

Patient information from the BMJ Group

Sickle cell disease

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Sickle cell disease

Sickle cell disease is a condition you're born with. Your blood doesn't carry oxygen as well as it should, and blood cells sometimes stick together and block blood vessels. If you have sickle cell disease, you may feel well most of the time. But you'll sometimes get pain in a part of your body.

Sickle cell disease can cause life-threatening problems. You'll probably need regular medical treatment. But many children and adults cope with this condition successfully. It doesn't usually stop them getting on with their lives.

We've brought together the best research about sickle cell disease and weighed up the evidence about how to treat it. You can use our information to talk to your doctor and decide which treatments are best for you.

What is sickle cell disease?

Sickle cell disease is a condition that affects your blood. Red blood cells carry oxygen around your body. But if you have sickle cell disease, these cells don't work properly. They can change from a disc to a sickle shape, which causes small blood clots.

Sickle cell disease is sometimes called **sickle cell anaemia**. That's because it causes a condition called **anaemia**, which can make you feel very tired.

Sickle cell disease affects a chemical in your body called **haemoglobin**. It's found in red blood cells. It's the haemoglobin in your red blood cells that carries oxygen around your body when you breathe. If you have sickle cell disease, your body makes a kind of haemoglobin that doesn't carry oxygen as well as it should.

Sickle cell disease and your genes

Sickle cell disease is a condition that you are born with. It's caused by an abnormal **gene** that can be passed down from your parents.^{[1] [2]} You get the disease only if you inherit this abnormal gene from both parents.

You inherit two genes that tell your body how to make haemoglobin, one from your mother and one from your father. A normal gene tells your body how to make normal haemoglobin, called **haemoglobin A**. An abnormal gene tells your body how to make sickle

Sickle cell disease

haemoglobin, or **haemoglobin S**. If you inherit haemoglobin S genes from both your parents, you get sickle cell disease.

Red blood cells that contain haemoglobin S don't work as well as ordinary red blood cells. Red blood cells are normally disc-shaped. They look a bit like a ring doughnut. They can move freely through your blood vessels. But red blood cells that contain haemoglobin S can change into a sickle, or crescent, shape. These sickle cells can get stuck in your small blood vessels. This stops blood getting to part of your body and causes pain.

Sickle cell trait

If you inherit one normal gene and one abnormal gene, you don't get sickle cell disease. But you do have something called **sickle cell trait**. This means that you still carry the gene, and can pass it on to your children. If your partner also carries the abnormal gene, your children could be born with sickle cell disease. If you and your partner both have sickle cell trait, you have a 1 in 4 chance of having a baby with sickle cell disease each time you're pregnant. ^[3]

There are other kinds of faulty haemoglobin genes. If you inherit a sickle cell gene and another kind of faulty gene, you can get a different type of sickle cell disease.

You can have a blood test to find out if you have sickle cell disease or sickle cell trait. ^[2] In the UK, most babies are tested for sickle cell disease soon after they're born. ^[4] If you're a parent, ask the doctor to explain exactly what the test results mean. If the test is positive, make sure your doctor tells you whether your child has sickle cell disease or just one abnormal gene (sickle cell trait).

Sickle cell crises

People with sickle cell disease can get severe pain. These attacks are often called **crises**. If your red blood cells don't have enough oxygen, they change from soft round discs into hard, brittle sickle shapes. These sickle-shaped red blood cells get stuck in your small blood vessels. This can cause pain.

Babies don't usually start to get attacks of sickle cell pain until they're 3 to 6 months old. ^[5] Another type of haemoglobin (called **fetal haemoglobin**, or **haemoglobin F**) protects babies in the womb and while they're very young.

We don't know for certain what brings on an attack of pain. But these things may play a part: ^[3]

- Infections
- Lack of oxygen
- Not drinking enough fluids (getting dehydrated)

Sickle cell disease

- Stress (such as major surgery or childbirth)
- Getting cold.

You may get more sickle pain if you're pregnant, especially during the last three months before the birth. ^[6]

Malaria (a condition spread by mosquito bites) can cause attacks of sickle cell pain. In tropical Africa, malaria is the most common trigger for pain. ^[7]

What are the symptoms of sickle cell disease?

Pain is the most common symptom of sickle cell disease. Attacks of pain happen to almost everyone with sickle cell disease at some point in their life. The pain can last for hours or days. It's sometimes only mild, but it can be extremely bad.

Normal **red blood cells** are disc-shaped. They look a bit like a ring doughnut. They can move freely through your blood vessels. If you have sickle cell disease, your red blood cells can change into a sickle, or crescent, shape. These sickle cells can get stuck in your small blood vessels. This stops blood getting to part of your body, and causes pain. You can get pain in any part of your body.

