

A parent's guide  
to managing  
sickle cell disease

If you are told your child has sickle cell disease you will probably have lots of questions. In this book we will describe what sickle cell disease is, the different types, treatments available and offer practical advice on living with and supporting a child with sickle cell disease.

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Copies of this book and other resources can be downloaded from the Sickle Cell Society [www.sicklecellsociety.org](http://www.sicklecellsociety.org)

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Picture drawn by Chakoura

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We hope the information contained in this book will continue to encourage and empower you as you continue to make efforts to look after a child with sickle cell disease.

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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 condition which affects the blood. Like most parents when given such news you may feel shock, and be confused, angry or afraid. Often parents cannot believe the news and may feel responsible or blame themselves for 'giving' their child a disease. They may not know what to say to relatives and friends. They are bewildered about how to care for their child and what to expect in future.

Such feelings are natural to all parents when their child has been found to have an inherited condition, whether it is sickle cell or any other inherited disease. 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 healthcare professionals may not have cared for many people with this condition. If that is the case, it is worth mentioning this book and other useful materials which can be accessed at: [www.sicklecellsociety.org](http://www.sicklecellsociety.org)

You may 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, born of the same parents, so do not assume that how it affected the person you know or knew is how it will affect your child. There are quite a lot of myths and popular misconceptions about sickle

cell disease. We hope that this handbook will dispel some of these by giving you clear, accurate information.

We have tried to make this book easy to understand and to use. Some of the scientific terms may be new to you. These are briefly explained in the glossary (page 107). You may want to read the book all the way through. You can also just dip into it to find out about particular aspects of sickle cell disease. The book is an introduction and a start of your learning about the condition. It is important to get more detailed information from the people caring for your child as they will be familiar with the specific ways in which your child experiences the disease.

See page 118 for a summary of the government's recommended national guidelines for the care and management of children with sickle cell disease.

Sickle cell disease affects both boys and girls. Apart from a medical condition called priapism, which only affects boys/men, all the things discussed in this book relate 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 things mentioned in this book will affect your child.**



Sickle cell disease is a term covering a number of different but similar conditions which affects haemoglobin. Haemoglobin gives blood its red colour and is responsible for carrying oxygen from the lungs to all parts of the body.

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## What is sickle cell disease?

Sickle cell disease is a term covering a number of different but similar conditions which affects 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 (HbSS), Sickle Haemoglobin C disease (HbSC) and various forms of sickle beta thalassemia, such as, sickle beta plus thalassemia (HbS/β+Thalassaemia) and sickle beta zero thalassemia (HbS/β<sup>0</sup>Thalassaemia). These are described further on page 14.

These conditions are called 'sickle' cell disease because the red blood cells, which are normally round and very flexible, become rigid and 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 very small blood vessels. If the sickled cells get trapped in the blood vessels, this reduces the blood supply to that part of the body and causes pain and sometimes damage. This is sometimes called a 'painful crisis'. It often comes on suddenly, and typically lasts several hours or days. Usually it does not cause any permanent damage.

### What causes the cells to sickle?

A normal red blood cell is round, soft, spongy and very flexible. It carries oxygen very well and is able to travel through the body without getting stuck in the blood vessels. The sickle red blood cell on the other hand is hard, rigid, breaks easily and is not able to keep its round shape.

Red blood cells change to a sickle shape when oxygen levels in the body are low. They usually return to their original round shape when they get more oxygen, often when they have passed through the lungs, but after a while they lose their ability to make this change and cease to carry oxygen.

Sickled red blood cells are not as flexible as normal red blood cells and cannot always pass through very small blood vessels.

Getting very cold or dehydrated tends to make red cells lose oxygen and can increase sickling. The things that can cause sickling of red cells and obstruction to blood flow include:

- Dehydration (lack of water in the body)
- Infections and fevers
- Sudden changes in body temperature, particularly skin cooling
- Excessive physical exertion
- Stress

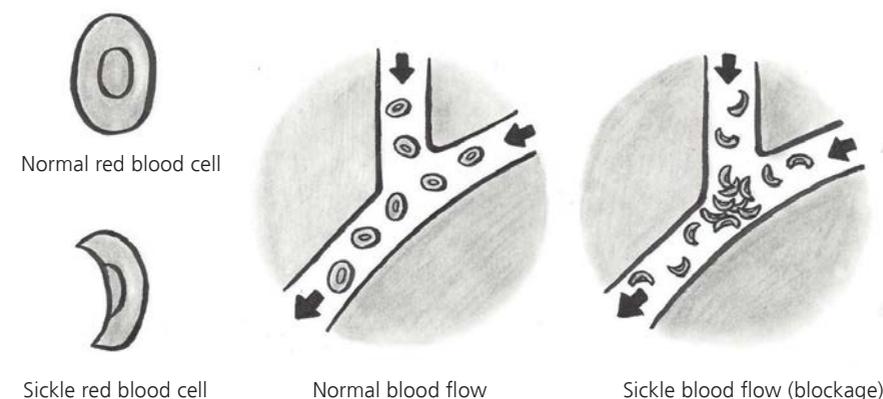
**See page 24 for more information on how to avoid some of the causes listed above.**

### How did my child get sickle cell disease?

Sickle cell disease is inherited. This means that your child inherited an unusual type of haemoglobin from both you and your partner. (See illustrations from page 77).

The normal and most common haemoglobin type is haemoglobin A. There are over 1000 different types of unusual haemoglobin but the ones that are commonly seen in the United Kingdom are haemoglobin S (sickle haemoglobin), haemoglobin C, haemoglobin D and beta thalassemia. 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 but this amount decreases gradually over the first few

**Figure 1** Normal and Sickle cell blood flow



years of life to reach adult levels of about 1%. Some children with sickle cell disease carry on producing small amounts of haemoglobin F, which may be beneficial.

An inherited condition like sickle cell disease remains with a person all their life.

Sickle cell disease varies in severity from one person to the next for reasons that are not clear. It is known that inheriting alpha thalassaemia trait (also known as being an alpha thalassaemia carrier) or having the ability to make lots of haemoglobin F tend to make sickle cell disease less severe.

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

## Types of sickle cell disease

All babies are now tested for sickle disease in their first week of life using a spot of blood taken from a heel prick. It usually takes a few weeks to get the results, and if the baby seems to have sickle cell disease a repeat test is necessary to confirm this and find out exactly which type of sickle cell disease he has inherited.

### Sickle cell anaemia (HbSS)

This is the commonest form of sickle cell disease and occurs if your child has inherited sickle haemoglobin (HbS) from both parents. Sickle cell anaemia is very variable and it is not possible to predict what sort of problems might occur.

The aim is that all children with sickle cell anaemia will live happy and normal lives, although medical problems will occur sometimes.

On average, people have about one significant health problem per year that is related to this form of sickle cell disease.

### Sickle haemoglobin C disease (HbSC)

This occurs if your child has inherited sickle haemoglobin from one parent and haemoglobin C from the other. In general, HbSC disease is less severe than sickle cell anaemia but the same health problems can occur.

### Sickle beta thalassaemia disease (HbS $\beta$ thal)

This occurs if your child has inherited sickle haemoglobin from one parent and beta thalassaemia from the other. There are many different types of beta thalassaemia mutation. A mild beta thalassaemia mutation in combination with sickle haemoglobin causes sickle beta plus thalassaemia, commonly written HbS/ $\beta$ +thalassaemia; this combination usually causes less severe disease. A severe form of the beta thalassaemia mutation in combination with sickle haemoglobin results in sickle beta zero thalassaemia, commonly written HbS/ $\beta^0$ thalassaemia, which is often more like HbSS.

For more information about beta thalassaemia contact the UK Thalassaemia Society (details on page 106) or one of the specialist centres listed on page 112.

### Sickle haemoglobin with hereditary persistence of fetal haemoglobin (HbS/HPFH)

This occurs if your child has inherited sickle haemoglobin from one parent and high fetal (baby) haemoglobin levels (HbF) from the other parent. Your child will continue to make significant amounts of fetal haemoglobin (approximately 30%) and should have very mild sickle cell disease with no significant problems. Penicillin and transcranial doppler scans are not usually recommended for those with S/HPFH.

### Sickle haemoglobin D Punjab disease (HbSDPunjab)

This is a rare form of sickle cell disease and occurs if your child has inherited sickle haemoglobin from one parent and haemoglobin D Punjab from the other parent. This is usually similar to sickle cell anaemia, causing similar health problems. Penicillin and transcranial doppler scans are usually recommended.

### Sickle haemoglobin E disease (HbSE)

Haemoglobin E (HbE) is very common in Southeast Asian countries, like Thailand and Vietnam, and also in parts of India and Bangladesh; it is a type of thalassaemia. Sickle haemoglobin E occurs if your child inherits sickle (HbS) from one parent and haemoglobin E from the other. It causes a mild form of sickle cell disease sometimes with problems such as acute pain, but does not usually cause major

health problems. Penicillin and transcranial doppler scans are usually recommended.

### Sickle haemoglobin O-Arab disease (HbS<sup>OArab</sup>)

This is a rare form of sickle cell disease, occurring when haemoglobin S is inherited from one parent and haemoglobin O-Arab from the other. Haemoglobin O-Arab occurs in the Middle East, but is also found in populations across the world. This combination usually causes a disease like sickle cell anaemia (HbSS), and is similarly variable, with some people getting more problems and others very few. Regular penicillin and transcranial doppler scans are usually recommended.

### Other unusual haemoglobin combinations

There are several other combinations of unusual haemoglobin which can cause sickle cell disease but these are rare.

In this book we write about sickle cell disease as if it is one condition although the different types are not all the same, and everyone's experience of an illness differs. Some experience milder symptoms than others and children with the same sort of sickle cell disease, even children with the same parents, may have different experiences and medical problems.

## Sickle haemoglobin carriers and those carrying other unusual haemoglobins

Being a 'carrier' of sickle haemoglobin is sometimes known as having sickle cell trait, and does not usually cause significant illness. It is not a form of sickle cell disease and will never change into sickle cell disease. If a person is born a carrier they are healthy and will always be a carrier.

Being a carrier of sickle haemoglobin means that you have inherited normal haemoglobin A from one parent and sickle haemoglobin S from the other; this is sometimes commonly written HbAS.

In extreme conditions where there is a lack of oxygen, such as when deep sea diving or being on top of a high mountain,

a carrier of the sickle cell gene may have some health problems, such as pain in the spleen. There is also evidence that kidney problems are a bit more common especially as people get older, and very rarely, extreme exercise may cause severe dehydration, heat exhaustion and severe illness.

People who are sickle cell carriers (HbAS) may need to train differently if they join the armed forces or take up sport professionally.

Other common haemoglobin carrier states seen in the UK include haemoglobin C carrier, where a person has inherited normal haemoglobin A from one parent and haemoglobin C from the other parent (HbAC). Beta thalassaemia carrier is when a person has inherited normal haemoglobin A from one parent and beta thalassaemia ( $\beta$ thalassaemia) from the

other parent (HbA $\beta$ thalassaemia). These do not cause health problems, but might make some of the person's blood/results slightly abnormal. For example, people who carry beta thalassaemia tend to have small, pale red cells and might be very slightly anaemic which can be mistaken for a condition called iron deficiency and treated with iron tablets unnecessarily.

The importance of knowing if you carry sickle cell or any other unusual haemoglobin is that it can be passed on to your children. How this inheritance works is explained further on pages 78-84.

### Why did sickle cell first occur and who is affected?

It is thought that the sickle cell haemoglobin first occurred thousands of years ago, probably in Africa and other tropical countries. Being a carrier for sickle haemoglobin seems to offer some protection against malaria, which is often fatal in young children. Over thousands of years, sickle cell trait has become increasingly common in areas of the world where malaria occurs, and more children are therefore born in these areas with sickle cell.

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 Afro-Caribbeans
- 1 in 12 Turks

- 1 in 20-50 Asians
- 1 in 100 Northern Greeks

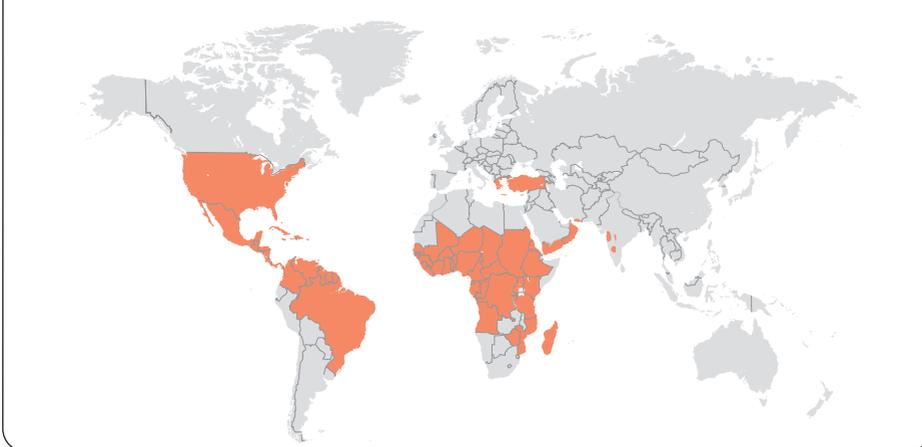
For information about malaria and the need for protection see page 35. All the other unusual haemoglobin types that have been described here probably also offer some protection against malaria.

## The effects of sickle cell disease

### How does sickle cell affect children?

During the first few 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 which is called Fetal haemoglobin (HbF), and usually a very low level of sickle haemoglobin S (HbSS) (about 5-10%) or any other unusual haemoglobin that your child has inherited. For example in the combination of HbSC, the child will make about 5-10% of S and C combined. Over the first year of life the haemoglobin F begins to reduce as your child starts making more haemoglobin S. The rate at which haemoglobin F drops can be linked to when the symptoms of sickle cell disease start. Some children have higher than usual amounts of haemoglobin even till adult hood, and this may be beneficial. 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 serious sickle cell complications or crises.

**Figure 2** Worldwide distribution of the Sickle Cell Gene



Children with sickle cell disease may be smaller than those who do not have the condition but they generally grow at a steady rate.

However, your child will still be at risk of serious infection and will need to be on penicillin (oral antibiotics) by 3 months of age. (See page 27.) One of the first signs of sickle cell complications, which may occur after the age of about 6 -18 months, is swelling of one or more fingers or other parts of the hand, or one or more toes or other parts of the foot. This is known as hand-foot syndrome, also called dactylitis. Although this may be distressing at the time it does not mean the child will necessarily have more problems in the future. This symptom of crisis is usually treated with simple pain medicine and the child is encouraged to drink extra oral fluids.

### Anaemia

When a child is making a lot more haemoglobin S, these red blood cells have a much briefer life span than the cells that contain the usual haemoglobin A. The body tries to compensate by making more red blood cells but it usually cannot compensate completely hence your child becomes anaemic. Your child may look pale and the palm of his hand and his 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. (See page 23.)

Folic acid tablets or other vitamin medications may be prescribed because

the body uses folic acid when making new red blood cells. However, in the United Kingdom most children get enough folic acid from their normal diet and extra folic acid is not necessary. There is no evidence to suggest that giving folic acid will improve your child's health, but it does no harm. Only give folic acids if directed to do so by your child's specialist doctor.

There are some rare additional complications of sickle cell disease which can lead to a worsening of the anaemia, such as acute splenic sequestration. (See page 47.)

### 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 produced during this process is a yellow pigment called bilirubin.

The liver clears the bilirubin from the body, but if there is a lot of bilirubin being produced 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 when they are in pain or have other ailments.

It can be a useful sign that your child is not as well as usual. There is no specific treatment for this sort of jaundice and the common practice of giving your child lots to drink will not make much difference. If they have very significant jaundice this may be associated with gallstones, and

in such cases your child needs to see a doctor. (See page 48.)

### 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 the condition but they generally grow at a steady rate. They tend to go through puberty later than average but they go on growing for a little bit longer to eventually reach their normal adult height.

### Enlarged spleen

The spleen is an organ that lies on the left side of the stomach under the rib cage. The spleen helps to clear infection from the body and also clears up old or damaged blood cells. 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 may continue to be enlarged for some time but then reduces in size and may stop working altogether. This is because it becomes filled with the sickled red blood cells that it is trying to clear from the body. If the spleen gets filled with sickle cells it cannot clear the body of infection. This is why we recommend that your child takes penicillin twice daily. (See page 27 for further information on penicillin). Sometimes a lot of blood gets trapped in the spleen and it gets very big worsening the child's anaemia. This is an acute splenic sequestration. (See page 47.)

Depending on the type of sickle cell disease your child has, you may be shown how to feel your child's spleen when he

**Figure 3** Spleen being examined



*You may be shown how to feel your child's spleen when he is well, so that when he is unwell you can tell. If it is bigger than usual he should be seen by a doctor.*

is well, so that you can tell if it is getting bigger and could need medical attention. We recommend that you feel your child's spleen once daily, especially when your child is unwell.

### Painful episode

Pain is a known and sometimes common symptom of sickle cell disease. The classic pain episode seen in sickle cell disease is often called a 'painful crisis', which occurs when the very small blood vessels become blocked by 'sickled' red blood cells. Usually the pain will last several days, although sometimes it lessens more quickly. When this occurs your child will need to be given regular painkillers and plenty of fluids and occasionally may need admission to hospital, depending on what is causing the painful episode. If the pain gets better quickly in a few hours, it may not be caused by sickle cell disease, and might be a simple headache or tummy pain which everybody gets sometimes.

Dactylitis, also known as 'hand-foot syndrome' may be the first painful episode that you see and usually occurs between the ages of 6 and 18 months when a finger or other parts of the hand, or a toe or other parts of the foot, become swollen and painful. After this age, pain may occur more commonly in the arms, legs or back.

For further information about pain and how to manage sickle cell pain at home see pages 44-45. It is worthwhile remembering that not all pain is due to sickle cell crisis or related to sickle cell disease.

### Bed wetting (Nocturnal Enuresis)

Bedwetting is normal in all children up until the age of about 7 years. It may take longer for a child with sickle cell disease to become dry at night. Because of tiredness from the anaemia the child may sleep very deeply at night and not wake up in time to go to the toilet. In addition, in sickle cell disease the kidneys are not able to produce concentrated urine. Urine in the bladder is very dilute. The bladder becomes very full and the child has to get up at night, sometimes several times, to go to the toilet. In addition he is encouraged to drink lots of water during the day so his kidneys will produce more urine. Wetting the bed is outside your child's control and he should never be punished.

Bedwetting is therefore not uncommon in children with sickle cell disease. Most achieve dryness eventually and there are certain techniques that can help. We suggest you talk to your specialist nurse or doctor if you are concerned.

