personal and social developmental opportunities, and promote self esteem and optimism. In particular hospital youth workers support young people in the move from child to adult services

## Ophthalmologist

A doctor who checks peoples' eyes and vision. Ophthalmologists are based in community health clinics or hospitals. People with sickle cell disease can develop changes in their eyes which could lead to disturbance of vision. The changes in children can occur as early as 6 years old. Children receiving desferrioxamine (see page 36) need their eyes tested regularly.

## Paediatrician (Hospital)

Your child may be looked after by a general paediatrician that is a doctor who specializes in looking after children's conditions, or a paediatric haematologist, who is a specialist in blood disorders affecting children. Who you see depends on the sort of doctors available in your hospital.

## Paediatrician (Community)

This is a doctor who is a specialist in managing long term conditions and disability in the community. They are responsible for coordinating all necessary care such as physiotherapy or speech and language therapy, and they liaise with social services, education and the voluntary sector as appropriate.

## Paediatric home care nurse

Some health districts employ community children's nurses. They are trained in nursing sick children in their own home and may visit after your child is discharged from hospital but still needs some nursing care or ongoing medication, for example intravenous antibiotics.

## Pharmacist/chemist

The pharmacist at your local chemist, dispenses prescribed medications, and can give advice on a variety of health issues and minor ailments. Some large chemists have a credit card scheme where your child's regular prescribed medication details are put on computer so that when you visit the chemist you can be given advice and prompt attention.



## Phlebotomist

A trained health worker who takes blood and send it to the laboratory for testing.

## Physiotherapist

A physiotherapist helps people do physical exercises to prevent any physical complications or worsening of a medical problem. Physiotherapists advise on how to keep the body working properly physically and your child may see one, for example, after having a stroke.

## Play specialist

Play has a special function in the hospital environment. Working closely as part of the multi-disciplinary team, hospital play specialists: organize daily play and art activities in the playroom or at the bedside; provide play to achieve developmental goals; help children master and cope with anxieties and feelings; use play to prepare children for hospital procedures; support families and siblings; contribute to clinical judgment through their play-based observations; teach the value of play for the sick child; encourage peer group friendships to develop; organize parties and special events

## Psychologists

**Clinical psychologist:** The sickle cell team may include a clinical psychologist, a specialist who can help in dealing with children's thoughts, feelings and behaviours, for example in managing pain and other symptoms, bedwetting, or strong fear of injection needles. They can also look at the child's learning ability particularly if the child has had a stroke or is not doing as well as expected in school. The clinical psychologist could do a neuropsychological assessment to help determine whether the child needs to be referred to the educational psychologist attached to their school.

**Educational psychologist:** An educational psychologist is a specialist in helping children with schooling and learning difficulties, will accept referrals from schoolteachers, clinical psychologists and others. They will do an educational psychological assessment and may recommend extra educational support or specialist education if the child needs it.

## School health services

Once your child starts school you will see less of the health visitor, and in most some areas you may not see them at all. Your child's development and health monitoring will be taken over by the school nurse and doctor. They will continue to see your child right through the school years and keep a record of your child's health, including immunizations and development from birth.

The school nurse and doctor can help you explain your child's condition to the teachers; they can also help educate them about sickle cell disease and how to best manage your child in school.

## Sickle cell nurse specialist/counsellor

Some health districts employ nurse specialists, sometimes called counsellors. These are nurses, midwives or health visitors who have had additional training in caring for people with sickle cell and other genetic disorders affecting haemoglobin, for example thalassaemia and G6PD. The nurse specialist also has training in genetic counselling for these conditions. Such specialists may be based in a sickle cell and thalassaemia Centre, community clinic, health centre, GP surgery or hospital. (See pages 87-91.)



## Social worker

A trained person who helps families cope with an illness in the family. A social worker can offer families health and social advice, including advice on housing, ill health prevention, self care, coping with an illness in the family, income and welfare benefit entitlements. A social worker is a family Carer, offering families support to help them prevent or deal with individual or family problems.

Some sickle cell and thalassaemia Centres have specialist social workers as part of their team and they work with parents of children with sickle cell disease and adult clients.

## ROLE OF SICKLE CELL & THALASSAEMIA CENTRES

Through the efforts of people with sickle cell disease, parents, health and other workers the first sickle cell centre was set up in Brent in 1979. Since then many more Centres have been set up and they provide services for people with and at-risk of sickle cell, thalassaemia, G6PD and related conditions. For list of Centres and services in the UK see pages 87-91.



Most Centres are part of and funded by the National Health Service (NHS) for short or long term. Some are jointly funded and managed by the NHS, the local authority or a voluntary organization. Many of these Centres are an important part of and work closely with the hospital haematology department.

The majority of Centres are run and managed by specialist nurses or counsellors who have many years experience as nurses, midwives and health visitors and been through specialist sickle cell and other training. The team of professionals in a centre is varied but may include:

- Specialist nurse
- Medical doctor (usually a haematologist)
- Social worker
- Psychologist

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**Centres offer advice or services to clients, professionals, and the general public on:**

- Screening and genetic counselling
- Support for people with sickle cell disease
- Education of the general public about sickle cell and related conditions
- Information about support groups and voluntary organizations
- Housing advice
- Social Services advice
- Welfare advice
- Health promotion activities through leaflets, posters, video, audio tapes and other materials
- Training and education of health, allied and non-health professionals
- Publication of guidelines, books and other resources for professionals and lay groups
- Influencing policy makers (e.g. government health ministers)

They also monitor the effectiveness of services provided for people with or 'at risk' of sickle cell to ensure they get appropriate care and services in the hospital and community.

If you do not have a specialist centre or service in your area, contact the nearest centre to where you live (see pages 87-92), or one of the voluntary organizations. They will be able to advise you as to how to get this service. There are no strict boundaries for giving people advice.

