North East & Yorkshire Haemoglobinopathy Coordinating Centre

Keeping well for Adults with Sickle Cell Disorder

Information for patients, families and carers

Haematology - Haemoglobinopathy Coordinating Centre

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Complications in Sickle Cell Disorder

The main complication of sickle cell disorder is a painful crisis, this is a result of the red blood cells changing shape and blocking the flow of blood to various parts of the body.

Other complications of sickle cell disorder (SCD) include:

- Acute chest syndrome (severe crisis in the chest which can affect breathing)
- Stroke
- Increased infections
- Leg ulcers
- Bone and joint damage especially of the shoulders and hips
- Gallstones
- Kidney damage and loss of water in the urine
- Priapism
- Blood blockage/pooling in the liver or spleen (sequestrations)
- Eye damage
- Delayed growth

Prevention of these complications

There are several things that can help prevent a crisis from developing.

These are listed below:

- **Taking folic acid daily -** This vitamin is used to make red blood cells and is used up as your body makes these red blood cells at a faster rate in sickle cell disorder. If the level of folic acid is low, then this can slow the red cell production down and cause you to become anaemic.
- **Regular use of prophylactic antibiotics -** The spleen is gland in the left side of the tummy which acts like a filter to remove any bacteria from the blood stream. The spleen does not work in sickle cell disorder and so you are more vulnerable to infections which can be quite serious. There are 2 ways in reducing the chances of this happening - The first is by using regular low dose antibiotics. The incidence of residence to these antibiotics in the UK is quite low. The other method is ensuring that you are vaccinated to protect you against the bacteria that most commonly caused the infections in SCD.
- **Staying hydrated -** Drinking plenty of water/juice (8-10 glasses for adults). In warmer weather you may need to increase the amount of water or juice. Tea, coffee and sugary drinks can act as a diuretic and make you pass lots of urine; these should be used in moderation.
- **Treat infection as soon as it occurs -** If in doubt, see your doctor.
- **Avoid too hot or cold temperatures -** Wear warm clothes outside in cold weather and inside air-conditioned rooms during hot weather. Also, try to avoid swimming in cold water.
- **Try to exercise regularly -** Try to avoid over exertion with exercise so much that you become really tired and when you do exercise drink lots of fluids to keep hydrated.

- **Reduce or avoid stress -** Talk to your doctor if you are depressed or have problems with your family or job.
- **Getting plenty of rest.**
- **Try to be positive about yourself.**
- **Tell you doctor if you think you may have a sleep problem-** These could include snoring, or if you sometimes stop breathing.
- **Get early antenatal care -** If you are planning a pregnancy or have become pregnant inform your GP and the haemoglobinopathy team so that we can discuss with you your options and help advise and support you.
- **Only travel in commercial airplanes -** If you travel in an unpressurised aircraft, talk to your doctor about extra precautions.
- **Smoking and tobacco use -** Try to avoid if possible. If you do smoke, try to quit. Advice and support can be given, speak with your GP for cessation support.
- **Alcohol -** This dehydrates you. Advice and help can be given if you have any concerns around alcohol. Please speak with your doctor.

Pain killers

People with sickle cell disorder will often experience both acute and chronic pain.

The majority of the time, pain can be managed at home following a detailed care plan given by your haemoglobinopathy team.

Below is a guide of how to manage pain at home and when to seek medical advice.

I am in pain, what analgesia should I take?

The first choice of painkiller (analgesia) is usually paracetamol. This should be taken as soon as you start getting pain; the adult dose is 2 tablets (1g) every 4-6 hours. However if you have previously been advised to only take 500 mg please stay with this dose and consult with your GP or medical team.

You should not take more than 8 tablets in 24 hours.

You may prefer to take or add one of a group of medicines called NSAID's (non-steroidal anti-inflammatory drugs). These tablets should not be taken long term as they can cause stomach problems. They work by reducing swelling, inflammation and therefore pain. NSAIDs are especially effective for bone pain.