### Will my child have all these signs and symptoms?

Not necessarily. The aim of this book is to provide you with advice and guidance.

Every child may not encounter the exact same experiences or complications. The advice provided is designed to guide you with the information on what to do, should you encounter some of the issues or complications outlined.

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 an enlarged spleen or dactylitis (hand-foot-syndrome). Some children rarely experience pain although this is the commonest symptom of sickle cell disease.

Children who have sickle – haemoglobin C disease (HbSC) or some types of sickle beta plus thalassaemia (HbS $\beta^{+}$ Thal) – tend to be only slightly anaemic and usually do not get jaundiced unless they have a sickle cell complication. An enlarged spleen in these forms of sickle cell disease is more common and does not usually cause any serious problems.

**For information on medical complications of sickle cell disease see pages 46-50.**

## 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 should not be affected by sickle cell disease. This is because he will still be producing a lot of baby haemoglobin F and not so much haemoglobin S. However, one of the main risks is serious infection due to 'pneumococci'. Even in the first few months the spleen may not be able to clear the body of this infection. The national standard for medical care of children with sickle cell disease is that your baby should take twice-daily penicillin (an antibiotic) from the age of 3 months so as to prevent this infection. This will be recommended strongly by your doctor. This and other standards of care can be found in the 'Sickle Cell Disease in Childhood: Standards and Recommendations for Clinical Care' publication available on the Sickle Cell



Society website at: [www.sicklecellsociety.org/paediatricstandards](http://www.sicklecellsociety.org/paediatricstandards).

After about 3 to 6 months of age it is possible that sickle cell problems may start to 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 pain episode.



For how to manage sickle cell pain and other complications see page 44.

### 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 from his diet as well as vitamins and minerals needed for healthy growth.

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

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 to a bone infection called osteomyelitis (see page 48).

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 thinner than others because most of the energy provided by the food goes towards making new red blood cells 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 and discussed with you. 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.

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 him health problems. Modifying your cultural and religious practice is a part of staying healthy.

Before your child reaches the age when he is expected to start fasting, it may be helpful to arrange a meeting with your religious leader and 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 supplements?

On the whole extra vitamins are not needed as your child will get sufficient from a normal balanced diet. The one vitamin that children do not get enough of from their diet is vitamin D. We make this vitamin from sunlight so it is important that your child gets enough sun on his skin. One of the symptoms of vitamin D deficiency is bone pain and this could get confused with sickle cell pain. If you are concerned about this your specialist doctor can do a blood test to check if your child is vitamin D deficient and may recommend taking supplements.

All babies are advised to take Abidec (multivitamins). It is important that your child continues to take this after the first 2 years of life.

Your child does not need other vitamins unless your family eats a special diet.

If your child becomes more anaemic than usual, and he is not already taking it, folic acid supplements may be prescribed by your doctor. This helps the body to make more red blood cells. Some clinics prescribe folic acid, 1—5 mg 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 sickle 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 traditional, herbal or complementary medicines do remember to tell his hospital doctor because it may be important to consider this when prescribing other treatments for your child.

Children should be encouraged to find their own level of tolerance and to take part in normal activities with other children.

### Avoiding things which may trigger an illness

#### Infection

Infection is an important trigger of sickle illness, 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. One of the most common is pneumococcal infection which can be avoided by taking the recommended penicillin twice daily and having regular pneumococcal immunisations (see page 29). Salmonella infection can be avoided by re-heating food thoroughly and ensuring

that eggs and chicken are properly cooked. All children should take advantage of the routine childhood immunisation programme, which will protect them from whooping cough, meningitis, haemophilus influenza, mumps, measles and German measles as well as the less common ones: polio, diphtheria and tetanus.

It is also advisable for your child to be immunised against influenza (flu) every year. If he is travelling, it is important to consider whether he needs any special medications, for example, anti-malaria drugs (see page 35).

#### Adequate fluids

It is important that your child drinks enough to maintain a steady fluid balance. In sickle cell disease the kidneys are not able to concentrate urine and so he will pass large quantities of dilute urine.

When your child is well, he will probably drink enough to make up for this loss in the urine but if he becomes unwell, for example with a fever or with diarrhoea and vomiting, he needs to drink more to avoid dehydration (see page 42 for how to manage fever). Insufficient water in the body (dehydration) can trigger a sickle cell pain episode. Plain water or diluted juice should be encouraged and fizzy drinks avoided because these may cause stomach pain in some children. It is not necessary to force your child to drink more than he wants when he is well and there is no evidence that it makes the yellowness in his eyes (jaundice) any better.

#### Extremes of cold and heat (temperatures)

Excessive chilling of the skin may trigger a painful episode, for example swimming in very cold water or getting wet from rain water. When in the house your child should not be overdressed and the heating should not be too high as there is then the risk that your child may get too hot and sweaty causing him to lose fluid and his skin become chilled. Make sure he has sufficient outer layers when he goes outside if it is cold or windy. 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 cold, he should be advised not to swim. It is important to discuss this with your child's school so that they are aware of the importance of this advice and the reason behind it.

#### Stress and anxiety

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 pain episode 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.

#### Physical exertion

Physical activity can reduce stress, so on the whole it should be encouraged. Children with sickle cell disease may find competitive sports more difficult because of their anaemia, which may lead to fatigue. Children should be encouraged to find their

own level of tolerance and to take part in normal activities with other children.

### Common ailments – effect on children with sickle cell disease

Most of the health problems affecting your child with sickle cell disease will probably be common things which can affect any child, including those who do not have sickle cell disease. These problems will usually get better quickly without any complications, but occasionally sickle cell disease can make things a bit more complicated. These common illnesses include:

#### Coughs and colds (upper respiratory infections)

All children get lots of coughs and colds, particularly in winter and when they first go to nursery or school. In general these should not cause complications related to sickle cell disease, and your child should recover in a week or so. It is important your child is kept warm and that he is given plenty to drink. Paracetamol is helpful if your child has a fever or feels unwell. He should continue to take his penicillin as usual, but extra antibiotics are not usually needed because most coughs and colds are caused by viral infections and antibiotics do not help. Occasionally, a cough or cold might lead on to acute pain from sickle cell disease, and giving paracetamol and ibuprofen should help. If the pain becomes very severe, admission to hospital may be needed.

## Influenza (flu) and Covid-19

Influenza is a severe viral infection which is commonest in winter and sometimes occurs in big outbreaks across the world, called pandemics. Symptoms include high fevers, muscle aches and pains, shaking ('rigors'), coughing and sickness. It is sometimes hard to know whether symptoms are due to flu or a different viral infection, but initially children should stay at home, drink plenty of fluid and continue their penicillin. In up to half of cases of actual influenza a significant sickle complication may develop, including acute chest complications, severe pain and anaemia.

If your child seems very unwell, then they may need admitting to hospital and treating with anti-viral medicines (such as Tamiflu), strong antibiotics and sometimes blood transfusion.

Vaccination against influenza is recommended each year (see page 29), and you should normally be contacted by your GP about this in autumn. If your child does not get invited for the vaccination contact your GP surgery.

Covid-19 is a severe viral infection which emerged in late 2019 and caused a global pandemic. At the time of writing this book we are still learning more about how Covid-19 affects people with sickle cell disease, but it seems that children usually have mild symptoms and complications are no more common than for influenza. It is important to follow official advice whilst the pandemic continues, particularly regarding the need to wash hands carefully and to stay at home as necessary.

Vaccination will become very important to prevent and control the virus long-term.

## Diarrhoea and vomiting

Children often get gastroenteritis, which is usually due to viral infections such as norovirus and rotavirus. This usually improves in a few days and antibiotics do not help, but your child should continue the recommended daily penicillin. It is important for your child to drink plenty of fluids and oral rehydration fluids may be helpful (Dioralyte, Electrolade, World Health Organisation (WHO) Oral Rehydration Salts). If the vomiting is very severe and your child is unable to keep fluids down, admission to hospital will be needed for intravenous fluids (a drip) to be given to prevent dehydration.

## Urine infection

Urine infections are more common in children with sickle cell disease, particularly in girls. Symptoms include burning and stinging on passing urine and needing to go frequently. It is important to see your doctor, who can test the urine and give antibiotics to treat the infection. If your child gets several urine infections, further tests may be useful to check the kidneys and bladder.

## Headaches

Headaches are common in children and are often caused by tiredness, stress, dehydration, constipation or viral infections. Sickle cell disease can also cause headaches due to sickling in the skull but this is uncommon.

Very rarely, sickle cell disease can damage the blood vessels in the head and cause severe headaches. Most normal headaches should be managed at home with paracetamol, plenty of fluids and rest. Try to identify if there is an underlying cause such as constipation or problems in school.

If headaches occur once a week or more, you should discuss this with your GP or sickle cell doctor. If your child has a very sudden or severe headache or has any difficulty speaking or moving, you should seek urgent medical advice from your nearest hospital Accident & Emergency (A&E) department.

## Rashes

Rashes are not usually due to sickle cell disease. Common causes of rashes in children include viral infections and allergies. In general you should see your GP if you are concerned.

## Asthma

Asthma is common among all children and it is not caused by sickle cell disease. It is important to treat asthma in the usual way, with inhalers and sometimes with oral medicines. Sometimes asthma attacks cause acute problems in sickle cell disease and asthma may increase the risk of sickle complications, so it is important to see your GP or practice nurse for advice about the best possible treatment for your child's asthma.

## Immunisations and preventative medications

### Penicillin

Making sure your child has 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 in a person with sickle cell disease.

The spleen is an important part of the body's defense 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 this can be fatal. You can reduce the risk of pneumococcus

**Making sure your child has penicillin twice a day is one of the most important things you can do for your child.**

by ensuring you child takes penicillin syrup or tablets twice daily as recommended. If your child is allergic to penicillin then another antibiotic, usually erythromycin, may be prescribed instead.

For the penicillin to be effective it must be taken twice every day at around the same time. Make sure you do not run out of supplies. If your child is unwell and has been put on another antibiotic by your GP or the hospital, check whether you need to stop the penicillin and remember to start it again once the other prescribed 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. The dose of penicillin is as follows

**The dose of penicillin is as follows:**

- 62.5mg twice a day until 1 year of age;
- 125mg twice a day from 1 until 5 years of age;
- 250mg twice a day from 5 years onwards.

It is strongly recommended that penicillin is given throughout childhood and carried on into adulthood. It is probably better to use 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. However it is now possible to obtain sugar free penicillin from your pharmacist. 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 to take.

**Routine childhood immunisations**

Your child should receive all the same childhood immunisations recommended for every child in the UK. These immunisations include: diphtheria, whooping cough, tetanus (DPT), polio, haemophilus influenza (Hib), PCV (conjugate pneumococcal vaccine), meningitis C and measles, mumps and rubella (MMR) and Hepatitis B. These are fully explained in the parent-held record (or baby book). If you are not

sure 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 immunisations. In fact it is perhaps even more important that your child should be fully protected because children with sickle cell disease are more susceptible to infections and infections can trigger sickle cell complications.

**Other immunisations**

**Pneumovax**

Pneumovax gives protection against more of the pneumococcal types that cause infection than the routine PCV vaccine. This vaccine is given at around two years of age and then every 5 years thereafter even throughout adulthood; it is given routinely to all children with sickle cell disease and your GP surgery or hospital specialist doctor will prescribe and give it.

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

**ACWY**

This gives protection against meningococcus types A and C, which cause meningitis. Even if your child has had the Men C vaccine, which protects against meningococcus type C, if you are travelling to some parts the world, e.g. Africa and Asia, he should also have ACWY as it will protect against meningitis type A.

**Hepatitis B**

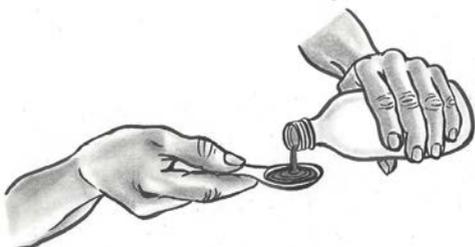
Your child will probably have a blood test in the outpatient department to check whether he has been in contact with hepatitis. Hepatitis B is occasionally passed from mother to baby in the womb. In the UK babies are now offered this vaccination as part of the national routine childhood immunisation programme. However it may need to be given to children who have recently moved to the UK and were not given it in their country of origin. Very occasionally hepatitis B can be passed on through a blood transfusion, but the blood transfusion services in the UK are extremely careful and screen all blood donors for hepatitis B (see page 55). If your child should need regular blood transfusions for any reason and has not already received a course of immunisations, his doctor may advise that a course should be given.

**Influenza (flu)**

The flu virus causes an infection which may lead to respiratory problems that can be serious in children with sickle cell disease. Your specialist doctor will advise and recommend that your child should be given the flu vaccine by your GP, from the first autumn after your child is 6 months old and then once a year thereafter.

Your child should have the same immunisations as other children. It is also advisable to ask your GP to give your child the flu vaccine each autumn.

**Figure 4** Medicine being poured



*Follow the instructions for giving any medication, you can ask your doctor, nurse or pharmacist for advice.*



# Management and treatment

As you learn more about sickle cell disease you will find that you become more knowledgeable on how to manage many aspects of the illness at home and when to seek medical help.

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## Managing your child when away from home

### Carers, child minders and nurseries

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

You can obtain leaflets and booklets and a copy of this book can be accessed on the Sickle Cell Society website [www.sicklecellsociety.org/resource/parentsguide/](http://www.sicklecellsociety.org/resource/parentsguide/). This 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.

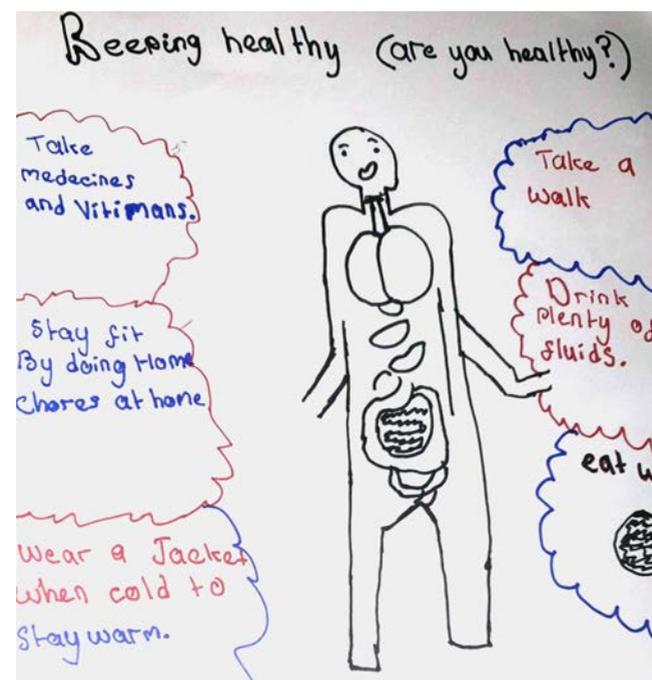
### Your child in school

Whilst he 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 he is not exposed to things which may trigger a

sickle cell pain episode. The following are examples of some things they could do:

- Make sure he is kept warm and away from draughty windows.
- Allow him to take part in physical activities but recognise that he may get tired more quickly than other children. Every child is different and the amount of exercise that he is able to cope with will need to be judged in school.
- Prevent chilling after physical exercise.
- Swimming does not usually cause problems for children with sickle cell disease, but should be avoided if the water or pool environment is very cold. As soon as he has finished swimming it is important for him to have a warm shower and get dressed immediately to prevent chilling of the body. Particular attention should be paid to wet hair because a lot of heat is lost through the scalp.
- Make sure he is able to drink enough in school, especially during the summer months.
- Be sympathetic when he 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 he starts school, it is worth making a specific appointment to meet with his school nurse. Discuss your child's sickle cell disease, how it affects him, what precautions need to be taken in the school environment to keep him well, how he



Picture drawn by Tiarrah

behaves when he is in pain, what teachers need to look out for and the actions to take when he 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 his teachers and other school members, whilst making sure that your child is not stigmatised or singled out from his classmates. If your child experiences bullying or teasing because of his 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.

The specialist nurse or school nurse will be able to visit the school with you to help the school learn about sickle cell disease and to complete a school health care plan which outlines your child's specific needs in the school environment. If this has not been arranged you can request it and be given a copy of the care plan for your records.

### Educational progress

Sickle cell disease may affect your child's educational progress. This may be because he is frequently unwell and has to miss

school. Children may suffer from stroke and this may affect their learning ability and their behaviour. Routine annual Transcranial Doppler (TCD) scans (see page 62) 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 can be normal, but may suggest that sickle cell disease has reduced the blood supply to parts of the brain, sometimes causing small scars (silent strokes). This can be diagnosed on an MRI scan (see page 62). A clinical psychologist will be able to assess your child and find in what areas he may need extra help with his school work.

**When travelling to malaria zones children with sickle cell disease must be protected against the possibility of getting malaria.**

Some hospitals have school rooms and can 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 him to do provided he is well enough.

All schools are able to provide some extra teaching, but if he 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. The statement defines what extra teaching support is required and will be provided in the 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. You can also find information from the document '*Sickle Cell and Thalassaemia: A guide to School Policy*' available at the Sickle Cell Society website – [www.sicklecellsociety.org/resource/supporting-young-people-sickle-cell-school-guide/](http://www.sicklecellsociety.org/resource/supporting-young-people-sickle-cell-school-guide/).

### **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 usually required as all modern aircraft are pressurised to maintain steady oxygen levels.

As soon as you know the date of your travel, talk to your pharmacist, GP or clinic doctor and tell them which country you are going to and seek their advice. They can tell you which vaccines, immunisations

or special drugs your child needs to take before going away, how soon the drug should be started, how much he 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 owing 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. Using insect repellent sprays and creams on exposed skin, especially when going out in the evening, is also useful.

Anti-malarial medications are recommended. Generally these are started at least 2–3 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.

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-malaria medication be continued for at least 2–4 weeks after returning from your journey. Some forms of malaria are resistant to the usual medications, such as 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-malarial medications, it is important to know if your child has a deficiency of an enzyme called G6PD (see page 61). 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 deficiency.

People with sickle cell trait growing up and living in a malarial area develop some natural resistance to malaria, but if they live away from the malarial 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-malarial medications when visiting a malarial area.

### **Travel vaccinations**

It may be necessary for your child to have anti-meningitis immunisations when going abroad (see page 27). In some countries, vaccination against hepatitis and yellow fever is also recommended. Talk to your GP, pharmacist or hospital doctor well in advance in order to leave enough time for your child to obtain the relevant immunisations before you travel.

