

Iron Overload and its Treatment in Patients with Sickle Cell Disease or Thalassaemia

This leaflet explains the condition and the treatment of iron overload. If you have any further questions, please speak to a doctor or nurse caring for you.

What is iron overload?

Patients who have regular or occasional blood transfusions for thalassaemia or sickle cell disease will get more iron than their body needs. Unfortunately this is an unavoidable consequence of transfusions. The body cannot get rid of excess iron itself and if this excess iron builds up it can cause damage to some parts of the body. Treatment for excess iron is known as chelation therapy and is used to remove the excess iron from the body.

Some patients with thalassaemia who do not receive blood transfusions may also develop iron overload due to the way their body absorbs more iron than is needed to try to make more red blood cells.

What are the symptoms of iron overload?

Symptoms of iron overload depend on where the excess iron is deposited in your body. People often have no symptoms at all until the iron overload is severe.

The internal organs where iron can damage include:

- The liver - Iron in the liver can cause scarring of the liver known as fibrosis which can then progress to cirrhosis (a severe form of liver damage that can eventually lead to liver failure). Symptoms of cirrhosis include loss of appetite, tiredness, very itchy skin and jaundice (yellowing of the skin). Significant iron overload in the liver for a long time can sometimes cause liver cancer.
- The heart – Iron in the heart can cause irregular heart rhythms and lead to heart failure.
- The pancreas – Iron in the pancreas can lead to diabetes. Some of the symptoms of diabetes include increased urination, feeling thirsty, very tired and unexplained weight loss.
- Hormone glands – The thyroid may slow down which can cause tiredness and the parathyroid gland may not work properly causing problems with bone health. The sex hormone glands in the brain that stimulate the release of the sex hormones can also be affected, causing problems with puberty, growth and fertility.

Many of these symptoms may not develop until the iron overload is quite severe. Feeling well with no symptoms is not a way to be sure that there is no iron overload. This is why regular monitoring is very important.

How is iron overload diagnosed and monitored?

Your risk and severity of iron overload can and will be measured by a combination of blood tests and scans as needed. Many of these will be done regularly when you are attending clinic or for a transfusion. Other tests will be arranged as they are needed and will be discussed with you. The tests combined will tell us if there is too much iron in your body, which organs in your body are affected by too much iron and, if you are on treatment for iron overload, the tests will tell us how well that treatment is working.

- Blood tests – The most common blood tests used for monitoring and identifying iron overload are ferritin level and iron studies. Ferritin is a protein that stores iron. These tests give us an estimation of how much extra iron is in your system. Ferritin levels are a simple blood test to take and can be done at any time (often done whenever attending for a blood transfusion) and the results are usually available within 24 hours. As ferritin levels can also be high when you have an infection or liver inflammation, additional tests are very useful to identify accurately the amount of iron in the body.
- Specialised MRI scans – These scans are done in the Atkinson Morley Wing at St George's Hospital and can measure the amount of iron in the heart and liver. They give an accurate result and are a very useful way of monitoring the iron levels in people with significant iron overload. If this is the case, you will have the scans every one to two years.
- Other blood tests – These can be used to monitor the effect of iron on the function of the organs. These tests may be for thyroid function, liver function, sex hormone levels and bone metabolism. We can also test for diabetes and these tests will all be done depending on how severe your iron overload is.
- Review by other specialists – Depending on the severity of the iron overload and the organs involved you may be reviewed by specialists in our cardiology (heart), hepatology (liver) and endocrinology (hormone) departments. These specialists may also ask for extra tests such as a fibroscan (an ultrasound looking for scarring in the liver) or an echocardiogram (an ultrasound of the heart). Any referrals and extra tests that are needed will be discussed with you.

If you are referred for scans or appointments with other doctors within the hospital it is very important that you attend these. They are an important part of monitoring your health and keeping you as well as possible.

How can iron overload be treated?