## VOLUNTARY ORGANIZATIONS

Sickle cell Centres and hospitals that provide care for people with sickle cell disease may have patient support groups. These are made up of parents, children, adults, family members and friends of people with sickle cell disease. Some interested health and social care providers are often invited to offer support if and when needed. To find out if there is one in your area talk to your nurse specialist, health visitor, social worker or doctor or contact one of the national voluntary organizations listed on pages 91-92.

There are several voluntary organizations available to help and advise people with sickle cell disease. The main focus of these organizations is to promote national awareness of sickle cell disease and influence health and social care purchasers and providers who buy or provide services for their local community. They also influence those who give you and your child care: for example, hospitals, GPs, dentists, social and welfare providers. Voluntary organizations do this in an effort to bring about positive change in caring for and helping people with sickle cell disease, and ultimately to find a cure for this condition.

Some voluntary organizations visit patients whilst they are in hospital or help them resettle when they are discharged home. They help new parents get in touch with other parents who have more experience of caring for a child with sickle cell disease.

The objectives of each organization vary at a local level.



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## Organization for Sickle Cell Anaemia Research (OSCAR)

This was the first voluntary organization, established in 1975 by a group of parents and people with sickle cell disease. Their initial aim was to help raise funds to find a cure for sickle cell disease. Although the national OSCAR no longer exists there are local OSCARs operating nationwide: OSCAR Bristol, OSCAR Birmingham, and others.

The members of the organization offer parents, individuals and families support. If requested, they visit people in hospital or at home. When necessary they act on behalf of and as the voice of individuals and families to make sure they get fair and equal health and social services.

## The Sickle Cell Society

The Sickle Cell Society was established in 1979 by a group of adults with sickle cell disease, parents, Carers and interested health and social care professionals. The aim is to help improve the poor standards of care provided for the growing number of people with sickle cell disease in the UK. The second aim is to educate health and social care professionals on how to care for their patients effectively.

The Sickle Cell Society is the only national sickle cell voluntary organization in the UK and recently appointed regional officers located in various parts of the country to offer support to their local population.

The Society influences policy makers and health and social care providers. It has a welfare fund that helps parents and adult members who are experiencing financial difficulties. Once a year the members organize a summer holiday for children with sickle cell disease. For families needing additional support or experiencing additional stresses, they have developed a unique scheme which enables parents to get respite care for their child with sickle cell disease. Such a support system is of great benefit to all members of the family.

## United Kingdom Thalassaemia Society

This is a national organization which offers advice and support to people with and at risk of thalassaemia.

## Membership of voluntary organizations

Membership of any of these voluntary organizations helps families and individuals with sickle cell disease to have a collective voice and influence health and social care services provided for them. Through these groups families get to know about any new developments and research. Without a collective voice it is very difficult for individuals to argue for what they need or to influence the people who make decisions about health and social care services provided for them.

Many of the voluntary organizations get a lot of support and encouragement from prominent members of the community, including politicians and celebrities, for example, Trevor Phillips OBE, Archbishop of York, Dr John Sentamu, Bishop Sir Wilfred Wood, the comedian Lenny Henry OBE, Trevor McDonald OBE, Floella Benjamin, Garth Crooks, Dawn Butler MP, and many others.

There are several other local sickle cell support groups and voluntary organizations. They generally provide support to their local community. For more information about these organizations contact them direct, (addresses on page 91-92) or contact one of the sickle cell and thalassaemia Centres listed on pages 87-91.

Remember there is strength in a collective voice support your local support group and voluntary organization and they will be able to help you ensure that you continue to get the health and social service and support that you need.

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## QUESTIONS PARENTS OFTEN ASK

### How will I know if my child is having a sickle cell crisis?

This is the question most commonly asked by parents, especially when their child is newly diagnosed. They are worried that they may miss the signs and their child will be in unnecessary pain. As a parent you get to know your child better than any one else. If and when she is in pain you will notice a change either in her behaviour or her mood, even when your child is still very young. There are also some tell-tale signs. For example, has she got a temperature, is she sweaty, has she got unusual swelling of any part of her body, is she hugging a part of her body, is she refusing to feed? (See page 21 for information about managing illness.)

### At what age will the sickle cell crisis start?

This is unpredictable, a small number of children will have a crisis by the time they are six months old, others between six to twelve months. Others may not experience a crisis until they become toddlers. It also depends on the type of sickle cell disease your child has and how it affects her as an individual. Some people have no crisis for several years, or rarely have crisis even into adulthood.

### Do I need to give my child a special diet?

Although your child has sickle cell disease she is able to eat the same foods as the rest of the family and does not need a special diet. As long as she is eating a balanced diet there is no need for you to worry. Because children with sickle cell disease use up a lot of their energy for keeping well they tend to be thinner than their peers, therefore, some specialists suggest increasing the calorie intake a little, for example giving her more protein rich foods such as meat, fish and other foods. Ask your health visitor or specialist nurse about this. Your child may have a smaller appetite than her brothers and sisters, but this is nothing to worry about. Most children will eat enough to keep themselves well.

Many parents suggest that it is best not to make too much fuss about diet because it often makes the situation worse. Talk to your health visitor, doctor or specialist nurse if you are worried about your child's eating habits. (See page 13 for information about diet.)

## If sickle cell is so common why have I never heard of it?

It is possible that no one has ever mentioned it to you before, or you have not paid much attention to it as it has not affected you directly before. It is more common in certain groups and depends on where your ancestors came from originally. For example, one in ten African-Caribbean, and one in four West Africans have sickle cell trait, which is a healthy carrier state.

## Is it only Black people who have sickle cell disease?

Having sickle cell disease or trait is not related to the colour of one's skin. It depends on where one's ancestors came from originally. This determines the genes which they can inherit from their parents. Although sickle cell is less common in some racial groups it does occur infrequently, for example, it can occur in white northern Europeans.

## Can my other children or any one else catch sickle cell disease from my daughter?

It is not possible to catch sickle cell disease, because it is not an infection. Sickle cell disease is inherited. Your child inherited her genes from you and her father. Genes are passed on from one generation to the next, from both sides of the family. (See page 52 for an explanation of inheritance.)

