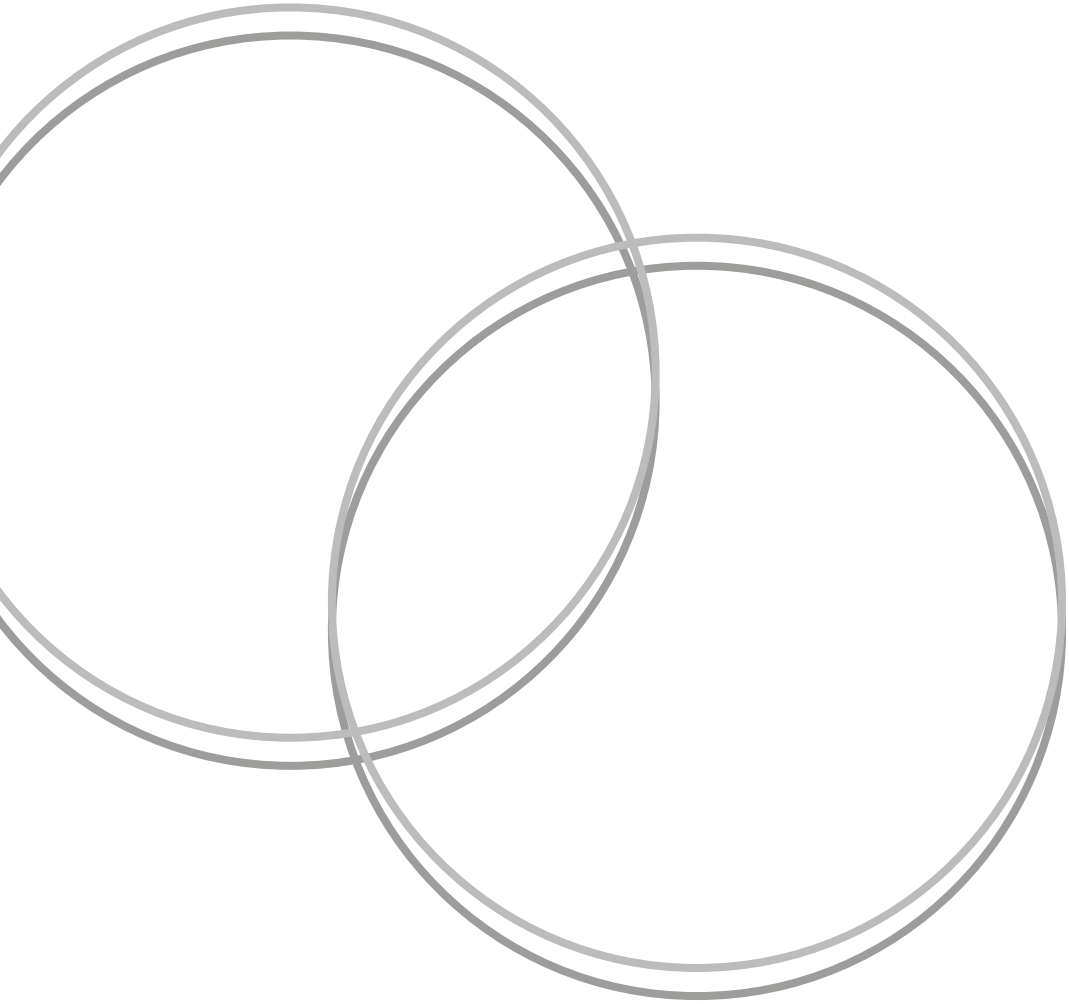
Oxford University Hospitals
NHS Foundation Trust

Autoimmune haemolytic anaemia

Information for patients



Your doctor has given you this leaflet because you have suspected autoimmune haemolytic anaemia. This leaflet will explain the condition, how it is diagnosed and how it is treated. If you have any questions, please do not hesitate to discuss this with your doctor.

What is autoimmune haemolytic anaemia (AIHA)?

Autoimmune haemolytic anaemia (AIHA) is a rare blood condition that causes the number of red blood cells in your body to be reduced.

How does this happen?

Antibodies are proteins which are made by the immune system. These proteins normally attach to the surface of bacteria. They act like labels to tell the immune system to destroy the bacteria. In AIHA, the immune system (which normally defends the body against infection) makes an antibody against its own red blood cells. This causes break down of the red blood cells called haemolysis.

Red blood cells carry oxygen from the lungs to the organs and usually live for 120 days. The body is constantly making new red blood cells to replace the old ones. In AIHA the immune system attacks healthy red blood cells in error. Red blood cells are broken down more quickly than they can be replaced, resulting in anaemia.

Anaemia is the condition of having less haemoglobin (the substance on the red blood cells which carries oxygen) than normal. This usually means a reduced number of red blood cells are present.

Why does this happen?