### **Other medications whilst abroad**

Your child will need to continue his penicillin medication. Depending on the level of health care available in the country you are visiting, consider whether you need to take a supply of the pain medication he normally uses, for example, paracetamol and ibuprofen. It is worth discussing this with your GP or hospital doctor.

### Travel insurance

Remember, most other countries do not have a free national 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 last there. 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 include information on:

- The type of sickle cell disease your child has.
- His usual blood levels (*as recently recorded*).
- Any operations or complications he has had.
- Medication that he takes regularly.
- Any treatment that your child is having, for example, regular blood transfusions or hydroxyurea.
- The type of analgesic (painkiller) which helps him 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.

It is also advisable to carry your medications and important documents such as your GP/ hospital doctor letter in your hand luggage in case your checked-in baggage is delayed or goes missing.

### Care whilst travelling

If travelling on long flights, it is important to inform the airline that your child has sickle cell disease beforehand. Some airlines suggest that all children with sickle cell disease should have oxygen whilst flying, but usually this is not essential. During the flight it will be necessary to give your child extra fluids and make sure he keeps warm because of the air conditioning. He should also move around regularly. Should he complain of pain, especially in the chest, please inform the air hostess/steward, as it may be necessary to give him oxygen. Airplanes can be quite cool, even whilst travelling in a hot country, because many use air coolers, so take some warm clothing with you. 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 winter periods, so the warm clothing may be useful when you arrive at your destination. If your child is unwell with severe pain or breathing problems before the flight, he should not fly until he has fully recovered.

### Preventing an illness whilst abroad

Whilst on holiday your child will need to keep taking his routine daily medications. Depending on which country you are visiting, it is worth asking your haematologist to find out the name of a specialist doctor or sickle cell centre in the area to which you are travelling. If you are unsure about the cleanliness of the water supply in the country you are visiting, it may be worth taking water sterilisation tablets with you – these 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 are using sterilisation tablets, you should read the instructions on the container very carefully and use exactly as instructed.** If your child becomes unwell with diarrhoea and/ or vomiting there is a simple recipe recommended by the World Health Organisation to prevent dehydration.

Mix the following for your child to sip/drink:

- Four-finger scoopful of sugar (about 1oz, which is 30g)
  - plus a thumb and finger pinch of salt (about half a teaspoon)
  - plus a full cup of water (about 6–8ozs, which is 150–250ml)

Alternatively, you can buy sachets of an oral rehydration powder mix such as Dioralyte or Electrolade to take with you. This is available from your local chemist.

### Checklist before you travel

For advice about travel planning, it is highly recommended that you talk to your health visitor, school nurse, specialist nurse or contact one of the voluntary organisations listed on page 112.

1. Letter from the doctor or specialist nurse.
2. Any extra vaccinations required.
3. Family doctor (GP) for anti-malarial medication and other routine medication, e.g. penicillin, 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.
7. Thermometer (you may need to check if your child has a fever).
8. Water sterilisation tablets.
9. Insect repellent spray or cream.
10. Clothing suitable for varying seasonal temperatures.

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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 specialises in sickle cell disease within a hospital. This clinic may be known as the Paediatric Haematology Clinic or Sickle Cell & Thalassaemia Clinic and is usually staffed by some or all of the following:

- Consultant Paediatrician
- Consultant Haematologist
- Consultant Paediatric Haematologist
- Clinical Nurse Specialist
- Community Specialist Nurse or Nurse Counsellor
- Psychologist or Specialist Clinical Psychologist
- Social Worker/ Support or Welfare Officer
- Receptionist/ Secretary/ Clinic Clerk
- Phlebotomist
- Radiologist/ Sonographer

Visits to the outpatient clinic are very 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. This, however, will depend on how well your child is.

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. However, this cannot always be guaranteed

It is a good idea to keep in contact with the clinic even if your child is well and let them know if you are unable to attend your appointment or if you change address or phone/ mobile number. 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 specialist team to monitor your child's health and development, for you to get information about sickle cell, learn how it may affect your child and learn how to safely manage symptoms at home. The clinic also provides an environment to 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 treatments/ vaccinations he has had in the hospital. If your child misses more than one appointment, it is important you contact your local sickle cell team urgently to book another appointment. Depending on your hospital's policy, missing lots of clinic



appointments may result in a referral being made to the child safe-guarding team.

### Getting information and support

The clinic also offers an opportunity for discussing 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 workers 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 your child is well a blood test once a year may be sufficient. (For an explanation about blood tests see page 60). In addition, from the age of 2 years, a special test to check the blood flow through the brain is carried out every year until the age of 16. This is known as a Transcranial Doppler (TCD) scan, and it is done to check whether your child might be at high risk of having a stroke (see page 49).

### Contact between the hospital clinic and your child's GP

The doctor at the clinic will keep your GP fully informed about your child's condition and the amount of antibiotic (penicillin) he needs to take daily. You should go to your GP in good time for renewed prescriptions.

It is important that you do not wait until you run out of the medication.

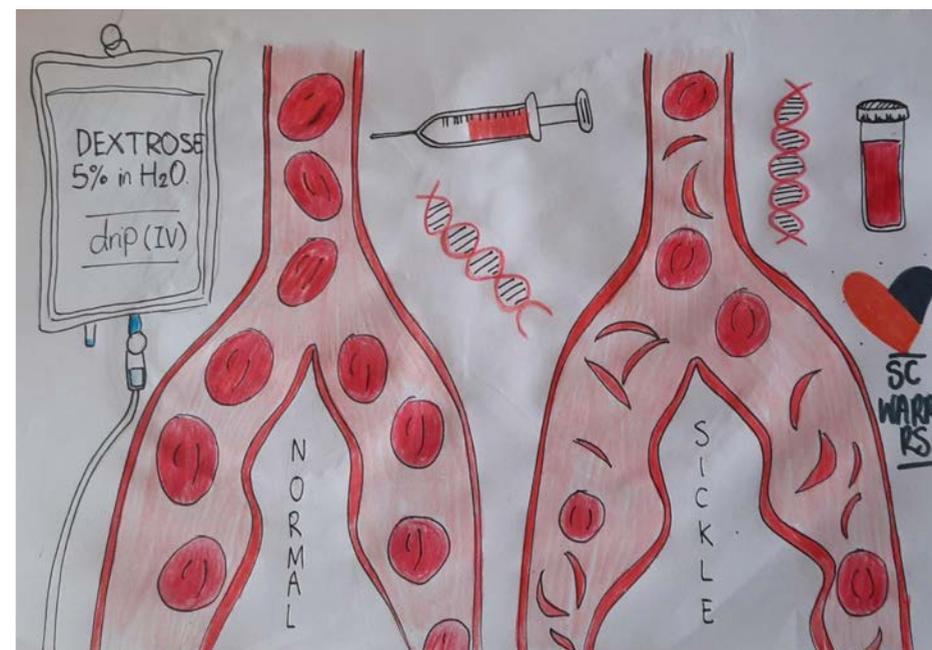
If your child is unwell, during working hours, it is best to contact your sickle cell nurse specialist for advice and guidance on whether to see your GP or be reviewed at the hospital. If there are specific concerns that you wish to discuss with your specialist team then it is usually possible to ask for an earlier appointment than your next routine visit.

If your child is unwell and you cannot manage the illness at home (see page 50 for information about medical emergencies), you should call to inform your specialist nurse or GP.

If you feel your child needs to be seen or treated urgently, you should go to A&E, or you may be able to go straight to a day unit or hospital ward for assessment, depending on your local hospital's policy.

### The National Haemoglobinopathy Registry

The doctor or nurse you see at the hospital will discuss the National Haemoglobinopathy Registry with you. This is a registry of all the patients in the UK with sickle cell disease, and is used to know how many patients there are, and in which areas of the country they live. This helps plan services for patients with sickle cell disease, to try and ensure that everyone can receive the best possible care.



Picture drawn by Asi

## Managing an illness at home

### Fever

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

Your child has a fever if her temperature stays above:

- 38°C, if taken in the mouth (if your child is above 8 years old)
- 37.5°C, if taken in the armpit (if she is younger than 8 years old)

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.

### Thermometers

Glass thermometers are not recommended for use in children for safety reasons and most parents prefer using digital thermometers because they are fast and easy to read.

Digital thermometers can be used in the mouth (oral), outer ear (tympanic) or under the armpit (axillary), or on the forehead. The digital thermometer will show you the exact temperature in numbers.



### How to use a digital rod thermometer

**Oral thermometer:** Place under the tongue and wait until the digital thermometer beeps. Do not give your child anything hot or cold to eat or drink for half an hour before taking an oral temperature. Food and drink can change the reading by warming or cooling the child's mouth.

**Ear thermometer:** Pull the top of your child's ear up and back; place the tip (covered with a probe cover) in the ear canal opening and press the button until it beeps. Be sure you are pointing the probe into the ear canal opening and not at the wall of the ear because if it is not used properly it can give an incorrect result. Use a clean probe tip each time.

**Axillary thermometer:** Place in the armpit with the tip in the deepest crease and wait until the digital thermometer beeps before reading. After use, wash the tip with warm water and soap and put it back in its case.

### How to use a forehead tape thermometer

Forehead tape thermometers are often preferred by parents for use with babies and young children and 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. A green colour signifies a normal temperature and red signifies that your

It is a good idea to keep a thermometer at home so that you can check her temperature.

child has a raised temperature. The tape is usually re-usable.

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 older than 8 years old, but make sure that her mouth is firmly closed when the thermometer is in place. If taking the temperature under the armpit, hold your child's arm firmly against the side of her body for at least 4–5 minutes.

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

- Give paracetamol or other prescribed pain medication (use as prescribed or follow the manufacturer's instructions)
- Give plenty of fluids to drink
- Remove most of her clothes

Don't chill your child's body too quickly. The room should be at normal temperature, approximately 21°C. Take her temperature about every half hour to check that it is going down. If her temperature does not come down and stays at 38°C or above when taken in the mouth or 37.5°C or above when taken in the armpit your child needs to see a doctor straight away, which maybe either your GP or in the hospital.

If you need further advice contact your hospital doctor, specialist nurse or call the NHS health advice line on telephone number 111.

## Managing sickle cell pain at home

Most episodes of 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. She may be fretful and miserable, persistently crying, or only crying when moved. As your child gets older, you will find that she 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 painkillers (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 4–6 hours as recommend on the bottle or by your doctor or pharmacist, but do not exceed the amount recommended.

Your doctor may also prescribe ibuprofen (Junifen), which helps relieve pain and inflammation. This can be given with the paracetamol. The doctor may prescribe other painkillers for your child to use at home and as your child gets older, she will know which one works best for her. 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 her to the Accident & Emergency department of your local hospital. 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. Dehydration (not enough water in the body) is known to be one of the causes of sickle cell 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 pain episode. 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

**Moist towels:** 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:** 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:** these 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.

Hot water bottles should not be used as they can get too hot and burn the skin. Also, you should not use ice to cool painful areas, as this can cause severe damage.

### 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, play computer games, watch television or play other games. Anything that can help distract your child's attention from the pain will be helpful (see information on self-help manual on page 111).

## When to seek medical and nursing help

If you have tried all the above remedies and your child still has a temperature or her pain is not relieved by the medications you have given her, 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 50.



In these situations you will need to get your child to the hospital straight away. Some hospitals have an arrangement so that you can take your child straight to the children's ward. If your hospital does not have this arrangement, take her to the Accident and Emergency department.

If you are unsure whether your child needs to go to hospital, always seek advice from your GP, specialist nurse or hospital doctor.

## Some medical problems

Please remember that sickle cell disease is very variable, and your child may never get any of the following problems. Sometimes it is possible to manage a medical problem at home either on your own or with the help of your specialist nurse or GP. Sometimes it will be necessary for your child to go to hospital because she needs medical attention. If you are unsure whether your child needs to go to hospital, always seek advice from your GP, specialist nurse or hospital doctor.

### Painful episodes

This is the most common medical problem in children with sickle cell disease and is caused by red blood cells blocking small blood vessels. One of the first signs of

sickle cell disease may be painful swelling of a finger (s) or the hand or toe (s) or foot.

This is known as dactylitis (hand-foot syndrome) and may occur from about 6 months of age. If your baby has been crawling or walking and then suddenly seems reluctant to do this, it may be because of dactylitis. She will need to be given regular painkillers and plenty of fluids.

However, if the pain remains severe and cannot be managed at home, or if she is not able to drink or is vomiting, she will need to go to hospital (see page 42 for how to manage sickle cell 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.

Blockage of blood vessels can occur in any part of the body – muscles and bones, stomach or chest – and will cause pain in that part.

Pain may be mild but sometimes can be very severe and frightening for your child. Pain can sometimes be triggered by your child getting cold or hurting herself, or by a viral infection, for example, flu, but many times there is no obvious cause. If the pain is severe and the painkiller you are using at home has not worked, it may be best for your child to go to hospital where she can be given stronger pain medications.

### Of course, like everyone else, she may have pain that is not due to sickle cell.

This can be confusing but as she grows up you and she will learn how to tell the difference.

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

In this condition the spleen gets very big over a few hours or days, 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 rapidly.

This complication is more common in children who are under 5 years of age. Once this has happened, it is possible for the spleen to enlarge again and the doctor may advise that your child has her spleen removed. This operation is called a splenectomy. If your child has had an episode of acute splenic sequestration, it may help if you learn how to feel the spleen for any enlargement in future. If your child appears pale, shows signs of being unwell and her spleen becomes much more enlarged, she will need to see a doctor straight away. Ask your doctor or nurse specialist to show you how to feel for your child's spleen.

## Parvovirus B19 infection

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 haemoglobin to fall to very low levels, requiring urgent treatment. A blood transfusion is usually necessary. Your child can only get this infection once because the body develops immunity.

## Chest infection and acute chest syndrome

Children with sickle cell disease are more likely to get problems with their chests, usually due to a combination of infection and sickled cells in the lungs. This is called acute chest syndrome and is similar to pneumonia in many cases.

Acute chest syndrome typically causes your child to have a cough, fever or chest pain and her breathing rate may be faster than normal. She may also feel pain over her back and abdomen.

Your child will need to be admitted to hospital if she has acute chest syndrome; a chest X-ray will be taken and your child will 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 55 for information on blood transfusion.) It is also important that your child breathes as deeply as possible. To encourage her to do so, she may be asked to blow/suck into a tube called an incentive spirometer. Occasionally children become more unwell and need help with breathing, and might be moved to the intensive care unit.

## Painful hip (avascular necrosis of the femoral head)

This is due to sickling in the topmost part of the thighbone. It is sometimes noticed by chance on an X-ray, but it can cause pain in the hip and may make your

child limp. This sort of pain 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 by using crutches. Avascular necrosis can also occasionally occur in other parts of the body, for example the shoulder joint or elbow.

### **Infection in the bone (osteomyelitis)**

Osteomyelitis can be difficult to distinguish from a sickle cell pain episode but it is quite rare. 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. Your doctor may ask the orthopaedic (bone specialist) doctor to explore the swelling to look for infection. If osteomyelitis is confirmed, antibiotics will be given for at least 6 weeks.

### **Blood in the urine (haematuria)**

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, the haemoglobin inside them is converted to bilirubin, a yellow pigment. This is responsible for the yellow colour of the whites of the eyes in many children with sickle cell disease. The bilirubin may also form gallstones within the gall bladder, a sac which produces bile and lies behind the liver.

Many children with sickle cell disease over the age of 10 years have gallstones and these can be shown by doing an ultrasound of the abdomen. Sometimes the stones get caught in the gallbladder and cause pain, particularly on the right side of the body. If gallstones start to cause problems, it is advisable to have an operation to remove the gallbladder, because the stones can sometimes cause a serious infection, known as cholecystitis. Your child can manage very well without a gall bladder and her health will not be affected.

### **Painful erection of the penis (priapism)**

Priapism is caused when sickled red blood cells block the blood vessels of the penis, causing a painful erection. This can occur at any age, but is less common in younger children. It is more common at night or in the early morning. Going to the toilet to pass urine sometimes helps to relieve priapism. Your child may need painkillers. A warm bath or shower and gentle walking may also 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 2 hours, your child should be seen in the hospital because treatment 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. There is a leaflet that discusses priapism in more detail. Ask your specialist nurse or doctor for a copy.

### **Stroke**

A small number 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 by a doctor. She may need special imaging tests of her brain and a blood transfusion (see page 50 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 because a stroke may follow. Without blood transfusion it is possible that the stroke may be more extensive and cause more permanent damage. It is usual for the initial signs of weakness to get better over a few months, but learning problems may persist.

Unfortunately, after a first stroke there is quite a high risk that another stroke may occur. To prevent this happening monthly blood transfusions are recommended. A

test called a Transcranial Doppler scan (TCD) can be carried out to see if your child may be at risk of having a stroke (see page 62). It is advisable that this is done yearly from the age of 2 years.

### **Silent stroke**

Sometimes the brain can be damaged without there being any obvious sign, such as weakness. This can lead to learning problems. It is important that your child's school teacher is aware of her diagnosis and can keep a close eye on her progress. If you or her teachers are concerned about her school performance, it is important to let your doctor know. A special brain scan can be carried out and other tests, such as a psychological assessment, can be undertaken to look at her learning abilities.

### **Eye problems**

Different problems can affect all parts of the eye and can sometimes affect vision. If your child complains of blurring of vision or pain in her eye, you should consult your doctor immediately.

It is important that your child's schoolteacher is aware of her diagnosis and can keep a close eye on her progress.

## Headaches

Although headache is mentioned earlier it is repeated here because if it is severe it can be a medical problem that needs attention. Headaches are quite common in children with sickle cell disease.

Headaches do not tend to last very long and can usually be treated with painkillers, for example paracetamol.

You should seek medical advice if your child's headaches are very frequent or persist for more than 2 hours at a time. Your child should be seen straight away if as well as a headache she has a temperature above 38.5°C, or a rash, is vomiting or has a stiff neck.

## Medical emergencies

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

**Fever** oral temperature of 38.5°C or above

**Very severe headache**, dizziness or stiff neck

**Breathing difficulty** with pain or trouble breathing, or breathing very quickly and noisily

**Pain**, if severe and not responding to the pain medicine you have at home

**Very pale colour**, particularly affecting the palms, or lips

**Severe abdominal pain**, particularly with increased jaundice or enlargement of the spleen

**Painful erection of the penis** lasting more than 2 hours

**Change in behaviour**, such as appearing confused, drowsy or unable to speak

**Fits, convulsions**, body spasms and loss of consciousness

**Weakness**, particularly if not associated with pain

If you have difficulty reaching your GP, take your child to the nearest Accident and Emergency Department and 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 into hospital because she is unwell or because she 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 her whilst she is in hospital. This person can be a family member or a friend, but they must be 16 years or older. 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 and some of the staff 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. Consider for example:

- Who will take your child to hospital?
- Who will stay with your child in hospital?
- Who will visit your child and how often?
- Who will look after your other children when you are at the hospital?
- What arrangements have you made with your employer if your child becomes ill?
- How will you deal with the cost of visiting the hospital, or loss of income if you are not able to go to work?