Some people get pain only every few years. But other people get pain quite often. ^[5] ^[8]
The pain in sickle cell disease is sometimes described as throbbing, sharp, or gnawing. ^[6]

If you're looking after a child who has sickle cell disease, it's important to learn how to spot the signs that they're in pain. Younger children can't always explain very well what they're feeling or where something hurts.

Other symptoms

Feeling very tired

Red blood cells carry oxygen around your body. Sickle cells can't carry oxygen as well as normal red blood cells. And they don't last as long. Normal red blood cells live about 120 days, but sickle cells live from only 10 to 20 days. ^[9] Having sickle cell disease means that your blood has less haemoglobin than normal, which is called **anaemia**. This means that your blood can't carry enough oxygen around your body. This means that you:

- Get tired easily
- Get breathless
- Have a fast heartbeat.

Sickle cell disease

Anaemia can get worse if you get an **infection**. It can also get worse if you have a problem with your **spleen**. Red blood cells can get stuck in your spleen so they can't travel round your body. This is called a **sequestration crisis**. You should see a doctor if you suddenly feel a lot worse than usual. If you have a child with sickle cell disease, make sure they see a doctor if they seem worse than usual or get tummy pain.

Infections

If you have sickle cell disease, you're more likely to catch infections, especially when you're young. This is partly because sickle cell disease can damage your spleen, which is part of your body's system for fighting infections.

Swollen hands and feet

Inflammation of the bones of the hands and feet can make them red, swollen, and painful. It's common in young children. It's known as **hand and foot syndrome**.

Jaundice

Your eyes or skin may look slightly yellow, even if you are well. This happens because your red blood cells get broken down more quickly than normal.

Sickle cell trait

If you have sickle cell trait, you won't get any of the symptoms of sickle cell disease. But you may need extra oxygen if you have a **general anaesthetic** for an operation. And you're advised against some sports where your oxygen supply may be less, such as scuba diving or climbing very high mountains. ^[2] To read more about sickle cell trait, see [What is sickle cell disease?](#)

How common is sickle cell disease?

Sickle cell disease is most common among people whose families come from Africa.

People whose families come from Mediterranean countries, the Caribbean, the Middle East, and Asian countries also can be affected. ^[10]

About 1 in every 2,000 babies born in England has sickle cell disease. ^[11]

Between 10 in 100 and 30 in 100 people living in tropical parts of Africa carry the sickle cell gene. ^[7] Between 1 in 100 and 2 in 100 people born each year in Africa probably have sickle cell disease.

What treatments work for sickle cell disease?

Sickle cell disease is serious, but lots of people live with it successfully. You should aim to live as normal a life as possible.

Sickle cell disease

There are things you can do to prevent the pain and other health problems sickle cell disease can cause. You should be ready to act straight away if you or your child becomes ill.

- Children with sickle cells are more likely to get **infections** . So doctors recommend they take an **antibiotic** called penicillin regularly. Doctors also recommend using **vaccines** to prevent **pneumonia** and other infections.
- Taking a drug called hydroxyurea regularly may prevent sickle cell pain and some other health problems.
- Some people with sickle cell disease are at a higher risk of having a **stroke** . You can have routine brain scans to check on your risk. If a scan shows you might have a stroke, there are treatments that can help. Some children may need to have regular **blood transfusions** to prevent a stroke.
- If you get mild or moderate pain, you may be able to cope at home by taking painkillers and drinking more fluids. But if you get a high temperature or severe pain, you should go to hospital for strong painkilling drugs. You might also need treatment with oxygen and antibiotics, and you might need to have fluids put into your body through a drip (also called an IV or an **intravenous infusion**).
- If you get severely **anaemic** (so your blood can't carry enough oxygen), you might need a blood transfusion.

We've looked at two kinds of treatments. Some treatments try to prevent the problems that sickle cell disease can cause. And there are treatments that can relieve the pain caused by sickle cell disease.

- [Treatments for pain caused by sickle cell disease](#)
- [Treatments to prevent complications of sickle cell disease](#) .

There are also things you can try to help yourself. To read more, see [Helping yourself with sickle cell disease](#) .

Treatment Group 1

Treatments for pain caused by sickle cell disease

There are several kinds of medicines that relieve pain. Doctors have guidelines about which drugs to use for sickle cell pain. They follow a three-step ladder for relieving mild, moderate, and severe pain. ^[19]

If your child has sickle cell disease, it's important to learn how to spot the signs that they're in pain. Younger children can't always express themselves very well or describe their symptoms clearly. For example, they might just go quiet if they're in pain.

