Pain management in sickle cell disease

A guide for parents

Pain and sickle cell disease

Pain is the most common problem experienced by people with sickle cell disease (Hb SS, Hb SC, HbS-beta thalassaemia).

What is a pain crisis?

Red blood cells are usually round. When they become sickled they become less bendable and can block small blood vessels. This stops blood from delivering oxygen to the tissues and causes pain in that part of the body. This is called a pain crisis or a vaso-occlusive crisis.

Pain crises in sickle cell disease can start from as early as three months of age. In this age group the hands and feet may become puffy and painful. This is known as dactylitis. As children become older the pain can occur in the arms, leg, back, chest or abdomen. The pain can start suddenly and last for hours to days.

Causes of pain crises

Pain crises often occur without warning and cannot be prevented. However certain situations can bring on crises such as infections, exposure to severe cold, dehydration (not drinking enough) and stress. You may have fewer and less painful crises if you do the following:

- drink plenty of fluids
- avoid swimming in cold pools (swim in heated swimming pools) and dry off quickly when coming out of the swimming pool
- dress warmly, including hats and gloves in cold weather
- limit exposure to people with viral illnesses, colds
- avoid stressful situations.

Pain management at home

Many pain crises can be managed at home with:
- pain medications
- fluids
• hot packs
• rest.

Pain killers that can be used for treatment of pain at home include paracetamol, ibuprofen and codeine phosphate. Prescriptions for these medications can be obtained from your GP or from the paediatric haematology clinic. The exact amount of medication to be given to your child depends on your child’s age and weight. The amounts (dose) will be noted on the medication bottles given to you.

**Mild to moderate pain**

If the pain is mild, paracetamol can be given every six hours. Ibuprofen can also be given every six hours. These two medications can be used together, often given in alternated doses, so that your child can receive pain medication every three hours. It is best to ensure that your child is eating some food when they are taking ibuprofen on a regular basis.

**Severe pain**

Codeine Phosphate can be taken in addition to paracetamol and ibuprofen if the pain is severe. When the pain is improving stop giving the Codeine Phosphate first.

**Additional measures**

• Ensure your child is drinking lots of fluids.
• Massage may be helpful.
• Heat is helpful (try heating pads, hot water bottles or warm baths).
• Distractions such as soothing music, TV, toys.
• Encourage your child to rest.

**General points**

It is important to keep a small supply of pain medications at home at all times as pain crises can occur at any time.

If you are giving your child pain medications, it is a good idea to write down:
• the amount of medicine
• the time that it was given to your child
• the effect it had on your child (for example, pain relief or no effect).
When to come to the hospital

Children with sickle cell disease have direct access to Frederick Hewitt Ward. If you feel that your child need to be assessed, please call the ward on 020 8725 2081 and tell them that you are coming in.

Reasons for immediate medical attention are:

- fever greater than 38.5 degrees Celsius
- severe chest pain or difficulty breathing
- severe abdominal (tummy) pain or swelling
- severe headaches/dizziness
- problems with seeing
- painful erection
- child is very pale
- child unable to move arm or leg
- seizures
- severe pain that is not responding to pain killers
- vomiting or diarrhoea.

Hospital management of pain

If the pain is not controlled with medications at home, your child may need to be admitted to hospital for further treatment. In the hospital your child may be given a stronger pain medication such as morphine to control the pain.

- Morphine can be give by mouth, but sometimes if the pain is severe the morphine may be given directly into the vein through a cannula (small tube inserted into a blood vessel). In some cases it is necessary to give the morphine by a continuous infusion in the vein or under the skin. When prescribed by doctors experienced in the management of pain crises, morphine does not cause dependence. Children are not discharged home while they are still taking morphine.

- While your child is in hospital they will be encouraged to drink fluids or they will be given fluids through the cannula to prevent dehydration.

- Many children that take strong pain medications such as codeine and morphine can develop constipation which can then cause abdominal pain. Children that are in hospital on pain medications are often prescribed a stool softener and laxatives to prevent this problem.

- Some children may become very tired and sleepy when receiving strong pain medications. They will be encouraged to perform breathing exercises (blowing bubbles for children under five years) or they will be given a blowing machine (incentive spirometer) and taught how to use
Some children may require extra oxygen and will be given this through a face mask.

- Some children who have a pain crisis may develop a fever. If this happens to your child, blood tests will be taken and your child will be given antibiotics.

You may stay with your child while they are in hospital. If you cannot do this, another close relative over 18 years of age may stay with them for support and comfort.

It is very distressing to see your child in pain. We hope that this guide gives you the information to manage painful episodes at home and helps you decide when to bring your child to hospital. Please contact us if you have further questions.

Contacts

Children’s outpatient department (Dragon) 020 8725 2953

Haemoglobinopathy counsellor 020 8700 0615/0616

Paediatric haematology secretary 020 8725 3921
If you wish to contact the paediatric haematology consultant please contact the paediatric haematology secretary.

Freddie Hewitt Ward 020 8725 2081