## Is there a cure for sickle cell disease?

There is a cure for sickle cell disease called 'bone marrow transplantation' or BMT for short, but this cannot be offered to all children with sickle cell disease for a number of health reasons (see page 63 for more details.)

## Will my child be able to have children?

Because your child has sickle cell disease does not mean that he or she will be infertile. Sickle cell disease in itself does not cause infertility except in cases where there is damage to the reproductive organs. For example, a small number of boys and men have repeated episodes of 'priapism', an involuntary painful erection of the penis. (See page 27.) On rare occasions priapism may cause impotence, and treatment may be required to try and correct the impotence in adulthood.

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## Will my child die before the age of 21?

People with sickle cell disease can live the same number of years as anyone else provided they maintain good health and receive appropriate health care when they are unwell. Sadly, due to complications of sickle cell disease a small proportion of people with this condition may die at a younger age than the average person, but these deaths are not necessarily related to age.

## Will giving my child antibiotics (penicillin) every day not be harmful to their immunity?

Children with SCD are prone to infections and especially pneumococcal infections. This is because their spleen does not work properly from very early childhood. Research has shown that giving a low dose of penicillin daily can prevent some pneumococcal infections and help prevent death. Even if the child develops another type of infection there are other antibiotics that can be prescribed by your doctor.

It is very important that you give your child penicillin daily as this can make a difference between life and death, always discuss this with your doctor before stopping it.

## Can herbs cure my child's sickle cell disease?

There is no known herb which has been proven through research to cure sickle cell disease. At the moment it is not clear which herbs or natural remedies may help relieve the symptoms of sickle cell crisis. For a drug or herb to be licensed by the government and recommended by health professionals it needs to be tested to make sure the product is safe, that it works and that it will not cause any harm or make a person's condition worse.

## Will my daughter have periods?

Most young girls with sickle cell disease may start their periods a little bit later than their friends, but this is nothing to worry about; they will start eventually. If your daughter does not start her periods by the age of about sixteen years or you are at all worried it is worth talking to your GP or specialist doctor at the hospital. Delay in starting menstrual periods may be due to other causes, which are not necessarily related to sickle cell disease.

## Will my daughter's periods be more painful because of her sickle cell disease?

Some women and young girls who do not have sickle cell disease experience mild, moderate or even severe period pain. If your daughter has this it is not necessarily due to her sickle cell disease. It is worth taking the general advice given to women who experience period pains. Your GP, practice nurse at the surgery or pharmacist at the chemist will be able to advise you about this. Avoid giving your daughter any drugs unless this has been checked with your doctor because your doctor will be able to check whether it will interfere with any of the drugs he or she has prescribed.

## I thought sickle cell protects against malaria, so why do I have to give my child an anti-malaria drug when we go to a malarial zone?

Those with sickle cell trait who live in malarial areas of the world develop some resistance to malaria. Even though they can get malaria it tends to be mild and will not be as severe as it is for those who do not have sickle cell trait. However, if a person moves away from a malarial area they quickly lose this partial resistance to malaria and need to take the same precautions as anyone else.

**Those with sickle cell disease react very badly to malaria.** Therefore, if your child is going to an area of the world where malaria is common, especially in tropical countries, it is important that she has adequate protection, because malaria can be fatal in people with sickle cell disease. (See page 39 for information about anti-malarial drugs.)

## Why is a sickle cell crisis more common in cold windy weather?

If the skin cools down very rapidly, for example after getting caught out in the rain without protection, or if it is a cold windy day and the skin is exposed the chilling of the skin can cause a painful crisis.

## Can sickle cell disease cause death?

The complications of sickle cell disease can cause death. This is why it is important for parents, Carers and individuals with the condition to learn as much as they can about it so that they can help reduce the likelihood of complications. Sadly, despite doing all the right things to prevent complications, they may still occur and result in death. It is important for parents to know that it will not necessarily be their fault or any one else's if their child dies.

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## Can my child be circumcised?

Doctors do not recommend circumcision for a boy unless there is a particular medical reason. Doctors usually advise against it in all children because of the risk of bleeding and infection. Your son's sickle cell disease will not be made worse if he is circumcised for religious or social reasons. Female circumcision is illegal in the UK.

## I have sickle cell trait can I donate blood and or bone marrow?

Yes, you can be a blood or bone marrow donor. Remember you are a healthy carrier. Sickle cell trait is in your red blood cells. There are other parts of the blood which are just as vital as the red blood cells: white blood cells which fight infection, platelets which help the blood to clot, plasma which is the blood's water, all these are still very useful. You could donate bone marrow to a brother or sister with sickle cell disease if there was a good 'match'.

Parents are often worried about their child receiving donated blood, but all donated blood is tested very carefully in the UK and other developed countries. In the UK blood transfusion Centres do not usually recommend using a parent or other family member's blood.

## Can I give my blood to cure my child's sickle cell disease?

Blood cannot cure sickle cell disease. It may help reduce symptoms provided it is given regularly, because blood cells have their own life span. Normal red blood cells live approximately 120 days before they are destroyed by the body, but the body is constantly making new cells to replace these. Blood needs to be carefully cross-matched to make sure the donor's and the recipient's blood matches exactly. If your blood matches your child's and you donated the blood which is then given to your child, that blood will still only live in your child's body for a maximum of 120 days, in addition you can only safely donate blood about twice a year, because you need your blood yourself. Meanwhile your child's bone marrow is constantly making sickle red blood cells which will eventually replace the blood which was transfused.

## Why does my son have more sickle cell crises than my daughter?

Sickle cell disease affects people differently, even two people with the same mother and father. It is important not to compare them. Manage them according to their individual experiences of their disease and their individual needs.

## **Could my child's sickle cell disease be due to someone putting a curse on my family through witchcraft?**

No, sickle cell disease is inherited through the genes and occurs even in countries where they do not believe in witchcraft. In some communities there are many myths and superstitions surrounding sickle cell disease, the majority of which are untrue. If you have a religious background it may be worth talking to your religious leader or your specialist nurse about it. Many specialist nurses in the UK share your cultural background and some may share the same religious background as you, they may be able to help you examine this aspect if it is worrying you. Talk to your specialist nurse or contact one of the voluntary organizations (see pages 91-92).