Sickle cell disease

The treatments we look at below can be used for children with sickle cell disease as well as adults. Talk to your doctor if you're worried about getting the right dose or choosing the right kind of painkiller for your child.

Treatments for pain caused by sickle cell disease

Usual treatments for pain

- [Step 1: mild pain](#)
- [Step 2: moderate pain](#)
- [Step 3: severe pain](#)

Other treatments for pain

- [Extra fluids](#)
- [Oxygen](#)
- [Blood transfusion](#)
- [Piracetam](#)

Treatment Group 2

Treatments to prevent complications of sickle cell disease

We've looked closely at the research and ranked the treatments into categories, according to whether they work.

Treatments to prevent complications of sickle cell disease

Treatments that work

- [Regular penicillin to prevent infections in children](#)

Treatments that are likely to work

- [Hydroxyurea](#)
- [Anti-malaria drugs](#)

Treatments that work but whose harms may outweigh benefits

- [Regular blood transfusions for children at high risk of stroke](#)

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Treatments that need further study

- [Vaccines to prevent infections in children](#)

Other treatments

We haven't looked at the research on these treatments in as much detail as we've looked at the research on most of the treatments we cover. (To read more, see Our method.) But we've included some information because you may have heard of them or be interested in them.

- [Bone marrow transplants](#)
- [Bottle blowing](#)

What will happen to me?

It is possible to get severe pain and a lot of complications. But lots of people live with sickle cell disease and don't get many painful attacks. And there are several effective treatments.



You or your child may go for years without having any problems from sickle cell disease.

There are some serious health problems that can affect children and adults with sickle cell disease. If you or your child has the disease, it may be worrying to read about these problems. But remember that each individual is affected differently.

Sickle cell disease

Here are some of the health problems that sickle cell disease can cause. We've tried to say how common these problems are, but the research doesn't always tell us.

- Severe infections: Some people get infections caused by bacteria . Severe infections such as pneumonia , meningitis , and septicaemia are common, especially among young children.^[12] It used to be that about a third of children with sickle cell disease got a serious infection before they were three years old.^[13] But this number is much lower today, as many children are now treated with antibiotics . (See [Treatments to prevent complications of sickle cell disease](#) .)
- A stroke: A stroke can happen when a blood vessel in your brain becomes blocked (to read more, see our information on [Strokes](#)). About 1 in 10 children with sickle cell disease have a stroke.^[14] A brain scan can be done to see if your child is at risk.^[15]
- Acute chest syndrome: This is a complication that affects your lungs. It can be life-threatening. You may get a temperature, a cough, chest pain, have difficulty breathing and your anaemia may get worse. About one-half of people with sickle cell disease get acute chest syndrome at some point.^[16] Younger children are more likely to get it.^[17] It's one of the most common reasons for someone with sickle cell disease needing to go to hospital.^[16]
- An enlarged spleen or liver: This happens when blood suddenly pools in your spleen or liver . You can become anaemic and get low blood pressure . Doctors call this a **sequestration crisis**. If you get an enlarged spleen, you may need an operation to remove it.
- Aplastic crisis: This means that your bone marrow stops making red blood cells . It can happen if you get an infection. You may need a blood transfusion to treat an aplastic crisis.
- Organ damage: Blood vessels blocked by sickle cells can cause damage to your organs. So can the infections you might get. You may get damage to your kidneys , spleen, liver, and lungs. This can make you feel ill, and it can even be life-threatening.
- In men, a painful, long-lasting erection: This is known as **priapism**. You might find it embarrassing, but it's important to get treatment. If you don't get treated, this condition can damage your penis.
- Joint pain: You may get more aches and pains.
- Eye problems: These are caused by abnormal blood vessels at the back of your eye.

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- Leg ulcers: You can get a leg **ulcer** when a wound doesn't heal well.

People with sickle cell disease also tend to weigh less, on average, than other people, and they usually go through **puberty** two or three years later.

No one can predict what will happen to someone with sickle cell disease. But, on average, people with sickle cell disease don't live as long as people who don't have the condition. If you want to find out more, see [Life expectancy for people with sickle cell disease](#) . But bear in mind that these numbers are averages, based on a large group of people. They may not apply to you or your child.

