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Haematology Department

**Sickle Cell Disease – Chronic Complications**

**Pulmonary Hypertension**

Information for parents and carers

**Introduction**

This leaflet is designed to explain about the long term (chronic) complications of sickle cell disease. The information does not cover everything. If you are ever worried about your child then please contact your Sickle Cell Team or take your child to Accident and Emergency Department.

**What is pulmonary hypertension?**

Pulmonary hypertension is high blood pressure in the blood vessels that carry blood from the heart to the lungs. This is a common complication in teenagers and adults with sickle cell disease.

**Is pulmonary hypertension the same as regular hypertension?**

No. Regular hypertension also known as high blood pressure is when a higher pressure is needed to pump the blood around your body. In pulmonary hypertension the blood vessels in the lungs have become narrower meaning that the heart has to work harder to pump blood through them increasing the pressure inside them.

**Is pulmonary hypertension serious?**

Pulmonary hypertension is a serious condition. It results in damage to the right-hand side of the heart making the heart less efficient at pumping blood around the body and getting oxygen to the muscles. If it is not treated, pulmonary hypertension can cause [heart failure](http://www.nhs.uk/conditions/heart-failure/pages/introduction.aspx), when the heart struggles to pump enough blood around the body. This can be fatal.

**How common is pulmonary hypertension?**

Approximately one third of adults with sickle cell disease develop mild pulmonary hypertension. About 10% of patients develop severe pulmonary hypertension. (Caboot & Allen, 2008)

**What are the symptoms of pulmonary hypertension?**

The symptoms of pulmonary hypertension are similar to symptoms of long term sickle cell problems, so can sometimes be overlooked. The symptoms of pulmonary hypertension are:

* [Shortness of breath](http://www.nhs.uk/conditions/shortness-of-breath/Pages/Introduction.aspx).
* ****[Fatigue](http://www.nhs.uk/livewell/tiredness-and-fatigue/pages/tiredness-and-fatigue.aspx) (tiredness).
* Feeling faint or dizzy.
* Decreased exercise tolerance.
* Unexplained swelling of the ankles.

The symptoms are very similar to those of anaemia.

**What should I do if I notice any of these symptoms in my child?**

It is important if you notice a change in your child or spot any of the symptoms, to let your Sickle Cell Team know.

**What is the treatment for pulmonary hypertension?**

Symptoms caused by sickle cell disease such as anaemia, low oxygen saturations and regular painful crisis increase the risks of pulmonary hypertension. Stopping these things can help prevent the early stages of pulmonary hypertension getting worse. This can be done by having regular blood transfusions or taking a medicine called Hydroxycarbamide.

For more advanced stages of pulmonary hypertension medication or an operation to widen blood vessels and reduce the pressure can be tried.

**How can pulmonary hypertension be prevented?**

From the age of 12 your child will have an echocardiogram every three years. An echocardiogram checks the pressure inside the heart vessels and will identify early stages of pulmonary hypertension.

**Further information**

If you have any questions or want any further information please contact your Sickle Cell Team on 0151 252 5070.

**References**

Caboot, J. & Allen, J. 2008. Pulmonary complications of sickle cell disease in children. Current Opinion in Pediatrics, Vol 20.

**Useful websites**

[www.alderhey.co.uk](http://www.alderhey.co.uk)

[www.sicklecellsociety.co.uk](http://www.sicklecellsociety.co.uk)

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****This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child’s treatment.

This information can be made available in other languages and formats if requested.

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