## **Should I allow my child to go swimming?**

Sickle cell disease affects individuals differently. Some people can cope with strenuous exercise, like going swimming; others find this may bring on a crisis, particularly if the water is cold. Always make sure that your child wraps up quickly with a large bath towel after swimming and avoid chilling of the body. Take extra care to make sure the hair is dry because babies and children lose a lot of heat from their scalp. It is important not to stop your child from living as normal a life as possible. Doing any activity which they enjoy makes life more pleasant, provided they don't take unnecessary risks and they respond to their body's messages, when it is telling them to slow down or stop doing something.

## **We already have one child with sickle cell disease surely we will not have another one?**

Since you and your partner carry the unusual gene then there is a chance with each and every pregnancy that you can have another child with sickle cell disease. (For more information and examples of how inheritance works see page 52.)

## **Do people with sickle cell disease live to old age?**

Although the complications of sickle cell disease can cause early death, people with sickle cell disease do live into adulthood, even old age. The type of sickle cell disease a person has can also influence whether they develop complications or not and the type of complications which they may develop.

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## My child still wets the bed. Is she just lazy?

Wetting the bed is a common problem for a lot of children, especially children with sickle cell disease. Your child will probably be just as upset about it as you are. The main cause of this problem is that her kidneys do not concentrate urine very well and she passes larger than normal amounts of diluted urine. In addition, you encourage her to drink a lot of fluid so as to prevent dehydration and this adds to her passing a lot more urine. If your child is over six years old and still wets the bed talk to your doctor or nurse specialist. (See page 12 for more information.)

## If I trust in God will my child's sickle cell disease be cured?

If you believe in God you may be of the opinion that God gave doctors and nurses the opportunity to train, gain knowledge and skills to be able to care for your child properly.

If however, you are worried that perhaps your religious beliefs and what you have been told to do in caring for your child with sickle cell disease appear to be in conflict you should talk to your specialist nurse, GP or hospital doctor. It may be useful to visit your religious leader with your specialist nurse, she or he will be able to help you explain about sickle cell to your religious leader/ elder and also learn from them how to best support you in caring for your child in future. Sometimes what we assume will not be permissible because of our religion is not always the case and your specialist nurse together with your religious leader will help you explore these issues.

Most important, it is advisable to talk to your doctor, GP, specialist nurse, psychologist or other health professional before deciding to stop any treatment or medication that has been recommended for your child. Giving your child daily penicillin is vital to prevent infection and stopping this medication may endanger your child's life.

You and your child should be able to practice your religion irrespective of the type of condition your child has. In most cases the medical treatment being given to your child does not prevent you from practising your religion. However, your religion should be practised in a way that does not endanger your child's life.

## GLOSSARY OF TERMS AND ABBREVIATIONS

**Alloimmunisation** Development of antibodies against foreign material entering the body: for example, transfused blood that is not correctly matched.

**Amniocentesis** A method of testing the fetus in the womb. A small sample of the fluid around the fetus is taken and tested to find out which haemoglobin type the baby has inherited. (See page 59.)

**Antibodies** These are produced by the body to fight infection or to destroy anything the body does not recognize: for example, substances in unmatched donated blood.

**Anaemia** Insufficient red blood cell haemoglobin.

**Analgesic** Painkillers for example, Paracetamol.

**Antipyretic** A drug given to reduce fever.

**Aplastic crisis** This is when the bone marrow stops making new red blood cells, usually as a result of a virus infection. (See page 26.)

**Bone marrow transplant (BMT)** Bone marrow is taken from a donor and transplanted into someone with a disease, for example sickle cell disease or leukaemia

**Carrier (see Trait)** A person who has inherited one usual haemoglobin gene and one unusual haemoglobin gene, for example, sickle cell trait.

**Chelation** Removal of iron from the body to prevent iron being deposited in the organs of the body and causing damage. It is usually given routinely if someone with sickle cell disease is having regular blood transfusions.

**Crisis** A term used to describe different complications of sickle cell disease but commonly used to describe the pain of sickle cell 'painful crisis'.

**Chromosome** The carrier of all the genetic codes in the body, each person has 22 pairs of non-sex chromosomes and 1 pair of sex chromosomes.

**Chorionic villus sample (CVS)** A method for testing the unborn baby in the womb, a small piece of chorion (after-birth) is taken and tested to find out which type of haemoglobin the baby has inherited. (See page 59.)

**Computerized tomography scan (CT scan)** Picture taken of tissues in any part of the body in order to detect damage, used especially to detect damage to the brain

**Dactylitis** Swelling of the fingers, hand, toes or feet. It is often the first sign that a child has sickle cell disease. It is more common in babies, but may occur in older children though rarely seen in adults. (See page 11.)

**Desferrioxamine** A drug used to help the body get rid of excess iron, usually given when a child is on long-term blood transfusion. (See page 36.)

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**Drip** Fluid is passed into the body slowly through a small plastic tube, also see intravenous.

**Enuresis** Bed wetting.

**Epistaxis** Nose bleed.

**Fetal blood sampling (FBS)** A method of testing the unborn baby in the womb where a small sample of blood is taken from the cord and tested to find out the type of haemoglobin the baby has inherited from the parents (see page 60).

**Gene** Human characteristics are coded on string-like structures called genes, which are inherited from both parents. Genes are arranged like steps on the chromosomes and they determine a person's genetic make up: for example, whether a person will be tall or short, black or white and whether he or she will inherit sickle cell from their parents.

**Haematuria** The presence of blood in the urine.

**Haemoglobin (Hb)** Red pigment in the red blood cells which enables the cells to carry oxygen (air) from the lungs to all the body parts to keeps the body alive.

**Haemoglobin electrophoresis** A blood test to determine the type of haemoglobin a person has inherited from their parents.