When to get help

It's important you get medical attention for the problems that sickle cell disease can cause. See a doctor if you get: ^[9]

- A temperature, especially if it lasts a long time
- Pain that doesn't go away if you take ordinary painkillers
- Abdominal pain that lasts a long time
- Pain or difficulty breathing
- Chest pain
- Trouble speaking
- Changes in your vision
- Weakness or numbness in your hands or feet
- Headaches that don't go away
- An erection that lasts a long time when you're not aroused.

Support groups

Sickle cell disease is a serious condition. But lots of people live with it successfully. It might help to talk to other people who've had experiences similar to yours. You can get more information from the Sickle Cell Society (<http://www.sicklecellsociety.org>).

Treatments:

Step 1: mild pain

In this section

Sickle cell disease

Your doctor may advise you to try an over-the-counter painkiller if your pain is mild. You may be able to cope yourself, at home. There's not much research on how well these drugs work for sickle cell pain. But you could try paracetamol or a type of painkiller called a nonsteroidal anti-inflammatory drug (NSAID).^[19]

- Paracetamol. People often take paracetamol to help with mild pain and fever. Paracetamol doesn't usually cause side effects. But you should be careful not to take too much, as an overdose can damage your **liver**. This damage can be bad enough to kill you.
- A type of painkiller called a nonsteroidal anti-inflammatory drug (**NSAID**). Ibuprofen is an NSAID that can be bought over-the-counter. Brand names include Nurofen. You can also get own-brand ibuprofen from supermarkets and pharmacies. A drawback of ibuprofen and other NSAIDs is that they can irritate the lining of your stomach. This may cause stomach **ulcers** or bleeding in your stomach. Taking high doses of some NSAIDs every day for a long time can increase your risk of a **heart attack** or **stroke**. This isn't likely to be a problem if you take an NSAID for a short time to treat pain.

Both of these drugs can be used for children. You can get them as syrups, such as Calpol. These are easier for young children to take.

Aspirin is a common painkiller, but it should not be given to children under 16.^[23] It can cause a dangerous problem called **Reye's syndrome**.

Step 2: moderate pain

In this section

Your doctor may give you or your child a weak opioid drug, such as codeine.^[19] Opioid drugs are strong painkillers. Or you may be prescribed a stronger nonsteroidal anti-inflammatory drug (**NSAID**). Doctors often prescribe a combination of painkillers.^[24]

- Codeine belongs to a group of drugs called opioids. You need a prescription for most opioids. These drugs can get less effective if you take them for a while. Codeine combined with paracetamol is often used. It's called co-codamol. Codeine shouldn't be given to children aged 12 and younger.^[25]
- Stronger NSAIDs can be taken by mouth or by an injection into your muscles.

Ketorolac and diflunisal are both strong NSAIDs. We found five small studies (called randomised controlled trials) that looked at using these drugs to treat sickle cell pain.^[26] ^[27] ^[28] ^[29] ^[30] They were compared with two opioid painkillers called pethidine and morphine.

Sickle cell disease

There wasn't enough evidence to say which type of painkiller worked best.^[31] And we need more research to be sure whether taking an NSAID on top of another strong painkiller can improve relief from pain.

NSAIDs have side effects. You can get stomach pains, diarrhoea, skin rashes, headaches, and dizziness.

Taking high doses of some NSAIDs every day for a long time can increase your risk of a heart attack or stroke. This isn't likely to be a problem if you take an NSAID for a short time to treat pain.

Very rarely, breastfeeding babies can get serious problems if their mother is taking codeine.^[32] There isn't a problem for most mothers, but a small minority of women absorb codeine much faster than normal. This means more of the drug gets into their breast milk, which can cause side effects for the baby. If your baby is sick, reluctant to feed, or sleeps more than usual, stop taking codeine and see your doctor straight away. If you become very sleepy yourself, it's also a good idea to talk to your doctor.

Step 3: severe pain

In this section

If you or your child gets very bad pain, your doctor may recommend a strong opioid painkiller, such as morphine. You'll have to go to hospital.^[19]

Morphine and diamorphine are the most commonly used drugs for bad sickle cell pain. You can take opioids by mouth or as an injection.

The research shows taking morphine by mouth works just as well as having an injection.^[33] We also found two small studies (randomised controlled trials) that looked at a system where people could press a button to release a dose of painkiller.^[34] ^[35] People who controlled their own dose of painkillers had just as much relief from pain as people who were given a dose by a nurse or doctor at regular intervals.

Almost one-half of the people in the studies we looked at had side effects. These included a fever, itchiness, nausea, vomiting, and constipation.