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

The key to preparing your child for hospital is to tailor your explanations and activities to your child's age and level of understanding. You know your child best, so you will be able to gauge what she will be able to understand and when.

### Some things that may help you

- Let your child know that you feel the hospital visit, procedure or test is the right thing to do. Children can usually sense how a parent feels so it is important to stay positive in front of your child as they will pick up on your worries/anxieties.
- If your child has something that they find comforting to have close by, it may be useful to bring it to the hospital. For example, she may have a favourite toy, book, game or blanket which makes her feel safe.
- While you are in hospital, keep to as normal a routine as possible. We know this can be hard at times but keeping to regular bedtimes and so on can add a touch of normality to a hospital stay.

- Most importantly, let your child know that it is OK to feel many different ways about coming to the hospital such as curious, worried, angry, scared or frustrated. These are perfectly normal feelings and nothing to worry about.

It's alright and good for your child to let you know if she is worried or in pain. Crying or yelling is allowed; 'being brave' is not always useful. Encourage your child to talk about how she is feeling.

### Common reasons for hospital admission

Your child may be admitted to hospital for:

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

### Pain relief

You will probably be able to manage painful episodes 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 she 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 she has already taken and at what time. Your child may know the best painkillers for her and which ones helped in previous painful episodes.

Some parents are concerned about using very strong painkillers, such as morphine, because they can be associated with drug dependency (when 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 painful episode 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 of 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 their parents will 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, she can control (within safe limits) the amount of painkiller she 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 6–8 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 she has some control over the pain and will make her 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 best to manage your child's painful episode. The team may advise on other ways to help your child cope with the pain, such as relaxation, which can be used with the medications. The play specialist can help you practice some of the relaxation and distraction methods with your child. (See page 111 for details of the Children's Self-help Manual.)

### Receiving other medications

Antibiotics are commonly given, particularly if your child has a high temperature.

The temperature may be caused by a painful episode 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 she is vomiting or not able to take fluids orally, the antibiotics may be given directly into the vein rather than by mouth.

### Intravenous fluids (drip)

It is important for children with sickle cell disease to drink regularly. If your child is not able to drink normally, she may need to be given fluids through a vein (intravenous), which is commonly called a drip, to ensure that she gets sufficient fluids to prevent or correct dehydration.

### Medical investigations

Sometimes children are admitted into hospital for medical investigations that cannot be done on an outpatient visit, such as investigating why a child snores and whether there is a medical reason that needs to be treated. Depending on the nature of the investigation the admission is usually overnight or for a few days.

### Blood transfusion

If your child needs regular planned blood transfusions, this is usually done on a day attendance but sometimes it may need to be done overnight. If this is the case it will be discussed and planned with you.

### 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 as well.

If your child goes back to school before finishing a course of antibiotics, it may be necessary for her 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 organisations listed on page 112.

You should expect:

- To be given an expected date of discharge within 24 hours of admission. The expected date of discharge will be reviewed on a regular basis to check that sufficient progress is being made to enable her to be discharged on the proposed day. The expected date may be adjusted during your child's admission depending on developments

in the care she is receiving. You should be informed of any developments as they happen

- To have follow-up arrangements in place prior to discharge. If your child needs a follow-up appointment, this will be made before you leave or the hospital will write to you with details of the appointment
- To be given any training or information to enable you to care for your child once she has been discharged

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

- 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 she 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 or after parvovirus infection (see page 47)
- Acute chest syndrome: to improve oxygen delivery to the body and reduce sickling in the lungs
- Stroke or other problems with the brain
- To prevent strokes happening: if the Transcranial Doppler (TCD) scan shows abnormal findings which suggest that there is a high risk of stroke
- To prevent problems before an operation
- To treat severe or worsening damage to organs such as the lungs, kidneys or liver

### Types of blood transfusion

**Top-up or simple transfusion:** one or more bags of blood are given through the veins in the arms over a few hours.

**Exchange transfusion:** blood is transfused into the child through one vein and removed at the same time through a different vein. This allows the amount of sickle haemoglobin in the blood to be reduced quickly without making the blood too thick. This is sometimes done when someone is very unwell or having a stroke.

**Long-term blood transfusion:** this involves giving regular blood transfusions every 3–4 weeks to keep the sickle haemoglobin low and improve anaemia. Regular transfusions eventually lead to iron overload, which can cause other health problems if it is not managed properly.

### Blood transfusion before an operation

Children with sickle cell disease may need surgical operations for problems unrelated to their sickle cell disease, such as removing the tonsils. Other operations which are fairly commonly performed in children with sickle cell disease include removing the spleen and removing the gall bladder. Sickle cell disease can complicate operations and blood transfusions are usually given before surgery to reduce this risk. The blood transfusion will usually be given a few days before the operation, as a day visit, in order to increase the haemoglobin.

### Blood safety

Every precaution is taken to ensure that the blood that your child is given is the correct match for her 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 107) being made, which might cause problems with future transfusions.

If you have strong objections to blood transfusions, it is useful to discuss this with your doctor, nurse specialist or religious leader before an emergency occurs.

**Reducing the risk of infection:** all donated blood in the United Kingdom is tested for HIV, hepatitis and other infections. Blood donors are also questioned to make sure they are not likely to have infections which might affect the blood.

### Religion and blood transfusion

Some people have strong personal or religious reasons for not wanting their child to have a blood transfusion. Medical staff will always try to respect your beliefs if at all possible. However, sometimes a blood transfusion is essential to save a child's life, such as when severe anaemia develops or a stroke occurs. By law medical staff must do everything possible

to save your child's life, and in extreme circumstances this may involve giving a blood transfusion against your wishes.

If you have strong objections to blood transfusions, it is useful to discuss this with your doctor, nurse specialist or religious leader before an emergency occurs.

### 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 organs such as the liver and heart, so it is important to take medicine to remove the excess iron. This is called iron chelation and is usually started about a year after starting regular transfusions. There are three medicines

which are used commonly to remove excess iron, and these are sometimes given together. The main problem with iron chelation is that people sometimes forget to take the drugs reliably, and dangerous levels of iron build up. It is very important to take these iron chelator medicines every day, as prescribed.

- **Desferrioxamine (Desferal):** desferrioxamine has been used for more than 30 years and is a safe and effective drug. The main problem is that it cannot be taken by mouth and has to be injected, either under the skin or into a vein. Usually a small needle is inserted under the skin and the medication is given slowly overnight using a small pump. Ideally it should be given at least five nights a week. Although desferrioxamine works well, the injections often make it difficult to use reliably.
- **Deferiprone (Ferriprox):** deferiprone can be taken by mouth, as a tablet or liquid three times per day. It is not often used in sickle cell disease, but may occasionally be used if there is a lot of iron in the heart or other treatments are not working. The main side-effect is to make the white cell count low, so weekly blood tests are recommended.
- **Deferasirox (Exjade):** deferasirox is probably the most commonly used iron chelator in the UK. It is taken once per day as tablets which can be crushed. It is found to be safe and effective. The main side-effect is stomach pains and kidney function has to be monitored carefully.

## 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, children who keep making lots of fetal (baby) haemoglobin (HbF) as they get older are at lower risk of some complications, such as acute pain. Some medicines and treatments are used to reduce some of the problems caused by sickle cell disease.

### Hydroxycarbamide (hydroxyurea)

Hydroxycarbamide is the only drug which has been shown to improve symptoms in sickle cell disease, and can also help reduce the risk of stroke. It might also help prevent damage to organs, such as the kidneys and lungs and reduce symptoms of anaemia. It has been used in people with sickle cell disease for about 30 years, and some studies suggest that taking it for a long time will also increase life expectancy. Hydroxycarbamide is taken once a day and blood tests are needed every 2 months to monitor its effects.

Hydroxycarbamide acts directly on the bone marrow, where blood is made, to reduce the amount of sickle haemoglobin. There were some concerns that this could damage the bone marrow and increase the risk of conditions like leukaemia. Long-



Picture drawn by Miai

term studies have not shown any evidence of this and hydroxycarbamide seems very safe, even in young children. It is also possible that hydroxycarbamide might reduce fertility in later life, although again there is no strong evidence to suggest this, and it does not seem to be a big problem.

Nearly all children with HbSS and HbS/ $\beta^0$ thalassaemia benefit from taking hydroxycarbamide, and it should be discussed with you in the first year of your baby's life, with a view to starting to take it.

In children, blood is usually taken from the back of the hand or in the arm. The doctor or nurse will explain the tests to you and why they are needed.

Currently in the UK more than 50% children with sickle cell disease take hydroxyurea, particularly those with the following complications:

- Serious episodes of pain occurring more than twice per year
- Reduced school attendance due to sickle cell disease
- Acute chest syndrome needing treatment with blood transfusion or intensive care
- Damaged arteries in the brain

- Large amounts of protein in the urine
- Severe anaemia (haemoglobin less than 70g/l)
- Other evidence that the sickle haemoglobin is causing damage to the body

### Bone marrow transplantation (BMT), (stem cell transplantation)

This is the only cure for sickle cell disease at the moment. Bone marrow transplantation (BMT) involves taking bone marrow or blood stem cells from a 'matched' brother or sister and giving it to the child with sickle cell disease. Strong chemotherapy has to be given first to remove the child's own bone marrow so that the donor stem cells will be able to grow successfully in the child's bone marrow.

Whilst this treatment has been done successfully on many patients with sickle cell disease, there are several problems.

It is best done using cells from a matched brother or sister when the child is very young, possibly before any complications have arisen and before the severity of their sickle cell disease is known. As the severity of sickle cell disease is so variable, it is difficult to justify a high-risk treatment like BMT in the majority of people with sickle cell disease. A small number of children get very ill and die during the transplant (less than 1%) and there may be several unwanted long-term side effects, including infertility. 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 transplant.

Researchers are working on ways of making bone marrow transplants safer and more widely available, including using donors who are not brothers or sisters. If your child is having very severe complications, you may be offered one of these experimental transplants.

**Although BMT cures a person's sickle cell disease it does not change their genes. When they come to have children they will still pass on the sickle cell gene to their offspring.**

### Gene therapy

Gene therapy involves taking some cells from your child's blood (stem cells), and then correcting the sickle mutation in these cells in the laboratory. Chemotherapy is then given to your child to get rid of the sickle bone marrow, and the new corrected cells are given back. These cells grow into new bone marrow and can potentially cure the sickle cell disease. This is still an experimental procedure and only a small number of children with sickle cell disease in the world have undergone gene therapy. Trials are now starting in the UK for severely affected children, but it is likely that over the next ten years or so this will become an important treatment option for some children.

### New drug treatments

New drugs are being developed to treat sickle cell disease. This usually involves clinical trials in which children are given a drug and monitored very closely. This will often involve some children being given the active drug, and others being given a placebo (inactive drug), to see the side-effects and benefits. In clinic, you may be given information about clinical trials to see if you and your child want to take part. We hope that in the next ten years lots of new treatments will become available.

At the moment, there are three new drugs which may become available for use in the UK, although it is not certain:

- Crizanlizumab: an injection given every few weeks which reduces the number of painful episodes
- Voxelotor: taken by mouth each day and reduces anaemia
- L-glutamine: taken by mouth each day, it may reduce the number of painful episodes

## Medical tests and what they mean

### Blood tests

In children, blood is usually taken from a vein in the back of the hand or in the arm. In babies it may be taken from the heel of a foot. The doctor or nurse will explain the tests to you and why they are needed. To make having blood tests less painful the nurse or phlebotomist may apply a small amount of local anaesthetic cream (Emla) or a cold spray onto your child's skin to make the area numb. The numbness will wear off after a few hours.

It is recommended that children with sickle cell disease have a TCD scan every year from the age of 2 years until 16 years of age.

### Haemoglobin electrophoresis

This test is done to identify and measure the different types of haemoglobin a person has inherited from their parents (see pages 78-84). It does not tell you the amount of haemoglobin in a person's blood; a separate test called full blood count (FBC) will provide this value (see page 60).

This test will also show if there is any fetal haemoglobin (HbF) present (see page 61 for further explanation).

### 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 (carrier) with her sickle cell disease, it will not cause extra problems and can make some complications less common. Finding out if the trait is present is done by DNA analysis using blood obtained from a simple blood test.

### Reticulocyte count

Reticulocytes are young red cells. The reticulocyte count helps to show how well the bone marrow is working. If the reticulocytes are low in number, it means the bone marrow is not working as well as it should be. This occurs following parvovirus infection, and is sometimes called an 'aplastic crisis'. High reticulocyte counts often mean that there is a lot of sickling going on.

### Full blood count (FBC)

FBC is one of the most commonly used tests. It measures all the cells in the blood including: red cells (which carry oxygen), white cells (which fight infection) and platelets (which help to form clots when bleeding). It also measures the haemoglobin level (Hb). This value lets you know how anaemic a person is. In sickle cell disease the normal level of Hb varies between 60g/l and 110g/l. It is important to know and record your child's normal Hb when she is well, because when your child

becomes unwell it is likely to fall. This will help your child's doctor to know whether she might need treatment to increase the haemoglobin, such as a blood transfusion.

### Haemoglobin S (HbS) level

This test is done to find out the percentage of sickle cell haemoglobin in a person's blood. This value is useful if your child is receiving regular blood transfusions. It helps guide the doctor on how much blood to give and how frequently.

### Haemoglobin F (HbF)

This is also known as fetal haemoglobin or baby haemoglobin. This type of haemoglobin is found in large quantities in the foetus and newborn baby. Once a baby is born the level of HbF starts to fall. HbF remains relatively high in babies but is almost completely replaced by the haemoglobin inherited from parents by the time a baby reaches the age of 1 year.

Everybody, including adults, has some HbF in their blood, about 1%, even those who do not have sickle cell disease. In the case of sickle cell disease this can vary between 1% to about 25%. In general, higher HbF provides some protection against sickling and reduces the risks of some sickle cell-related complications. If your child is started on hydroxycarbamide then your child's HbF is likely to increase. This will be checked regularly to monitor its effects.

### Blood chemistry

A number of tests are done to check the level of various substances in the blood. These are particularly used to show how well the liver and kidneys are working, and

to measure levels of calcium and vitamin D. The tests also measure bilirubin, which is the substance that causes jaundice and can make the eyes look yellow as a result of the rapid breakdown of red cells.

### Glucose 6 phosphate dehydrogenase (G6PD) deficiency test

G6PD is an enzyme which protects the red blood cells against chemicals. It is very common for people to inherit low levels of this enzyme along with sickle cell disease, which means the red cells are easily broken down (haemolytic anaemia) leading to jaundice (yellowing of the skin and whites around the eyes).

All babies with sickle cell disease should be tested for G6PD deficiency. If your baby has G6PD deficiency it should not cause many problems, but it is important to avoid giving your child broad beans (also called fava beans) and certain medications, such as certain antibiotics and antimalarials. Your doctor or nurse should discuss this with you and give you an information leaflet.

### Urine test

Urine testing can show whether there is a urine infection, which might need treatment with antibiotics. It is also used to see if there is too much protein in the urine, which can be a sign of early kidney damage.

### X-rays and scans

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

### **Chest X-ray**

This is usually done to look for infection.

### **Bone X-ray**

This is done if there is a history of injury and the bone could be broken. It is sometimes used to look for infection in the bone, but is only useful if the bone has been painful for at least 4 or 5 days. Other bone X-rays are done to look for damage to joints, such as the hip, and to monitor growth.

### **Ultrasound scan of the abdomen**

This can show the size and shape of the kidneys, the liver and spleen, and is sometimes done if someone is getting abdominal pain. It can also show gallstones, which are more common in sickle cell disease and can cause abdominal pain.

### **Echocardiogram**

This test looks at the heart to see how well it is working.

### **CT scan**

This is a type of X-ray which can show details of the inside of the body. It involves exposure to some radiation and is usually only done when there are particular worries about the brain, lungs or abdomen.

### **MRI scan**

MRI scans use a very powerful magnet to show the inside of the body and are particularly useful for looking at the brain and bones. The scan is very safe and does not involve use of needles or exposure to

radiation. An MRI scan can take up to 30 minutes to finish, so young children under the age of 7 years usually need a general anaesthetic to make sure that they lie still.

### **MRA scan**

MRA scans use a powerful magnetic field, radio waves and a computer to evaluate blood vessels and identify abnormalities. This scan is safe and does not use radiation. It may require an injection of contrast material (dye) into a vein to help visualise the blood vessels.

### **Transcranial Doppler scan**

Transcranial Doppler (TCD) is a safe, non-invasive ultrasound to check the circulation in a person's head. It measures the speed and direction of blood flow in the blood vessels of the brain.

It is recommended that children with sickle cell disease have a TCD scan every year from the ages of 2 years, until 16 years of age. This scan helps to identify any narrowing or blockages in the blood vessels supplying blood to the brain. Narrowing or blockages in the vessels shows an increased risk of stroke. This occurs in a small number of children with sickle cell disease. If the scan shows any abnormality it is usually repeated a week or so later. If the abnormality persists, doctors will discuss how to prevent further damage to the arteries. This will usually involve starting regular blood transfusions or medication. Other scans or special tests may be requested and these will be explained to you by your doctor.





# Living with Sickle Cell Disease

Giving your child space to grow and become independent is an important part of helping her to cope with her sickle cell disease.

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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 emotions when they are first told that their newborn child has sickle cell disease. It is similar to grief; denial, anger, sadness, and fear. All of these are normal.

If you did not know that you and your partner are carriers of sickle or any other unusual haemoglobin 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 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 anymore. 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. 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 will think 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 illness. Do not be alarmed if this happens. Most of the time emotions are beyond our control; we just feel them and have to accept them when they occur. People have different ways of coping with their feelings, but the first important step is to recognise how you feel and go through the experience without feeling guilty.

### Public attitudes 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 and how they respond to you and your child's illness. Many people have little or no knowledge of sickle cell disease, and there are a lot of myths, taboos, wrong beliefs, and stigma. Some people think that it is a condition that affects only black people – this is not true. Although it is more common in black people, it can occur in any racial group and depends on the haemoglobin an individual has inherited from both parents. Some people hold strong cultural or religious beliefs. For example, some people believe that a child has been born with this disease because God is punishing the child's parents or family.

Unfortunately, these myths, cultural beliefs, and stigma from the public, friends, or even family members may lead to a negative attitude towards children with sickle cell disease or to the labelling 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.

These attitudes 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 and help dispel such wrong beliefs.

