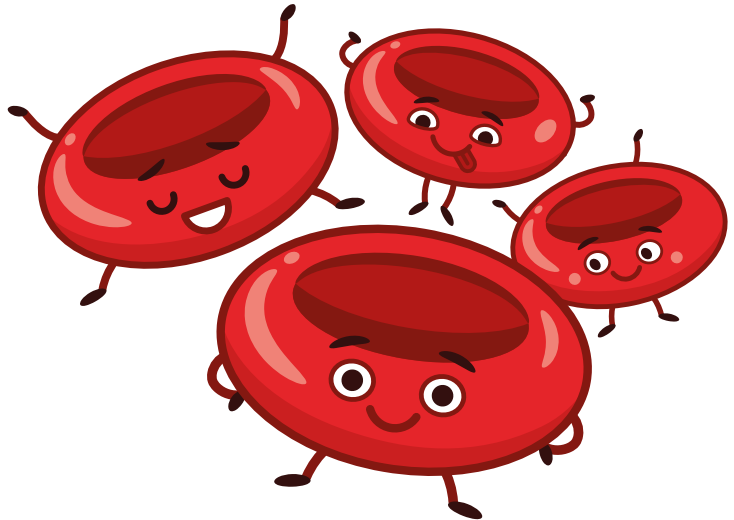


Hereditary Spherocytosis

Information for
parents



leeds children's
hospital

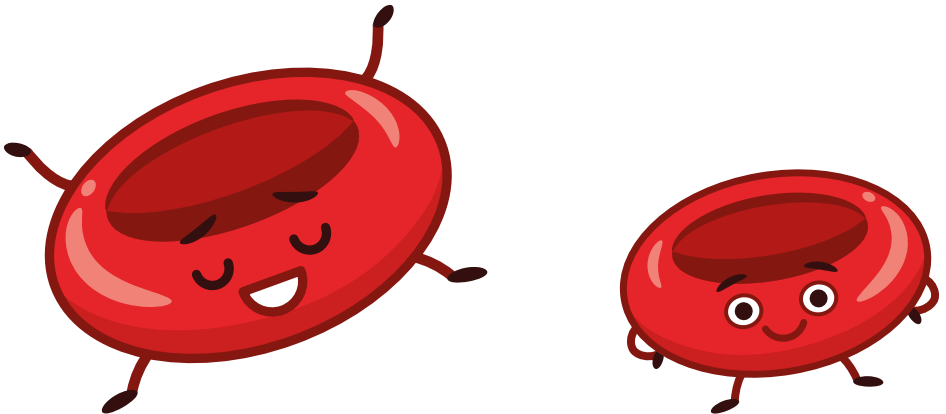
caring about children

What is Hereditary Spherocytosis?

Hereditary Spherocytosis (HS) is a medical condition caused by a problem with cells in the blood called red blood cells. Red blood cells are very important because they carry the oxygen which is essential for life. They are made in bone marrow, which is inside your child's long bones (like the thigh bone).

Red blood cells are usually shaped like a flattened circle so that they can easily squeeze through the smallest blood vessels but in people with HS, they become round, like a football. This means that they cannot flow through smaller blood vessels, which can become blocked. They are also removed from your child's blood too quickly and destroyed by their spleen, an organ of the left side of the abdomen (tummy). One of the jobs the spleen does is recycle old red blood cells.

When your child's bone marrow can't produce red blood cells as fast as their spleen is destroying them, there aren't enough red blood cells in the blood flowing around your child's organs. This is a medical condition called anaemia (a-nee-mee-ah). Because of the increased work your spleen must do, it gets bigger, which can also cause problems.



What are the symptoms of HS?

Anaemia

A condition caused by a decreased number of red blood cells. People with anaemia look pale and may feel tired, sleepy and lack energy.

Jaundice

People with jaundice have slightly yellow skin and the whites of the eyes are also slightly yellow. This is caused by the increase in breakdown of red blood cells and can get worse with infection.

Abdominal pain

This is less common, but when it does happen, it may be because of an enlarged spleen or because of gallstones. Gallstones are caused by the increased breakdown of red blood cells and can cause inflammation in the gall bladder.

If you think your child has symptoms of any of the conditions listed above, it is very important that you discuss these with your haematology doctor or nurse.



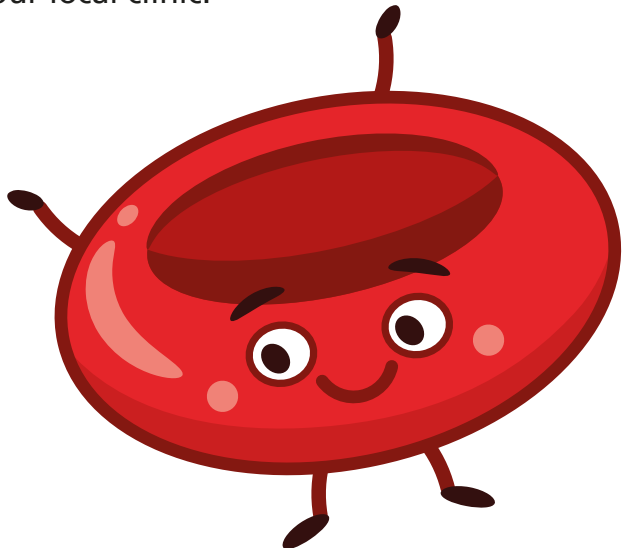
Why does my child have HS?

