

Warm autoimmune haemolytic anaemia

This leaflet is for adult patients diagnosed with warm autoimmune haemolytic anaemia (AIHA). If you have any questions or concerns after reading this leaflet, please ask your doctor or nurse.

What is warm autoimmune haemolytic anaemia?

Warm autoimmune haemolytic anaemia (AIHA) is a condition in which the body's immune system breaks down red blood cells.

Red blood cells contain haemoglobin; this carries oxygen from the lungs to the rest of the body. The normal level of haemoglobin in the blood is more than 115g/L for women and more than 120 g/L for men.

When a patient has AIHA the bone marrow tries to compensate for the broken down red cells by making more, but with time the level of haemoglobin drops (anaemia) and patients typically feel tired or breathless. When red cells break down, the products from them may cause patients to appear jaundiced (a yellow colour in the eyes or skin). The condition is described as 'warm' because the red cell breakdown occurs at normal body temperature rather than in the cold.

What causes warm autoimmune haemolytic anaemia?

In 50% of patients there is no apparent cause for AIHA, in which case it is described as 'idiopathic'. In the other 50% of patients there may be an associated condition, often another autoimmune disease such as Rheumatoid Arthritis or Systemic Lupus Erythematosus (SLE) or a disorder of the white cells that make up the immune system such as lymphoma. Occasionally, some medications can be associated with the condition.

What tests are used to diagnose and monitor AIHA?

AIHA is diagnosed from blood tests; these allow us to measure the haemoglobin level and to detect the antibodies the immune system makes, which are the cause of the red cell breakdown. Blood tests also allow us to measure the extent of red cell breakdown, which we call haemolysis. The red cell breakdown produces bilirubin (which causes the yellow colour) and a protein called LDH (lactate dehydrogenase) give a useful measure of how much haemolysis is occurring. The reticulocyte count measures the youngest red cells and is a measure of how quickly the body is making new red cells to make up for the breakdown. Patients with AIHA normally have a combination of blood tests, scans and sometimes biopsies to establish if there is an associated underlying cause.

What is the treatment for AIHA?

Treatments in AIHA are primarily directed to suppress or alter the body's immune system to prevent breakdown of red cells. In addition, a vitamin called folic acid is important for helping

the body to make new red blood cells. The most suitable treatment depends to some extent on whether there is an underlying condition. The following describes treatments commonly used in AIHA.

- **Steroids:** The most common first treatment for AIHA is with steroids, for example prednisolone, which is taken as several tablets once a day in the morning. Steroids are an effective treatment and improve the haemoglobin in 8 out of 10 patients, although this may take a period of 1-2 weeks. Often steroids are given at relatively high doses initially then gradually reduced over time. For most (again 8 out of 10) of these patients the haemoglobin will fall again in the future, often after steroid treatment has stopped or reduced to levels close to the body's normal steroid production. If treatment is needed again then steroids are often used initially but high doses cannot be used for too long periods due to the side effects, so alternative treatments are usually considered.

Steroid side effects: As with any medication different people experience different side effects to different degrees depending on the doses used. The most common issues with steroids are:

- Raised blood sugars, which may require medication.
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- Increased risk of infections.
- Changes in mood and poor sleep.
- Irritation of the stomach (medication is normally used to reduce this).
- Thinning of the bones (osteopenia) or weakened muscles, if long courses are used.

Patients are commonly prescribed other medications with the steroids to reduce these risks.

If you have taken steroids for more than 2 weeks is important not to stop them suddenly without the advice of a doctor or you may become unwell.

- **Intravenous immunoglobulin (IVIG):** IVIG is a medicine containing antibodies (immunoglobulin) which is given into a vein, usually in your arm, through a drip (intravenously) over a few hours. Antibodies are produced by white blood cells to fight infections. IVIG is made from donations from numerous blood donors. IVIG may be used if red cell breakdown is very severe, particularly if blood transfusion is needed.

IVIG side effects: The most common side effects of IVIG are headaches, muscle aches or fevers. There is a small risk of a reaction (such as a fast heart rate or breathlessness) while the IVIG is being given, so you will be monitored closely by a nurse. IVIG can very rarely cause kidney damage or blood clots. If you have new symptoms after IVIG then it is important to discuss them with your treating doctors and nurses.