### Your emotional wellbeing

Tackling your feelings about your child's sickle cell disease directly is the best way to deal with your emotional wellbeing. Do not deny or avoid the situation, this will help you feel less anxious, stressed, or sad. Parents can feel a loss of control, for example, when they are unable to help their child feel better, or are uncertain about what will happen in future. It's important to manage your feelings.

Build a social support network you can rely on to help ease your load, don't be isolated. Also, make time to take care of yourself, both physically and emotionally. Eating well, exercising and having hobbies or other interests can help keep stress in check. Don't feel guilty to take some time for your own health and emotional wellbeing. In the long run, doing so will help both you and your child.

Dealing with your emotional wellbeing is an ongoing process, everybody handles

As you learn more about your child's condition it will become less frightening or worrying.

it in a different way. However, seek professional help when you need it, don't be afraid to say you are struggling.

### Who can help?

It may help to talk about your 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 to 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 and concerns. You can meet other parents at sickle cell support groups listed at: <https://www.sicklecellsociety.org/supportgroups/>.

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 healthcare professional who helps people deal with personal issues and emotional problems affecting their lives. A psychologist is not a psychiatrist.

They will deal with your emotional wellbeing, 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 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 and plan your child's care and where to get help and support when you need it. Most importantly, your child will be developing her own individual personality to become that little person you get to know and love.

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

### **How to tell your child about sickle cell disease**

Talking to your child for the first time about their sickle cell disease is very difficult but it is the best way to move forward. Try and do this as soon as they can understand that they have a chronic illness that they have been born with. Many parents struggle with how to speak to their child about her illness. Be sure you're sharing age-appropriate information. Don't give too much information, but also don't try to hide the facts. If a child overhears a doctor or nurse and doesn't understand what's happening, she may begin to imagine the worst.

Children don't process bad news in the same way as adults do. Young children may not understand right away, so they may not have a strong initial reaction to bad news. Parents may be confused by this. Allow them to ask questions at any time.

### **Your young child with sickle cell disease**

Children with sickle cell disease are often very strong psychologically and emotionally and are able to cope with their illness. Your child's ability to cope

with her illness depends on a number of things, such as the severity of the illness, her personality, your attitude to the illness and your ability to handle your child's and other family members' feelings.

Your child may feel guilty because of her illness and that she may be causing a lot of worry for the family emotionally and possibly financially. 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 6–7 years, when they may realise 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, including her teachers at school.

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 and punishment when they misbehave. Your child has to learn the difference between what is 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

possible parenthood. On the other hand she may be more emotionally mature than her friends.

Sometimes the frequency of painful episodes and other sickle-related illnesses increases at this time because of hormonal changes going on in the body and changes in social lifestyle. Adolescents may find themselves spending more time in hospital, which can be very disruptive to their lives. They may find it hard to keep up with work at school, college or university or to take part in sports and social activities. If your child has frequent episodes of pain, illness and hospitalisation 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. Like any other teenager, your child can find adolescence a distressing time, and she will need a lot more encouragement and support. It is sometimes helpful to talk to someone who knows about and has lived with sickle cell disease and how it can affect a person's life, an older person with sickle cell disease perhaps.

### **Coping with sickle cell disease and pain**

As your child gets older she needs to learn to cope with the illness effectively and to recognise that she can play an active part in keeping herself well, and reducing the chances of getting ill and pain. They should be encouraged to talk about their sickle cell disease, pain and their daily concerns or worries. This may help them feel better.

It is common for children and adolescents to have frequent pain at home. This is usually mild to moderate in intensity but may lead to other problems including low mood, anxiety, considerable loss of schooling, and working time for parents. Some children may choose to keep minor pain to themselves, and feel that it is a bother to others. It is important for older children and adolescents to know how to manage pain at home with appropriate analgesia for the level of intensity. In addition, how they deal with pain has to be looked at since these have been shown to predict how pain is felt, and how health services are used.

Talk to your child about what she is feeling. Parents may be surprised by which aspects of an illness are most difficult for a child.

Children and adolescents should be encouraged to take their mind off illness and pain, with distraction including games, computers, music and television. Also, it is okay for them to have support from a psychologist when they find it

difficult to deal with anything including their feelings, and pain.

### **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 that they will not have the fear of catching it.

#### **How to manage 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 if and when appropriate. Try to share your time equally with all your children, recognising that each child has different needs at different times.

Share how you feel so 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**

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.

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 when you are not there. Otherwise they will worry that they would not know what to do during a painful episode or 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:

- How many children in your family and how old?
- 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 circumstance. 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.

### 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 as stated above. Also, you need to consider the skills that you have to offer employers and the distance between your place of employment and your home. You may be able to negotiate with your employer to have flexible working hours, work part-time, job share or work from home.

#### ***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 or 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 work 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 you have already and help you consider whether you should continue in the same job or career. It may be possible to do the

Share how you feel so your family know that you all have needs, including you and your partner.

same job but working more flexibly. The Skills Funding Agency may be able to help and 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 part-time or full-time.

Health/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 person 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 what you want to do with your life. Talk to an independent financial adviser who will help you explore your personal situation and reach a decision that suits you 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 111. 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 Citizens Advice Bureau (CAB).

There are a number of useful practical guidebooks from reputable bookshops which may help you plan and organise your family life so that having a child with sickle cell disease does not create undue stress for you and the rest of the family.



## 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. As a result they 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 Pensions, job centre plus and the Pension Service's websites.

The Citizen's Advice Bureau (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 means and non means-tested benefits.

For information on most government services across the UK access <https://www.gov.uk>.

## Universal Credit

Universal Credit replaces 6 benefits (**housing benefit, income support, job seekers allowance, employment and support allowance, working tax credit and child tax credit**), with a single monthly payment if you're out of work or on a low income. It is the main benefit in some parts of the UK. The best way to find out what is available in an area is to use a free, independent calculator. Visit: <https://www.gov.uk/universal-credit>.

## Benefits Calculators

Use an independent benefits calculator to find out:

- what benefits you could get
- how to claim
- how your benefits will be affected if you start work

<https://www.gov.uk/benefits-calculators>

## Links to the calculators

- entitled to  
[https://www.entitledto.co.uk/?utm\\_source=BADviser&utm\\_medium=referral&utm\\_campaign=GovUK](https://www.entitledto.co.uk/?utm_source=BADviser&utm_medium=referral&utm_campaign=GovUK)
- Turn2us  
[https://benefits-calculator.turn2us.org.uk/AboutYou?utm\\_source=BADviser&utm\\_medium=referral&utm\\_campaign=GovUK](https://benefits-calculator.turn2us.org.uk/AboutYou?utm_source=BADviser&utm_medium=referral&utm_campaign=GovUK)

- Policy In Practice

<https://www.betteroffcalculator.co.uk/free>

### Benefits for families

Families can apply for a range of benefits. Follow the link for a full list.

<https://www.gov.uk/browse/benefits/families>

### Website for advice on employment issues

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

### Specific Benefits

#### Disability Living Allowance (DLA) for children under 16

<https://www.gov.uk/disability-living-allowance-children>

#### Disability Living Allowance helpline

Telephone: 0800 121 4600  
Textphone: 0800 121 4523  
Disability Benefit Centre 4  
Post Handling Site B  
Wolverhampton WV99 1BY

#### Disability Living Allowance (DLA) for children (claim form)

<https://www.gov.uk/government/publications/disability-living-allowance-for-children-claim-form>

#### Personal Independence Payment (PIP) for those 16 to 64 years of age

PIP replaced DLA in 2013 for people with disabilities or long term health conditions,

age 16 to 64. It is a point based entitlement and claimants may be asked to take part in an assessment to ascertain how the disability or illness affects their ability to start and complete daily and mobility activities. The outcome of the assessment would determine the level of award <https://www.gov.uk/pip>.

#### Carer's Allowance

Carers may be eligible for Carer's Allowance if they, the person they care for and the type of care they provide meets the qualifying criteria. <https://www.gov.uk/carers-allowance>.

#### Disabled Students' Allowances (DSA)

<https://www.gov.uk/disabled-students-allowances-dsas>

#### Sources of further information

[www.gov.uk](http://www.gov.uk)  
[www.turn2us.org.uk](http://www.turn2us.org.uk)  
[www.citizensadvice.org.uk](http://www.citizensadvice.org.uk)  
[www.disabilityrightsuk.org](http://www.disabilityrightsuk.org)  
[www.moneyadviceservice.org.uk](http://www.moneyadviceservice.org.uk)  
[www.cpag.org.uk](http://www.cpag.org.uk)  
[www.capuk.org](http://www.capuk.org)  
<https://england.shelter.org.uk/>  
[www.carers.org](http://www.carers.org)  
<http://sicklecellwork.dmu.ac.uk/>

#### Carers UK

[www.carersuk.org](http://www.carersuk.org)  
[info@carersuk.org](mailto:info@carersuk.org)

#### England, Scotland and Wales

Telephone: 0808 808 7777

#### Additional information about benefits and support for carers

<https://www.nhs.uk/conditions/social-care-and-support-guide/support-and-benefits-for-carers/>

Some local sickle cell & thalassaemia centres (see page 112) also offer welfare advice and or are able to direct you to local areas for support.

### Pregnancy and future births

If you already have a 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 any 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 we 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 contact a sickle cell and thalassaemia centre, see page 112. If there is no specialist centre in your area talk to your GP.

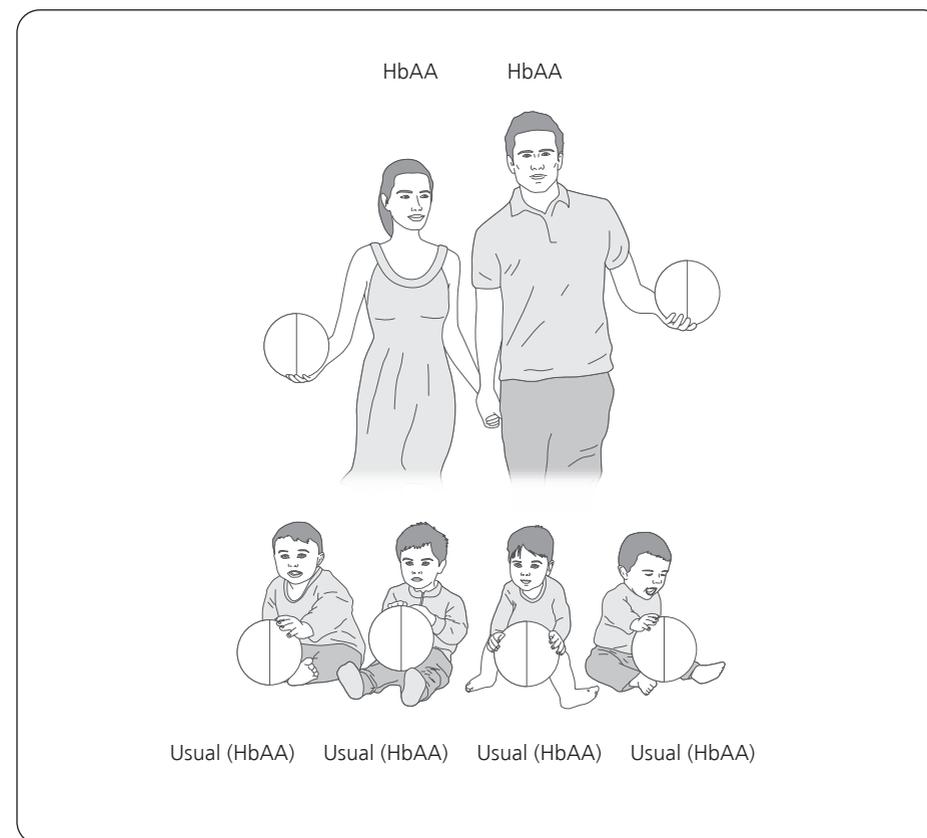
### What can our child inherit?

Your child will inherit any one of your two haemoglobins as well as one of your partner's two haemoglobins, which one of each is not predictable, it is a matter of chance. Below are some examples and the chances are the same for each and every pregnancy.

### Example 1

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

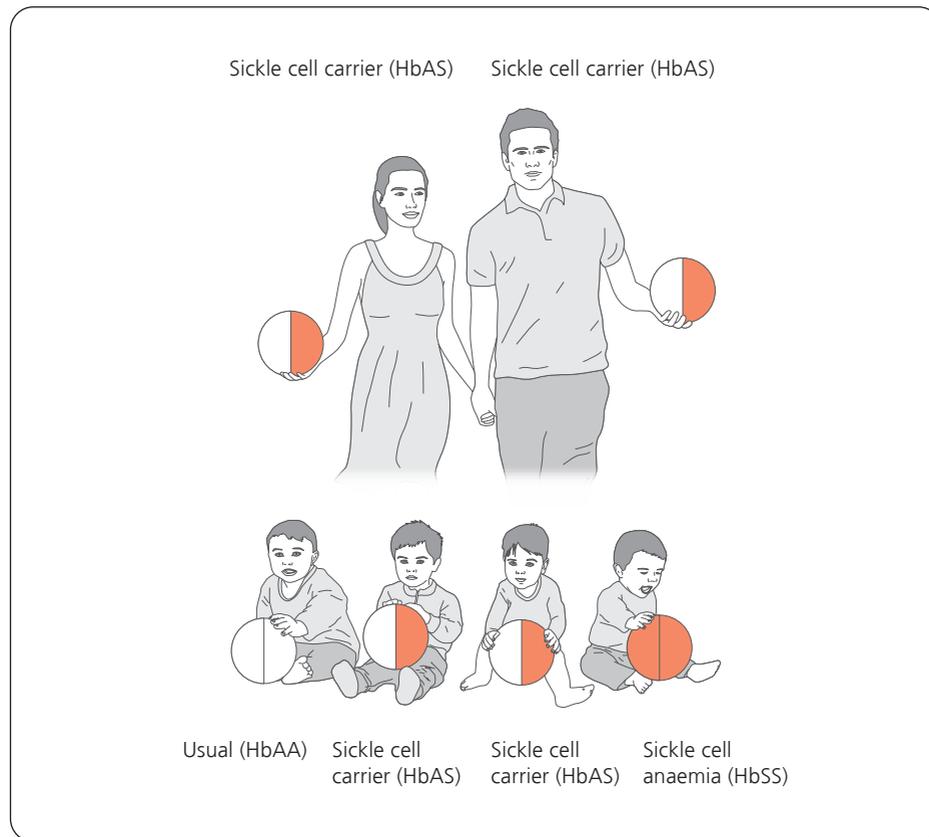
There is a 4 in 4 chance (100%) that their child will inherit the usual and most common haemoglobin combination (HbAA).



### Example 2

If both parents are sickle cell carriers of haemoglobin S (HbAS), **EACH and EVERY time they are expecting a child:**

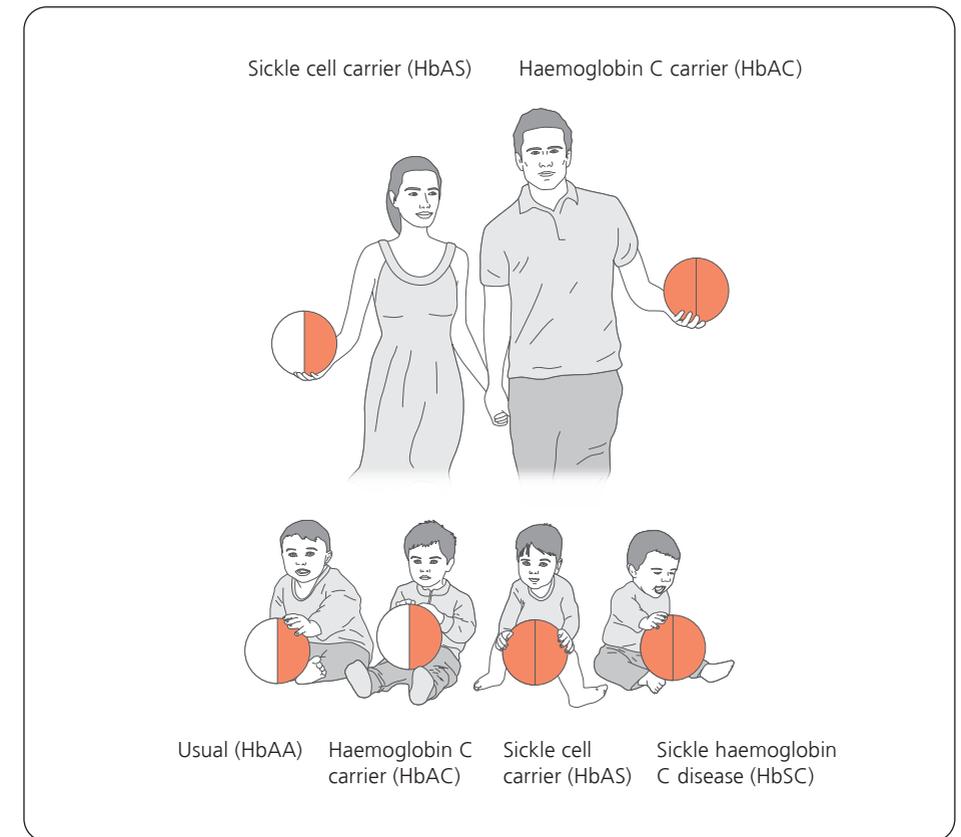
There is a 1 in 4 chance (25%) that their child could inherit the usual haemoglobin (HbAA), a 2 in 4 chance (50%) that their child could be a sickle cell carrier (HbAS) and a 1 in 4 chance (25%) that their child could inherit sickle cell anaemia (HbSS).



