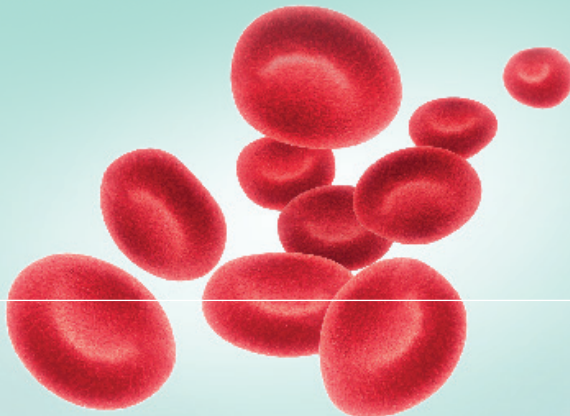


# University College Hospital

## Treatment options for iron overload

North Central London Haemoglobinopathy Network jointly with Whittington Health, Royal Free London, and Luton and Dunstable NHS Foundation Trust

Joint Red Cell Unit



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## What is iron overload?

Iron overload is when there is excess iron in the body. You can have iron overload if you:

- Are receiving regular transfusions with red blood cells – this is called transfusional iron overload and occurs because red cells contain iron. You are at risk of transfusional iron overload particularly if you are receiving regular top-up transfusions where you only receive blood and blood is not taken out at the same time. If you have exchange transfusions (usually only suitable for those with sickle cell disease) where blood is taken out and given at the same time, the iron may not build up as fast or even build up at all.
- Have a genetic disorder that causes you to absorb more iron from your diet than normal. This is common in genetic haemochromatosis, but can also be caused by a red cell disorder.
- Have been taking too many iron supplements – this is called iatrogenic iron overload.

The body has no natural way of getting rid of the excess iron, which then deposits in the organs and can cause organ damage.

## What are the symptoms of untreated 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 excess iron can damage:

- The heart – iron in the heart can cause irregular heart rhythms and lead to heart failure. This is less common in sickle cell disease, but was the reason why, in the past, thalassaemia patients who did not take their iron overload treatment properly died in their early 20s. Excess iron in the heart can also be a problem in hereditary haemochromatosis.
- The liver – iron in the liver can cause scarring of the liver known as fibrosis that 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 pancreas – iron in the pancreas can lead to diabetes. Some of the symptoms of diabetes include increased urination, feeling thirsty and 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.

## Measuring iron overload

There are different types of tests that may be performed to measure the level of iron in your blood and within certain organs. They are outlined below.

- Counting the amount of blood the person has received will give a good idea whether transfusional iron overload is likely and should be looked for.
- Ferritin and iron studies – ferritin is a protein that stores iron. Both ferritin and iron studies involve taking a blood sample to estimate the amount of iron in the body. Ferritin studies can be performed at any time, whereas iron studies are only performed when you have not had any iron-containing food for about 12 hours. The results from both studies are usually available within a few hours. As ferritin levels can also be high when you have an infection or liver inflammation, these studies may not always be enough to understand what is going on.
- Specialised MRI scans – these measure iron in the heart and liver and are an excellent way of monitoring the iron levels in people with significant iron overload. If this is the case, you will have the scans every year or so.
- Monitoring the effect of iron on the function of the organs – other blood tests can also be performed to detect how organs are functioning and to see if a build-up of iron has occurred in them. These tests may be for thyroid function, liver function, sex hormone levels and bone metabolism, all of which are done every three months in patients with iron overload. There is also a screening test for diabetes (glucose tolerance test) that is performed once a year.
- Occasionally, it is necessary to perform a liver biopsy. This is where a piece of liver tissue is taken and used to measure the iron content. Liver biopsies are rarely performed to assess iron levels as the MRI scans can provide this information. However, they may be performed for other reasons, for example if cirrhosis is suspected or if you have a liver virus such as hepatitis C.

We will give you information about any tests you will have.

## When is iron overload treated?

If you are receiving regular transfusions, the treatment for excess iron is usually started as the ferritin nears 1000ug/l.

If you have a red cell disorder and have a high ferritin but you are not receiving regular transfusions, you will usually have an MRI scan to calculate the amount of iron in your liver and heart. Once the team looking after you has the relevant information, they will agree on the most appropriate treatment for you.

