

Chelation therapy for iron overload

Information and advice for patients

Sickle Cell & Thalassaemia

What is iron overload?

Patients who have regular blood transfusions to treat their thalassaemia or sickle cell disease will get more iron than their body needs. This excess iron can cause damage to different parts of the body. Treatment for excess iron is known as chelation therapy and is usually started a year after regular transfusions

What are the symptoms of iron overload?

Symptoms of iron overload depend upon where the excess iron is deposited in your body. Often no symptoms are felt until the iron overload is severe. The excess iron can damage:

- **The heart:** Iron in the heart can lead to heart failure and irregular heart rhythms.
- **The liver:** Iron in the liver can cause scarring of the liver known as cirrhosis. Symptoms for cirrhosis include loss of appetite, tiredness, very itchy skin, jaundice (yellowing of the skin) as well as others. (Please note that this is not a full list of symptoms, and if you require further advice, please contact the SCaT centre.)
- **The pancreas:** Iron in the pancreas can lead to diabetes. Some of the symptoms of diabetes include increased urination, feeling thirstier, feeling very tired and unexplained weight loss. (Please note that this is not a full list of symptoms, and if you require further advice, please contact the SCaT centre.)
- **Hormone glands:** The thyroid may slow down which can cause tiredness. The sex hormone glands can also be affected, causing problems with fertility and developing secondary sexual characteristics (these are features you develop during puberty such as for females enlarged breasts and for males facial hair).

It is important iron overload is treated as early as possible to reduce or prevent damage from the excess iron building up.

Measuring iron overload

There are different types of tests that can be performed to measure the level of iron in your blood and within certain organs. These include:

- One blood test that you may have measures an iron storage protein called ferritin. If the ferritin amount is constantly raised it indicates that a build-up of iron has occurred. The ferritin level can be reduced to normal with medication.
- Other blood tests can also be performed to detect how other 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 level, certain vitamin and mineral levels as well as screening tests for diabetes (glucose tolerance test) and adrenal glands.

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- Occasionally, it is necessary to perform a liver biopsy. This is where a piece of liver tissue is used to measure the iron content.
- In certain cases, special scans of your heart may be required to assess whether a build-up of iron has occurred and to see how your heart is functioning. The best way of looking for iron in the heart is by MRI scans which measures the amount of iron in the heart and liver.

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

How is iron overload treated?

Chelation therapy is the term used to describe the process of removing extra iron from the body. The iron-removing medicine (the iron chelator) works by binding to the extra iron so that it can be removed from the body.

What are the benefits of chelation?

The benefit of using chelation therapy to get rid of excess iron will help to prevent damage to organs (including the heart, liver, pancreas and different glands).

What are the risks and side-effects of chelation?

There are different types of iron chelation medications and their risks and side-effects vary. Please read the whole leaflet for risks of each medication.

What are the risks of not having the treatment?

As blood contains iron which can build up in different parts of the body, if you do not take this medication, it can cause damage to these parts:

- **The heart:** This can be mild, moderate or severe and is picked up using a special type of scan called an MRI scan. Large amounts of iron in the heart can lead to heart failure and irregular heart rhythms. Iron in the heart is dangerous but it can be removed by using strong iron-removing medications.
- **The liver:** Iron overload in the liver can result in scarring of the liver which is also known as cirrhosis
- **Pancreas:** A large amount of iron in the pancreas can lead to diabetes. Diabetes is irreversible and is treated with insulin
- **Hormone glands:**
 - Underactive thyroid which can cause tiredness.
 - Sex hormone glands: In women periods may be delayed or irregular or stop. Later on this can affect fertility. In men, less testosterone is made, (testosterone is needed for muscle bulk, secondary sexual characteristics such as facial hair etc.).

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Are there any alternatives to this treatment?

There are no alternatives to iron chelation.

What are the different medications available for chelation therapy?

There are 3 types of iron-removing medication which can be used in chelation therapy:

- Desferrioxamine (also called Desferal)
- Deferiprone
- Deferisarox (also called Exjade)

Your doctor will decide which type of therapy is best for you depending on where the excess iron has built up in your body and what organs it is affecting. It is important that you use only the dose prescribed and check the expiry date.

Desferrioxamine (also called Desferal®)

How to take it?

This medicine is given either under the skin (subcutaneously) or into a vein (intravenously). When the medicine is given under the skin, special small and easy needles are used called Thalassets. If a needle into the skin is not suitable for you, then a long-term intravenous line can be used. These types of lines include hickman® lines and groshong® lines or PICCs and Portacaths need close monitoring. You and your nurse will develop a plan of care for looking after your long-term intravenous line. Nowadays, treatment has been simplified and desferrioxamine is available in more convenient ways for usage.

How often should I be using desferrioxamine?

Your doctor will determine the dose and frequency of your treatment. For treatment to be effective, you must make sure that the pump is properly connected and turned on as discussed with your doctor. To remove the extra iron and reduce the problems in the body, you need to use their pump continuously for at least 10-12 hours. Some patients prefer to be attached to their pump until it finishes; this takes 48 hours. An occasional missed dose will not cause a problem but frequent missed doses will cause long-term problems.

