NHS
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NHS Trust

Managing the pain of an acute sickle cell crisis

Information for patients experiencing an acute sickle cell crisis

Sickle Cell and Thalassaemia Centre

What is a painful sickle cell crisis?

The sickle cell crisis or vaso occlusive crisis occurs when the red blood cells form a sickle shape and block small blood vessels.

This can cause extreme pain in the affected areas of the body because the tissues are not receiving enough oxygen.

What causes a painful crisis?

- The cold or sudden change in weather
- Dehydration
- Infection
- Overexertion
- Stress
- Lack of oxygen
- No cause

Different types of pain; What type of pain do I have?

Acute pain

Acute pain lasts for a relatively short time and then resolves. This is typical of sickle cell crisis pain that can start mildly and then become very intense. This pain usually resolves with a week though can vary if affected by other conditions that cause sickling.

Chronic pain

Chronic pain is persistent and long term, lasting over 3 months. In sickle cell chronic pain can result from damage to the body from repeated sickling.

Neuropathic pain

Sometimes in sickle cell disease people can develop persistent nerve or neuropathic pain. The exact causes are complex but may be caused by damage to the nerve or changes to the way the brain processes pain.

How to manage an acute painful episode?

You probably already have some strategies for managing pain at home from a vaso occlusive crisis. Some suggestions are:

- Drink plenty of fluid, up to 3 litres, to prevent dehydration
- Keep warm but do not get too hot.
- Heat rubs or heat pads to the affected painful area can be soothing
- Think about the distraction techniques that work for you
- Take oral painkillers for at least 24 hours when the pain is mild to moderate
- If the pain does not settle or is severe seek medical attention as soon as possible.

What painkiller (analgesia) should I take?

The first choice of painkillers (analgesia) should be Paracetamol. This should be taken as soon as you start getting pain. The adult dose is two tablets (1g) every four to six hours. You should not take more than eight tablets in a 24 hour period.

What if this does not work?

The next part of step 1 for many patients is a group of medicines called NSAIDs (non-steroidal anti-inflammatory drugs). There are two common choices in this group, either Ibuprofen or Diclofenac.

Ibuprofen can be bought over the counter. The dose you take is usually 200- 400 mg three times a day. Diclofenac is available only on prescription. The dose is usually 50 mg three times per day. Diclofenac is not recommended if you have heart disease or if you have had a stroke.

NSAIDS work by reducing swelling (inflammation) and therefore pain. NSAIDs are especially effective for bony pain and pain that is throbbing in nature. They work well in combination with paracetamol.

There are risks of taking NSAIDS so they should not be taken on a long-term basis. They can cause problems, as below:

- NSAIDs can cause stomach irritation and stomach ulcers and **should not be taken on an empty stomach**.
- NSAIDs may also cause kidney damage, and fluid retention.
- You should consult your doctor before taking any of these tablets if you have any stomach, heart, liver or kidney problems, asthma or if you are pregnant or breast-feeding.

Are there any other options? The next step

If you have been advised not to take the NSAIDs or the pain is still not resolving, you may be able to take a codeine-based /weak opiate drug. The common choices are either dihydrocodeine, tramadol or combinations with codeine and paracetamol (such as Co-codamol). These are taken for moderate to severe pain.

Opiate preparations should be taken only after consultation with your doctor if you have asthma, kidney or liver problems. Constipation, nausea, dizziness are some of the possible side effects of codeine.

Please refer to the table below for more information:

Pain relief drug name	Dosage and other information
Dihydrocodeine	This is available only on prescription; the dose is 30-60 mg every four to six hours.
Codeine phosphate	This is available only on prescription; the dose is 30-60 mg every four to six hours.
Co-codamol	This is a pre-prepared drug that mixes codeine and paracetamol; the dose is 1-2 tablets up to 4 times a day. It is important not to use paracetamol when using Co-codamol.
Tramadol	This is an alternative to codeine preparations and is another opiate available on prescription. The dose is 50-100mg every 4-6 hours. Do not exceed 400mg in 24 hours.