**Haemoglobin type** The type of haemoglobin inherited from both parents, this is determined by the genes.

**Haemolytic anaemia** The type of anaemia seen in sickle cell disease caused by excessive and rapid breakdown of the red blood cells, but it is not the same as iron deficiency.

**Hepatitis** A viral infection of the liver.

**In Vitro Fertilization (IVF)** Fertilization of an egg by a sperm outside of the body. Children born in this way are sometimes referred to as test tube babies

**Intravenous** This means 'through the vein'. Fluid or medication may be given through a vein. When a small tube is left in the vein in order to give fluids this is commonly called a 'drip' because the fluid drips slowly into the vein.

**Jaundice** Yellow pigment in the skin or eyes caused by excessive breakdown of red blood cells and production of bile.

**Malaria** A disease commonly found in tropical countries, carried by some mosquitoes. It can be fatal in both people with normal haemoglobin and in those with sickle cell disease.

**Magnetic resonance imaging (MRI) scans** Pictures taken of any part of the body to look at tissues or organs for any damage.

**Opiates** Drugs obtained from the opium poppy seed and used to relieve severe pain: for example, morphine and Pethidine. These drugs can be addictive.

**Osteomyelitis** Infection of the bone.

**Patient-controlled analgesia (PCA)** A method of giving pain relief which gives the person in pain some control over how their pain is managed. (See page 32.)

**Pre implantation genetic diagnosis (PID)**

Method used with in vitro fertilization (see IVF). A woman's egg is fertilized with a man's sperm in the laboratory. A few days later the developing egg is tested for the genetic condition, like sickle cell anaemia and if it does not have the genetic condition the fertilized egg is placed in the woman's womb to grow into a baby.

**Prenatal diagnosis (PND)** Testing an unborn baby in the womb.

**Priapism** A painful involuntary and persistent erection of the penis.

**Pump** A device which pumps medicine into the body. For example, it is used to give desferal and for giving painkillers.

**Red blood cell** This is the part of the blood which acts as a storehouse for haemoglobin. Haemoglobin is required for carrying oxygen around the body.

**Screening** A test performed to assess whether a person has a particular disorder or is a carrier for that disorder.

**Sequestration** Trapping and pooling of blood in a body organ for example the spleen or liver.

**Sickle** A sickle is a farming tool with a curved blade. The term is used to describe 'sickled' red blood cells that have changed from a normal round shape to a banana shape.

**Spleen** A small organ on the left hand-side, just under the rib cage. Its function is to filter the blood and help protect the body from infection, but it may have problems working properly in people with sickle cell disease.

**Splenectomy** An operation to remove the spleen.

**Trait** Carrier of a genetic condition: for example, sickle cell trait. People with sickle cell trait do not have a disease therefore they do not have any symptoms.

**Transcranial Doppler Study (TCD)** A special scan to see how well blood is flowing through the blood vessels in the brain (see page 66).

**Vaso-occlusion** Blockage of the blood vessels due to sluggishness of blood flow. It causes mild to severe pain and is the most common form of sickle cell crisis pain.

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# USEFUL ADDRESSES

## Specialist sickle cell & thalassaemia centres and services in London

### Centres in London

#### Barking and Dagenham

Havering and Redbridge  
Haemoglobin Disorders Service  
Cedar Centre- Unit Management Office  
King George's Hospital  
Barley Lane, Goodmayes, Essex IG3 8YB  
Phone/Fax: 020 8983 8781

#### Bexley and Bromley

Sickle Cell & Thalassaemia Service  
Antenatal Clinic  
Beckenham Hospital  
379 Croydon Road  
Beckenham, Kent BR3 3QL  
Phone: 01689 866 811  
Fax: 01689866773

#### Brent

Sickle Cell & Thalassaemia Centre  
122 High Street  
Harlesden, London NW10 4SP  
Phone: 020 8961 9005  
Fax: 020 8453 0681  
Website: [www.sickle-thalassaemia.org](http://www.sickle-thalassaemia.org)  
Email: [brent@sickle-thalassaemia.org](mailto:brent@sickle-thalassaemia.org)

#### Camden and Islington

Sickle Cell & Thalassaemia Centre  
The Northern Health Centre  
580 Holloway Road  
London N7 6LB  
Phone: 020 7445 8035 / 8036  
Fax: 020 7445 8037

#### City and Hackney

Sickle Cell & Thalassaemia Centre  
457 Queensbridge Road  
Hackney, London E8 3TS  
Phone: (Switchboard) 020 7683 4570  
Fax: 020 7853 6709

#### Croydon

Sickle Cell & Thalassaemia Centre  
316 - 320 Whitehorse Road  
Croydon CR0 2LE  
Phone: 020 8251 7229  
Fax: 020 8251 7248  
Email: [sickle.cell@virgin.net](mailto:sickle.cell@virgin.net)

#### Ealing

Sickle Cell & Thalassaemia Centre  
Windmill Lodge  
West London Healthcare NHS Trust  
Uxbridge Road  
Southall UB1 3EU  
Phone: 020 8354 8022  
Fax: 020 8354 8948

#### Greenwich

Sickle Cell & Thalassaemia Service  
Gallions Reach Health Centre  
Bentham Road  
Thamesmead SE28 8BE  
Phone: 020 8320 5712/3  
Fax: 020 8311 8895

#### Hammersmith and Fulham

Sickle Cell & Thalassaemia Centre  
Richford Gate Primary Care Centre  
Richford Street  
London W6 7HY  
Phone: 020 8237 2980  
Fax: 020 8237 2986

### **Haringey**

The George Marsh Sickle Cell & Thalassaemia Centre  
St Ann's Hospital  
St Ann's Road  
London N15 3TH  
Phone: 020 8442 6230  
Fax: 020 8442 6575  
Email: gmc.office@haringey.nhs.uk

### **Hounslow**

Sickle Cell & Thalassaemia Service  
92 Bath Road Hounslow TW3 3EL  
Phone: 020 8321 2449  
Fax: 020 8321 2467

### **Newham**

Sickle Cell & Thalassaemia Centre  
Plaistow Hospital  
Samson Street  
London E13 9EH  
Phone: 020 8586 6262 / 6386  
Fax: 020 8586 6396

