NHS
Sandwell and West Birmingham
NHS Trust

Iron Overload and Chelation Therapy

Information and advice for patients

Sickle Cell and Thalassaemia Centre, City Hospital

What is iron overload?

Patients who have regular blood transfusions to treat their thalassaemia or sickle cell disease will receive 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 commenced a year after regular transfusions.

What are the symptoms of iron overload?

Symptoms of iron overload depend upon where the excess iron is deposited. Often no symptoms are felt until the iron overload is severe. Do not wait for symptoms. It is important that you take your chelation therapy so that you do not get damage from the excess iron building up. 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.
- **The pancreas:** Iron in the pancreas can lead to diabetes. Diabetes is irreversible and is treated with insulin.
- **Hormone glands:** The thyroid may slow down which can cause tiredness. The Sex hormone glands can also be affected, causing problems with developing secondary sexual characteristics and perhaps fertility.

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. To decide if you have a build-up of iron in the body, different tests can be performed. 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. At SCAT we only check this every 3 months for thalassaemia patients. We do not check it for sickle cell patients as it is unreliable. The ferritin level can be reduced to normal with medication. It is important to point out that the ferritin level is a less reliable test for patients with sickle cell disease, please ask your nurse to explain why. If you have sickle cell disease and have iron overload it is important that you have MRI scans of your heart and liver – further information is below.
- 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.
- For all haemoglobinopathy patients having blood transfusions the best way of looking for iron in the heart is by MRI scans which measure the amount of iron in the heart and liver. It is important that you attend these appointments. We can try to request theses appointments to fit around your lifestyle. The MRI scan results can then be discussed with you in your clinic appointment.
- Very occasionally, it is sometimes necessary to perform a liver biopsy. This is where a piece of liver tissue is used to measure the iron content.

You will be given further information regarding the test you will receive.

How is iron overload treated?

Chelation is the term used to describe the process of removing the 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?

Using iron chelation medication 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 see below 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.).

Are there any alternatives to this treatment?

There are no alternatives to iron chelation.

Iron removing medications (iron chelators)

There are three types of medication available: they are called desferrioxamine (also called Desferal®), deferiprone and deferasirox (also called Exjade®). Your doctor will decide which type of therapy is best for you depending on where the excess iron has built up 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. In addition, PICCs (peripherally inserted central catheter) and Portacaths (implanted port) 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 Desferal® is available in more convenient ways for usage.

How often should I be using desferrioxamine (Desferal®)?
Your doctor will determine the dose and frequency of your treatment. For treatment to be effective, you must make sure that the pump or infuser is properly connected and turned on as discussed with your doctor/nurse. To remove the extra iron and reduce the problems in the body, you need to use your pump as directed. The infuser will take 48 hours to empty. You may have a schedule of 2 days on, 1 day off or you may need 1 infuser a week. Your nurse will help you fit your schedule around your life. An occasional missed dose will not cause a problem but frequent missed doses will cause long-term problems.

Possible side-effects
Desferal® 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. You can change the needle and site half way through the infuser (at 24hours) to reduce this problem. 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.
- Ringing of the ears (tinnitus) and a decrease in night vision. It is important that patients on Desferal® have regular hearing and eye monitoring every year.
- Certain bacteria grow on the iron 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. If you are having Desferal® through a long-term intravenous line then strict cleanliness is really important.

Storing Desferal®
Keep in a 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 Desferal® administration so please ask if you require it.

Oral iron chelators

Other drugs that are available that can remove iron are deferiprone and deferasirox. 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 usually given 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 ED (emergency department) if out of hours as prompt treatment with antibiotics may be needed.
- Reddish brown colour of your urine, which may look alarming but will not cause long-term problem
- 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

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

How often should I be using deferasirox?

Deferasirox it is now a film coated tablet, it was previously a tablet you had to dissolve in water. The tablet can be taken with a light meal or on an empty stomach. 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 Deferasirox (Exjade®)
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. There are 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 or nurse. Remember to ask if there is anything that you are unsure of; the team are always to happy to explain. .