Some painkillers may not work as well if you use them every day. And it's possible to become addicted to strong painkillers such as morphine. But this isn't common for people with sickle cell disease.^[36]

Another painkiller called pethidine isn't used much to control severe sickle cell pain. There's a risk of seizures (fits) and breathing difficulties if you take this medicine.^[37]

Extra fluids

In this section

Sickle cell disease

Doctors may give you extra fluids either as a drink or from a drip (also known as an IV or intravenous infusion). They'll do this if you haven't got enough fluids in your body (you're dehydrated). But there's no good research to show whether giving extra fluids helps with sickle cell pain.

Oxygen

In this section

If you haven't been getting enough oxygen, your red blood cells can change from being disc-shaped to sickle-shaped. Sickle cells can stick together and block your blood vessels, which causes pain. So you may need extra oxygen if the oxygen levels in your blood are low. Oxygen treatment is always recommended if you have acute chest syndrome. Acute chest syndrome is a dangerous problem with your lungs. It can cause difficulty breathing. (To learn more, see [What will happen to me?](#)) However, we don't know if giving oxygen treatment will help your pain. ^[38]

Blood transfusion

In this section

You may need a blood transfusion if you have acute chest syndrome, a stroke, or severe anaemia. Most people with sickle cell disease will need a blood transfusion at some point. ^[39] But we don't know whether a blood transfusion will help your pain in a sickle cell crisis.

Piracetam

In this section

We found a review of three studies of a drug called piracetam in sickle cell disease, in which 169 people took part. Piracetam is known to have effects that reduce the formation of blood clots. However the studies weren't very good and didn't provide any convincing reasons to use piracetam in sickle cell disease. ^[40]

Regular penicillin to prevent infections in children

In this section

Sickle cell disease makes people more likely to catch infections, such as pneumonia (an infection of the lungs). Young children with sickle cell disease are less likely to get infections if they take penicillin regularly. Doctors recommend taking penicillin throughout childhood. ^[41]

Penicillin is a type of drug called an antibiotic. It kills the bacteria that can cause infections. In the studies we looked at, some children took penicillin tablets twice a day. Other children had penicillin injections every month.

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One summary of the research (a **systematic review**) looked at more than 800 children under the age of 5.^[42] Children who took penicillin were less likely to get infections like pneumonia.

- About 4 in 100 children got pneumonia if they took penicillin regularly.
- About 9 in 100 children got pneumonia if they didn't take penicillin.

But we found one study (a **randomised controlled trial**) that looked at children aged 5 or older.^[43] Among these older children, those who continued to take penicillin twice a day were just as likely to get ill as those who did not. So taking antibiotics regularly might benefit younger children only.

You can get mild side effects from taking antibiotics, but they're not common with penicillin.^[44] None of the children in the studies got serious side effects.

Some children are **allergic** to penicillin, and get a bad reaction if they take it. If your child is allergic, your doctor may suggest they take a similar drug called erythromycin instead.

Hydroxyurea

In this section

Hydroxyurea is a drug that helps prevent attacks of sickle cell pain and reduces the need for hospital treatment and blood transfusions. It may also prevent acute chest syndrome, which is a dangerous problem with your lungs (to learn more, see [What will happen to me?](#)).

Hydroxyurea was originally used to treat some kinds of cancer. It also seems to make sickle-shaped **red blood cells** more flexible. This may mean they're less likely to block your blood vessels and cause pain. The brand name is Hydrea.

Some people take hydroxyurea for a long time.

We found two summaries of the research (**systematic reviews**).^[45] One, which looked at children, found that hydroxyurea increased haemoglobin levels, reduced the number of times children had to be admitted to hospital, and reduced how often children had a pain crisis.^[46]

The second summary looked at adults with sickle cell disease.^[45] It found that people taking hydroxyurea had higher haemoglobin levels, and were much less likely to be admitted to hospital. Hydroxyurea also reduced the number of painful attacks in adults.

The research also shows that adults taking hydroxyurea were at less risk of getting acute chest syndrome and of needing a blood transfusion.^[45] But we need more research to know if taking hydroxyurea also lowers your chance of having other problems, such as a **stroke** or an enlarged **liver**.

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Hydroxyurea can have side effects. It was originally made to kill cancer cells, and can also damage some healthy cells, such as [white blood cells](#) .

In one of the studies we looked at, almost 8 in 10 people taking hydroxyurea got a condition called **neutropenia**.^[45] This is when your blood has a low number of a type of white blood cells called **neutrophils**, which fight infections. But no one in the study went on to get an infection. Other studies have found a reduction in levels of another type of white blood cells, called **leukocytes**, resulting in a condition called **leukopenia**.^[47]

Side effects reported in other studies include severe anaemia, a lower level of blood platelets (thrombocytopenia), rashes, and nail changes.^[48] There is also a weak link between the use of hydroxyurea and an increased risk of superficial skin cancer. This possible long-term side effect of hydroxyurea is uncommon, and may or may not turn out to be caused by treatment.