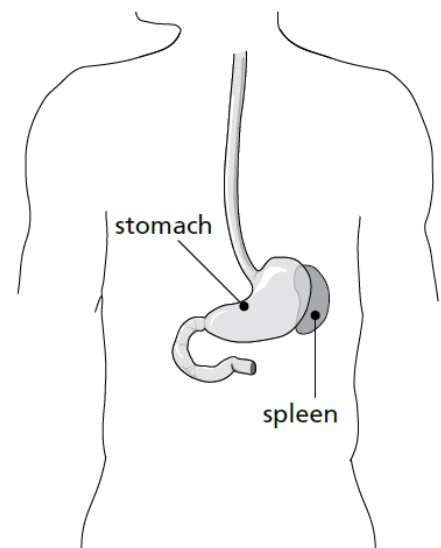
- **Rituximab:** Rituximab is a drug which was first used to treat cancers of the immune system, but has also been used for many years to treat AIHA. It is a manufactured antibody made in a laboratory rather than from donated human blood. Rituximab is given as an infusion through a drip (a small tube into a vein in your arm), once a week for four

weeks. Each infusion takes a few hours. It usually takes a few weeks for rituximab to work, although in some people it takes longer than this.

At least 3 out of 4 people given rituximab will have an improvement in their anaemia and on average this lasts around 2 to 3 years. In time, the haemoglobin drops again for most patients. If rituximab works well for you, the treatment can be repeated if needed.

Rituximab side effects: Most people who are treated with rituximab for AIHA experience few side effects. The most common problem is a reaction to the infusion at the time (such as a fast heart rate or breathlessness), but you will be monitored closely while it is given. There is an increased risk of infections, although serious problems are rare. Sometimes viruses a patient has had in the past can become active again. For example, the chicken pox virus may become active again as shingles, and your doctors will check you have not had some particular viruses such as hepatitis B before giving you the treatment.

- **Other immunosuppressants:** Other medications that suppress the immune system can also be used to treat AIHA. Examples of these drugs are azathioprine and mycophenolate mofetil (MMF), which are taken as tablets. These drugs are mainly used to reduce the doses of steroids required to minimise steroid side effects. Their main side effects are the risk of infections and they require monitoring of blood tests to be used safely. They may also cause other side effects such as rashes, sensitivity to sunlight, gastrointestinal symptoms or headaches. If your doctors think one of these drugs might be helpful for you they will discuss it in more detail.
- **Splenectomy (removal of the spleen):** Removing the spleen in an operation has been used as a treatment for AIHA for a long time, although it is performed less often in recent years due to the development of new treatments such as rituximab. It is effective for around two-thirds of patients in improving their anaemia, although sometimes only after a period of several months. For one half of these patients who improve with splenectomy the effect is long lasting, but for the other half the anaemia returns, although lower doses of steroids or immune suppressing drugs may then be needed compared to before splenectomy. The operation can usually be done laparoscopically (using very small cuts to carry out keyhole surgery), which means you should recover more quickly. Sometimes, the operation needs to be carried out using open surgery (a larger cut). Your surgeon will discuss this with you if they think you are likely to need this type of operation.



Risks of splenectomy: Like any surgical operation, splenectomy has risks which should be discussed with the surgeon. Based on previous patients, we know on average that if 500 people have the operation by keyhole surgery, one may die because of the operation;

either at the time of surgery or from complications happening afterwards. This is nearer to 1 in 100 people who have open surgery.

Other risks include:

- Reaction to the general anaesthetic.
- Excessive bleeding at the time of surgery which may require blood transfusion.
- Damage to other organs during the operation.
- Infection.

Your doctor will discuss your own situation and specific risks with you.

To reduce the risk of long term infection we advise patients to have vaccinations and to take long-term low dose antibiotics afterwards to help prevent infection. This is because the immune system is less able to fight infections without the spleen.

What happens now?

This leaflet tells you about different treatments for AIHA. Your doctor may have recommended one or more of these treatments. You should discuss any questions you might have about these treatments with your doctor, so you can make a decision together about which ones are appropriate for you.

Contact us:

If you have queries about your medication or issues such as blood tests please contact the Haematology West Ward 0118 322 6632 (Monday-Friday 8.30am-5pm). The nurses will either answer your query or pass a message to one of the haematology doctors.

Outside of these times if you are unwell and require medical attention contact your GP out of hours service or visit the Emergency Department (A&E). Take your tablets or a record of them with you. The GP or emergency doctor will be able to contact a haematologist for advice if required.

If you have queries about clinic appointments, please phone the Haematology Clinical Admin Team (CAT 8): 0118 322 8145 option 2 or email rbbh.CAT8@nhs.net (9am-5pm), Monday to Friday.

Important phone numbers:

West Ward Haematology Day Unit: 0118 322 6632 (nurses station)

West Ward Reception Desk / enquiries: 0118 322 7464

Haematology clinical nurse specialist: 0118 322 7689

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