How to contact the SCAT centre

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

Sickle Cell & Thalassemia Centre
Sandwell & West Birmingham Hospitals
City Hospital, Dudley Road
Birmingham B18 7QH
Tel: 0121 507 6040
City Hospital : 0121 554 3801

Opening Hours

Monday, 9am – 5pm
Tuesday, 9am – 6pm
Wednesday, Thursday and 9am – 5pm
Friday, 9am – 4pm
Saturday blood transfusion service only

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.

More information:

For more information please see our regional website:
www.westmidsstn.nhs.uk

For more information about our hospitals and services

Sandwell and West Birmingham Hospitals NHS Trust; website *www.swbh.nhs.uk* you can also follow us on twitter *@SWBHnhs* and like us on facebook *www.facebook.com/SWBHnhs*.

OSCAR
Organisation for sickle cell and anaemia research and thalassaemia support (OSCAR)

Birmingham: *www.oscarbirmingham.org*
Sandwell: *www.oscarsandwell.org.uk*

UK Thalassaemia Society 19 The Broadway, Southgate Circus, London, N14 6PH.Telephone: 020 8882 0011. *office@ukts.org*

Thalassaemia International Federation, PO Box 28807, 2083 Acropolis – Strovolos, Nicosia, Cyprus. *www.thalassaemia.org.cy*

For more information about sickle cell disease and the support available to you, visit the sickle cell society website: *www.sicklecellsociety.org*

You can also read more about sickle cell disease on the NHS Choices website: NHS Choices: *www.nhs.uk/conditions/sickle-cell-anaemia*

Sources used for the information in this leaflet:

- Sickle Cell Society (2018). *Standards for the Clinical Care of Adults with Sickle Cell Disease in the UK*. [Online]. Available at: https://www.sicklecellsociety.org/wp-content/uploads/2018/05/Standards-for-the-Clinical-Care-of-Adults-with-Sickle-Cell-in-the-UK-2018.pdf [Accessed 20th March 2024].
- National Institute for Health and Care Excellence (2012). *Sickle cell disease*. [Clinical Guideline 143]. [Online]. London: NICE. Available at: https://www.nice.org.uk/guidance/cg143/ [Accessed 20th March 2024].
- Medicines and Healthcare products Regulatory Agency (2017). *Public Assessment Report Desferrioxamine Mesilate 500mg and 2g powder for injection*. London: MHRA. Available at: https://mhraproducts4853.blob.core.windows.net/docs/e98f1f301ef437315e9e4b2ab1686d4e9b0962d6 [Accessed 20th March 2024].
- British National Formulary (2024). *Deferiprone*. [Online]. London: BNF. Available at: https://bnf.nice.org.uk/drugs/deferiprone/ [Accessed 20th March 2024].
- Apotex Europe (2010). Package Leaflet: Information for the user – Ferriprox 500 mg film-coated tablets – Deferiprone.
- British National Formulary (2024). *Deferasirox.* [Online]. London: BNF. Available at: https://bnf.nice.org.uk/drugs/deferasirox/ [Accessed 20th March 2024].
- Novartis Europharm Limited (2013). *Package leaflet: Information for the user – EXJADE 125/250/500mg dispersible tablets – Deferasirox*.
- Sickle Cell Society (2021). *A parent's guide to managing sickle cell diseases*. [Online]. Available at: https://www.sicklecellsociety.org/wp-content/uploads/2019/06/15121-Sickle-Cell-A-Parents-Guide-v7-OPT.pdf [Accessed 20th March 2024].
- United Kingdom Thalassaemia Society (2016). *Standards for the clinical care of children and adults with Thalassaemia in the UK*. [Online]. 3rd ed. Available at: https://ukts.org/3d-flip-book/standards/ [Accessed 20th March 2024].

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