### **South East London**

Sickle Cell & Thalassaemia Centre  
Wooden Spoon House  
5 Dugard Way  
Off Renfrew Road  
Kennington  
London SE11 4TH  
Phone: 020 7414 1363  
Fax: 020 7414 1357

### **South West London**

Sickle Cell & Thalassaemia Service  
Balham Health Centre  
120 Bedford Hill  
London SW12 9HP  
Phone: 020 8700 0615/ 0616 / 0643  
Fax: 020 8700 0615  
Email: sicklethal.balham@wpct.nhs.uk

### **Sutton & Merton**

Sickle Cell & Thalassaemia Service  
St. Helier Hospital  
Wrythe Lane  
Carshalton, Surrey SM5 1AA  
Phone: 020 8296 3371/ 2000

### **Waltham Forest**

Sickle Cell & Thalassaemia Centre  
St James' Health Centre  
St James' Street  
London E17 7PJ  
Phone: 020 8520 0921  
Fax: 020 8928 2476

## **Centres Outside London**

### **Airedale (WEST YORKSHIRE)**

Sickle Cell & Thalassaemia Service  
Keighley Health Centre  
Oakworth Road  
Keighley  
West Yorkshire BD21 1SA  
Phone: 01535 295684

### **Birmingham**

Sickle Cell & Thalassaemia Centre  
Ladywood Community Health Centre  
St Vincent Street  
West Birmingham  
West Midlands B16 8RP  
Phone: 0121 465 4201/2/3  
Fax: 0121 465 4212

### **Bradford**

Sickle Cell & Thalassaemia Centre  
Manningham Clinic  
Lumb Lane  
Bradford BD8 7SY  
Phone: 01274 730836

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**Bristol**

Sickle Cell & Thalassaemia Centre  
Central Health Clinic  
Tower Hill  
Bristol BS5 0JD  
Phone: 0117 922 7571  
Fax: 0117 927 2180

**Cardiff**

Sickle Cell & Thalassaemia Centre  
Butetown Health Centre  
Loudoun Square  
Cardiff CF1 5UZ  
Phone: 02920 471055  
Fax: 02920 482674  
Email: Sickle.Cell@cardiffandvale.wales.nhs.uk

**Coventry**

Sickle Cell & Thalassaemia Service  
25 Warwick Road  
Coventry CV1 2EZ  
Phone: 024 7696 1332

**Derby (South)**

Sickle Cell & Thalassaemia Service  
Haematology Department  
Derby City General Hospital  
Uttoxeter Road  
Derby DE22 3NE  
Phone: 01332 785372

**East Lancaster and Cumbria**

Sickle Cell & Thalassaemia Service  
Edith Watson Unit  
Burnley General Hospital  
Casterton Ave  
Burnley BB10 2PQ  
Phone: 01282 474487

**Gloucester**

Sickle Cell & Thalassaemia Centre  
The Edward Jenner Clinical Unit,  
Gloucestershire Hospitals Foundation  
NHS Trust  
Greater Western Road  
Gloucester GL1 3NN  
Phone: 08454 225224  
Fax: 08454 225273

**Huddersfield**

Sickle Cell & Thalassaemia Service  
Princess Royal Community Health Centre  
Greenhead Road  
Huddersfield  
HD1 4EW  
Phone: 01484 344321

**Leeds**

Sickle Cell & Thalassaemia Centre  
Chapelton Health Centre  
Spencer Place  
Leeds LS7 4BB  
Phone: 0113 295 1016 / 1017  
Fax: 0113 2951018

**Leicester**

Sickle & Thalassaemia Centre  
Charnwood Health Centre  
1 Spinney Hill Road  
Leicester LE5 3GH  
Phone: 0116 2425663 / 0116 2538031  
Fax: 0116 2531568

**Liverpool**

Sickle Cell & Thalassaemia Centre  
Abercromby Health Centre  
Grove Street  
Liverpool L7 7HG  
Phone: 0151 708 9370

### **Luton**

Sickle Cell & Thalassaemia Service  
The Lodge  
4 George Street  
West Luton  
Bedfordshire LU1 2BJ  
Phone: 01582 708312  
Fax: 01582 511001

### **Manchester**

Sickle Cell & Thalassaemia Centre  
352 Oxford Road  
Junction of Denmark Road  
Manchester M13 9NL  
Phone: 0161 274 3322  
Fax: 0161 273 7490

### **Milton Keynes**

Sickle Cell & Thalassaemia Service  
Milton Keynes NHS Trust  
Whalley Drive Clinic  
Whalley Drive Bletchey  
Milton Keynes MK3 6EN  
Phone: 01908 660033  
Fax: 01908 365501

### **Nottingham**

Sickle Cell & Thalassaemia Service  
Mary Potter Hostel  
Gregory Boulevard  
Hyson Green  
Nottingham NG7 7HY  
Phone: 0115 970 5844  
Fax: 0115 979 1718

### **Preston**

Sickle & Thalassaemia Service  
Saul Street Clinic  
Saul Street  
Preston PR1 2QU  
Phone: 01772 401185  
Fax: 01772 883 502

### **Reading**

Sickle & Thalassaemia Service  
Haematology Department  
Royal Berkshire Hospital  
London Road  
Reading  
Berks RG1 5AN  
Phone: 0118 987 7689  
Fax: 0118 987 7755

### **Sandwell**

Sickle Cell & Thalassaemia Centre  
Haematology Department  
Sandwell Healthcare Trust  
Lyndon, West Bromwich  
West Midlands B71 4HJ  
Phone: 0121 553 1831, bleep 376

### **Scotland (West of Scotland)**

Sickle Cell & Thalassaemia Genetic  
Counselling Service  
Ferguson-Smith Centre for Clinical Genetics  
Regional Genetic Service  
Yorkhill Hospital  
Glasgow, Scotland G3 8SJ  
Phone: 0141 201 0808  
Fax: 0141 201 0361