Some people in studies got hair loss, a rash, or an upset stomach.^[45] But we don't know whether these problems were caused by hydroxyurea. We need more research to be sure.

Anti-malaria drugs

In this section

Malaria is a disease that's passed on by mosquitoes. It can trigger sickle cell pain and increase the risk of children dying. Doctors recommend you take drugs to prevent malaria if you live in or visit a tropical country.

There's not much research on whether taking anti-malaria drugs helps prevent problems caused by sickle cell disease. The studies that have been done are small.^[49] But because malaria can be dangerous if you have sickle cell disease, doctors usually recommend this treatment if you live in or visit a tropical country.^[7]

There are several different malaria drugs, and the side effects will depend on which ones you take.^[44] To learn more, see [Malaria prevention](#) .

Regular blood transfusions for children at high risk of stroke

In this section

About 10 in 100 children with sickle cell disease will have a stroke.^[50] This happens when a blood vessel in the brain becomes blocked by sickle cells that have stuck together. If a brain scan shows abnormal blood vessels in your child's brain, your doctor may recommend having blood transfusions every few months. This increases the number of normal cells in your child's blood.

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A summary of the research (a [systematic review](#)) found that children having blood transfusions every three to five months were less likely to have a stroke than children who didn't. ^[51] Without transfusions, 11 in 100 children had a stroke. This fell to just 1 in 100 children if they had blood transfusions.

But regular blood transfusions can cause problems. Some children get too much iron in their body, which can damage their heart, liver and other organs. Their immune system might also start attacking the new blood cells from the transfusion. Your doctor will help you weigh up the risks and benefits of this treatment to decide if it's right for your child.

Vaccines to prevent infections in children

In this section

A type of bacteria called pneumococcal bacteria can cause illnesses such as [pneumonia](#) and meningitis. Children with sickle cell disease are more likely to get these infections, so doctors recommend they have a vaccine to protect them. ^[41] But the research doesn't tell us whether vaccines can prevent problems caused by sickle cell disease. ^[52] ^[53]

A summary of the research (a [systematic review](#)) found no serious side effects from the vaccines. ^[52] Your child may get a mild temperature at first and some pain and swelling where they are injected.

Bone marrow transplants

In this section

A [bone marrow](#) transplant is the only treatment that offers the chance of a cure for sickle cell disease. But it may be suitable only for a small number of people. ^[54] This treatment is still being developed. There's a risk of serious side effects because the drugs needed to make the transplant possible can be poisonous. It may also be hard to find someone to donate the bone marrow for the transplant.

Bottle blowing

In this section

One small study has looked at a treatment called bottle blowing for acute chest syndrome. ^[55] Acute chest syndrome is a problem with your lungs caused by sickle cell disease. It causes difficulty breathing. (To learn more, see [What will happen to me?](#)) Blowing into a bottle filled with water can exercise your lungs. People who had this treatment were less likely to get chest problems such as blocked air passages in their lungs.

This treatment is also used to treat people with [pneumonia](#) . You might hear your doctor call it **incentive spirometry**.

Further informations:

Life expectancy for people with sickle cell disease

Sickle cell disease can cause serious health problems. Some of these can be life-threatening. This means that people with sickle cell disease don't live as long, on average, as people who don't have the condition.

However, when you read these numbers, bear in mind that statistics are based on lots of people. You are an individual. No one can predict what will happen to you. It's also important to remember that treatments for sickle cell disease are getting better. People are likely to live longer now than when these statistics were collected.

These numbers are the averages for men and women in the US. ^[18]

- Men with sickle cell disease live for about 42 years, on average.
- For women, the average is 48 years.

In some parts of Africa, where good health care isn't always available, about one-half of children with sickle cell disease die before their first birthday. ^[7] But the life expectancy in the UK is improving all the time.

Helping yourself with sickle cell disease

There isn't much research on things you can do to prevent sickle cell pain. Some doctors think that keeping warm, taking moderate exercise, and drinking plenty of fluids may help. Here are some things that are recommended for people with sickle cell disease: ^[9]

- Make sure you get enough rest ^[19]
- Avoid stress
- Don't smoke
- Make sure you drink enough fluids
- Try to avoid very hot or cold temperatures
- Try to avoid high altitudes (where there's less oxygen in the air).