There are two common choices this group either Ibuprofen or Diclofenac. Ibuprofen can be brought over the counter at chemists (Nurofen). The dose you take is usually 200-400mg three times a day. Diclofenac is available only on prescription. The dose is usually 50mg three times a day unless agreed otherwise by your GP or the Haemoglobinopathy team.

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 or kidney problems, asthma or if you are pregnant or breast-feeding.

Please consult with the Haemoglobinopathy team if you are unsure if you can take these or you have any questions.

Are there any other options?

If you have been advised not to take NSAIDs or the pain is still not resolving, you may be able to take a codeine-based painkiller. The common choices are Dihydrocodeine, tramadol or combinations with codeine and paracetamol. These are taken for moderate to severe pain and should only be used on a short-term basis.

Codeine phosphate is available only on prescription, the dose is usually 30-60mg every 4-6 hours. Co-codamol is a pre-prepared painkiller that mixes codeine and paracetamol. The dose is usually 1-2 tablets up to 4 times a day.

It is important not to use paracetamol when using co-codamol.

Codeine preparations should be taken only with consultation with your doctor if you have asthma, kidney or liver problems.

Constipation, nausea, dizziness are some of the possible side effects of codeine.

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 Haematology department or go via A+E (follow guidance from the Haematology team). The next step would usually be the introduction of opioid painkillers.

Opioids are stronger painkillers that are available. There are several different opioids painkillers which work in similar ways.

The most common side effects of this group of painkillers are drowsiness, dizziness, nausea, vomiting and constipation (anti-sickness medication and laxatives can be used to reduce this). 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 to be monitored closely especially if you have never had or rarely have this type of painkiller. You should seek advice from your doctor if you are pregnant, breast-feeding or have any kidney problems before taking this medication.

Choice of opiates for severe pain

Usually Morphine or Oxycodone preparations which can be given, orally or by injection. Other opiate painkillers include Fentanyl patches which are are available if required.

We will regularly discuss your options and preferences in your clinic appointments and these will be documented on your Individualised Care Plans (ICP).

If you need to come into hospital other medicines may be added to help with the pain such as Gabapentin or Pregabalin, if needed this will be discussed further between yourself and the Multi-disciplinary team and you may be referred to the Palliative care team for further advice.

When pain settles

A painful episode may last several days.

If strong opioids have been used it is necessary to taper these medicines over a few days rather than stopping abruptly since the body may become used to these painkillers.

Sudden withdrawal may itself cause pain.

Points to remember:

- If your pain is persisting at home and 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, you may be supplied with a short term supply after discharge, the prescription should not be repeated unless this has been discussed and advised to by the Haemoglobinopathy team.
- It is recommended that you do not drive or operate machinery as you may feel drowsy after taking the medication.
- Different analgesia is effective for acute and chronic pain. If you have concerns about your medication, please discuss with your doctor or nurse.

Hydroxycarbamide (Hydroxyurea) use in Sickle Cell Disease

What is Hydroxycarbamide?

Hydroxycarbamide is regularly used in the treatment of different blood disorders.

Adults with sickle cell disorder who experience regular or severe Vaso-occlusive crisis (VOC) are usually started on hydroxycarbamide to reduce the frequency and severity of a VOC.

Why is it used?

Hydroxycarbamide can reduce the complications of sickle cell disorder.

The best evidence comes from a large trial involving several centres in America. A large group of identical patients were given either Hydroxycarbamide or a placebo medicine.

Hydroxycarbamide was shown to reduce the number and severity of painful crises, the number and severity of chest crises (lung sickling), the number of blood transfusions, and the number of admissions to hospital.

Long term follow up has shown that patients taking this treatment are in better physical health.

How does it work?