Iron overload in patients with sickle cell disease, thalassaemia or due to blood transfusions for other reasons can be treated by medication that removes the iron from the body. Usually the body cannot get rid of extra iron by itself so the medication binds to the iron and allows the body to remove it in urine or faeces.

This is called **iron chelation**. There are different medications available that can be used for iron chelation. These are

- Desferrioxamine (also called Desferal®)
- Deferiprone (also called Ferriprox®)
- Deferasirox (also called Exjade®).

Your doctor or specialist nurse will discuss with you which type of therapy is recommended for you and why.

The decision to start iron chelation therapy will depend on many things, including how many units of red blood cells you have received if you are receiving regular or occasional transfusions, your ferritin level and your MRI results. All these will be discussed with you.

If you have blood transfusions for the treatment of sickle cell disease then exchange transfusions are usually recommended over 'top up' transfusions. This is a way to have the benefit of blood transfusions with less iron overload as the iron is exchanged out in the blood cells. We can also remove some iron in some patients with sickle cell disease by the way we do the exchange transfusions (red cell depletion.) Please see our leaflet on exchange blood transfusions in sickle cell disease for further information.

Iron chelation medications

Below is a brief description of the three different types of medications that can be used to treat iron overload. When your treatment is being decided the doctors or nurses will explain which medication you are being prescribed, how to take it and any possible side effects. When you receive your medication you will also be told how to take the medication and how to store it. Please also read the patient leaflet that comes with the medication.

Please do ask your doctors or nurses any other questions you have or concerns whilst you are taking the medication.

Desferrioxamine (Desferal®)

How desferrioxamine is given

This medicine is usually given into fat tissue (subcutaneously) or into a vein (intravenously) as an infusion over several hours. This is usually for 8 to 12 hours each day but some people have the infusion running for 24 hours.

If the decision is for you to have desferrioxamine subcutaneously, then your specialist nurse will work with you and the day unit so that you can be taught to self-administer the medication if possible and who to contact for questions.

Occasionally it is recommended desferrioxamine should be given directly into the vein (intravenously), either because the subcutaneous route is not suitable or because more intensive iron chelation is required. If this is the case you will need a long term intravenous line. Again your specialist nurse and the day unit will work with you so you can get all the information you need before the line is inserted. You will be taught how to self-administer the medication and look after the line.

Dose and frequency of desferrioxamine

Your haematology doctor will advise you on the dose and frequency of your treatment but it is often at least five days a week.

Possible side effects

Desferrioxamine is widely used and some people have no side effects. However, some possible side effects include:

- Irritation or blisters on the skin where the needle is placed – you can avoid this if you rotate the site of injections. It is also important to ensure that the needle is properly positioned under the skin. Your nurse will give you information on what to look out for if your treatment involves a long-term intravenous line. Please do speak to your specialist nurse or to the day unit if you notice skin reactions.
- Vision and hearing problems can occur such as ringing of the ears (tinnitus) and a reduction in night vision. Eye and hearing tests should be done before starting treatment and during therapy. These issues usually go away upon stopping treatment.
- Abdominal pain, fever, diarrhoea and vomiting – these may indicate an infection caused by bacteria called Yersinia. This can be a particular problem when you are having chelation therapy as the iron-removing medication can feed the bacteria with the iron it has picked up, worsening the infection. If any of these symptoms occur, stop your treatment and call your specialist nurse for advice.

Deferiprone (Ferriprox®)

How do I take deferiprone?

Deferiprone is a tablet (usually more than one) that is taken three times a day.

Possible side effects

- Infections - this medicine can lower the number of neutrophils, a type of white blood cell that helps to fight infections. Your doctor will ask for a blood test every two weeks to check your neutrophil levels. If you get a sore throat, a temperature over 38°C, feel shivery or have any other signs of infection, contact your specialist nurse.

If it is after hours, go to the Emergency Department (A&E) as you may need antibiotics right away.

