

Treating acute painful sickle cell episodes in hospital

Information for the public

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About this information

NICE clinical guidelines advise the NHS on caring for people with specific conditions or diseases and the treatments they should receive. The information applies to people using the NHS in England and Wales.

This information explains the advice about treating acute painful sickle cell episodes (also sometimes known as 'painful crises') in hospital that is set out in NICE clinical guideline 143.

Does this information apply to me?

Yes, if you are:

- an adult with sickle cell disease who is treated in hospital for an acute painful sickle cell episode
- a parent or carer of a child or young person who is treated in hospital for an acute painful sickle cell episode (although it is recognised that many children and young people will want to know for themselves what the NICE guidance says).

No, if you:

- are a sickle cell carrier

- have sickle cell disease and have types of episodes or crisis that are not acute painful episodes.

Your care

In the NHS, patients and healthcare professionals have rights and responsibilities as set out in the NHS Constitution (www.dh.gov.uk/en/DH_132961). All NICE guidance is written to reflect these. You have the right to be involved in discussions and make informed decisions about your treatment and care with your healthcare team. Your choices are important and healthcare professionals should support these wherever possible. You should be treated with dignity and respect.

To help you make decisions, healthcare professionals should explain acute painful sickle cell episodes and the possible treatments for them. They should cover possible benefits and risks related to your personal circumstances. You should be given relevant information that is suitable for you and reflects any religious, ethnic or cultural needs you have. It should also take into account whether you have any physical or learning disability, sight or hearing problem or language difficulties. You should have access to an interpreter or advocate (someone who helps you put your views across) if needed.

Your family and carers should be given their own information and support. If you agree, they should also have the chance to be involved in decisions about your care.

You should be able to discuss or review your care as your treatment progresses, or your circumstances change. This may include changing your mind about your treatment or care. If you have made an 'advance decision' (known as a 'living will' in the past) in which you have already given instructions about any treatments that you do not wish to have, your healthcare professionals have a legal obligation to take this into account.

All treatment and care should be given with your informed consent. If, during the course of your illness, you are not able to make decisions about your care, your healthcare professionals have a duty to talk to your family or carers unless you have specifically asked them not to. Healthcare professionals should follow the Department of Health's advice on consent (www.dh.gov.uk/en/DH_103643) and the code of practice for the Mental Capacity Act. Information about the Act and consent issues is available from www.nhs.uk/CarersDirect/moneyandlegal/legal. In Wales healthcare professionals should follow advice on consent from the Welsh Government (www.wales.nhs.uk/consent).

In an emergency, healthcare professionals may give treatment immediately, without obtaining your informed consent, when it is in your best interests.

If you are under 16, your parents or carers will need to agree to your treatment, unless it is clear that you fully understand the treatment and can give your own consent. In an emergency, if your parents or carers cannot be contacted, healthcare professionals may give treatment immediately when it is in your best interests.

Acute painful sickle cell episodes

Acute painful sickle cell episodes (also sometimes known as painful crises) are one of the most common and upsetting effects of sickle cell disease.

These painful episodes are often unpredictable, but they can have different causes, such as infection, lack of fluids (dehydration) or low levels of oxygen in the blood. The episodes happen when the sickle-shaped red blood cells block small blood vessels so that the blood does not flow normally. This damages the tissue, which causes pain. Some painful episodes may be worse than others, but the pain can be very severe.

A painful episode usually lasts for a few days, but it may be shorter or longer. Most painful episodes can be treated at home, but if the pain is very bad the person may have to go to hospital. The NICE guidance is about treating painful episodes in hospital.

What can I expect when I first get to hospital?

An acute painful sickle cell episode should be treated as a medical emergency. There should be procedures in place to make sure that you receive the care you need.

The healthcare professionals caring for you should respect the fact that you are likely to be an expert in your condition, as is your carer if you have one. They should ask about and listen to your views, and talk with you about the planned treatment for the current painful episode as well as any treatment you have had for previous episodes. They should also ask you whether you have any concerns about how you are feeling, and whether you need any extra support.

Your healthcare professional should do the following:

- Ask you to describe how bad your pain is using a [pain scoring tool](#). If you are a child or young person, the type of tool used should be suitable for someone of your age.
- Offer you pain relief within 30 minutes of arriving at hospital. (See '[Early pain relief](#)' and '[Ongoing pain relief and monitoring](#)' for further details about pain relief.)

- Regularly check your blood pressure, blood oxygen levels, heart rate, breathing rate and temperature.

Your healthcare team should also check whether a problem other than an acute painful sickle cell episode could be causing your pain, particularly if it feels different from the pain you have experienced before during a painful episode.

Questions you might like to ask your healthcare team

- Please tell me more about acute painful sickle cell episodes
- How long might I have to stay in hospital?
- Can you provide any information for my family/carers?

Questions that family members, friends or carers might like to ask

- What types of treatment are available for a child who is having an acute painful sickle cell episode?
- What can I/we do to help and support the person with sickle cell disease?
- Is there any additional support that I/we as carer(s) might benefit from or be entitled to?

Early pain relief

Some treatments may not be suitable for you, depending on your exact circumstances. If you have questions about specific treatments and options, please talk to a member of your healthcare team.

The type of pain relief you are offered will depend on how bad the pain is, which painkillers are suitable for you, whether you have taken any painkillers before arriving at hospital, and your individual care plan (if you have one).