Hydroxycarbamide has several effects on sickle cell disorder:

- It increases the amount of foetal (baby) haemoglobin (HbF). HbF is the baby haemoglobin which all infants are born with, and which normally disappears by 6 months of age.
- hydroxycarbamide increases the water content of the sickle cell which makes it more difficult for red blood cells to develop sickle shape.
- It reduces the ability of the red blood cell to stick to the lining of the blood vessels, helping to prevent a crisis.
- Hydroxycarbamide reduces the white blood (neutrophil) count, which is often raised in patients with sickle cell disease.
- Hydroxycarbamide also increases the levels of a substance called nitric acid which helps the blood flow through tissues of the body.

Your haemoglobinopathy team can provide more information around hydroxycarbamide if you are using or have questions around the use of this medicine.

There are also separate information leaflets available should you require them.

Risks of infection

The spleen is a gland which sits in the left hand of the tummy under the rib cage.

It does not work effectively in patients with sickle cell disorder.

What does the spleen do?

The spleen forms part of the body's defence against certain infections.

If the spleen does not work correctly, you will still be able to cope with most infections, but in some cases serious infection may develop quickly.

The risk of this happening is higher in children, however there is still a risk in adults.

What kind of infections am I susceptible to?

The most common infection that patients will experience will be simple viral infections such as coughs and colds.

However, some infections due to bacteria can be quite serious and may cause blood stream infections, pneumonia and meningitis.

What precautions should be taken to minimise the risk of infection?

For some patient's regular low doses of antibiotics are given to help prevent the onset of infections. If your doctor has recommended antibiotics, then it is advisable that these are taken daily. The usual recommended antibiotics are either penicillin V 250mg twice daily, or erythromycin 250mg twice daily if you are penicillin allergic.

However if you do not want to take daily antibiotics, we can prescribe you a short supply of prophylactic antibiotics so you can keep a supply at home in case of fever or infection.

Please note medical help should always be sought promptly if unwell.

It is important to regularly check with your GP that your vaccines are up to date.

The following vaccines are recommended:

- Pneumococcal (every 5 years)
- Conjugated Meningococcal C Vaccine (a single dose should be given if not already received as part of childhood immunisations)
- Haemophilus influenzae type b (a single dose should be given if not already received as part of childhood immunisations)
- Influenza – Flu jab (every year if you are eligible)
- Covid vaccine (every year)
- Other vaccines may be recommended if you are travelling to high-risk areas (please check with your GP or a specialised local travel clinic)
- If travel is being planned to areas with Malaria, then medicines to prevent Malaria may be needed. Please contact your local travel clinic for more information.

What are the warning signs of infection?

- Fever – if you feel hot you should always check your temperature? Temperatures of 38^{C} and above always requires prompt action (normal body temperature is around 37^{C}). However, if you are unwell and your temperature is lower then please seek advice.
- Rigors – violent shivering that you cannot control
- Severe sore throat
- Cough – with discoloured phlegm
- Pain or burning when passing urine
- Sudden severe headache, neck stiffness or intolerance of bright light

In the event of any of the above or if you feel unwell, please seek medical attention as soon as possible. Early diagnosis and treatment are essential and may be lifesaving.

Please contact your doctor or the Haematology department via the contact details provided by your haemoglobinopathy team.

Priapism

What is priapism? A MEDICAL EMERGENCY

Priapism is a complication of sickle cell disorder and is a form of sickle cell crisis. It is a prolonged painful erection of the penis which may occur without prior sexual stimulation. The true incidence is unknown but is likely to affect as many as 50% of men with sickle cell disorder.

What causes priapism?

It is caused by blockage of the blood vessels in the penis by sickle cells, and a failure of blood flow back into the body. Blood remains trapped in the penis causing painful enlargement and priapism.

The following factors may trigger an attack:

- Infection
- Dehydration
- Alcohol consumption
- Occasionally following sexual activity

Types of priapism

- **Stuttering priapism** – The episodes of priapism are short attacks the subside spontaneously. These attacks can occur with varying frequency and patients may not seek medical advice.
- **Fulminant priapism** – This is a major episode of priapism which does not subside after several hours and is an emergency that requires urgent medical attention. The longer episode of priapism lasts, the greater the risk is of long-term damage to the penis which can cause impotence (failure to have an erection).

