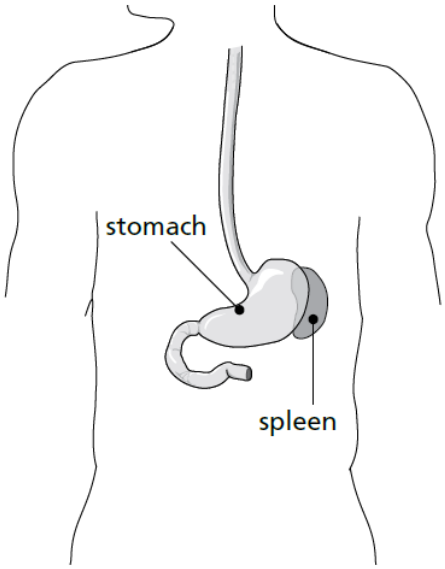
Immune Thrombocytopenic Purpura (ITP)

This leaflet is for adult patients diagnosed with Immune Thrombocytopenic Purpura also known as Immune Thrombocytopenia or ITP, a condition with an increased risk of bleeding due to low levels of platelets caused by the body's immune system. If you have any questions or concerns after reading this leaflet please ask your doctor or nurse.

What is ITP?

Immune thrombocytopenic purpura (ITP) is a condition which causes the number of platelets in your blood to be reduced (thrombocytopenia). Platelets are small cells in the blood which work as the first line of defence to stop bleeding and bruising (purpura) after an injury. If you do not have enough platelets in your blood, you are likely to bruise easily or may be unable to stop bleeding without medical help if you cut yourself. Other common symptoms are nose bleeds or bleeding gums and for women to have heavy periods.



In ITP the platelets are reduced because of the body's immune system. White blood cells in your blood, bone marrow and spleen (an organ in your abdomen) are part of your immune system. If you develop an autoimmune condition, your immune system can become overactive and destroy things it shouldn't, such as your own platelets. This often happens mainly in the spleen. ITP is a type of autoimmune condition (auto means 'against yourself').

ITP behaves differently in different people. For example, children usually get ITP after a viral infection and normally get better on their own without treatment, whereas ITP in adults more frequently needs treatment to stop or prevent bleeding. Some people with ITP have other conditions including immune conditions like rheumatoid arthritis, infections such as hepatitis or HIV, or other blood disorders. If you have any of these medical issues, your ITP may be treated differently.

The platelet count

A normal platelet count is between 150 to 450 thousand million platelets per litre of blood. This is usually referred to by doctors and on test results just using the first numbers, such as '150' or '450'. You are unlikely to get bleeding or bruising symptoms unless your platelet count is below 30, so treatment isn't necessarily needed just because the platelet count is low. If a medical procedure or operation is planned it may be necessary to give treatment to raise the platelet count to reduce the risk of bleeding even if you have no symptoms.

What tests are used to diagnose ITP?

There is no single test which can diagnose ITP. The diagnosis is made mainly by a combination of the symptoms you have had, a doctor examining you and blood tests. For some patients further tests may be advised such as scans or taking a sample from the bone marrow. This may be recommended when you are first diagnosed or later on if there is a change in your condition.

What is the treatment for ITP?

Treatments in ITP are primarily directed to suppress or alter the body's immune system to prevent destruction of platelets, or in some cases to increase production of platelets. In addition, a drug called tranexamic acid is often used to help the body form stronger blood clots and particularly to help with bleeding symptoms such as gum bleeding, nose bleeds or heavy periods. Tranexamic acid is a tablet that is normally taken three times a day and is a very safe drug when used at standard doses but is generally avoided when there is visible blood in the urine due to the risk of blood clots blocking the drainage of the urine.

Steroids:

The most common first treatment for ITP is with steroids, for example prednisolone which is taken as several tablets once a day in the morning. Steroids work by damping down the immune system, reducing the destruction of platelets. Steroids are an effective treatment and improve the platelet count in 8 out of 10 patients. Often they are given at relatively high doses initially then gradually reduced over time. For most (8 out of 10) patients the platelet count will fall again in the future, although treatment may not be needed. If treatment is needed again then steroids are often used initially but high doses cannot be used for long periods due to the side effects of steroids 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.

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 immunoglobin (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 is a safe product but due to restrictions that apply to all blood products if you receive IVIG you will not be able to donate blood in the UK, even when you recover from ITP.

IVIG works quite quickly, usually within a few days and more than 8 out of 10 patients will have a good response, but the effect doesn't last long (a few weeks at most) and so it will not cure your ITP. It is generally given in more urgent situations or before surgery or procedures.

IVIG side effects:

The most common side effect of IVIG is headaches and 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. There is a risk of developing short-lasting anaemia after the infusion and 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.

Splenectomy (removal of the spleen)

Removing the spleen in an operation has been used as a treatment for ITP for a long time, although it is performed less often in recent years due to the development of new treatments. It is effective for around two thirds of patients, who then do not need any more treatment. Sometimes a special scan called an Indium Platelet scan is used to help predict how likely it would be to be effective. 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. You may need some treatment to increase your platelet count before the 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 if 500 people have the operation by keyhole surgery, 1 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 (non-keyhole) surgery.

Other risks include:

- Reaction to the general anaesthetic.
- Excessive bleeding at the time of surgery (which may happen even if you have a normal platelet count).
- 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.

Rituximab

Rituximab is a drug which was first used to treat cancers of the immune system, but has also been used for nearly 20 years to treat ITP. It is a manufactured antibody (developed by a medicines company), 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. Each infusion takes a few hours. It usually takes a few weeks for rituximab to work, although some people respond months after treatment.

Around two out of three people given rituximab will have an improvement in their platelet count. In time, the platelet count will usually drop again but often the platelet count stays at a good level for at least a year. If rituximab works well for you, the treatment can be repeated later if needed.