For many people, the cause of AIHA is not obvious. This is called idiopathic or primary AIHA.

Some people develop AIHA due to other factors; these people have secondary AIHA. Secondary AIHA may be caused by:

- infection e.g. Hepatitis C, HIV, cytomegalovirus (CMV), Epstein-Barr virus (EBV), tuberculosis
- cancer e.g. lymphoma, chronic lymphocytic leukaemia (CLL)
- other autoimmune conditions e.g. SLE (systemic lupus erythromatosus), scleroderma
- certain drugs.

What are the symptoms of autoimmune haemolytic anaemia (AIHA)?

Normal haemoglobin levels are about 130-180 grams per litre of blood in men and 120-170g per litre in women. When haemoglobin drops below this level, it may cause a number of symptoms, such as:

- breathlessness
- tiredness
- heart palpitations
- chest pain
- headache
- pale skin.

AIHA may also have other effects on the body, due to the products of broken-down red cells.

You may also experience:

- yellowing of skin and whites of the eyes (jaundice)
- dark urine,
- gallstones
- abdominal fullness due to an enlarged spleen (splenomegaly).

Diagnosing AIHA

As your doctor suspects that you have anaemia, you will have a blood test to look at your 'full blood count'. This checks the number of red blood cells and amount of haemoglobin in your blood.

If the full blood count shows anaemia (low red blood cells and low haemoglobin), your doctor may do more blood tests.

Further blood tests for AIHA are:

- Raised bilirubin and lactate dehydrogenase (LDH) levels. This looks for evidence that your red blood cells are being broken down.
- Looking at your blood film under microscope for changes in red blood cell shape.
- Direct antiglobulin test (DAT) also called Coombs' test (DCT). This test will identify whether your red blood cells are covered in antibodies produced by your body, which occurs in AIHA.

In most cases of AIHA there is no underlying disease, but your doctor may want to order further tests, such as a CT body scan or a bone marrow biopsy, to exclude an underlying cause. Further blood tests may also be ordered to look for infections.

Treating AIHA

There are a number of different treatments for managing AIHA. If you are found to have AIHA, the type of treatment your doctor will recommend will depend on how bad your symptoms are, your overall health and whether you have primary or secondary AIHA.

If your AIHA has been caused by an underlying infection or cancer (secondary AIHA), these problems may be treated by your doctor first. If your doctor believes a medication is causing your AIHA, you may need to change to an alternative drug.

Treatments for AIHA

Steroids

Most people are prescribed steroids such as prednisolone to treat their AIHA. These drugs work by dampening down your immune response which will slow down the breakdown of red blood cells.

Steroids are a good treatment for AIHA and most patients only need a short course of treatment.

What are the side effects of steroids?

Steroids normally cause no problems if they are given for a short period of time. People often feel hungrier when taking steroids and may put on weight.

If you need repeated courses of treatment or have to take steroids for a long time, you may have other side effects, such as:

- Increased risk of bacterial and viral infections (such as colds). This is because steroids weaken your immune system.
- Thinning of the bones (osteoporosis)
- Stomach ulcers
- High blood sugar levels (diabetes)
- A change in your facial appearance
- Thinning and bruising of the skin.

These side effects usually reverse when the steroids are stopped. Your doctor may also prescribe you medications to prevent stomach ulcers and bone thinning if you are taking steroids for a long time.

Can I change my steroid dose?

It is very important that you do not stop taking steroids without advice from your doctor, as your body starts to rely on them. They need to be cut down slowly to so that your body has time to adjust, otherwise you experience weakness and fatigue. If you are worried about possible side effects, please discuss your treatment with your doctor before making any changes to your medication.

What are the alternatives to steroids?

Although steroids are an effective treatment for many patients with AIHA, they are not right for everyone. Your doctor may recommend that you try another treatment for your AIHA. This could be because:

- Your did not respond to steroids at all.
- You responded but then 'relapsed'. This means your red blood cell count fell back down once your steroid dose was cut.
- You have a medical condition (e.g. diabetes) that could be made worse with steroid treatment.
- Some people also choose not to take steroids because of side effects they have had before.

There are several options for treating AIHA if steroids aren't right for you. The other possible treatments are:

- rituximab
- splenectomy
- intravenous immunoglobulin (IVIG)
- other immunosuppressant medications.

Rituximab

Rituximab is a drug that was first used to treat cancer, but is now used to treat AIHA along with several other autoimmune conditions. Like steroids, rituximab dampens down the immune response and stops red blood cells from being broken down too quickly. It is an antibody (developed by a medicines company) that affects your white blood cells. It is not made from donated human blood.

Rituximab is given as an infusion through a drip (a small tube into a vein in your arm) once a week for four weeks. It takes a couple of hours for the dose to be given. It usually takes a few weeks for rituximab to work, although some people respond many months after treatment.

What are the advantages of rituximab?