Possible side-effects

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

- Irritation or blisters on the skin where the needle is placed. Rotating the site of injections can avoid these problems. It is also important to ensure that the needle is properly positioned under the skin. You will have information from your nurse about what to look for if your treatment involves a long-term intravenous line.

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- Ringing of the ears (tinnitus) and a decrease in night vision. It is important that patients on desferrioxamine have regular hearing and eye monitoring every year.
- Certain bacteria grow on the excess iron that is removed; the most significant is a bacteria called Yersinia which can cause abdominal pain, fever, diarrhoea and vomiting.

If any of these symptoms occur, stop your treatment and seek medical help urgently.

Storing desferrioxamine

- Keep in fridge in the packaging.
- Do not freeze.
- Keep out of reach of children.
- Keep a check on the expiry dates.

There is another leaflet that explains how to take desferrioxamine so please ask if you require it.

Oral iron chelators

Other drugs that are available that can remove iron are deferiprone and desferasirox. Both these medicines can be taken by mouth (orally).

Deferiprone

Deferiprone is especially effective in removing iron from the heart.

How often should I be using deferiprone?

It is taken three times daily. It is important to use only the dose prescribed and to check the expiry date of the medication. An occasional missed dose will not cause a problem but frequent missed doses will cause long-term problems. If you take more than you should, you should contact your doctor immediately.

Possible side-effects

This medicine can reduce the body's ability to fight infection by lowering one of the types of white blood cells that fight infection. Your doctor will ask you to have a blood test performed every week to check that the cells that fight infection are not affected.

If you have a sore throat, temperature above 38°C, shakes or any symptoms suggestive of infection please contact either your doctor, or the SCaT centre (within working hours) or the Emergency Department (ED) if out of hours as prompt treatment with antibiotics may be needed.

Other possible side-effects include:

- Reddish brown colour of your urine, which may look alarming but will not cause long-term problem

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- Nausea and sickness (often reduced by taking the tablets along with meals)
- Increased appetite
- Stomach pain
- Joint pain

Storing deferiprone

You should keep this out of reach of children. Keep medication stored above 30°C.

Desferasirox

Desferasirox, also known as Exjade®, is particularly good at removing iron from the liver.

How often should I be using desferasirox?

It is given once a day as a tablet which dissolves in water. It is important to use only the dose prescribed and to check the expiry date of the medication. An occasional missed dose will not cause a problem but frequent missed doses will cause long-term problems. If you take more than you should, you should contact your doctor immediately.

Possible side-effects

- Nausea, sickness and diarrhoea; these usually improve over time.
- Stomach pain and indigestion
- Kidneys problems: Your doctor will ask you to have a blood test performed regularly to check that the kidneys are working properly.
- Skin rashes
- Blurred vision
- Hearing problems

If you develop these problems please contact your doctor. Please attend your appointments for your blood tests as these are to check whether the medicines are causing any problems with your liver or kidney.

Storing desferasirox

You should keep the medication in its original packaging and keep it 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 nurses and doctors at SCaT about ways you can achieve this. We have charts that we can print off to 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 doctor and remember to ask if there is anything that you are not sure of.

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Contact details

If you have any questions or concerns please contact the Sickle Cell and Thalassaemia Centre.

Sickle Cell & Thalassaemia Centre

Sandwell & West Birmingham Hospitals

City Hospital

Dudley Road

Birmingham

B18 7QH

Tel: 0121 507 6040

Monday, Wednesday & Thursday 9am – 5pm

Tuesday 9am – 6pm

Friday 9am – 4pm

The information in this leaflet is general and is intended to be a guide only. Please discuss the specific details of your treatment with your GP/doctor.

Further information for patients

Birmingham Sickle Cell & Thalassaemia Service

Soho Health Centre

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Sickle Cell Society

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For more information about our hospitals and services please see our websites *www.swbh.nhs.uk* and *www.swbhengage.com*, follow us on Twitter @SWBHnhs and like us on Facebook *www.facebook.com/SWBHnhs*.

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Sources used for the information in this leaflet

- Sickle Cell Society, 'Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK', 2008.
- National Institute for Health and Care Excellence, 'Sickle cell disease', 2010.
- Medicines and Healthcare products Regulatory Agency, 'Public Assessment Report Desferrioxamine Mesilate 500mg and 2g powder for injection,' 2008.
- British National Formulary, 'Deferiprone,' accessed April 2014.
- Apotex Europe, 'Package Leaflet: Information for the user – Ferriprox 500 mg film-coated tablets – Deferiprone', September 2010.
- British National Formulary, 'Deferasirox,' accessed April 2014.
- Novartis Europharm Limited, 'Package leaflet: Information for the user – EXJADE 125/250/500mg dispersible tablets – Deferasirox,' September 2013.
- Brent Sickle Cell and Thalassaemia Centre. 'A parents guide to managing sickle cell disease. Third edition. 2012

If you would like to suggest any amendments or improvements to this leaflet please contact the communications department on 0121 507 5495 or email: swb-tr.swbh-gm-patient-information@nhs.net



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