Sickle cell disease

We couldn't find any good studies about avoiding the cold to prevent sickle cell pain. But a study that looked back over 10 years shows people with sickle cell disease were more likely to go to hospital if the weather was cold. ^[20]

We also couldn't find any good studies to show whether exercise can trigger sickle cell pain. Doctors think moderate exercise is unlikely to cause harm in people with sickle cell disease, and it may reduce the risk of heart disease. But very strenuous exercise might bring on an attack of sickle cell pain. This is because it lowers the amount of oxygen in your blood.

We couldn't find any good studies about drinking plenty of fluids. ^[21] But we know that people with sickle cell disease are more likely to have health problems caused by dehydration. ^[22] So people with sickle cell disease are advised to drink plenty of fluids to keep hydrated.

Glossary:

anaemia

Anaemia is when you have too few red blood cells. Anaemia can make you get tired and breathless easily. It can also make you look pale. Anaemia can be caused by a number of different things, including problems with your diet, blood loss and some diseases.

red blood cells

Red blood cells are the part of your blood that makes it red. Their main job is to carry oxygen from your heart and lungs to the tissues of your body. Once these cells unload oxygen, they pick up carbon dioxide. They take carbon dioxide back to your lungs so it can be breathed out of your body.

genes

Your genes are the parts of your cells that contain instructions for how your body works. Genes are found on chromosomes, structures that sit in the nucleus at the middle of each of your cells. You have 23 pairs of chromosomes in your normal cells, each of which has thousands of genes. You get one set of chromosomes, and all of the genes that are on them, from each of your parents.

infection

You get an infection when bacteria, a fungus, or a virus get into a part of your body where it shouldn't be. For example, an infection in your nose and airways causes the common cold. An infection in your skin can cause rashes such as athlete's foot. The organisms that cause infections are so tiny that you can't see them without a microscope.

dehydrated

When you're dehydrated, you don't have enough fluid in your blood. This could be because you're not drinking enough or because you're losing water by sweating or having diarrhoea.

malaria

Malaria is a dangerous but treatable disease caused by a parasite. This parasite lives in mosquitoes in parts of Asia, Africa and South America. You can catch it if you are bitten by an infected mosquito. Malaria can give you symptoms similar to flu (influenza). The most common symptom is a fever.

spleen

Your spleen is an organ that sits on the left side of your body just below your ribs. It helps your body fight infections.

inflammation

Inflammation is when your skin or some other part of your body becomes red, swollen, hot, and sore. Inflammation happens because your body is trying to protect you from germs, from something that's in your body and could harm you (like a splinter) or from things that cause allergies (these things are called allergens). Inflammation is one of the ways in which your body heals an infection or an injury.

general anaesthetic

You may have a type of medicine called a general anaesthetic when you have surgery. It is given to make you unconscious so you don't feel pain when you have surgery.

bacteria

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Bacteria are tiny organisms. There are lots of different types. Some are harmful and can cause disease. But some bacteria live in your body without causing any harm.

pneumonia

Pneumonia is an infection in your lungs. Anything that causes infections (bacteria, viruses or fungi, for example) can give you pneumonia.

meningitis

Meningitis is a swelling in the thin layers of tissue that surround your brain and your spinal cord. It usually happens because of an infection with certain kinds of bacteria or viruses. Meningitis can give you a severe headache and a stiff neck. And you may find it difficult to keep your eyes open in the light. Meningitis is a life-threatening disease. If you have these symptoms, you should go to hospital straight away.

septic shock

Septic shock is a serious condition caused by a large number of bacteria getting into your blood. It's also called septicaemia, sepsis or blood poisoning. Usually, when bacteria get into your blood, your immune system kills them. But if your immune system isn't working well, it can get overwhelmed. Then, the bacteria multiply and start to release poisonous chemicals (called toxins) into your blood. These chemicals cause your blood pressure to drop massively. When this happens, organs such as your brain, heart, kidneys and liver may not be able to work properly because they aren't getting enough blood. Septic shock needs to be treated urgently, usually with antibiotics, to stop these vital organs failing and to prevent death.

antibiotics

These medicines are used to help your immune system fight infection. There are a number of different types of antibiotics that work in different ways to get rid of bacteria, parasites, and other infectious agents. Antibiotics do not work against viruses.

stroke

You have a stroke when the blood supply to a part of your brain is cut off. This damages your brain and can cause symptoms like weakness or numbness on one side of your body. You may also find it hard to speak if you've had a stroke.

liver

Your liver is on the right side of your body, just below your ribcage. Your liver does several things in your body, including processing and storing nutrients from food, and breaking down chemicals, such as alcohol.