Many patients have a good response to rituximab infusions, and if relapse occurs, the treatment can be repeated months or years later if needed.

What are the side effects and risks of rituximab?

Most people who are treated with rituximab for AIHA have no side effects. The most common problem is a reaction to the infusion (such as a fast heart rate or breathlessness), but you will be monitored closely while it is given.

Rituximab works by stopping your body making antibodies and may potentially increase the risk of infections for the next 12 months. Most patients do not have problems. There is an extremely rare viral infection that can affect the nervous system, which a few people treated with rituximab have had. Before receiving rituximab, you must also be screened for hepatitis B (a viral infection), as rituximab can make this infection more serious.

Splenectomy

Splenectomy is the removal of the spleen. As red blood cells are mainly destroyed in the spleen, splenectomy can cure the AIHA.

Splenectomy is carried out in an operation under general anaesthesia (where you are made to be unconscious). The operation is normally done laparoscopically using very small cuts in the skin (sometimes called 'keyhole surgery'). With this type of operation, you should recover quickly, with only a few days spent in hospital.

Sometimes the operation needs to be carried out using an open surgery, which uses a larger cut. Your surgeon will discuss this with you if they think you are likely to need an open operation.

What are the advantages of splenectomy?

Splenectomy offers the best chance of curing AIHA. Around 2 out of 3 patients do not need any additional therapy after their spleen has been removed. In most people, red blood cell levels in the blood start to increase within the first 2 months after surgery.

What are the risks of splenectomy?

As with any surgical operation, splenectomy has risks. You may have a higher risk of complications from surgery if you have other medical conditions or are very overweight. Your doctor will discuss your own situation and specific risks with you.

Possible complications or risks of splenectomy include:

- reaction to the general anaesthetic
- bleeding during surgery
- damage to other organs
- infection
- death. If 500 people have a laparoscopic splenectomy, 1 may die from the operation, either at the time of surgery or from complications happening afterwards. This risk is nearer to 1 in 100 for the open (non-keyhole) surgery.

Splenectomy can increase your risk of infection as some of the white blood cells that help the body to fight bacteria and viruses are made in the spleen. To reduce the risk of infection in the long term,

you will need to have a number of vaccinations (immunisations or 'jabs') before having surgery. After the surgery, you may have to take long-term low doses of antibiotics to help prevent infection, or you may be given a packet of antibiotics to keep at home in case you become unwell.

You must seek medical advice quickly if you develop symptoms of an infection and you should carry a card to say that you have had your spleen removed in case you are in an accident. Your doctor and surgeon will discuss these details with you.

Intravenous immunoglobulin (IVIG)

This is a medicine containing antibodies (immunoglobulin). Antibodies are produced by white blood cells to fight infections. Immunoglobulins are a human product, which means that they have been collected from numerous blood donors. This medicine is given into a vein using a drip (intravenously). It is given as an infusion (liquid mixture) over a few hours. Nobody understands exactly how IVIG works to treat AIHA.

What are the advantages of IVIG?

IVIG works quickly, usually within a few days. For this reason, it is sometimes used as an emergency treatment for people with very severe anaemia. Unfortunately, the effect of IVIG doesn't last long (a few weeks at most), so it will not cure your AIHA.

What are the risks of IVIG?

There is a small risk of a reaction (such as fast heart rate or breathlessness) while the IVIG is being given, so you will be monitored closely by a nurse.

There is also small risk of developing a rare complication called aseptic meningitis. This causes headache, neck stiffness and a dislike of bright lights. This condition usually gets better on its own, but if you develop these symptoms after treatment with IVIG you should contact the triage number at the end of this leaflet so they can assess you.

IVIG can very rarely cause kidney damage and blood clots. There is an extremely small risk (less than one in many millions) of infections

such as hepatitis and HIV (as it is made from donated blood). Once you have received IVIG you will not be able to donate blood in the UK, even when you recover from AIHA.

Other immunosuppressant medications

A range of other medications that suppress the immune system can also be used to treat AIHA. Some examples of these are: azathioprine, cyclosporine, cyclophosphamide and mycophenolate. These medications work in slightly different ways, but all reduce the number of white blood cells in your body. Like steroids, these medicines increase your chance of getting infections.

How to contact us

Please speak to your doctor in clinic if you have any concerns or questions about this leaflet or would like further explanations.

If you have problems after any of the treatments please call Haematology Triage at the Churchill Hospital. If you are worried that your symptoms have become worse, you can ask your GP to repeat your blood count, or contact the Haematology Secretary.

Cancer and Haematology Centre Triage

Tel: **01865 572 192**
(24 hours)

Haematology Secretary

Tel: **01865 235 880**
(9am-5pm)

If you need an interpreter or would like this information leaflet in another format, such as Easy Read, large print, Braille, audio, electronically or another language, please speak to the department where you are being seen. You will find their contact details on your appointment letter.

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