If you received blood for a period of time but no longer need it, for example during treatment for a haematological cancer or during a bone marrow transplant, you will often have your iron overload treated when you are better and out of hospital.

If you do not have an inherited red cell disorder but have become blood transfusion-dependent later in life (for example due to bone marrow disease), your treatment will be tailored to your particular needs and circumstances. Research suggests that treatment does not always improve symptoms in this patient group and so the benefits of treatment will be carefully weighed against disadvantages to ensure that it is right for you.

If you have genetic haemochromatosis and are found to have significant iron loading, you will receive treatment to bring the ferritin level to about 30ug/l. If your iron overload is not high at the time of diagnosis, the team looking after you may suggest monitoring.

## How can iron overload be treated?

There are two ways to treat iron overload:

- Using medicine that removes extra iron from the body (chelation therapy)
- Removing blood (venesection).

## What is venesection?

Venesection involves taking blood out of your vein, done in a similar way to blood donation.

A nurse will identify a suitable vein in your arm and place a cuff around the top of your arm. After your skin has been cleaned with an antiseptic wipe, they will insert a needle or cannula (a small plastic tube) into the vein and attach a venesection bag. The bag is used to collect blood and is usually placed on weighing scales, so that we can monitor how much blood comes out.

To make up for the blood that was removed, the body makes new blood using the existing iron stores. If this happens repeatedly, the amount of iron in your blood is gradually depleted.

How much blood we take out, and how often and how many times you will need to have venesection, varies from person to person and will depend on your individual circumstances. Your doctor will discuss this with you in more detail.

Venesection is usually performed in the haematology, adolescent or paediatric day unit.

Please note that venesection will only be suitable for you if you are not currently receiving regular blood transfusions.

### Side effects include:

- Feeling faint – to help with it, please drink plenty of fluids before and after the venesection. If you feel faint despite this, we can give you IV fluids (fluids in your vein) or take less blood, or both.
- Bruising – you may get a bruise where needle was inserted.
- Anaemia (low red cell count) – this can happen if your venesections are frequent. If this happens, you may need to have a break from venesections or have them less often.

## What is chelation?

Chelation therapy is used to remove extra iron from the body. It uses a medicine that binds to iron and together they are released into the urine or faeces, or both.

There are three types of iron-removing medication used in chelation therapy:

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

Your doctor will discuss with you which type of therapy is best for you.

## Desferrioxamine (Desferal®)

There is a separate leaflet about this, **Desferal® treatment for iron overload** – please ask a member of staff for a copy if you have not received one.

### How do I take desferrioxamine?

This medicine can be given either under the skin (subcutaneously) or into a vein (intravenously). When it is given under the skin, special small needles are used, either Thalasset® or butterfly needles. If a needle under the skin is not suitable for you, a long-term intravenous line can be used. You and your nurse will develop a plan of care for looking after your long-term intravenous line.

### How often should I be using 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. Please ensure that you follow the instructions on how to take this treatment.



## 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.
- Ringing of the ears (tinnitus) and a decrease in night vision – it is important that you have hearing and eye monitoring every year.
- 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 the haematology advice line immediately.

## Storing desferrioxamine

- Keep it in the original packaging in the fridge.
- Don't freeze.
- Keep out of reach of children.
- Keep a check on the expiry dates.

## Deferiprone (Ferriprox®)

### How do I take deferiprone?

Deferiprone is a tablet that is taken three times a day. It is important that you use only the prescribed dose and check the expiry date. An occasional missed dose will not lead to complications but frequent missed doses will cause long-term problems, such as iron overload. If you take more than you should, please call the haematology advice line immediately.

## Possible side effects

- Infection – this medicine can reduce the body's ability to fight infection by lowering the levels of one of the types of white blood cells that respond to infection (neutrophils). Your doctor will ask you to have a blood test every two weeks to check that neutrophils are not affected. If you have a sore throat, temperature above 38°C, feel shivery or have any other symptoms that may suggest an infection, please contact the haematology advice line (contact details are on page 12). If it is out of hours, go to the Emergency Department (A&E) as you may need prompt treatment with antibiotics.
- Reddish brown colour of your urine, which may look alarming but will not cause long-term problems.
- Nausea and sickness – this can often be reduced by taking the tablets with meals.
- Increased appetite.
- Stomach pain.
- Joint pain.
- 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. However, this is most seen in patients taking desferrioxamine and is less common if you take deferiprone. If you experience any of these symptoms, stop your treatment and call the haematology advice line immediately.

