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The Royal Liverpool
and Broadgreen
University Hospitals
NHS Trust

Patient information

Sickle Disorders

When Should Adults With Sickle Cell Disease Attend The Emergency Department (A&E)

Haematology Department

This leaflet provides you with information about when adults with Sickle Cell Disease (SCD) need to go to the Emergency Department (A&E) and what will happen when you are there.

If you recognise your condition is getting worse at home, and you have time, ask a friend or carer to pack a bag of essentials such as overnight clothing and toiletries and any medications you are taking at the moment.

How do I get to the Emergency Department?

In an extreme emergency it is essential that you dial 999 and call an ambulance. However, in most cases the ambulance will only take you to your nearest Emergency department to ensure that you are treated as quickly and safely as possible.

If it is not an extreme emergency and your condition allows, you should travel to the Emergency Department using your usual means of transport, or public transport.

What should I do when I get to Emergency Department?

- 1) If you have called an ambulance then you will be taken directly through to an assessment area. It is always important to tell the ambulance crew and the hospital staff that you have sickle cell disease and tell them your current symptoms.
- 2) If you have made your own way to the Emergency Department report to reception. Tell them immediately that you have sickle cell disease. Tell them if you are having a crisis and pain, and you are unwell, you should be sent straight to the treatment area. This will help speed up your assessment so you can receive painkillers in good time. The receptionist will complete your registration form.

When should I go to the Emergency Department?

You need to go to the Emergency Department if you recognise any of these signs.

Pain

Pain is the most common symptom of SCD and you should have painkillers at home. You should attend the Emergency Department if you have any of the following symptoms.

- Pain which does not settle with your usual painkillers.
- Pain which is more severe than usual or is getting more severe despite the painkillers.
- Pain which is severe and is different to your usual pain.
- A high fever.
- Shortness of breath or difficulty in breathing.

Acute chest syndrome

Acute chest syndrome is a very serious complication of SCD and prompt hospital treatment is required. The symptoms and signs can develop very rapidly over a short period of time (i.e. within a few hours). Don't wait for all the symptoms listed below to develop.

You should go to the Emergency Department if you develop any of these symptoms:

- Shortness of breath.
- Difficulty in breathing and coughing.
- Rapid/fast breathing.
- Severe pain in the chest wall or upper abdomen.
- Severe pain in the upper part of the spine.
- A high fever with a temperature above 39.0° C.
- Rapid/fast pulse rate.

Stroke

Sickling in the brain can cause a stroke. A stroke happens due to a blood clot or bleeding into the brain, which causes brain cells to die. The signs of a stroke may only last a few hours but must not be ignored. Early treatment saves lives and increases the chance of making a better recovery.

What are the signs of a stroke? The Face-Arm-Speech-Test (FAST)

Three simple checks can help you recognise whether you or someone has had a stroke or mini-stroke:

- **Facial Weakness:** Can you / the person smile? Has their mouth or an eye drooped?
- **Arm Weakness:** Can you / the person raise both arms?
- **Speech problems:** Can you / the person speak clearly and understand what you say?
- **Test these symptoms.**

If you see any of these signs, call 999 immediately.

Priapism

Priapism is a painful erection of the penis which often starts in the early hours of the morning. It is a common problem in male patients with SCD, especially in teenagers and young adults.

It occurs in two forms:

1. **Stuttering episodes** which last two to four hours but often come back and may turn into a severe attack. Recurrent stuttering episodes should be evaluated by haematology in outpatients. If you are concerned about stuttering priapism please contact the clinical nurse specialist who can help arrange a medical review if required.

2. **Severe attacks** lasting longer than four hours which can result in impotence (difficulties in getting a normal erection). If you experience priapism lasting longer than two hours and if you notice associated symptoms such as fever, difficulty passing urine, dehydration or pain in other parts of your body you need to go to the Emergency Department.

If you are embarrassed or worried about privacy please ask for a male doctor or nurse to examine you. If you would like more information about priapism please ask for the Priapism patient information leaflet.

What happens normally when I attend the Emergency Department?

You will be seen initially by a nurse who will triage you and assess your pain and physical condition. The nurse will also complete vital signs such as blood pressure, pulse, temperature and oxygen levels. This is essential and your co-operation is required so the doctors can diagnose and treat you promptly and safely.

Once you have been seen by a nurse, a doctor working in the Emergency Department will need to take your history and examine you and give you any immediate treatment required. This may include fluids, pain relief, oxygen and antibiotics.

It is important that you allow the doctor to examine you and complete investigations such as bloods tests, as they can give indications of the cause of your SCD crisis and detect major complications quickly. The doctors and nurses in the Emergency Department will follow guidelines created by your sickle cell team to help them manage your sickle cell complications.

It is helpful if you know the following information

- Your sickle cell diagnosis (e.g. HbSS).
- What pain killers you have taken and the last time you took them.
- Details of your current medications.
- Your normal haemoglobin level and oxygen levels.
- What you normally take as pain relief in hospital.
- What complications you have experienced in the past, both sickle cell and non sickle cell.

How quickly will I get pain relief?

Our standard is to get your pain relief within 30 minutes of arriving and pain control within two hours for acute sickle cell pain.

If you experience a problem in the Emergency Department please ensure you discuss this with a member of the sickle cell team so that they can assist in addressing your concerns.

Will I see a member of the sickle cell team in the Emergency Department?

Most of the time whilst you are in the Emergency Department you will be looked after by the department's team of doctors and nurses. However, if you have a major complication or problems with your pain control a member of the SCD/Haematology team can be contacted by the Emergency Department day or night for advice and assessment.

How and when will I be admitted to hospital?

In most cases you will be admitted if your pain fails to settle quickly or if you have a fever, dehydration, low oxygen levels or because you need specific treatment such as strong pain relief, oxygen or intravenous antibiotics or fluids.

You will also be admitted if the doctor is worried that you are at risk or have developed a major complication of SCD. Normally you will be admitted by the attending medical or surgical team and they will normally contact the sickle cell/Haematology team to take over your care if appropriate the next day.

Haemoglobinopathy / Sickle Cell Team:

Dr D Simcox	Consultant Haematologist
Prof CH Toh	Consultant Haematologist
Dr V J Martlew	Consultant Haematologist
Dr T Dutt	Consultant Haematologist
Haematology Specialist Registrar (Rotational Position)	
Francesca Murphy	Clinical Nurse Specialist

Feedback

Your feedback is important to us and helps us influence care in the future.

Following your discharge from hospital or attendance at your outpatient appointment you will receive a text asking if you would recommend our service to others. Please take the time to text back, you will not be charged for the text and can opt out at any point. Your co-operation is greatly appreciated.

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