### **Sheffield**

Sickle Cell & Thalassaemia Service  
Park Health Centre  
190 Duke Street  
Sheffield S2 5QQ  
Phone: 0114 226 1744  
Fax: 0114 226 1742

### **Southampton**

Sickle & Thalassaemia Service  
Newton Health Clinic  
24-26 Lyon Street  
Southampton SO4 0LF  
Phone: 02380 900222  
Fax: 02380 900213

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## **Walsall**

Sickle & Thalassaemia Service  
Moat Road Clinic  
Moat Road  
Walsall  
West Midlands WS2 2PS  
Phone: 01922 775079/84

## **Wolverhampton**

Sickle Cell & Thalassaemia Support Project  
Ground Floor Office  
St John's House  
St John's Square  
Wolverhampton WV2 4BH  
Phone: 01902 444 076  
Fax: 01902 445 322  
Email: info@sctsp.org.uk

## **Voluntary Organizations**

### **SICKLE CELL SOCIETY**

54 Station Road  
Harlesden  
London NW10 4UA  
Phone: 020 8961 7795  
Fax: 020 8961 8346  
Email: info@sicklecellsociety.org  
Website: www.sicklecellsociety.org

This is the only national sickle cell organization in the UK. They employ regional officers in various locations in England. They offer a range of services, including educational materials (leaflets, books, posters, videos); advice on health, education, employment, welfare, travel, insurance; talks/ training for health, allied professionals and lay public; grants to those in financial hardship; education/achievement awards; recreational activities/holidays for children.

## **UK THALASSEAMIA SOCIETY**

19 The Broadway  
Southgate  
London N14 6PH  
Phone: 020 8882 0011  
Fax: 020 8882 8618  
Email: office@ukts.org  
Website: <http://www.ukts.org>

This is the only thalassaemia organization in the UK. They offer a range of services, including educational materials (leaflets, books, posters, videos); advice on health, education, employment, welfare, travel, insurance; talks/ training for health, allied professionals and lay public, they support development and publication of clinical management guidelines for health and allied professionals.

### **ORGANIZATION FOR SICKLE CELL ANAEMIA RESEARCH (OSCAR)**

The branches of OSCAR offer a range of services. One or two offer clinical health and social care and genetic counselling services, in-patient and out-patient support. All branches offer public health promotion, education of health allied professionals and lay groups and organize public awareness campaigns, produce and disseminate booklets, leaflets, posters, videos and other resources.

### **OSCAR BRISTOL**

Sickle Cell & Thalassaemia Centre  
256 Stapleton Road  
Easton  
Bristol BS5 0NP  
Phone: 0117 951 2200  
Fax: 0117 951 9570  
Email: oscarbristol@sicklecell.fsnet.co.uk  
Website: [www.bristoloscar.org](http://www.bristoloscar.org)

### **OSCAR TRUST LIMITED**

5 Lauderdale house  
Gosling Way  
Brixton  
London SW9 6JN  
Tel: 020 7735 4166  
Fax: 020 7820 9594

### **MIDLANDS SICKLE CELL & THALASSAEMIA SOCIETY**

(Formerly OSCAR Birmingham)  
251 - 253 Rookery Road  
Handsworth, Birmingham  
B21 9PU  
Phone: 0121 551 6553

### **SICKLE WATCH**

African Caribbean Cultural Centre  
9 Clarendon Road  
Hornsey  
London N8 0DJ  
Phone: 020 8888 2148  
Fax: 020 8881 5204

### **SICKLE CELL ANAEMIA RELIEF FOUNDATION (SCARF)**

C/O 114 Batman Close  
White City  
London W12 7NX  
Phone: 020 8248 8316

A local organization offering a range of services, including education materials (leaflets, books, posters, videos); advice; talks for professionals and lay public.

### **SICKLE CELL ANAEMIA RESEARCH (SCAR)**

PO Box 88  
Barking, Essex 1T11 8PH

An organization developed by the famous footballer Garth Crooks, dedicated to raising and funding research and studies into sickle cell disease. They offer an education grant to children who have learning difficulties due to frequent illness or hospitalization as a result of their sickle cell disease.

### **SICKLE CELL WOMEN'S WELFARE ASSOCIATION (AYOKA)**

Sickle Cell Project  
St Margaret's House  
15a Old Ford Road  
London E2 9PL  
Phone: 020 8981 9603

### **SICKLE CELL & YOUNG STROKE SURVIVORS**

801 Old Kent Road.  
London. SE15 1NX  
Phone: 020 7635 9810  
Email: [Info@scyss.org](mailto:Info@scyss.org)

### **THE SHEFFIELD SICKLE CELL & THALASSAEMIA FOUNDATION (SSCAT)**

Ace Business Centre  
110-120 The Wicker  
Sheffield S3 8JD  
Phone: 0114 275 3209  
Fax: 0114 279 6870  
Email: [sscatf1@btconnect.com](mailto:sscatf1@btconnect.com)

Apart from the voluntary organizations listed here there are several local patient/parent support groups all over the UK. To find out if there is one near you contact the sickle cell & thalassaemia Centre or Service in your area or contact any of the voluntary organizations listed.

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## Regional Genetic Centres in the UK

This list of regional genetic centres (RGCs) is included because not all areas of the UK have specialist sickle cell Centres or services. RGCs are specialist units often placed within National Health Service hospitals. They offer information, advice and counselling for families with or 'at risk' of any genetic condition. They enable the individual, couple and family to explore the likelihood of a condition occurring in their family. Where possible they explain how the condition is inherited, the possible health and social implications if the condition does occur and the options available when planning to have children.

RGCs generally only accept referrals from GPs, maternity units, other health care professionals and health specialists. However, clients who have used the service before, for example, during a previous pregnancy, are encouraged to contact the genetic centre directly when planning a subsequent pregnancy or at least as early as possible when the pregnancy is confirmed.

Because each RGC operates differently it is worth discussing this with your specialist nurse/doctor, GP or health visitor, or write to the RGC nearest to where you live and ask them to send you information about the type of services they provide and how they get referrals.