- Reddish brown colour of your urine, which may look alarming but will not cause long term problems. This is the iron coming out of your body.
- Nausea and sickness – This can often be reduced by taking the tablets with meals.
- Increased appetite.
- Stomach pain - This is more frequent at the beginning of therapy and tends to resolve within a few weeks without stopping treatment.
- Joint pain.

Deferasirox (Exjade®)

How do I take deferasirox?

Deferasirox is taken once a day as a tablet. You should take it at about the same time each day with some water. Deferasirox should be taken on an empty stomach or with a light meal. Taking your medication at the same time each day will also help you remember when to take your tablets.

For patients who are unable to swallow whole tablets, EXJADE film-coated tablets may be crushed and taken by sprinkling the full dose onto soft food such as yogurt or apple sauce (pureed apple). The food should be eaten immediately and completely consumed. Do not store it for future use.

Possible side effects

- Nausea, sickness and diarrhoea which usually improve over time.
- Stomach pain and indigestion.
- Kidney function – You will have a regular blood test to check that the kidneys are working properly and you may need urine tests.
- Abnormal liver function – You will have a regular blood test to check that your liver is working properly.
- Skin rashes.
- Blurred vision or reduction in hearing – You will have a regular ear and eye tests to check your hearing and sight.

If you develop any of these side effects or other symptoms which you think may be related to your medication, please call your specialist nurse.

It is important that you attend your appointments for your blood tests as these are to check whether deferasirox is causing any problems with your liver or kidneys.

Difficulties taking your medications

We know it can be difficult to take medication every day, especially if you are not having any symptoms from excess iron. However, it is very important to look after your health, as the damage caused by excess iron cannot always be reversed. If you are finding it difficult

to take your medication as often as you should then please do discuss this with someone from the haematology team, your doctor, nurse or psychologist. This is important so that we can understand why the medicine isn't working as well as we might expect it to and so we can work with you to try and make it easier for you to be able to take it.

Contact us

If you have any questions or concerns about iron overload or iron chelation.

Nazik Osei (clinical nurse specialist) Tel: 07825 978812

EnaAbena Akomah-Barnier (clinical nurse specialist) Tel: 07825 978812

Haematology Day Unit: 020 8725 1680

Apheresis Unit: 020 8725 0612

Dr Julia Sikorska (Lead sickle cell and thalassaemia consultant) Tel: 020 8725 0885

Dr Elizabeth Rhodes (Sickle cell and thalassaemia consultant): Tel: 020 8725 0885

Dr James Masters (Sickle cell and thalassaemia consultant): Tel: 020 8725 0885

Sickle cell and thalassaemia secretary Tel: 020 8725 0885

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.stgeorges.nhs.uk

Additional services

Patient Advice and Liaison Service (PALS)

PALS can offer you advice and information when you have comments or concerns about our services or care. You can contact the PALS team on the advisory telephone line Monday, Tuesday, Thursday and Friday from 2pm to 5pm.

A Walk-in service is available:

Monday, Tuesday and Thursday between 10am and 4pm

Friday between 10am and 2pm.

The Walk-in and Advisory telephone services are closed on Wednesdays.

Please contact PALS in advance to check if there are any changes to opening times.

PALS is based within the hospital in the ground floor main corridor between Grosvenor and Lanesborough wings.

Tel: 020 8725 2453 **Email:** pals@stgeorges.nhs.uk

NHS UK

The NHS provides online information and guidance on all aspects of health and healthcare, to help you make decisions about your health.

Web: www.nhs.uk

NHS 111

You can call 111 when you need medical help fast but it's not a 999 emergency. NHS 111 is available 24 hours a day, 365 days a year. Calls are free from landlines and mobile phones.

Tel: 111

AccessAble

You can download accessibility guides for all our services by searching 'St George's Hospital' on the AccessAble website (www.accessable.co.uk). The guides are designed to ensure everyone – including those with accessibility needs – can access our hospital and community sites with confidence.



Reference: HAE_IRO_01 **Published:** March 2025 **Review date:** March 2027