Measures to be taken during an episode

- Try to empty the bladder
- Drink lots of fluid
- Take painkillers
- Take a walk, try to move around if you can

Different patients have reported different methods to overcome acute episodes such as warm baths, hot water bottles (following the instructions of the water bottle and not placing directly onto the skin).

If the episode of priapism does not settle in 1 hour, please contact your haematology team for urgent review.

DO NOT DELAY

LONG TERM COMPLICATIONS MAY OCCURE IF YOU DO

What are the treatments for priapism?

Specialists called urologists would attempt to empty the blood and remove the trapped red blood cells. In some situations, surgery to drain blood from the penis and/or blood transfusions may be necessary; in this situation the urologists would discuss this further with you.

Other measures to help with sickle cell disorder would include fluids (by mouth or a drip) and pain medications which are like those used to manage a sickle cell crisis (tablets or injections).

What are the risks of not getting treatment?

Early intervention is important to prevent longer term complications such as impotence.

Preventative measures

Things you can do to reduce your chances of having an episode of priapism are:

- Exercise regularly
- Avoid late nights – make sure you get enough sleep
- Avoid excessive alcohol consumption
- Avoid smoking

In clinic, your consultant will discuss medications that may be used to prevent further episodes for happening.

What are some of the challenges?

The onset of priapism is a frightening experience and is often associated with embarrassment and guilt. There is no need to suffer in silence because help is just a phone call away.

Below is a link to a informative video about Priapism on our HCC site.

Leg Ulcers

Leg ulcers can occur in people already diagnosed with sickle cell disorder.

These can add to the pain and debilitation that having sickle cell disorder brings and can also be seen as painful open wounds.

Symptoms

Sickle cell ulcers usually present around or behind the ankle, they can also present around the base of the toes in more extreme cases.

They can be single ulcers or multiple and can occur in both legs at once. They are usually painful, vary in size and shape and severity.

Sickle cell ulcers often deteriorate during a sickle cell crisis due to the general reduction in oxygen to the skin.

Tolerance to treatments can be difficult. Neuropathic pain and hypersensitivity around the ulcer may also be extreme.

Treatment

Treatment can be trickier simply because you will also have other side effects of sickle cell disease that you are trying to manage. It is likely you will also be taking various other medication, and you may also be trying to minimise the impact of the ulceration on your employment.

Please talk to your healthcare professional about these complexities so they can be considered for your management plan. It is very important that you also inform your haematology team so they can have input on the care plan and offer advice.

Treatment plans will include medical management with a haematologist, pain management, compression therapy (depending on area of ulcer), walking review and footwear.

Prevention

If you have sickle cell disorder, make sure you look after your legs and avoid lower limb trauma where possible.

Seek medical attention as soon as you notice a break in your skin to the lower limb so treatment can commence and prevent further breakdown.

Elevate legs regularly to prevent venous complications and where possible wear compression hosiery as a prevention.

Things to consider:

- **Dietary intake and hydration - we have published a specialised leaflet for nutrition in sickle cell.**
- **Seeking help when needed, if that is financial or psychological please talk to the Haemoglobinopathy team. There may be a dedicated psychology team and a benefits advisor from Citizens Advice Bureau who can provide you with advice and support**
- **Remember, If you are struggling you are not alone the haemoglobinopathy team are here and help and support you.**

Please use the QR code or link below to take you to the Patient Information Leaflet (PIL) section of the North East and Yorkshire Haemoglobinopathy Co-ordinating Centre Website.

- www.ney-hcc.co.uk/patient-information/

Notes

Further information

More information on sickle cell disease can be found in various places as listed below.

- The Sickle Cell Society website - www.sicklecellsociety.org

- Sickle cell anaemia news website – www.sicklecellanemianews.com

- NHS – www.nhs.uk, ringing 111 or your local GP surgery

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