### Centres in London

#### **North West Thames Regional Genetics Centre**

The Kennedy Galton Centre  
Northwick Park Hospital  
Watford Road  
Harrow, Middlesex HA1 3UJ

#### **North East Thames Regional Genetics centre**

Institute of Child Health  
The Hospital for Sick Children  
Great Ormond Street  
London WC1N 3JH

#### **South East Thames Regional Genetics Centre**

Paediatric Research Unit  
Guy's Tower Block  
Guy's Hospital  
St Thomas' Street  
London SE1 9RT

#### **South West Thames Regional Genetics Centre**

Genetics Centre  
St Georges' Hospital  
Blackshaw Road  
London SW17 0QT

### Centres outside London

#### **Northern Region Genetics Service**

19/20 Claremont Place  
Newcastle-upon-Tyne NE2 4AA

#### **West of Scotland Region**

Genetics Service  
Duncan Guthrie Institute of Medical Genetics  
Yorkhill  
Glasgow G3 8SJ

#### **Regional Genetics**

Counselling Service  
Royal Liverpool Hospital  
Prescott Street  
Liverpool L7 8XP

#### **Regional Cytogenetics Laboratory**

Birmingham Heartlands Hospital  
Bordsley Green East  
Birmingham B9 5ST

## **East Anglia Regional**

Genetics Service  
Department of Clinical Genetics  
Box 134  
Addenbrooke's Hospital NHS Trust  
Hills Road  
Cambridge CB2 2QQ

## **Government agencies**

### **The National Association of Health Authorities (NAHAT)**

Head Office, Birmingham Research Park  
Vincent Drive  
Birmingham B15 2SQ  
Phone: 0121 471 4444

Will tell you which Primary Care Trust is responsible for providing health care services in your area and the local hospitals and community Trusts caring for your community.

### **NHS Departmental Equality Adviser**

Department of Health Equality & Human Rights Group  
Department of Health  
6th Floor  
Skipton House  
80 London Road  
London SE1 6LH

This is the governmental representative who advises the government about equality issues including minority ethnic health issues. The adviser seeks the views of communities and feeds these back to the Minister of Health, so that decisions can be made about the best way to meet the specific needs of all sections of the population, including minority ethnic communities.

## **National Health Services Executive**

Headquarters  
Quarry House, Quarry Hill  
Leeds LS2 7UE  
Phone: 0113 254 5000  
Website: [www.open.gov.uk](http://www.open.gov.uk)

The NHS executive influences government minister's decisions about health services provision nationally and is responsible for securing resources and allocating these to Primary Health Care Trusts (PCTs). PCTs are responsible for buying health services for their local population and for monitoring the performance of hospital and community service providers.

## **Other useful contacts**

### **Antenatal Results & Choices (ARC)**

73 Charlotte Street  
London W1P 1LB  
Phone: 020 7631 0285  
Website: [www.arc-uk.org](http://www.arc-uk.org)

ARC provides support for women and couples going through the experience of making choices about an at-risk pregnancy. Dealing, for example with issues surrounding having prenatal diagnosis (testing the baby in the womb), making a decision about an affected pregnancy; deciding to terminate or not terminate an affected pregnancy.

### **Anthony Nolan Bone Marrow Trust**

Unit 2 – 3, Heathgate Place  
75 Agincourt Road  
London NW3 2NU  
Phone: 0207 284 1234  
Website: [www.anthonynolan.org.uk](http://www.anthonynolan.org.uk)  
Email: [eastgate@anthonynolan.com](mailto:eastgate@anthonynolan.com)

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When a child with sickle cell disease is going for bone marrow transplant there needs to be a perfect match. This organization keeps a vast register of bone marrow donors nationwide and may be able to help find a matching donor not just for sickle cell but other genetic conditions such as leukaemia.

**African-Caribbean Leukaemia Trust (ACLT)**

PO Box 670  
Croydon CR9 5DP  
Phone: 0208 667 1122  
Fax: 0208 667 1626  
Email: [aclt1@aol.com](mailto:aclt1@aol.com)  
Website: [www.cursitor.com/aclt](http://www.cursitor.com/aclt)

This organization was set up by the parents of Daniel DeGaille a black child with leukaemia who could not find a matched donor because there were few Black people on the register. The couple campaign to encourage more Black people to become bone marrow donors so that it will become possible for more black people to get a perfect match. They are linked to the Anthony Nolan Trust and help maintain a national register of ethnic minority donors.

**Useful Reading**

Anie, K and Fotopoulos, C (1998) *Coping with Sickle Cell Disease and Pain - A self-help Manual for Children* London: Brent Sickle Cell & Thalassaemia Centre (ISBN: 0 9531902 3 4)

Anie, K and Fotopoulos, C (1998) *Coping with Sickle Cell Disease and Pain - A self-help Manual for Adults* (ISBN: 0 95319021 8)

**Useful websites**

**For Advice on employment issues**

[www.33max.co.uk/government-education-employment-agencies.htm](http://www.33max.co.uk/government-education-employment-agencies.htm)  
[www.33max.xo.uk/careers-advice-training.htm](http://www.33max.xo.uk/careers-advice-training.htm)  
[www.gaapweb.com](http://www.gaapweb.com)  
[www.dti.gov.ph](http://www.dti.gov.ph)  
[www.careerbuilder.com](http://www.careerbuilder.com)

**Genetic Alliance**

(A consumer organization with a directory of support groups catalogued according to organization, disease or service)  
[www.geneticalliance.org](http://www.geneticalliance.org)

**Health Protection Agency**

[www.hpa.org.uk](http://www.hpa.org.uk)

**How stuff works (Life Sciences Section)**

[www.howstuffworks.com](http://www.howstuffworks.com)

**National Travel Health Network and Centre (NATHNAC)**

[www.nathnac.org](http://www.nathnac.org)

**Other**

**NHS Sickle Cell & Thalassaemia Screening Programme**

[www.screening.nhs.uk/sickleandthal](http://www.screening.nhs.uk/sickleandthal)

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