SPAH
SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

Patient Information

Acute chest syndrome in sickle cell disease

This document has been prepared by NHS National Services Scotland (NSS) on behalf of SPAH. Accountable to Scottish Government, NSS works at the heart of the health service providing national strategic services to the rest of NHS Scotland and other public sector organisations to help them deliver their services more efficiently and effectively. The SPAH Network is a collaboration of stakeholders involved in care of patients with haemoglobinopathies throughout Scotland, who are supported by an NSS Programme Team to drive improvement across the care pathway.

Sickle cell disease (SCD) is an inherited blood condition. It affects how red blood cells work and can cause problems in many 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 thought that ACS develops when sickle shaped blood cells clump together in the small blood vessels in the lungs or move to the lungs from somewhere else in the body.
ACS can develop:

- due to a lung infection like pneumonia.
- before, during or after an episode of pain in the abdomen or bones. This may be due to shallow breathing caused by the pain or large doses of strong painkillers.
- 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?

Symptoms of ACS can look like pneumonia. This is why you may be treated like you have both.

Symptoms include:

- pain in the chest, ribs or back
- fever (temperature over 38 degrees
- changes to breathing (fast, shallow or difficulty breathing)
- pale skin or bluish colour to lips or fingers
- coughing or wheezing
- increased heart rate.

If you develop these symptoms, you should go to your local hospital or emergency department immediately. If you need an ambulance do not delay and call an ambulance by dialling **999**.

How will I be treated for ACS?

At hospital you will be assessed to see what treatment you need including:

- checking your blood pressure, pulse, temperature, breathing rate and oxygen levels
- assessing your pain levels
- blood tests taken

- swabs taken from your nose and throat
- a chest x-ray may be taken.

Immediate treatment which may be given can include:

- intravenous (IV) fluids (a way to give fluids or medicine directly into a vein using a small tube)
- pain relief
- oxygen
- IV antibiotics
- inhaled medication (nebuliser)
- a blood transfusion or exchange transfusion (blood is slowly removed and replaced with donor blood).

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

What other treatment might I have?

You may also be seen by a physiotherapist who will give advice on deep breathing exercises. You may also be given an incentive spirometer to help with deep breathing. It’s important to follow the advice of the physiotherapist as this can help stop ACS from getting worse and help you to recover.

What happens after I have recovered from ACS?

Recovery time can vary. Some people feel better in just a few days, while others may take much longer depending on how sick they were. Once you’re feeling better, it‘s important to talk with your sickle cell team about how to reduce the chances of ACS happening again.

Further information

Can be found on the Scottish Paediatric and Adult Haemoglobinopathies Network (SPAH) website: spah.scot.nhs.uk

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