## Storing deferiprone

You should keep this medicine out of reach of children and store it above 30°C.

## Deferasirox (Exjade®)

There is a separate leaflet about this, **Exjade® treatment for iron overload** – please ask a member of staff for a copy if you have not received one.

### How do I take deferasirox?

Deferasirox is taken once a day as a tablet which dissolves in water. It is important that you use only the dose prescribed and check the expiry date. An occasional missed dose will not lead to complications but frequent missed doses will cause long-term problems, such as iron overload. If you take more than you should, please call the haematology advice line immediately.

### Possible side effects

- Nausea, sickness and diarrhoea – these usually improve over time.
- Stomach pain and indigestion.
- Kidney problems – you will have a regular blood test to check that the kidneys are working properly.
- Abnormal liver function – you will have a regular blood test to check that your liver is working properly.
- Skin rashes.
- Blurred vision.
- Hearing problems.
- 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. However, this is most seen in patients taking desferrioxamine and is less common if you take deferasirox. If you experience any of these symptoms, stop your treatment and call the haematology advice line immediately.

If you develop any of these side effects, please call the haematology advice line. 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 kidney.

### **Storing deferasirox**

You should keep the medication in its original packaging and away from moisture. If the packaging has been damaged, you should not use the medication.

**It can be difficult to remember to take your iron chelators every single day so talk to the team looking after you about ways to help you with this. We have charts that may help you – let us know if you would like one.**

**The information in this leaflet is intended to be a guide only. Please discuss the specific details of your treatment with your haematology doctor and feel free to ask questions if there is anything that you are not sure of.**

## Contact details

### Haematology advice line (office hours, adults and children):

020 3447 7359

### Adult haematology advice line (out of hours):

07852 220 900

### Paediatric helpline (out of hours):

- nurse in charge 07961 081 645
- ward T11 south 020 3456 7890 ext. 71103 or 71143

### Apheresis:

020 3447 1803

**Address:** The Joint Red Cell Unit  
Department of Haematology  
3rd Floor West, 250 Euston Rd  
London NW1 2PG

**Website:** [www.uclh.nhs.uk/JRCU](http://www.uclh.nhs.uk/JRCU)

### Haematology consultants:

Professor John Porter  
Dr Sara Trompeter  
Dr Perla Eleftheriou  
Dr Emma Drasar  
Dr Bernard Davis  
Dr Farrukh Shah

### Specialist nurses:

Bernadette Hylton (adults)  
Nancy Huntley (apheresis)  
Nina Gorman (children)

## Where can I get more information?

### **Manufacturers' leaflets:**

Website: [www.novartisoncology.com/products/exjade.jsp](http://www.novartisoncology.com/products/exjade.jsp)

Website: [www.ferriproxtotalcare.com/docs/PatResPatientBooklet.pdf](http://www.ferriproxtotalcare.com/docs/PatResPatientBooklet.pdf)

### **Thalassaemia International Federation:**

Website: [www.thalassaemia.org.cy/wp-content/uploads/pdf/about-hb-disorders/About%20Thal\\_Chapt%204\\_Iron.pdf](http://www.thalassaemia.org.cy/wp-content/uploads/pdf/about-hb-disorders/About%20Thal_Chapt%204_Iron.pdf)

### **Sickle Cell Society**

Tel: 020 8861 7795

Website: [www.sicklecellsociety.org](http://www.sicklecellsociety.org)

### **NHS Sickle Cell and Thalassaemia Screening Programme**

Website: [www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview](http://www.gov.uk/guidance/sickle-cell-and-thalassaemia-screening-programme-overview)

### **UK Thalassaemia Society**

Tel: 020 8882 0011

Fax: 020 8882 8618

Email: [office@ukts.org](mailto:office@ukts.org)

Website: [www.ukts.org](http://www.ukts.org)

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**Space for notes and questions**

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