You should be offered a strong opioid if your pain is very intense ('severe'). You should also be offered a strong opioid if your pain is still fairly bad ('moderate') even though you have already taken some pain relief. The strong opioid should be given as a single dose over a short period of time, so that it works quickly. This is known as a bolus dose.

If your pain is moderate and you have not yet taken any pain relief, you may be offered a weak opioid as an alternative to a strong opioid.

You should also be offered regular paracetamol and NSAIDs (non-steroidal anti-inflammatory drugs), as well as an opioid, to help with pain relief. However, there may be times when these are not suitable for you – for example, NSAIDs should usually be avoided if you are pregnant.

You should not be offered a drug called pethidine for pain relief.

Ongoing pain relief and monitoring

If you think that your care does not match what is described in this information, please talk to a member of your healthcare team in the first instance.

Healthcare professionals should check how well your pain relief is working. This should be done every 30 minutes until you are comfortable, and then at least every 4 hours after that. They should ask you how well the last painkiller you had worked and whether you feel you need more pain relief. They should also use a pain scoring tool that is suitable for you.

If you are still in severe pain despite having had a first dose of a strong opioid, you should be offered a second dose. If you had a weak opioid for moderate pain at first but your pain has got worse, you should now be offered a strong opioid.

If you need repeated doses of a strong opioid within a 2-hour period, you may be able to use a device that allows you to control the amount of medication you have yourself. This is called patient-controlled analgesia, or PCA for short.

You should be offered medication to help with the side effects of opioids, such as regular laxatives to avoid constipation, and anti-sickness and anti-itch drugs if you need them. If you are taking a strong opioid, you should be monitored regularly for possible side effects. You should have an assessment (for example, to see how alert you are) every hour for the first 6 hours, and then at least every 4 hours after that.

Healthcare professionals should encourage you to use your own ways of coping with the pain if these help you. Examples include techniques to help you relax, such as massage or reflexology.

If the usual treatments for an acute painful sickle cell episode don't seem to be working, you should be reassessed to see if something else might be causing your symptoms.

As your pain improves, your healthcare team should gradually reduce the amount of pain relief you have, after discussing this with you.

Questions you might like to ask about pain relief

- Why have you decided to offer me this particular painkiller?
- How long will it take to have an effect?
- I've already taken some painkillers. Will this affect my treatment?
- What side effects are associated with this medication?
- Are there any long-term effects of taking this medication?
- What other painkillers are available?
- Why are you offering me patient-controlled analgesia, and what does it involve?

Possible complications

Your healthcare team should be aware of possible complications that can occur at any time in people having an acute painful sickle cell episode. In particular, they should look out for a serious lung condition called acute chest syndrome that can affect people with sickle cell disease. Symptoms include chest pain, breathing problems and fever. Other possible complications they should be aware of include stroke, aplastic crisis (a severe type of anaemia that usually requires a blood transfusion), infections, osteomyelitis (a bone infection) and splenic sequestration (where red blood cells become trapped in the spleen).

Your healthcare team

Everyone in the healthcare team that is looking after you should have regular training in pain monitoring, pain relief and identifying possible complications of an acute painful sickle cell episode. They should also be aware that preconceptions about people with sickle cell disease seeking drugs and/or attention are not acceptable.

If one is available, you should be cared for in a specialist sickle cell daycare centre where staff have expert knowledge and training. Staff in specialist centres should also provide support to any healthcare professionals who care for you in hospital emergency departments.

You should be cared for in a place that is suitable for you. For example, children should be cared for in a dedicated children's ward or unit.

If you are pregnant, your healthcare team should get advice from an obstetrics team – that is, a team specialising in the care of pregnant women.

When you go home

Before you leave hospital, you (and/or your carer) should be given information on how to continue to manage your current painful episode. This should include how to get specialist support and more medication if you need it, and how to cope with any possible side effects of the treatment you have had.

Questions you might like to ask before you leave hospital

- Who should I contact if I need further support or medication after I get home?
- Are there any support organisations in my local area?

Explanation of terms

NSAIDs (non-steroidal anti-inflammatory drugs)

An NSAID is a type of drug that reduces inflammation and pain. Examples of NSAIDs used for treating acute painful sickle cell episodes include aspirin, diclofenac and ibuprofen.

Opioid

A type of painkiller used for moderate to severe pain. Morphine is an example of a strong opioid that is used for treating severe pain in people with an acute painful sickle cell episode. Weak opioids used for treating moderate pain include codeine and dihydrocodeine.

Pain scoring tool

A way of measuring how severe a person's pain is. There are several types, including number scales and questionnaires. A version for children (called the FACES scale) involves asking the child to look at pictures of faces and point to the one that best reflects how their pain makes them feel.

Sickle cell carriers

Sickle cell disease is caused by an inherited change to a gene (a 'genetic mutation') that affects the structure of haemoglobin (a protein found in red blood cells). People with sickle cell disease have two copies of the changed gene. A person that has one copy of the changed gene is called a sickle cell carrier, and does not experience acute painful sickle cell episodes.

More information

The organisation below can provide more information and support for people with sickle cell disease. NICE is not responsible for the quality or accuracy of any information or advice provided by this organisation.

- Sickle Cell Society, 020 8961 7795 www.sicklecellsociety.org

The NHS Sickle Cell and Thalassaemia Screening Programme website (<http://sct.screening.nhs.uk/>) contains information about sickle cell disease, including the tests offered at different stages of life, as well as screening and care.

You can also go to NHS Choices (www.nhs.uk) for more information.

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Accreditation

