NHS SCOTLAND

SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Patient Information

Acute chest syndrome in sickle cell disease

Sickle cell disease (SCD) is an inherited blood condition which can affect various parts of the body. This leaflet provides information about a serious complication of SCD called **acute chest syndrome (ACS).**

What is ACS?

Although the reasons are not clearly understood, it is believed that 'sickled' cells (blood cells that change shape) clump together in the small blood vessels in the lungs or move to the lungs from somewhere else in the body. Sometimes this is triggered by a lung infection such as pneumonia.

ACS can also develop before, during or after an episode of pain in the abdomen or bones. This may be because of shallow breathing due to pain or large doses of strong painkillers. It can also develop after a general anaesthetic.

Keeping well

There are some things that you can do to reduce the chance of ACS developing.

- Take your regular antibiotics as advised by your doctor.
- Keep up to date with your vaccinations.
- Do any breathing exercises which may have been advised by your physiotherapist or doctor.

What are the symptoms of ACS?

The symptoms of ACS can be similar to pneumonia. This is why you may be treated like you have both. Symptoms include:

- **chest, rib or back pain;**
- **a fever (a temperature of 38ºC or above);**
- **changes to breathing – difficulty breathing or rapid, shallow breathing;**
- **pale skin or blue lips or fingers;**
- **a cough or wheeze; and**
- **an increased heart rate.**

If you develop these symptoms you should go to your local centre or go to the Accident and Emergency Department as soon as possible. If transport is difficult, do not delay. You should call an ambulance by dialling **999**.

How will I be treated for ACS?

If you go to hospital, when you arrive you will be assessed to see what treatment you need.

- Your blood pressure, pulse rate, temperature, breathing rate and oxygen level will be measured and recorded.
- Your pain will be assessed.
- You will be examined by a doctor who will take your medical history.
- You will have bloods taken and swabs will be taken from your nose and throat.
- You will have a chest x-ray.

Immediate treatment may include:

- intravenous fluids;
- pain relief;
- oxygen;
- intravenous antibiotics;
- a nebuliser; and
- a blood transfusion or red cell exchange.

You will be closely monitored until there are signs that you are improving.

What other treatment might I have?

You may be seen by a physiotherapist and given advice on deep-breathing exercises. You may also be given an incentive spirometer, which helps with deep breathing. It is important to follow this guidance as this may help to stop ACS from getting worse and help you to recover.

What happens after I have recovered from ACS?

The time it takes for you to recover varies, and depends on how unwell you have been. It may be a few days but can be significantly longer. Once you have recovered it is really important to discuss with your sickle cell team how to reduce the chances of ACS happening again.

For more information about SPAH:
SPAH Website: www.spah.scot.nhs.uk
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If you need this leaflet in another format, please contact NSS.EqualityDiversity@nhs.scot, 0131 275 6000

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