What is Thalassaemia and how is it treated?

Sickle Cell and Thalassaemia Service

Information for Patients

Produced: May 2025
Review: May 2028
Leaflet number: 1615 Version: 1

What is Thalassaemia?

Thalassaemia is an inherited blood disorder. You are born with thalassaemia. The body does not make enough or any haemoglobin. Haemoglobin is a protein found in the blood. It carries oxygen around the body. People with this have genes that make less normal haemoglobin. Having thalassaemia can make you feel tired, weak and breathless.

What are the different types?

There are many different types of thalassaemia. They need to be treated in different ways.

- **Transfusion dependent Beta thalassaemia:** The amount of haemoglobin made is very low. You need regular blood transfusions from infancy so that you grow, develop and have quality of life.
- **Non-transfusion dependent Beta thalassaemia**: A lower amount of haemoglobin is made. There is enough for growth and development. You do not need blood transfusions. Growth may fail or there may be other complications. People may need blood transfusions as a child or adult.
- **Haemoglobin H disease:** Often a mild health problem. It is like long lasting anaemia, but you may occasionally need blood transfusions.

How is it diagnosed?

We diagnose it through a blood test. The NHS in England test all pregnant women. This is through is the NHS Sickle Cell and Thalassaemia Screening Programme. The father of the baby is tested if a woman is found to be a carrier. This is to find out the risk of the having baby with it.

Blood transfusions

Some patients will need regular blood transfusions. These treat and prevent anaemia. How often you need transfusions is different from person to person.

We offer blood transfusions to thalassaemia patients at the East Midlands Planned Care Centre (EMPCC) Day Unit at Leicester General Hospital. Visit the link:

www.leicestershospitals.nhs.uk/aboutus/our-hospitals/east-midlands-planned-care-centre-empcc/

Blood transfusion risks

Sudden transfusion reaction:

- high temperature, hot clammy, cold, shivery
- itchy skin and / or rash
- difficulty breathing
- chest pain
- wheezing
- swelling of lips, tongue or face
- not passing pee (urine)

This is rare. We will monitor you closely during your transfusion. If you do feel unwell let your nurse know.

Transfusion associated circulatory overload (TACO)

This means there is a chance of fluid being stored in the body (fluid overload). This is because of the blood transfusions. We will see how much of a risk you have of getting this. We may examine you to look for symptoms of fluid overload.

Infection

We test blood donations to reduce the risks of spreading infection through blood transfusion. There may be infectious risks we cannot test for or do not know about. We test patients every year for viral infections such as HIV, hepatitis B and hepatitis C.

Antibody formation

When you get a blood transfusion, your body might not know some proteins (antigens) on the donated blood cells. This is because they are new to your blood. Your body will create antibodies against them. These antibodies can then attack and destroy transfused blood cells.

There are many different antigens on red blood cells. It is impossible to perfectly match donated blood to every single one. People who get transfusions often are more likely to start to get these antibodies. Their immune system is exposed to different blood types more often.

If you start to get these antibodies, it can be harder to find compatible blood for future transfusions. That's why before each transfusion we check your blood for any new antibodies your body may have made.

Delayed transfusion reaction

After a transfusion, there is a small risk of having a late reaction. This can happen between 24 hours and 14 days after a transfusion. You may have:

- high temperature, hot clammy, cold, shivery
- itchy skin and/or rash
- yellow skin or eyes (jaundice)
- dark urine, or not passing much urine

Too much iron

Patients who have regular blood transfusions are at risk of having too much iron. Transfusions give the body extra iron. This can collect and cause iron overload.

Symptoms of iron overload are different for each person. It depends where the iron collects. Many people will not have symptoms until the iron overload is bad. It is important to have your blood test and scan and go to clinic appointments. This will help us know if you have iron overload.

Too much iron can:

- damage the heart leading to heart failure and irregular rhythms.
- damage the liver leading to cirrhosis.
- damage the pancreas which can cause irreversible diabetes.
- damage to hormone glands causing underactive thyroid (hypothyroidism), tiredness, and affecting sex hormones.

How do we check for iron overload:

- Blood tests to look at iron levels (ferritin). We see how well the liver is working (liver function tests (LFTs), and endocrine function tests (thyroid, sex hormones, adrenal gland).
- Glucose tolerance test (GTT) every year.
- MRI scans of the liver (Ferriscan) and the heart (Cardiac T2*).
- ECG and ECHO of the heart.

Treating too much iron

Treatment for too much iron is called iron chelation. It works by attaching to the excess iron. This helps the body remove it. There are 3 different types of medicine you can take. The type you are we give you depends on your needs and kidney function. You may have to take more than 1 type of iron chelation together.

Getting pregnant or getting someone pregnant while on these medications may affect your baby. Use birth control while on this medication to prevent pregnancy.

1. Deferiprone (Ferriprox)

This is a tablet. It works well to reduce iron in the heart. You will need to get regular blood tests to check your White Cell Count (WCC). This is because this medication can cause it to drop. If your WCC drops, it lowers your body's ability to fight infection.

Risks and side effects:

- Risk of infection. You may have temperature above 38, feeling shivery, sore throat, or other signs of infection.
- Your urine may turn red / brown. This is not likely to cause any problems.
- Feeling sick and being sick. Take the medication with food. It can help reduce this.

2. Deferasirox (Exjade)

This is a tablet. It works well to remove excess iron from the liver. You need to have regular blood tests for kidney and liver function (LFTs) while on this medication.

Risks and side effects:

- Damage to kidneys and liver. You will have regular blood tests to check this.
- Feeling sick and being sick and diarrhoea. This gets better over time.
- Urine tests are needed every month to check for high protein.
- Blurred vision. You will need eye tests every year to check for early changes to eye health.
- Hearing loss. You will need hearing tests every year to find any hearing damage.

3. Desferrioxamine (Desferal)

This is an injection. We give this just under the skin (subcutaneously) or into the vein (IV).
You take it at home. We will teach you how to do this. This medication needs to be stored in the fridge.

Risks and side effects:

- Yersinia infection. This can cause tummy (abdominal) pain, diarrhoea, being sick, fever. If you have any of these call the haematology helpline.

- Ringing in ears (tinnitus), hearing loss. You will need hearing tests every year to find any hearing damage.
- Sight impairment. You will need eye tests every year to check for early changes to eye health.
- Irritation or blisters may happen where the needle is inserted. Do not inject in the same area you injected before. Place the needle as directed to reduce the risk of this.
- Your urine can turn red / brown. This is not harmful.

Who do I call if I am unwell?

If you feel unwell with:

- symptoms of infection and are taking iron chelation,
- think you may have had a delayed transfusion reaction, or
- have symptoms of anaemia

You can contact the clinical nurse specialists or the haematology helpline.

Haematology Helpline (24 hours a day, 7 days a week)

Phone: 0808 178 2212

Adult Haemoglobinopathy Clinical Nurse Specialists (CNS) (Monday to Friday, 9am to 5pm)

Phone: 0795 089 1490

If the nurse or doctor feels you need to be seen, you may be asked to rearrange your blood transfusions either at:

East Midlands Planned Care Centre
Leicester General Hospital,
Coleman Rd,
Leicester
LE5 4PW
Phone: 0300 303 1573

Osbourne Treatment Centre
Leicester Royal Infirmary
Infirmary square
Leicester
Le5 1WW
Phone : 0300 303 1573

If you have any questions, write them down here to remind you what to ask:

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