### Example 3

If one parent is a sickle cell carrier (HbAS) and the other parent is a carrier of haemoglobin C (HbAC), **EACH time they are expecting a child:**

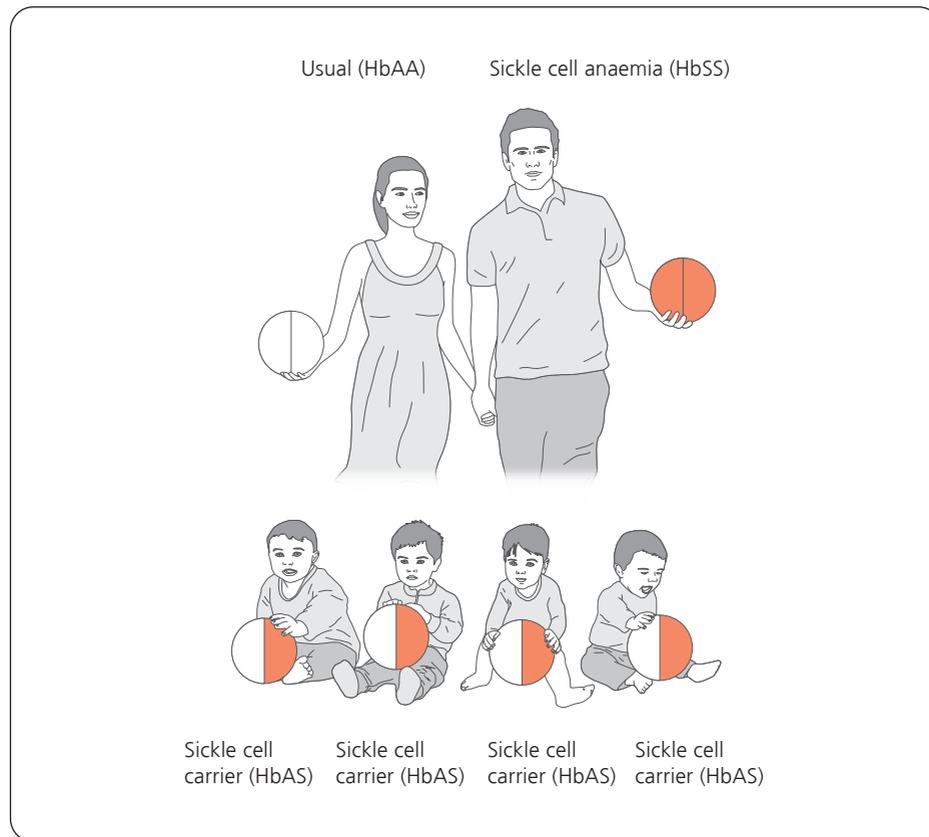
There is a 1 in 4 chance (25%) that their child could inherit the usual haemoglobin (HbAA), a 1 in 4 chance (25%) that their child could be a haemoglobin C carrier (HbAC), a 1 in 4 chance (25%) that their child could be a sickle cell carrier (HbAS) and a 1 in 4 chance (25%) that their child could inherit 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:**

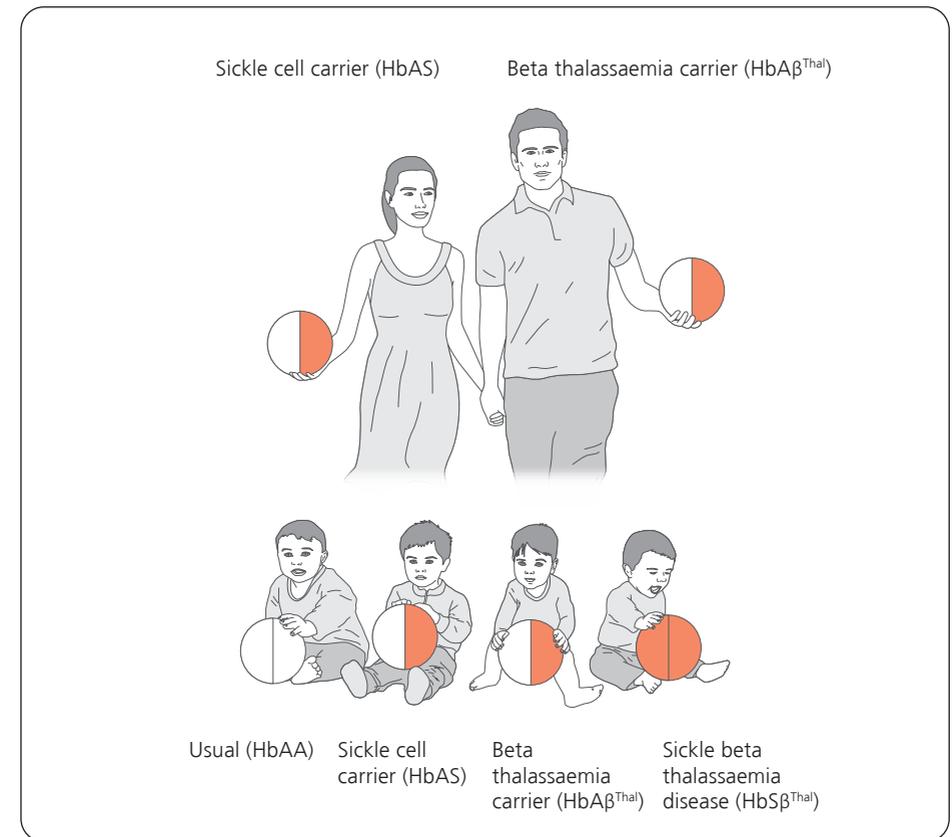
There is a 4 in 4 chance (100%) their child will be a sickle cell carrier (HbAS).



#### Example 5

If one parent is a sickle cell carrier (HbAS) and the other parent is a beta thalassaemia carrier (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 be a sickle cell carrier (HbAS), a 1 in 4 chance (25%) that their child could be a beta thalassaemia carrier (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>).

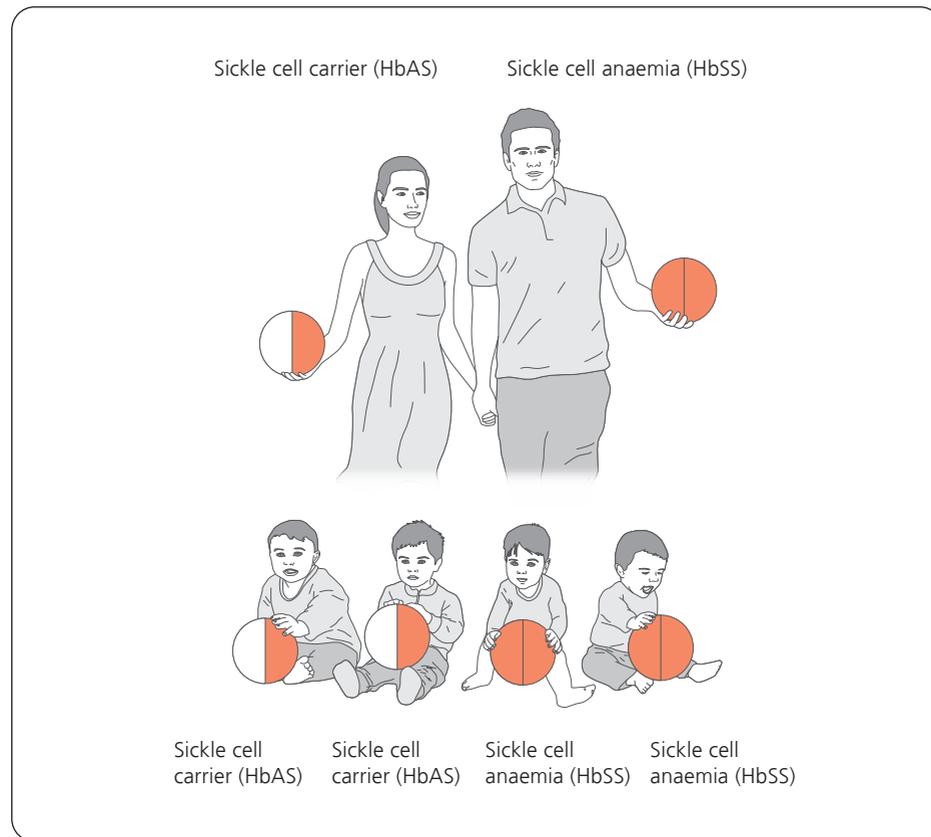


### Example 6

If one parent is a sickle cell carrier (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 be a sickle cell carrier (HbAS) and a 2 in 4 chance (50%) that their child could inherit sickle cell anaemia (HbSS).

**Remember that in all these examples, the four possible chances are the same for each and every pregnancy.**



### Can a pregnancy be tested before birth?

There are a number of ways of testing to find out if the foetus growing in the womb has a genetic condition. These tests are called pre-natal diagnosis (PND). They will tell you which haemoglobin type the foetus 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 two types used commonly: chorionic villus sample (CVS) and amniocentesis. Most women will attend a regional fetal medicine centre in a hospital as an outpatient. The procedure may require local anaesthetic but not always.

### Types of prenatal diagnosis (PND) tests

#### Chorionic villus sample (CVS)

This test is offered between the 11th and 13th week of pregnancy. The vast majority are done through the abdomen and a few are done via the vagina. A small piece of the placenta (this is what will develop to become the afterbirth later in pregnancy) is taken and sent for testing. The result is usually available within one week.

#### Amniocentesis

This test is offered after the 15th week of pregnancy and can be done at any later stage of the pregnancy. A small

amount of the liquid around the foetus, called amniotic fluid, is taken and sent for testing. The result is usually available within one week. The material for chorionic villus sample and amniocentesis is not taken from any part of the growing foetus. There is an increased risk of miscarriage, of less than 1%, as a result of having these diagnostic tests.

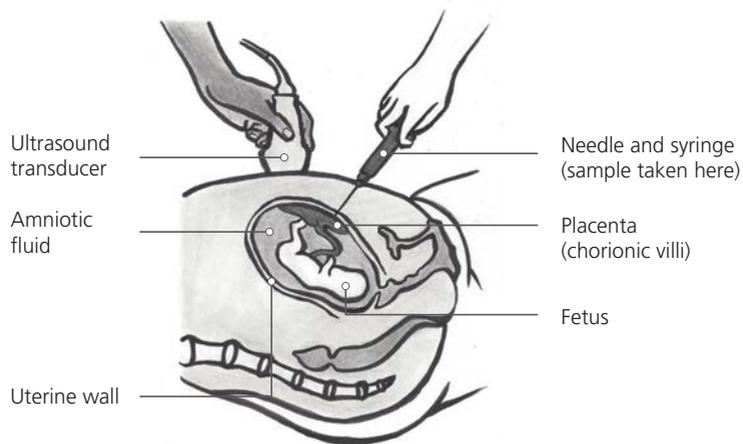
The risk calculation does not take into account the miscarriage rate that can occur in some pregnancies even where no test is done.

When planning to have another baby, if you wish to consider having any of these tests or you want to find out more about them talk to your GP, nurse specialist/counsellor, or someone at any of the specialist centres listed on page 112. The decision to have a pre-natal test rests with you and your partner. The healthcare 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.

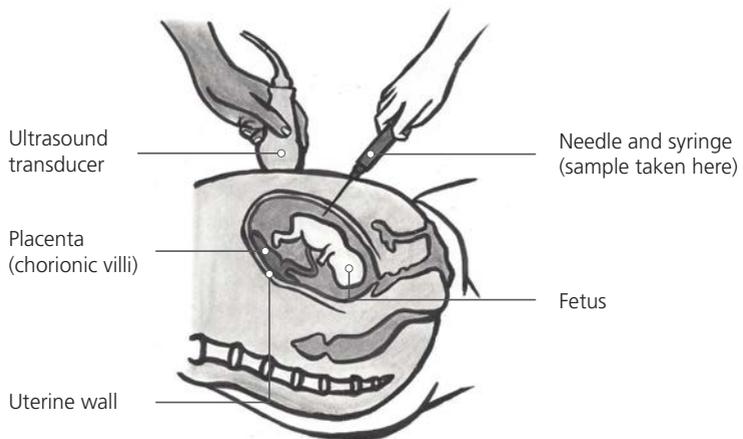
### What if the result shows that the foetus has sickle cell disease?

You may have thought about what you would do if the result shows that the foetus 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

**Figure 5** Chorionic villus sampling (CVS)



**Figure 6** Amniocentesis



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. If you decide to continue the pregnancy you will be offered support right through the pregnancy.

There is an organisation called Antenatal Results and Choices (ARC) who offer additional support to parents who are making choices about an at-risk pregnancy (see page 117).

### What will happen after the baby's birth?

Whether you chose PND or not, or if having had PND you decided not to terminate an affected pregnancy, you will be supported in your decision as you prepare for the arrival of your baby. If you have not had a child with sickle cell disease before this gives you an opportunity to find out as much as you can about sickle cell disease, perhaps talk to parents who have looked after a child with the condition.

It is common practice that where a couple is known to be at risk of having an affected child the baby is tested as soon as possible after birth, and this is done in most hospitals usually on the day of birth

or within a day or two but before the baby goes home.

All babies born in England are tested for sickle cell disease as part of the national routine heel-prick blood test offered on day 5 after birth. This test will show whether the baby has sickle cell disease or not, and, if the result indicates the condition, will be referred for specialist care.

### Pre-implantation genetic diagnosis

Pre-implantation genetic diagnosis (PGD) 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. At the moment PGD can only be offered to couples who are 'at risk' of having a child with sickle cell anaemia (HbSS) or sickle haemoglobin C disease (HbSC). The couple must also meet a number of criteria, for example, be non-/smokers and the woman must not be under or over-weight or over 40 years of age; and they must not have an unaffected child already.

The process involves giving the woman drugs over several months to stimulate her ovaries to produce mature eggs which are then removed and fertilised in the laboratory with sperm obtained from her partner or donor if she wishes to use this alternative. The developing embryo (fertilised egg) is then tested for sickle cell anaemia or sickle haemoglobin C disease, but not for other types of sickle cell disease or other genetic conditions.

If the embryo does not have these sickle cell diseases the fertilised egg is placed in the woman's womb with the aim of establishing a pregnancy.

This process is often referred to as 'in vitro fertilisation' (IVF) or 'test tube baby'. The success rate, sometimes referred to as the 'take home a baby' rate is about 33% in other words about 1 in 3 attempts will result in a baby being born.

IVF has been done for over thirty years and PGD for over twenty years.

Few hospitals offer this service because they need a PGD licence to operate and not all IVF centres will have this. PGD is centrally funded by the NHS therefore if you fulfil the criteria for PGD then you are automatically eligible for up to 3 cycles of PGD funding. If you do not fulfil the funding criteria and want to opt for private funding then PGD costs £12,000 per cycle (in 2021), which includes all the drugs required for the treatment.

When planning to have another baby, if you are interested in PGD discuss this option with your doctor, specialist nurse, or contact one of the specialist centres listed from page 112, you can also get more information from the following website: [www.guysandstthomas.nhs.uk/ourservices/pgd/](http://www.guysandstthomas.nhs.uk/ourservices/pgd/) or <https://www.hfea.gov.uk>.

## Questions parents often ask

### ***How will I know if my child is having sickle cell pain?***

This is the question most commonly asked by parents, especially when their child is a newly diagnosed baby. 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 anyone 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 or not allowing you to touch a part that may be causing her pain, is she refusing to feed or eat? (See page 44 for information about managing illness).

### ***At what age will sickle cell-related health problems start?***

This is unpredictable. A very small number of children will have a sickle-related illness by the time they are 6 months old, others between 6 and 12 months. Others may not experience any symptoms until they become toddlers or older. 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 sickle-related episode for several years, even into adulthood, especially those with milder forms of sickle cell disease, such as HbSC disease.

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

No. 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 in 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 22 for information about diet).

### ***Should I give her iron tablets, iron tonics and vitamins?***

The type of anaemia that your child has is not the same as iron deficiency anaemia. It is advisable NOT to give your child iron supplements or tonics unless it is prescribed by her doctor. Some specialists give children with sickle cell disease daily folic acid – this mineral will do no harm and helps blood production. A simple daily dose of multivitamins will do no harm but check with your specialist doctor or nurse.

### ***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 can be inherited. Although sickle cell is less common in some racial groups it does occur infrequently. For example, it can occur in white northern Europeans.

### ***Is there a cure for sickle cell disease?***

There is a cure for sickle cell disease called 'bone marrow transplantation' or BMT for short. There is also another possible cure called gene therapy but these cannot be offered to all children with sickle cell disease for a number of health reasons. (See page 58 for more detail).

### ***Will my daughter have menstrual periods?***

Most young girls with sickle cell disease may start their menstrual periods a little bit later than their friends, but this is nothing to worry about; they will start eventually. If your daughter has not started her menstrual periods by the age of about 16 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.

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***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 child's GP, practice nurse at the surgery or pharmacist at the chemist will be able to advise you about period pains.

***Will my child be able to have children?***

Yes. Because your child has sickle cell disease it 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 48). In rare cases priapism may cause impotence and treatment may be required to try and correct the impotence in adulthood.

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

In the UK it is very unusual for someone with sickle cell disease to die in childhood, and your child will almost certainly live to the age of 21. Life-expectancy for people with sickle cell disease is improving all the time, and the aim is that your child should live to be old and have a happy, normal life. Sadly, at the moment, sickle cell disease causes far more problems in many African countries, with the majority of patients dying in childhood.

***Will giving my child antibiotics (penicillin) everyday harm her immunity?***

Antibiotics will not harm your child's immune system and are very important. Children with sickle cell disease 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 infections and help prevent death. 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. Talk to your specialist nurse or 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 disease. 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.

***I thought sickle cell protects against malaria, so why do I have to give my child anti-malaria drugs when we go to a malaria 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 milder 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 35 for information about anti-malaria drugs).

***Why is sickle cell pain more common in cold, wet and or windy weather?***

It is not really known, but many people find that they get more episodes of pain when it is cold or windy. If the skin cools down very rapidly, for example after getting caught out in the rain, or if it is a cold windy day and the skin is exposed, the chilling of the skin can cause a painful episode. It may also be that infections with coughs, colds and influenza are more common in winter and these then trigger attacks of pain.

***Can sickle cell disease cause death?***

The complications of sickle cell disease can cause death, although this is unusual. 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, even if you do all the right things to prevent complications, they may still occur and very rarely result in death. It is important for parents to know that it will not necessarily be their fault or anyone else's if their child dies.

***Can my son be circumcised?***

In the UK 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 harmful and illegal.

***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 can be separated out and 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 you were 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 recommend using a parent or other family member's blood for an individual but they can donate to the general blood bank.

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

No. 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 blood which is then given to your child, that blood would still only live

in your child's body for less than about 120 days. (Donated blood cells do not last as long as a person's own blood cells). 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 red blood cells that can sickle and these will eventually replace the blood which was transfused.

***Why does my son have more symptoms of sickle cell disease 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 untrue myths and superstitions surrounding sickle cell disease. If you worry about witchcraft, it may be worth talking to your religious leader or 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. You can also contact one of the voluntary organisations listed on pages 115-116.

***Should I allow my child to go swimming and play sports?***

Yes. Exercise is important to help develop healthy muscles, lungs and heart. Sickle cell disease affects individuals differently. Some people can cope with strenuous exercise like swimming or football; others find this may bring on a painful episode, particularly if swimming in cool water. Always make sure that your child wraps up quickly with a large bath towel after swimming or after a shower to avoid chilling of the body. Take extra care to make sure her hair is dry because babies and children lose a lot of heat from their scalp. It is important not to prevent your child 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 child with sickle cell disease?***

Because you and your partner are carriers of an unusual haemoglobin gene and one or both of you carry the sickle haemoglobin gene, 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 pages 78-84).

