Iron overload from blood transfusions:information for patients and parents

What is transfusional overload?

Every unit of transfused blood contains iron, held inside the red blood cells. That is entirely normal – it is how blood carries oxygen. The difficulty is that the human body has no way of getting rid of iron it does not need. We absorb iron, we use it, we store it, but we cannot excrete it.

So when someone receives blood transfusions regularly, the iron from those transfusions builds up, year after year. Over time it is deposited in organs – particularly the liver, the heart, and the glands that make hormones – where it can cause damage.

Iron overload is expected in anyone having regular transfusions. It is not a sign that something has gone wrong. It is a known, predictable part of transfusion treatment, and it is managed.

Who does this affect?

Anyone receiving regular blood transfusions is at risk, including people with thalassaemia major or intermedia, sickle cell disease, congenital dyserythropoietic anaemias, Pyruvate Kinase Deficiency, Diamond-Blackfan anaemia, and other inherited anaemias where transfusions are needed long term.

The more transfusions, and the longer the period over which they are given, the more iron accumulates. This is why the counting starts early: your team will begin monitoring iron before it has caused any problem at all.

Why does it matter if I feel fine?

This is the hardest part of iron overload to take seriously, and the most important.
Iron overload causes no symptoms for years. By the time it does cause symptoms, damage has usually already been done to the heart or the liver, and some of that damage may not be reversible. The whole strategy is therefore to find and treat iron build-up long before you would ever notice it.

That means taking a treatment you may not feel any benefit from, to prevent a problem you cannot feel coming. It is genuinely a lot to ask, and it is completely understandable that chelation (the treatment given to remove iron from the body) is one of the hardest treatments to keep going with. If you are struggling with it, please say so – there are usually options.

If iron overload is left untreated over many years, it can cause heart rhythm problems and

heart failure, liver damage and scarring, diabetes, and problems with growth, puberty and fertility. It is a major cause of people with inherited anaemias dying younger than they need to.

How is it monitored?

Serum ferritin is a blood test that gives a rough measure of how much iron is stored in the body. It is easy to repeat, so it is usually checked often. It is not perfect – it also rises with infection or inflammation – so it is read as a trend over time rather than a single number.Chelation is usually started once ferritin is >1000. For people on chelation, ferritin should usually not be allowed to fall <500.

Transferrin saturation measures how much iron is circulating in the blood. It will almost always be about 100% for someone on transfusions.

MRI scans measure iron in the liver and heart directly. These are much more accurate than ferritin, and importantly, liver iron and heart iron do not always go up and down together – you can have a reassuring liver scan and still be accumulating iron in the heart. This is why heart MRI is done as well, usually every one to two years, and more often if the level is high. The scan is painless and does not use radiation. However, for some people with severe claustrophobia it can be difficult to sit through. Ferriscans measure only iron in the liver and T2* MRIs measure iron in the liver and the heart.

Liver function tests and hormone tests are also checked periodically as well as looking for diabetes (eg fasting glucose test, oral glucose tolerance test or other).This is because There is no scan that can measure iron in the pancreas or the glands, so the only way to monitor is see if these are working normally is measuring their function.

How is it treated?

The main treatment is **iron chelation therapy**. Chelation medicines bind to iron and carry it out of the body in urine or stool. They are the only way to remove the excess.

Three chelation medicines are used in the UK:

Deferasirox (Exjade, Jadenu) is taken by mouth once a day as a tablet. It is the one most people start with.

Deferiprone (Ferriprox) is taken by mouth as a tablet or liquid, usually three times a day. It is particularly good at removing iron from the heart, so it is sometimes chosen or added for that reason. It requires regular blood tests to check the white cell count, as it can rarely cause a serious drop.

Desferrioxamine (Desferal) is given as an infusion under the skin, usually overnight over several hours, several nights a week. It was the original chelator and remains very effective. It is used less often now that tablets are available, but it is still used when the tablets are unsuitable, or alongside them when iron levels are very high.

Sometimes two chelators are used together, if iron levels are high or the heart is affected.

Which medicine suits you depends on your iron levels, which organs are affected, your age, other medical conditions, and – legitimately – what you can actually live with. A chelator you take is better than a better chelator you do not take. Side effects should be reported rather than endured, because there is usually something that can be adjusted.

Never take iron supplements or multivitamins containing iron unless your haematologist has specifically told you to. Check the labels of over-the-counter supplements, as iron appears in many of them.

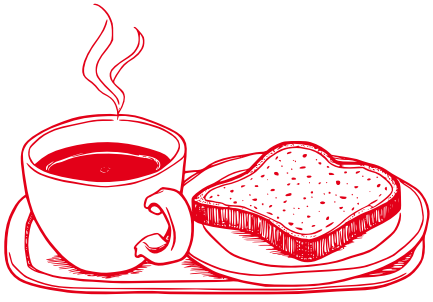
Vitamin C affects how iron is handled and can increase the amount of iron released into the blood, so ask your team before taking vitamin C supplements. This does not mean avoiding fruit and vegetables.

What can I do to help myself or my child?

Take the chelation. That is the single biggest thing, and it is the thing most worth asking for help with if it is not happening. If the tablets make you feel sick, if the routine has fallen apart, if a teenager has quietly stopped taking them – all of these are common, and all of them are fixable. They are not fixable if nobody knows.

Attend the monitoring, including the MRI scans, even when you feel well. Especially when you feel well.

Tea with meals slightly reduces iron absorption from food. This is a small effect and no substitute for chelation, but it does no harm.



Alcohol adds to the strain on a liver that is already handling excess iron, so it is worth moderating.

Ask your team what your current ferritin and scan results are, and what they are aiming for. Many people find that knowing the numbers, and seeing them move, makes the treatment feel less like a task with no feedback.

Iron overload is manageable. People who chelate consistently can expect their iron levels to come down and their organs to be protected.

When to seek medical advice

Contact your team if you notice:

- Palpitations, an irregular heartbeat, or breathlessness
- Increasing tiredness that is different from your usual
- Abdominal pain, especially on the upper right side
- Joint pain
- Increased thirst or passing a lot of urine (possible diabetes)
- In children or teenagers: delayed growth or delayed puberty
- Any side effect from chelation that is making it hard to take
- Fever or infection, if you are taking deferiprone – this needs a blood test promptly

Can we help?

CAN (Congenital Anaemia Network) is a charity specifically for patients with inherited anaemias. It was started by a group of patients and doctors, working together to tackle some of the problems of having an inherited anaemia – the lack of information and the loneliness of not knowing anyone else with your condition. We run events for patients to meet each other, as well as provide some additional information, support and services for patients who need that extra bit of help.

If you want to know more visit www.togetherwecan.uk or email us: info@togetherwecan.uk

Do you know your care should involve a specialist centre for inherited anaemias? These conditions are rare enough that few doctors see many of them and all patients with rare inherited anaemias should be under the care of a haemoglobinopathy specialist. Please read our 'How red cell services are organised in England' information sheet. If you don't know who your specialist service should be, please get in touch with us so we can help you find out.