



**SPAHA**

SCOTTISH PAEDIATRIC AND ADULT HAEMOGLOBINOPATHIES NETWORK

# Acute Chest Syndrome in Sickle Cell Disease

## Information for parents and carers

This document has been prepared by NHS National Services Scotland (NSS) on behalf of SPAHA. 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 SPAHA Network is a collaboration of stakeholders involved in care of patients with haemoglobinopathies, who are supported by an NSS Programme Team to drive improvement across the care pathway.

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# Scottish Paediatric and Adult Haemoglobinopathies Network

## Acute Chest Syndrome in Sickle Cell Disease - Information for parents and carers

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)**. ACS is one of the most common reasons children with SCD need to go to hospital. **It needs urgent hospital care.**

### 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 elsewhere 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 pain killers.
- after a general anaesthetic.

### What are the signs and symptoms of ACS?

Symptoms of ACS can look like pneumonia. This is why your child might be treated like they 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

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

### How will my child be treated for ACS?

At hospital you will be asked about your child's health, and they will be examined as follows:

- checking their blood pressure, pulse, temperature, breathing rate and oxygen levels
- checking their pain levels
- blood tests taken
- swabs taken from their throat and nose
- a chest x-ray may be taken

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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)

Your child will be closely monitored until they begin to improve.

#### What other treatment will my child have?

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

If you need an alternative format, contact [nss.equalitydiversity@nhs.scot](mailto:nss.equalitydiversity@nhs.scot) or Scotland BSL [contactscotland-bsl.org](http://contactscotland-bsl.org) for British Sign Language.

More information can be found on the Scottish Paediatric and Adult Haemoglobinopathies Network (SPAHS) website [spah.scot.nhs.uk](http://spah.scot.nhs.uk)