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

Yes. Major advances in healthcare, treatments, drugs and better management of sickle cell disease worldwide has helped to reduce some of the complications seen in the past. Some severe complications can cause early death but many people with sickle cell disease can and do live into adulthood and old age.

The type of sickle cell disease a person has can influence whether they develop complications which may have an impact on their lifespan.

***Will it be safe for my child to be in a school, college or other learning environment?***

Yes. It is important for your child to attend mainstream school, to mix with their peers and learn in an environment that will help them to reach their full potential socially and academically.

Many parents worry about infection, school environment, sports and other aspects they feel may pose a risk to their child who has sickle cell disease. Your specialist nurse can help by visiting the school to educate teachers and support staff, and with your involvement develop a 'School Healthcare Plan' to help the school know how to look after your child and what to do if she becomes unwell.

***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 need to encourage her to drink a lot of fluid so as to prevent dehydration which can cause sickling crisis, and this adds to her passing a lot more urine. If your child is over 6 years old and still wets the bed, talk to your doctor or nurse specialist. (See page 20 for more information).

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

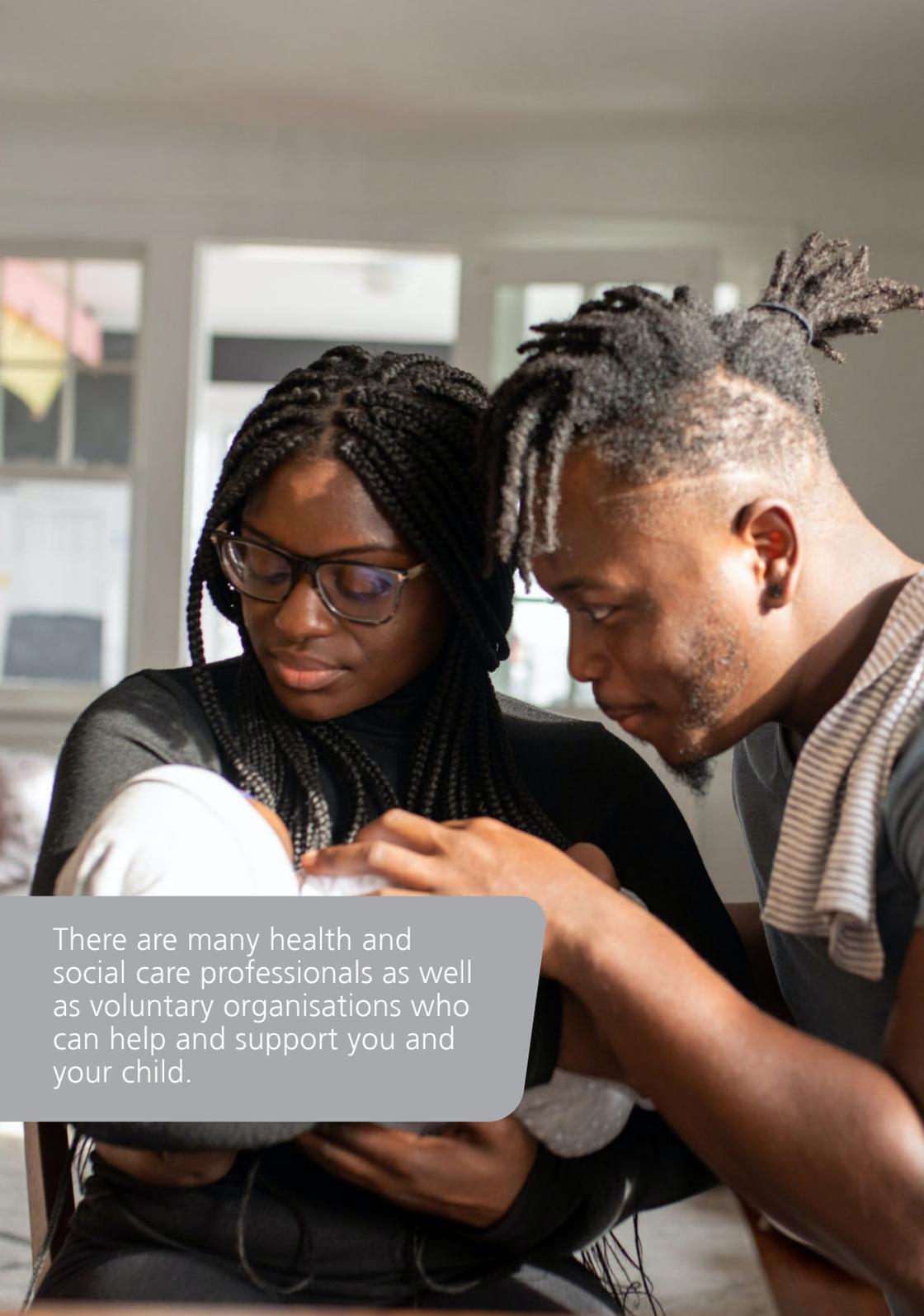
If you believe in God, you may be of the opinion that God gave doctors and nurses the opportunity to train, gain knowledge and skills to be able to care for your child properly. If however, you are worried that perhaps your religious beliefs and what you have been told to do in caring for your child 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 or spiritual 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 importantly, 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 practise your religion irrespective of the type of medical condition your child has.

In most cases the medical treatment being given to your child does not prevent practising your religion. However, your religion should be practiced in a way that does not endanger your child's life.





There are many health and social care professionals as well as voluntary organisations who can help and support you and your child.

# Other information

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Always take your child's red baby book or patient held record with you when you go to see a health professional.

## 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-related pain.

### Audiologist

Audiologists are specialists who check hearing; they are usually based in community centres or hospitals. One of the rare complications of giving desferrioxamine (see page 57) 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 and young people's nurse (paediatric nurse)

Paediatric nurses are specially trained to look after children and young people (from premature birth up to the age of 18) with acute and chronic illness. They are involved in assessing the patient's nursing needs and considering their medical, emotional and family circumstances. They plan and deliver 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, which is 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.

### Clinical Nurse Specialist for children and adolescents

These are paediatric nurses who are specially trained to look after children and young people up to age 18 years. The CNS has additional training, specialist knowledge, skill and experience in looking after children with sickle cell disease.

### 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. It is advisable for your child to see a dentist by the time she is 1 year old.

### General practitioner

Your child's general medical practitioner (GP) needs to know that your child has sickle cell disease, because he/she is responsible for your child's medical care in the community and will refer your child 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. The recommended daily preventive antibiotic is usually prescribed by your GP. Do

remember that if your child is ill, it is always advisable to see your GP first. Do not take your child to the hospital except in an emergency.

If you move to another area, it is important to get your child re-registered as soon as possible with a 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, together with the paediatrician and your GP, usually manage your child's sickle cell disease. Some hospitals employ specialist paediatric haematologists.

### Health visitor

Your health visitor (HV) offers health advice to help your family stay healthy. All children under the age of 5 years are seen by a HV regularly or occasionally, depending on the service provided in your area. They give advice on family health matters, including diet, childhood illness, immunisations, 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 will advise you about your family's

general health and well-being, but most can and do advise on sickle cell and other health issues affecting any member of the family. Some areas of their work overlap with the specialist nurse/counsellor's role. The hospital clinic doctor and nurse specialist 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 baby book or records with you to the clinic. If you are using an electronic baby book your health care professional can be given access to enter relevant healthcare or other information.

### Hospital youth worker

Some hospitals employ 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. Hospital youth workers support young people in their move from child to adult services.

### Ophthalmologist

This is a doctor who checks people's 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. Your child may be referred for regular eye checks or only referred if there is a concern. This depends on the policy of the hospital that is looking after her. The changes in children's eyes can occur as early as 6 years of age especially in children receiving desferrioxamine (see page 57).

#### **Paediatrician (hospital)**

Your child may be looked after by a general paediatrician, a doctor who specialises in looking after children's conditions, or a paediatric haematologist, who is a specialist in blood disorders affecting children. This depends on which doctors are 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 and speech and language therapy, and they communicate 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 on-going 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 phlebotomist is a trained health worker who takes blood and sends 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 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 team, hospital play specialists will do the following: organise daily play and art activities in the playroom or at the child's 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.

### **Psychologist (clinical)**

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 a 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 special tests (neuropsychological assessment) to help determine whether the child needs to be referred to the educational psychologist attached to their school.

### **Psychologist (educational)**

An educational psychologist is a specialist in helping children with schooling and learning difficulties. They 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 areas you will 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, such as immunisations and development, including the information collected since your child was born.

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.

Many boroughs also employ *Specialist Educational Needs Coordinators (SENCO)*, who work alongside schools and healthcare workers to identify the support your child needs and can be offered in the school.

### **Sickle Cell Nurse Specialist / Counsellor (Community)**

Some health districts employ community specialist nurses in some areas they are called counsellors. These are nurses, midwives and health visitors who have had additional training in caring for people with sickle cell and other genetic disorders affecting haemoglobin.

These specialists have training in genetic counselling and can advise on the possible chances of having another child with sickle cell disease. Such specialists may be based in a sickle cell and thalassaemia centre in the hospital or in the community. (See page 112).

### **Social Worker / Welfare Officer**

Social workers help families learn how to 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 and or welfare advisers as part of their team.

### **Sonographer**

A sonographer works in the x-ray department and is specially trained to take specialist images, for example, scanning the brain using a Transcranial Doppler for children with sickle cell disease.

## **National Health Service (NHS) specialist provision**

### **Local Haemoglobinopathy Teams (LHT) and Specialist Haemoglobinopathy Teams (SHT)**

In England there are Local Haemoglobinopathy Teams (LHT) providing sickle cell and thalassaemia services for their local patient population.

NHS England has designated 24 hospitals as Specialist Haemoglobinopathy Teams (SHT). These are responsible for specialist management of sickle and thalassaemia patients in their own hospital and in the LHT hospitals that are linked to them. The national standards state that every patient must be seen by a specialist (SHT) at least once a year and for management of specialist treatments such as blood transfusion and any sickle-related complications.

### **NHS England Haemoglobinopathy Coordinating Centre (HCC) and National Haemoglobinopathy Panel (NHP)**

Some of the SHTs, mostly as collaborative groups within a region, have been designated and commissioned by NHS England to be Haemoglobinopathy Coordinating Centres (HCC) for an identified geographical area.

In 2020 ten sickle cell HCCs and four thalassaemia and rare anaemias HCCs were created in England.

HCCs are responsible for coordination and monitoring specialist care and services provided within the designated region. They provide clinical leadership and ensure all patients have access to quality care and services irrespective of where they live in the region.

The National Haemoglobinopathy Panel (NHP) is commissioned by NHS England. Members of the steering group are taken from the regional HCCs and they are responsible for making decisions about top level specialist treatments, such as who should be considered for bone marrow transplant. The NHP members provide clinical leadership and national direction to ensure services are good, equitable and fair across the whole of England.

For further information about the HCC, NHP and other developments talk to your specialist nurse, doctor or other health and allied care professional.

## Sickle cell and 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. Many more centres, now known as sickle cell and thalassaemia centres, have been set up all over the UK and they provide services for people with and 'at risk' of sickle cell, thalassaemia, G6PD and related conditions. For a list of centres and services in the UK see page 112.

Most centres are funded by the National Health Service (NHS). Some are jointly funded and managed by the NHS, the local authority or a local voluntary organisation. Many of these centres are part of, or work closely with, the hospital haematology department.

The majority of centres are managed by specialist nurses or counsellors who have many years' experience as nurses and may also have midwifery and health visiting qualifications. All of these staff have had specialist training in care and management of people with sickle cell and related conditions.

The team of professionals in a centre varies but often includes:

- Community specialist nurses
- Clinical Nurse Specialists
- Medical doctors
- Social workers/ welfare officer

- Psychologists

Services offered include:

- Screening and genetic counselling
- Health maintenance and ill health prevention counselling
- Support for people with sickle cell, thalassaemia and related conditions
- Education for the general public about these conditions
- Development and distribution of educational resources and materials for patients, professionals and the general public
- Information about support groups and voluntary organisations
- Housing advice
- Social services advice
- Welfare advice
- Health promotion activities and events
- Training and education of health, allied and non-health professionals
- Influencing policy-decision makers (e.g. government health and education ministers)

They also monitor the effectiveness of services provided for people with, or at risk of, sickle cell and related conditions in order to ensure people 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 centre nearest to where you live (see page 112) or one of the voluntary organisations (see page 115) for advice on how to get access to these services. There are no strict geographical boundaries for giving people guidance, advice or information.

## Local patient support groups

Sickle cell and thalassaemia centres and hospitals that provide care for people with sickle cell disease may have patient support groups. These are made up of parents, children, young people, 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 a group in your area, talk to your nurse specialist, health visitor, social worker or doctor or contact one of the national voluntary organisations.

They help new parents get in touch with other parents who have more experience of caring for a child with sickle cell disease. Some take children on fun trips and organise fun events, such as Christmas parties for the children and fundraising dances. The objectives of each organisation vary at a local level. To find out if there is a local support group in your area speak to your hospital or community healthcare team.

## Organisation for Sickle Cell Anaemia Research (OSCAR)

This was the first national voluntary organisation to be established in the UK. It was established in 1975 by a group of parents and people with sickle cell disease. Their aim was to raise funds for research into finding a cure for sickle cell disease. Although the national OSCAR no longer exists there are a few local OSCARs still operating but in a similar way to local support groups.

The members 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.

## National voluntary organisations

### The Sickle Cell Society

The Sickle Cell Society is currently the only national voluntary organisation in the UK and is providing national influence and support. It was established in 1979 by a group of adults with sickle cell disease, parents, carers and interested health and social care professionals. The Society's main mission is to support people living with sickle cell to have the right quality of care and to empower them to achieve their full potential in all areas of life.

The Society raises awareness of sickle cell to the general public and helps educate health, social and other care professionals on how to provide effective and equitable care to people with the condition. They also take part in clinical and social research on sickle cell. They contribute to and influence national health and social policy decisions and have representation as part of The All Party Parliamentary Group (APPG) for Sickle Cell & Thalassaemia.

The Sickle Cell Society organise patient/carer meetings, educational programmes and events to help its members gain knowledge of clinical developments and to discuss health and social policies that may affect services provided for individuals and families affected by sickle cell. They organise fund-raising events and engage local, national and other organisations in order to promote general awareness and support for those with the condition.

A national helpline has been established which offers support and advice for patients, families and the general public. The Society organises an annual summer holiday for children with sickle cell disease, which gives parents a week's break from their caring roles.

### **United Kingdom Thalassaemia Society (UKTS)**

Some people inherit sickle haemoglobin with beta thalassaemia haemoglobin.

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

thalassaemia inherited with other unusual haemoglobins.

### **Membership of support groups and voluntary organisations**

Membership of a support group or voluntary organisation is open to people with sickle cell disease, their families, friends, health and allied care professionals and members of the public who wish to offer their support in promoting the activity of the organisation.

Through these groups families get to know about any new developments and research.

Membership provides an opportunity for affected individuals and families to have an understanding of how to influence the care provided for them and enable them to have a collective voice which is more powerful than the voice of a lone individual and usually more effective in influencing policy decision-makers.

There are local sickle cell support groups and voluntary organisations. They generally provide support to their local community. For more information about these organisations contact them directly (addresses on page 115) or contact one of the sickle cell and thalassaemia centres listed on page 112.

Remember there is strength in numbers and in the collective voice. Support your local support group and/or voluntary organisation and they will be able to support you, your child and those with sickle cell disease.

## **Sickle cell and thalassaemia all party parliamentary group (APPG)**

The All Party Parliamentary Group (APPG) for sickle cell and thalassaemia, aims to influence government policy and ensure appropriate health and social services are provided for people with or at-risk of sickle cell and thalassaemia. There is representation from all the main political parties and the key voluntary organisations.

## **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 foetus in the womb. A small sample of the fluid around the foetus is taken and tested to find out which haemoglobin type the foetus has inherited.

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

**Anaemia** Insufficient red blood cells or 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 viral infection.

### **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 (also see trait)** A person who has inherited one usual haemoglobin and one unusual haemoglobin is said to be a carrier or to have a trait, for example sickle cell carrier.

**Chelation** A method used for removing iron from the body in order to prevent the iron from 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 most commonly used to describe the pain caused by blockage of blood vessels, which is called a 'painful crisis'.

**Chromosome** This is a string-like structure which carries 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 foetus in the womb: a small piece of chorion (which will later develop into the afterbirth) is taken and tested to find out which type of haemoglobin the foetus has inherited.

**Computerised tomography (CT) scan** A picture is taken of tissues in any part of the body in order to detect damage, used especially to detect damage to the brain.

**Dactylitis** This is a complication involving swelling of one finger, several fingers or other parts of the hand; one toe, several toes or other parts of a foot. 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. It is rarely seen in adults.

**Desferrioxamine** A drug used to help the body get rid of excess iron, usually given when a child is on long-term blood transfusions.

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

**Enuresis** Bed wetting.

**Epistaxis** Nose bleeds.

**Foetus** This is the baby developing in the womb before it is born.

**Gene** 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.

**g/l** Grams per litre.

**Haematuria** This is the presence of blood in the urine.

**Haemoglobin (Hb)** Red pigment in the red blood cells which enables the cells to carry oxygen from the lungs to all the body parts to keep 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, it 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. It is not the same as iron deficiency.

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

**In vitro fertilisation (IVF)** Fertilisation 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 bilirubin.

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

**Magnetic resonance imaging (MRI)** 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. Opiate drugs are very useful for relieving moderate to severe pain and are used with care for relieving sickle cell pain effectively. They can be addictive if they are used inappropriately, for example for recreational purposes.

**Osteomyelitis** Infection of the bone.

**Patient-controlled analgesia (PCA)** A small pump is used to deliver pain medication into the body usually through a vein. Medication is given continuously through the PCA pump; where additional pain relief is required the patient presses a button on the pump allowing a small controlled dose of medication to enter the body. This allows the patient to take part in managing their pain effectively.

**Pre-implantation genetic diagnosis (PGD)** A method used with invitro fertilisation to enable an infertile couple to have a child. It is also used for couples who wish to avoid having a child without a named genetic condition, such as sickle.

A woman's egg is fertilised with a man's sperm in the laboratory. A few days later the developing egg is tested and if it does not have the genetic condition the egg is placed in the woman's womb with the hope that that it will mature into a baby.

**Prenatal diagnosis (PND)** A method of testing a foetus in the womb.

**Priapism** A painful, involuntary and persistent erection of the penis caused by sickled red blood cells blocking blood vessels in the penis and stopping blood flow.