low blood pressure

If your blood pressure is about 100/60 or less, your doctor may say that you have low blood pressure. Low blood pressure is usually not a problem unless it becomes too low to push blood to your brain and the rest of the body. If you have low blood pressure, you may sometimes feel dizzy when you stand up.

bone marrow

Your bone marrow is the soft material inside your bones. Bone marrow makes and stores blood cells.

blood transfusion

If you've lost too much blood from your body, you may need a blood transfusion to replace it. People with diseases of their blood, like sickle cell anaemia, sometimes need blood transfusions to replace blood that doesn't work properly.

kidney

Your kidneys are organs that filter your blood to make urine. You have two kidneys, on either side of your body. They are underneath your ribcage, near your back.

ulcer

An ulcer is an open sore. Ulcers can happen in many parts of your body, such as in your stomach, and the skin of your legs, mouth, or genitals.

puberty

Puberty is the time when boys and girls develop secondary sexual characteristics. For boys, the major changes include pubic hair, a deeper voice, and growth of their penis and testicles. For girls, major changes include pubic hair, breasts and starting to have periods. After puberty, girls are able to become pregnant and boys are able to father children.

heart disease

You get heart disease when your heart isn't able to pump blood as well as it should. This can happen for a variety of reasons.

vaccination

A vaccination is an injection a doctor can give you to protect you from getting an infectious illness (an illness that spreads between people).

intravenous infusion

When a medicine or a fluid, such as blood, is fed directly into a vein, it's called an intravenous infusion (or IV). To give you an intravenous infusion, a nurse, technician or a doctor places a narrow plastic tube into a vein (usually in your arm) using a needle. The needle is then removed and the fluid is infused (or dripped) through the tube into the vein.

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NSAIDs

NSAID stands for nonsteroidal anti-inflammatory drug. NSAIDs help with pain, inflammation and fever. They are called 'nonsteroidal' because they don't contain any steroids. Aspirin and ibuprofen are both NSAIDs.

acute myocardial infarction

Acute myocardial infarction is what doctors call a heart attack. A heart attack is when your heart muscle gets damaged because it isn't getting enough blood and oxygen. This can happen if a branch of your coronary arteries becomes blocked. During a heart attack, you may have pain or heaviness over your chest, and pain, numbness or tingling in your jaw and left arm.

randomised controlled trials

Randomised controlled trials are medical studies designed to test whether a treatment works. Patients are split into groups. One group is given the treatment being tested (for example, an antidepressant drug) while another group (called the comparison or control group) is given an alternative treatment. This could be a different type of drug or a dummy treatment (a placebo). Researchers then compare the effects of the different treatments.

diarrhoea

Diarrhoea is when you have loose, watery stools and you need to go to the toilet far more often than usual. Doctors say you have diarrhoea if you need to go to the toilet more than three times a day.

randomised controlled trials

Randomised controlled trials are medical studies designed to test whether a treatment works. Patients are split into groups. One group is given the treatment being tested (for example, an antidepressant drug) while another group (called the comparison or control group) is given an alternative treatment. This could be a different type of drug or a dummy treatment (a placebo). Researchers then compare the effects of the different treatments.

constipated

When you're constipated, you have difficulty passing stools (faeces). Your bowel movements may be dry and hard. You may have fewer bowel movements than usual, and it may be a strain when you try to go.

seizure

A seizure (or fit) is when there is too much electrical activity in your brain, which results in muscle twitching and other symptoms.

systematic reviews

A systematic review is a thorough look through published research on a particular topic. Only studies that have been carried out to a high standard are included. A systematic review may or may not include a meta-analysis, which is when the results from individual studies are put together.

allergic reaction

You have an allergic reaction when your immune system overreacts to a substance that is normally harmless. You can be allergic to particles in the air you are breathing, like pollen (which causes hay fever) or to chemicals on your skin, like detergents (which can cause a rash). People can also have an allergic reaction to drugs, like penicillin.

white blood cells

White blood cells are the cells in your blood that help your body fight infections. They are part of your immune system. The other cells in your blood, red blood cells, carry oxygen around your body.

sickle cell disease

Sickle cell disease is a blood disorder that runs in families. Under certain conditions, the red blood cells of people with this disease change shape to look like a sickle. This causes a lot of pain and a range of other symptoms, including infections in the lungs.

immune system

Your immune system is made up of the parts of your body that fight infection. When bacteria or viruses get into your body, it's your immune system that kills them. Antibodies and white blood cells are part of your immune system. They travel in your blood and attack bacteria, viruses and other things that could damage your body.

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