Most children who have HS inherit it from a parent. It is most likely that one of your child's parents has HS. About a third of children with HS don't have a parent with the condition. This time, HS is caused by a change in the genes that affect the red blood cells and has not been passed from parent to child.

How will HS affect my baby?

Babies with HS are more at risk of jaundice. The baby doctors looking after your new born should be told if your baby might have HS.

If you have a baby with HS, it is difficult to tell if they are paler than normal. You might notice a change in their sleeping and feeding routine. They might be more sleepy than usual or feed less well. These things suggest that anaemia could be increasing so if you notice either of these things, you should take your baby to your local clinic.



How will HS affect my child as they grow?

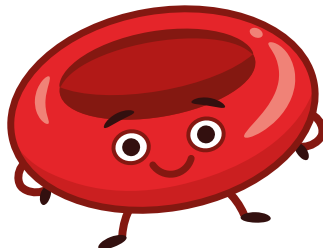
They may always have anaemia and look paler than most people. They may also have a degree of jaundice as described earlier.

Particular infections with a virus call parvovirus (slapped cheek syndrome) can slow the production of red blood cells by the bone marrow and lead to a more serious anaemia, which may occasionally require a blood transfusion.

In general though, your child should cope very well with fewer red blood cells (this is called a lower blood count). You should encourage your child to live as normal a life as possible. There is no reason why they cannot take part in most activities; however, if they want to take part in contact sports that have a risk of injury to the abdomen such as rugby, you should discuss this with your child's doctor because there is an increased risk of damaging a spleen that is larger than normal.

What is the treatment for HS?

Often, your child will be given folic acid tablets or liquid to take. Folic acid is a vitamin, which is important for making red blood cells. Rarely, children might need a blood transfusion; this is usually only necessary in the first year of life, or after a parvovirus infection and is much more unlikely in older children and adults.



If anaemia and other symptoms are quite severe, over many months or years, it might be necessary for your child to have an operation to remove the spleen (called a splenectomy); however, this is quite uncommon now. A splenectomy is usually done after the age of 6 years old and most are done around the age of moving to secondary school. Removal of the spleen increases the chances of certain bacterial infections. Should your child require a splenectomy, the haematology doctors will take care to protect them from such infections. It will be necessary for your child to have the appropriate vaccinations, before and after the operation. They would also need to take penicillin or another antibiotic twice per day.

Is HS a serious problem?

HS is usually only a mild medical condition. Many people go through life without knowing that they have HS.

How often will my child have to attend clinics?

With children, this depends on the age and whether their symptoms are severe or not. Babies diagnosed with HS have varying, unstable symptoms during the first year of their life. Because of this, doctors like to see a new baby perhaps every month at first. As the symptoms become more stable, visits are less frequent. Older children who do not have any problems will be seen every 6 - 12 months. Your haematology doctor / nurse will guide you through the types of appointments needed.

Many adults with HS that is stable do not regularly attend clinics.

Important contact information

Children's Haematology Clinical Nurse Specialists
(available Monday - Friday, 8.00 am - 5.00 pm):

Suzie Preston

Tel: **0113 3926867 / 07775 228860**

Ben Sykes

Tel: **07787 266096**

Children's Haematology Day Unit
(open Monday - Friday, 8.00 am - 5.30 pm)

Tel: **0113 3927179**

Ward 31

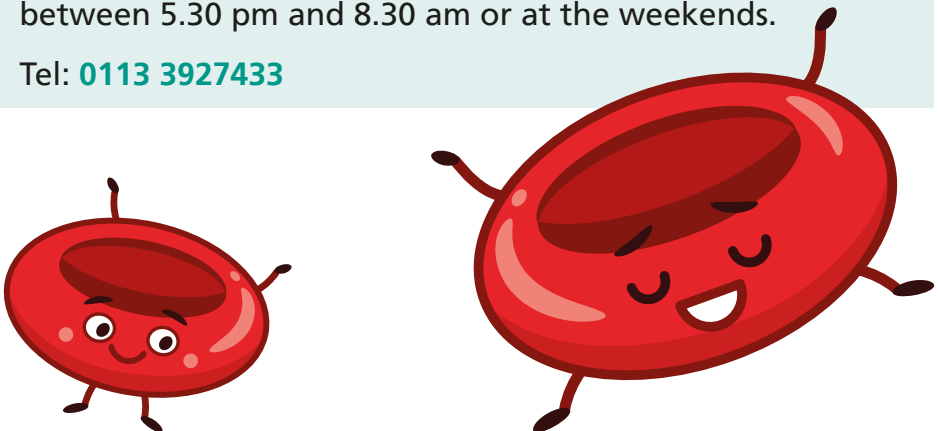
(birth - 13 years) - open 24 hours. Please only contact the ward between 5.30 pm and 8.30 am or at the weekends.

Tel: **0113 3927431**

Ward 33

(13 - 18 years) open 24 hours. Please only contact the ward between 5.30 pm and 8.30 am or at the weekends.

Tel: **0113 3927433**





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