But I'm still in pain, what should I do?

Most patients at this point would need to seek medical advice, and either come to the SCAT centre or go to ED (Emergency Department / A and E). The next step would usually be the introduction of opioid painkillers. Opioids (or opiates) are the strongest painkillers available. There are several different opoid painkillers which work in similar ways.

Possible side effects

The commonest side effects of this group of painkillers are:

- Drowsiness and dizziness
- Constipation, nausea and vomiting. There are medications that can help with the constipation and there are anti-sickness medications that can be used to reduce nausea and vomiting.
- Itching. This can be relieved by medication so please tell your doctors if morphine makes you itch.
- One of the most important side effects of opioid painkillers is a reduced breathing rate, which can lead to low oxygen levels. The breathing or respiratory rate needs monitoring closely especially if you have never had or rarely have this type of painkiller.

If you are pregnant, breast-feeding or have any kidney problems before taking this group of medications, you should seek advice from your doctor.

Choice of opiates

For most patients the preference is a morphine-based drug. This can be given in the following ways: an injection of Morphine Sulphate subcutaneously (given underneath the skin), as a tablet (Sevredol) or syrup form (Oromorph). Morphine or diamorphine may also be given as a subcutaneous patient-controlled analgesia (PCA). Other effective opiates include Oxycodone, Diamorphine, Buprenorphine, Fentanyl and Methadone.

Managing pain is like climbing the steps of the ladder. With an acute sickle cell crisis most people can do the first 2 steps at home and only need to come to a hospital for step 3. When you need strong opiates, the hospital is the safest place you can be, so you can be monitored.

The Pain Ladder

Mid pain ➡ Moderate to strong pain ➡ Strong to severe pain

STEP 1 ⤴ Mid pain.	**STEP 2** ⤴ Moderate to strong pain.	**STEP 3** Strong to severe pain
Start with Paracetamol, Aspirin or NSAID's	Add Codeine, Tramadol etc	Add Morphine, Fentanyl etc

When your pain is improving, and you are no longer requiring the strong opiates you can then go home and continue to manage the pain at home. Remember the pain ladder and that you have to take it a step at a time. Do not stop all your pain killers, reduce each step at a time.

The SCAT team can give you a pain plan for going home to follow.

What are the problems with long term opiate use?

There is a growing awareness of the risks associated with long term opiate use.

Side effects can include:

- Increased pain and headaches
- Constipation
- Sleep disordered breathing.
- Brain damage due to shallow breathing
- Overdose
- Mood changes muscle weakness
- Weight changes
- Hormonal changes
- Risk of fracture

In view of the harmful effects we recommend long term use of opiates is avoided if at all possible.

Tolerance

If you have been taking opiates for a long time your body will get used to them and they will become less effective. Increasingly higher doses will be needed to achieve the same pain relief.

Withdrawal

Once your body is used to opiates you will notice unpleasant symptoms, known as withdrawal symptoms, when you stop taking them. These symptoms include feeling tense or nervous, feeling sick, increased pain and low mood. In order to minimise these, it is important to reduce your usage gradually. You can ask your team for help.

Dependence

When you depend on opiates in order to feel well, or not feel sick or in pain you are said to be dependent. Sometimes people distinguish between physical dependence when you take opiates to avoid physical symptoms and psychological dependence when your perception is that you need opiates to function. Anyone taking opiates for a long time can be expected to develop some degree of dependence and it is ok to talk with your team if you have concerns about this.

Managing the pain of a sickle cell crisis without medication

Painkillers are a common method of managing the sickle cell pain and this can be effective, especially in the short term. There are reasons why you might want to try alternative treatment, for example:

- You have developed an allergy or do not like the side effects.
- You have another condition that stops you from using the medication.
- You are pregnant.
- Your medication does not work well anymore, or you may want to avoid a situation where it may stop working.
- You have previously experienced dependence or addiction to painkillers or other substances and you do not want to risk having similar problems again.

