NHS
National Institute for Health and Clinical Excellence

Understanding NICE guidance

Information for people who use NHS services

Treating acute painful sickle cell episodes in hospital

NICE 'clinical guidelines' advise the NHS on caring for people with specific conditions or diseases and the treatments they should receive.

This booklet is about the care and treatment in hospital of people with acute painful sickle cell episodes (also sometimes known as 'painful crises') in the NHS in England and Wales. It explains guidance (advice) from NICE (the National Institute for Health and Clinical Excellence). It is written for adults with sickle cell disease who experience acute painful episodes, and for the parents or carers of children and young people who have acute painful sickle cell episodes. However, it is recognised that many children and young people will want to know for themselves what the NICE guidance says. This information may also be useful for them and for anyone with an interest in the condition.

The booklet is to help you understand the care and treatment options that should be available in the NHS. It does not describe acute painful sickle cell episodes or the tests or treatments for them in detail. A member of your healthcare team should discuss these with you. There are examples of questions you could ask throughout this booklet to help you with this. You can get more information from the organisations listed on page 10. Medical terms printed in **bold** type are explained on page 9.

Information about NICE clinical guideline 143
Issue date: June 2012

Contents

The advice in the NICE guideline covers:

- Adults, young people and children with sickle cell disease who are treated in hospital for an acute painful sickle cell episode.

It does not specifically look at:

- People who are **sickle cell carriers**.
- People with sickle cell disease having a type of episode or crisis that is not an acute painful episode.

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_113613**). 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 pages 6–7 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?

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

Early pain relief

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

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.

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

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).

If you have talked to your healthcare team, and you think that a treatment is suitable for you but it is not available, you can contact your local patient advice and liaison service ('PALS') or NHS Direct Wales.

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, diclenofac 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. NHS Choices (**www.nhs.uk**) may also be a good place to find out more.

Your local patient advice and liaison service (usually known as 'PALS') may be able to give you more information and support. You should also contact PALS if you are unhappy with the treatment you are offered, but you should talk about your care with a member of your healthcare team first. If your local PALS is not able to help you, they should refer you to your local independent complaints advocacy service. If you live in Wales you should speak to NHS Direct Wales for information on who to contact.

About NICE

NICE produces guidance (advice) for the NHS about preventing, diagnosing and treating medical conditions. The guidance is written by independent experts including healthcare professionals and people representing patients and carers. They consider the evidence on the disease and treatments, the views of patients and carers and the experiences of doctors, nurses and other healthcare professionals. Staff working in the NHS are expected to follow this guidance.

To find out more about NICE, its work and how it reaches decisions, see ***www.nice.org.uk/AboutGuidance***

This booklet and other versions of the guideline aimed at healthcare professionals are available at ***www.nice.org.uk/guidance/CG143***

You can order printed copies of this booklet from NICE publications (phone 0845 003 7783 or email publications@nice.org.uk and quote reference N2749). The NICE website has a screen reader service called Browsealoud, which allows you to listen to our guidance. Click on the Browsealoud logo on the NICE website to use this service.

We encourage NHS and voluntary organisations to use text from this booklet in their own information about sickle cell disease.

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ISBN 978-1-84936-868-1
N2749 1k 1P Jun 12