Rituximab side effects:

Most people who are treated with rituximab for ITP have very 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.

TPO receptor agonists: romiplostim (Nplate™) and eltrombopag (Revolade™)

Thrombopoietin (TPO) is a hormone made by the liver which tells the bone marrow to make more platelets. TPO receptor agonists work in a similar way on the bone marrow. At present these drugs can only be used if you have already had other treatments for ITP and you have had your spleen removed (a splenectomy) or if you cannot have a splenectomy. Most patients have an improved platelet count with these drugs, although only around half with have a lasting response. Monitoring with blood tests is required to check the platelet count is not too high and guide the doses. Once you have started these treatments you will need to continue taking them for as long as your ITP persists, which may be many years. They do not cure the underlying problem, they just tell your body to make more platelets to replace the ones that are being destroyed.

Romiplostim (Nplate™) is given by an injection under the skin, usually once per week. You can be taught to give this injection yourself.

Eltrombopag (Revolade™) is a tablet which is taken once a day. It can't be absorbed by the gut if there is calcium nearby, so you must not eat foods high in calcium for two hours before or 4 hours after you take it. Most people find it easiest to take the medication early in the morning or just before going to sleep. Foods high in calcium include dairy products, cereals, tinned fish with bones and green leafy vegetables. Your pharmacist can give you more information about which foods to avoid.

TPO receptor agonist side effects:

Most people have few side effects with these drugs. The most common side effects are headaches, nausea or diarrhoea and altered liver blood tests. There may be a small risk of blood clots (which can be in the legs or lungs, or cause heart attacks or strokes) in people whose platelet count goes up to high levels. Your doctor will monitor your platelet count carefully while you are on this medication, to help avoid this. Sometimes your platelet count can go up and down a lot when you start these medications. This means that you will need quite frequent blood tests and clinic visits when you start to take this medicine.

Other immunosuppressants

Other medications that supress the immune system can also be used to treat ITP. Before rituximab and the TPO receptor agonists became available they were often used. They are used less often now, as they are thought to have more side effects than the newer treatments. They may be tried instead of, or alongside, the newer drugs if you don’t have a satisfactory response. Examples of these drugs are azathioprine, mycophenolate mofetil (MMF), cyclophosphamide, vincristine or ciclosporin. All are given as a tablet, except vincristine, which is given as an injection into a vein. If your doctor recommends one of these drugs they will tell you more about it.

Helicobacter Pylori treatment

Some people with ITP have an infection in their stomach, known as Helicobacter pylori. Sometimes, treating this infection with antibiotics and antacids for two weeks can improve the ITP. Helicobacter Pylori is diagnosed using a blood test, breath test or stool sample. Improvements of platelet count following treatment of the infection are not always permanent, but the treatment is very safe and so may be recommended by your doctor.

Dapsone

Dapsone is an antibiotic, but it can also be used to treat ITP. About half of people who have already had steroids and rituximab will have an improved platelet count with dapsone, although the effect often only lasts a few months. Before you are given dapsone, your doctor will check that you don't have a condition called G6PD deficiency, which is a rare inherited condition that affects red blood cells. Dapsone (and some other drugs) cannot be given to people who have G6PD deficiency, as it can cause severe anaemia from damage to the red blood cells.

Anti-D

Like IVIG (intravenous immunoglobulin), anti-D is a collection of antibodies from blood donors. However, it can only work for people who are 'rhesus positive' or more correctly, 'D positive' and who have not had a splenectomy. Anti-D is given through a drip over a few minutes. Anti-D is effective for about 7 out of 10 patients but the responses only last a few weeks and a side effect of the treatment is to cause red cells to break down, which may cause anaemia or rarely kidney problems.

Why can’t I have platelet transfusions to treat my ITP?

The platelets made by your bone marrow are healthy and it is only because your immune system is destroying them that you have a low platelet count. If you were to receive other people's platelets (given by transfusion) they would also be destroyed by your immune system. Platelets transfused to you would only last minutes or hours before being destroyed. Platelet transfusions can be useful as an emergency treatment if you have severe bleeding, as they can help you to form a clot, but they are not useful for long term prevention of bleeding.

What happens now?

This leaflet tells you about different treatments for ITP. 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 one would be appropriate for you. If you need to make an appointment to discuss this information with your doctor, please phone the Haematology Clinical Admin Team (CAT 8):

Tel: 0118 322 8145 or email CAT8@royalberkshire.nhs.uk
(9.00am to 5.00pm, Monday to Friday)
Further sources of information can be found at the end of this leaflet.

Contact us

If you have bleeding symptoms please ask your GP to check your platelet count urgently, or contact: West Ward 0118 322 6632 (Monday-Friday 8.30am-5pm)

If you have excessive bleeding or bruising, you must go to the Emergency Department (A&E) at your nearest hospital.

Bleeding symptoms may include fresh bleeding from your nose, mouth, in vomit, stools (faeces) or urine, or passing black sticky stools. A purple rash, usually on the ankles and legs, which does not fade when you press it, can also be a sign of a low platelet count.

Further information

- The ITP Support Association has a very good website. You can also use it to request leaflets on specific topics (such as splenectomy). Website: www.itpsupport.org.uk/ Email: itpsupport.org.uk
- NICE is the organisation that advises doctors which treatments they can prescribe. They have produced guidance on some treatments, which are available on:
  Romiplostim: www.nice.org.uk/guidance/ta221
  Eltrombopag: www.nice.org.uk/guidance/ta293.
  Rituximab: www.nice.org.uk/advice/esuom35/chapter/key-points-from-the-evidence

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

For more information about the Trust, visit www.royalberkshire.nhs.uk

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