Massage

Massage can get the blood flowing again or the stimulation can disrupt the pain signals. Ask someone to gently massage the affected area or try self-massage. There are devices available for self-massage for hard to reach areas.

Relaxation

Practicing relaxation can reduce your pain. Specific exercises that cause you to become deeply relaxed, such as progressive muscle relaxation are recommended.

Relaxation makes you feel good and reduces stress. It can also cause the blood vessels to widen (dilate) and relieve the sickle cell crisis blockage.

There are free apps you can download to your smartphone.

Emotional wellbeing

Managing your emotions and how you fell can help you with your pain. Emotions do not cause pain in sickle cell disease but a long-term illness together with its associated issues can leave you feeling down. Unfortunately, negative emotions like sadness, anger or worry amplify pain.

Remember the activities that bring you enjoyment. Try to find the time to phone a friend, pamper yourself with a face mask or watch that film you keep meaning to. Getting involved in activities or hobbies which take the focus of attention away from your pain can be helpful. A hobby like sewing, photography or model-making, which you can do even when activity is restricted, can be a good distraction. Hobbies can also give you something to talk about with other people, other than your pain.

Socialising and continuing to have contact with people can play an enormous part in helping you feel better. Pain can make it hard to get out to see people or to join in with what they are doing, but social contact can be at home, over a cup of hot chocolate, within whatever limits you have.

Staying Active

Staying active with mild to moderate exercise is very important for general health and preventing pain. Regular exercise promotes the release of endorphins your body's natural pain killer, which can reduce pain.

If you have developed a chronic condition like avascular necrosis you may need exercise to strengthen the muscles in that particular part of your body. When your muscles are stronger, they support your weight in everyday activities and put less strain on your joints or body.

Pacing yourself and setting goals

Getting on with your usual day can be either necessary or a good distraction. Taking regular breaks in activities, changing position, resting briefly between activities or doing stretches throughout the day can all help. Practising and regularly setting goals and using pacing techniques can together help you to achieve many of the activities that you want to each day or longer term.

With all these approaches, you will need to find what work for you. Different things work for different people.

Pain Management Points to Remember:

- Please discuss the most suitable choice of pain management with your team. Remember to consider all the things that you find helpful when in pain.
- Individual pain plans are available to all, speak to your team.
- If your pain is persisting at home, you develop other symptoms and/or you are concerned always seek medical advice.
- Never take more medication than prescribed or medication that has not been prescribed for you.
- Strong opioid medication such as Morphine should not be taken without careful monitoring and should only be used in hospital
- It is recommended that you do not drive or operate machinery if you feel drowsy after taking your medication. It is illegal to drive if you are impaired for any reason, even if you have a prescription.
- Work and activity; You may want to take some time off work, or you may want to continue as usual. If you are taking opiates at work, please ask the occupational health department if you can continue to work whilst doing so. Pregnancy: please tell your doctors that you are pregnant or might be.
- There are differences in the way that the pain of an acute sickle crisis and chronic pain (such as that due to long-term bony damage) are treated. Treatment choices are decided on an individual basis.
- If you have concerns about your medication, please, please discuss with your doctor / nurse.

Contact details

If you have any questions or concerns, please contact the Sickle Cell and Thalassaemia Centre.

Sickle Cell and Thalassaemia Centre

Sandwell & West Birmingham Hospital
City Hospital, Dudley Road
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Tel: 0121 507 6040
City Hospital : 0121 554 3801

Opening Hours

Monday, 9am – 5pm
Tuesday, 9am – 6pm
Wednesday, Thursday and 9am – 5pm
Friday, 9am – 4pm
Saturday blood transfusion service only

The information in this leaflet is general and is intended to be a guide only. Please discuss the specific details of your treatment with your GP/doctor.

More information

For more information please see our regional website:
www.westmidsstn.nhs.uk

For more information about our hospitals and services

Sandwell and West Birmingham Hospitals NHS Trust
www.swbh.nhs.uk

Further information

Sickle Cell Society,
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Email: **info@sicklecellsociety.org** **www.sicklecellsociety.org**

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