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

**Red blood cell** The part of the blood which acts as a storehouse for haemoglobin. Haemoglobin carries oxygen around the body and keeps the body alive.

**Screening** A test performed on large populations to see 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, used in the past for cutting wheat and other crops and is still used in many developing countries. The term is used to describe 'sickled' red blood cells because under a microscope, instead of being round, these cells are often curved like the shape of a farmer's sickle.

**Spleen** A small organ on the left-hand side of the body, 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 (also see carrier)** Carrier of a genetic condition: for example, sickle cell trait (carrier) do not have a disease; the vast majority do not have any symptoms or sickle-related ailments.

**Transcranial Doppler (TCD) scan**

A special scan to see how well blood is flowing through the blood vessels in the brain and help identify those children who are at risk of having a stroke.

**Vaso-occlusion** Blockage of the blood vessels by sickled red blood cells which flow sluggishly in a part of the body. It causes mild to severe pain and is the most common cause of pain in sickle cell disease.

**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 (available from Brent Sickle Cell & Thalassaemia Centre – see addresses list on page 112)

Sickle Cell Society and Public Health England (2019) 3<sup>rd</sup> edition *Sickle cell disease in childhood: standards and recommendations for clinical care*

Can be downloaded at:  
[www.sicklecellsociety.org](http://www.sicklecellsociety.org)

**Useful websites**

- De Montfort University Social Study of Thalassaemia and Sickle cell  
<https://www.dmu.ac.uk/research/research-faculties-and-institutes/health-and-life-sciences/social-study-of-thalassaemia--sickle-cell/social-study-of-thalassaemia--sickle-cell.aspx>
- Sickle Cell & Thalassaemia Screening Programme  
<https://www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview>

- Genetic Alliance [www.geneticalliance.org](http://www.geneticalliance.org)
- Public Health England:  
<https://www.gov.uk/government/organisations/public-health-england>
- 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)
- National Haemoglobinopathy Panel: [www.nationalhaempanel-nhs.net](http://www.nationalhaempanel-nhs.net)

## Sickle cell and thalassaemia centres and services

### Greater London

#### Barking & Dagenham, Havering & Redbridge

##### Sickle Cell & Thalassaemia Service

Department of Haematology  
Ground Floor (Orange Zone)  
Queen's Hospital  
Rom Valley Way  
Romford  
Essex RM7 0AG  
**Tel:** 01708 43 5000 ext 3677  
**Email:** bhrut.cvphaem@nhs.net

#### Brent (Brent, Harrow, Hillingdon, Bedford)

##### Brent Sickle Cell & Thalassaemia Centre

London Northwest University Healthcare NHS Trust  
Central Middlesex Hospital  
Acton Lane,  
London NW10 7NS  
**Tel:** 020 8453 2050/52  
**Email:** LNWH-tr.BrentSickleThal@nhs.net

#### Bromley

##### Community Children's Nursing Team

Phoenix CRC  
40 Masons Hill  
Bromley  
Kent BR2 9JG  
**Tel:** 0300 330 5777

#### Camden & Islington

##### Sickle Cell & Thalassaemia Centre

17a Hornsey Street  
London N7 8GG  
**Tel:** 020 3316 8853/8854  
**Email:** sicklethalcandi@nhs.net

#### Central London Community Healthcare NHS Trust (CLCH) (Hammersmith, Fulham, Kensington Chelsea & Westminster)

##### Sickle & Thalassaemia Service

The Medical Centre  
7E Woodfield Road  
London W9 3XZ  
**Tel:** 0207 266 8892  
**Email:** CLCHT.sicklethal@nhs.net

#### City & Hackney

##### The Sickle Cell & Thalassaemia Centre

457 Queensbridge Road  
Hackney  
London E8 3AS  
**Tel:** 020 7683 4570

#### Croydon

##### Sickle Cell & Thalassaemia Centre

316-320 Whitehorse Road  
Croydon, CR0 2LE  
**Tel:** 020 8251 7229  
**Email:** mhn-tr.sicklethal@nhs.net

#### Ealing

##### Sickle Cell & Thalassaemia Service

Carmelita House  
21-22 The Mall  
London W5 2PJ  
**Tel:** 020 8967 5694  
**Email:** WLM-tr.paedspecialistcommunitynursing@nhs.net

#### Eastham

##### Sickle & Thalassaemia Centre

19 –21 High Street South  
East Ham  
London E6 6EN  
**Tel:** 020 8821 0800  
**Email:** elt-tr.sickleandthal@nhs.net

#### Haringey

##### George Marsh Sickle & Thalassaemia Centre

St Ann's Hospital (part of North Middlesex University Hospital NHS Trust)  
St Ann's Road  
London N15 3TH  
**Tel:** 0208 702 6630  
**Email:** northmid.sicklecellservice@nhs.net

#### Hounslow & Richmond

##### Sickle Cell & Thalassaemia Service

Whitton Corner Health & Social Care Centre  
Percy Road  
Hounslow TW3 6JL  
**Tel:** 0203 771 6092

#### North East London

##### Sickle & Thalassaemia Service

Wood Street Health Centre  
3 Linford Road  
Walthamstow  
London E17 3LA  
**Tel:** 020 8430 7639  
**Email:** Nem-tr.sicklecellservice@nhs.net

#### South East London

##### Sickle Cell & Thalassaemia Centre

Wooden Spoon House  
5 Dugard Way  
London SE11 4TH  
**Tel:** 020 3049 5993  
**Email:** Gst-tr.referralstosickle@nhs.net

#### Wandsworth

##### Wandsworth Sickle Cell & Thalassaemia Service

160 Falcon Road  
London SW11 2LN  
**Tel:** 0330 058 1679  
**Email:** clcht.wandsworthsickle-thal@nhs.net

#### Waltham Forest Haemoglobinopathy Service

##### Sickle Cell & Thalassaemia Service

Wood Street Health Centre  
6 Linford Road  
London E17 3LA  
**Tel:** 020 8430 7639  
**Email:** Nem-tr.sicklecellservice@nhs.net

## Outside London

### Birmingham

#### Sickle Cell & Thalassaemia Service

St Stephens Community Hub  
171 Nineveh Road  
Birmingham B21 0SY  
Tel: 0121 466 3667  
Email: Bchc.sicklecellresults@nhs.net

### Cardiff

#### Sickle & Thalassaemia Centre

Butetown Health Centre  
Plas Iona, Butetown  
Cardiff CF10 5HW  
Tel: 029 20471055 / 029 2183 3283

### Coventry

#### Sickle Cell & Thalassaemia Service

University Hospital & Warwickshire NHS  
Trust  
Clifford Bridge Road  
Walsgrave  
Coventry CV2 2DX  
Email: paediatricaemoglobinopathy@  
uhcw.nhs.uk

### Dudley

#### Sickle Cell & Thalassaemia Service

Children, Young People & Families Division  
Brierley Hill Health & Social Centre – 3rd Fl  
Venture Way  
Brierley Hill  
Dudley DY5 1RU  
Mob: 07766 542721  
Email: BCHFT.Haemoglobinopathy@nhs.  
net

### Essex

#### Sickle Cell & Thalassaemia Service

Grays Health Centre  
Brooke Road  
Grays  
Essex RM17 5BY  
Tel: 0300 300 1521 Option 3

### Gloucester

#### Sickle & Thalassaemia Service

Gloucestershire Hospitals NHS Foundation  
Trust  
Edward Jenner Clinical Unit  
Greater Western Road  
Gloucester GL1 3NN  
Tel: 0300 422 5224

### Hertfordshire

#### Children's Sickle Cell Service

Peace Children's Centre  
Peace Prospect  
Watford WD17 3EW  
Tel: 01923 470680

### Leeds

#### Sickle & Thalassaemia Centre

Haematology Department  
Bexley Wing  
St James University Hospital  
Beckett Street  
Leeds LS 7TF  
Tel: 0113 843 4350

### Leicestershire

#### Sickle & Thalassaemia Service

Haemoglobinopathies Hub  
Ground Floor  
Osborne Building  
Leicester Royal Infirmary  
Leicester LE1 5WW  
Tel: 0116 258 6081

### Luton

#### Sickle Cell & Thalassaemia Service

Cambridgeshire Community Services-Luton  
The Poynt  
2-4 Poynters Road  
Luton LU4 0LA  
Tel: 0333 4050070  
Email: ccs-tr.lutonhaemoglobinopathy.nhs.  
net

### Manchester

#### Sickle & Thalassaemia Centre

352 Oxford Road  
Manchester M13 9NL  
Tel: 0161 274 3322  
Email: Mst-manchestersicklecell@nhs.net

### Milton Keynes

#### Sickle & Thalassaemia Service

Milton Keynes Community Health Services  
Children with Complex Needs Team  
Trust Headquarters  
Hospital Campus  
Standing Way  
Milton Keynes MK6 5NG  
Tel: 01908 724511 Option: 4

### Wolverhampton

#### Sickle Cell & Thalassaemia Service

Paycare House  
George Street  
Wolverhampton WV2 4DX  
Tel: 01902 444 076/77  
Email: info@sctsp.org.uk  
Website: www.sctsp.org.uk

## Patient support groups and voluntary organisations

### Sickle Cell Society

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

### UK Thalassaemia Society

19 The Broadway  
Southgate Circus  
London N14 6PH  
Tel: 020 8882 0011  
Email: office@ukts.org / admin@ukts.org  
Website: www.ukts.org

### **OSCAR Birmingham**

OSCAR Birmingham Ltd  
22 Regent Place  
Jewellery Quarter  
Birmingham B1 3NJ  
Tel: 0121 212 9547  
Email: [admin@oscarbirmingham.org.uk](mailto:admin@oscarbirmingham.org.uk)

### **OSCAR Sandwell**

120 Lodge Road  
West Bromwich  
West Midlands B70 8PL  
Tel: 0121 525 0556  
Email: [Oscar-sandwell@hotmail.org.uk](mailto:Oscar-sandwell@hotmail.org.uk)  
Website: [www.oscarsandwell.org.uk](http://www.oscarsandwell.org.uk)

## **Other useful contacts**

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

73 Charlotte Street  
London W1T 4PN  
Tel: 020 7631 0285  
Website: [www.arc-uk.org](http://www.arc-uk.org)

ARC provides support for women and couples making choices about an at-risk pregnancy considering such issues as 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  
Agincourt Road  
London NW3 2NU  
Tel: 020 7284 1234  
Website: [anthonymolan.org](http://anthonymolan.org)

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

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

2A Garret Road  
Thornton Heath  
Surrey CR7 8RD  
Tel: 0203 757 7700  
Email: [info@aclt.org](mailto:info@aclt.org)  
Website: <https://aclt.org>

This organisation was set up by the parents of Daniel DeGaille, a black child who developed leukaemia and could not find a matched donor because there were few black people on the register. The couple campaigned to encourage black people to become bone marrow donors so that it would 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.

## National Newborn and Children's Standards

<b>(SCT-S08): Sickle cell and thalassaemia screening: reporting new-born screen-positive results to parents</b>			
<b>Standard 1</b>	<b>DEFINITION</b>	<b>PERFORMANCE MEASUREMENT</b>	<b>NOTE</b>
	<p>Number of new-born infants with screen positive results for whom parents receive results by 28 days of baby's age</p> <p><i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b></p> <p>New-born infants born within the reporting period with screen-positive results</p> <p>Specified conditions to be detected in new-born screening are: HbSS, HbSC, HbS/β thalassaemia (S/β+, S/β0, Hb S/Lepore), HbS/DPunjab, HbS/E, HbS/OArab, HbS/HPFH, HbS with any other variant and no HbA, and other clinically significant haemoglobinopathies likely to be detected by chance in the newborn screening programme for example, beta (β) thalassaemia major, HbE/β thalassaemia and β thalassaemia intermedia.</p> <p>Carrier results need to be given to parents by six weeks after testing but these are excluded from this standard.</p>	<p>Acceptable: 90%</p> <p>Achievable: 95%</p>	<p>New-born screening extends to infants under aged one year who have not been tested since birth, for example, those newly migrated to England.</p> <p>Detection of thalassaemia is not part of the new-born screening programme but we expect beta (β) thalassaemia major to be detected and the same standards for communicating results to parents and enrolment into care applies</p>

<b>(SCT-09): Sickle cell and thalassaemia screening: timely follow up, diagnosis and treatment of newborn infants with a positive screening result</b>			
<b>Standard 2</b>	<b>DEFINITION</b>	<b>PERFORMANCE MEASUREMENT</b>	<b>NOTE</b>
	<p><b>Number of new-born infants:</b></p> <p>a) with clinically significant results who are seen by a paediatrician by 90 days of age; and</p> <p>b) with insignificant results who are discharged by 90 days of age</p> <p><i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b></p> <p>New-born infants with screen positive result born within the reporting period</p> <p>Specified conditions to be detected in new-born screening are: HbSS, HbSC, HbS/β thalassaemia (S/β+, S/β0, Hb S/Lepore), HbS/DPunjab, HbS/E, HbS/OArab, HbS/HPFH, HbS with any other variant and no HbA, and other clinically significant haemoglobinopathies likely to be detected by chance in the newborn screening programme for example, beta (β) thalassaemia major, HbE/β thalassaemia and β thalassaemia intermedia.</p> <p>Exclusions include: infants born outside of England who are over age one year before migrating to England to live, infants who die or move abroad before 90 days of age</p>	<p>Acceptable: 90%</p> <p>Achievable: 95%</p>	<p>Detection of thalassaemia is not part of the programme but we expect beta (β) thalassaemia major to be detected and the same standards for communicating results to parents and enrolment into care applies</p>

<b>Standard 3</b>		
<b>Timeliness of penicillin prophylaxis</b>	<b>DEFINITION</b>	<b>PERFORMANCE MEASUREMENT</b>
	<p>Number of infants with SCD offered penicillin (or equivalent antibiotic) prophylaxis by age 90 days <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b></p> <p>The number of infants born with SCD and who are eligible* to have prophylactic antibiotic</p> <p>* Infants with HbSC disease are excluded because the evidence is only available for giving penicillin prophylaxis to children with HbSS and HbS/β0 thalassaemia. However, most centres will offer penicillin prophylaxis to children with HbSC disease because the spleen of children with HbSC may not function properly and their immunity may not be sufficient at a later age</p>	<p>Acceptable: 95% Achievable: 99%</p>
		<p><b>NOTE</b></p> <p>If a child's parents refuse to give their child the recommended prophylactic medication, even after advice and support to understand the importance of this medication, their refusal must be recorded in the child's medical records.</p>

<b>Standard 4</b>		
<b>Coverage of pneumococcal immunisation at 2 years</b>	<b>DEFINITION</b>	<b>PERFORMANCE MEASUREMENT</b>
	<p>Number of children with SCD given PPV (Pneumovax ) at 24– 27 months <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b></p> <p>Children born with SCD aged 24– 27 months</p>	<p>Acceptable: 95% Achievable: 99%</p>
		<p><b>NOTE</b></p> <p>If the parents refuse to give their child this recommended vaccination this must be recorded in the child's medical records.</p> <p>The hospital paediatric service is responsible for monitoring that this vaccine has been given irrespective of where the vaccine is given including if given at the GP surgery.</p>

Coverage of Transcranial Doppler (TCD) scanning		
DEFINITION	PERFORMANCE THRESHOLDS	NOTE
<p><b>Standard 5</b></p> <p><b>Definition 1</b> Number of children with HbSS and HbS/<math>\beta</math>0 thalassaemia who have their first TCD at aged 24 and 36 months <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b> Children with HbSS and HbS/<math>\beta</math>0 thalassaemia aged 24 and 36 months</p> <p><b>Definition 2</b> Number of children with HbSS and HbS/<math>\beta</math>0 thalassaemia who are aged 3 to 16 years who have had a TCD test in the last 12 months <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b> Children with HbSS and HbS/<math>\beta</math>0 thalassaemia aged 3 and 16 years registered with the service</p>	Acceptable: 99%	<p>If the child's parents refuse for the child to have a TCD test this must be recorded in the child's medical records.</p> <p>The service must also record other reasons why the TCD was not done, for example, technical difficulty in doing the test or other means of testing e.g. if child is having regular MRI scan</p>

Coverage of hydroxycarbamide (hydroxyurea) therapy		
DEFINITION	PERFORMANCE MEASUREMENT	NOTE
<p><b>Standard 6</b></p> <p>Number of children with HbSS and HbS<math>\beta</math>0 thalassaemia aged 9 to 42 months where there is documented evidence of a discussion about hydroxycarbamide <i>Expressed as a percentage</i></p> <p>Number of children with HbSS and HbS<math>\beta</math>0 thalassaemia aged 9 to 42 months</p> <p>Number of children with HbSS and HbS<math>\beta</math>0 thalassaemia aged 2 to 16 years prescribed hydroxycarbamide <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b> Children with HbSS and HbS<math>\beta</math>0 thalassaemia aged 2 to 16 years</p>	<p>Acceptable (for children aged 9 months to 42 months): 99%</p> <p>Acceptable (aged 2 to 16 years): not yet determined</p>	Hydroxycarbamide is only licensed for use in children over the age of 2 years

<b>Standard 7</b>		
<b>Coverage of children identified through the screening programmed subsequently registered on the national haemoglobinopathy registry (NHR)</b>		
<b>DEFINITION</b>	<b>PERFORMANCE THRESHOLDS</b>	<b>NOTE</b>
<p>Number of infants with SCD registered on the new-born outcomes system where data has been pulled through to the National Haemoglobinopathy Register (NHR) <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b> Infants with SCD registered on the new-born outcomes system</p>	<p>Acceptable: not yet determined Achievable: not yet determined</p>	<p>From 2019, all infants identified as having SCD will be referred from the newborn screening laboratory to paediatric care using the newborn outcomes system. Data can be pulled through to the NHR when key data and parental consent are recorded on the newborn outcomes system</p>

<b>Standard 8</b>		
<b>Coverage of children who have had an annual review</b>		
<b>DEFINITION</b>	<b>PERFORMANCE THRESHOLDS</b>	<b>NOTE</b>
<p>Number of children with SCD aged 1 to 16 years who have started having an annual clinical review <i>Expressed as a percentage</i></p> <p><b>Who is included in measurement?</b> Children with SCD aged 1 to 16 years</p>	<p>Acceptable: 85%</p>	<p>None</p>

**Adapted from:**

Sickle Cell Society (2019) Sickle Cell Disease in Childhood: Standards and Recommendations for Clinical Care

London: Sickle Cell Society

[https://www.sicklecellsociety.org/wp-content/uploads/2019/11/SCD-in-Childhood\\_Final-version-1.pdf](https://www.sicklecellsociety.org/wp-content/uploads/2019/11/SCD-in-Childhood_Final-version-1.pdf)

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