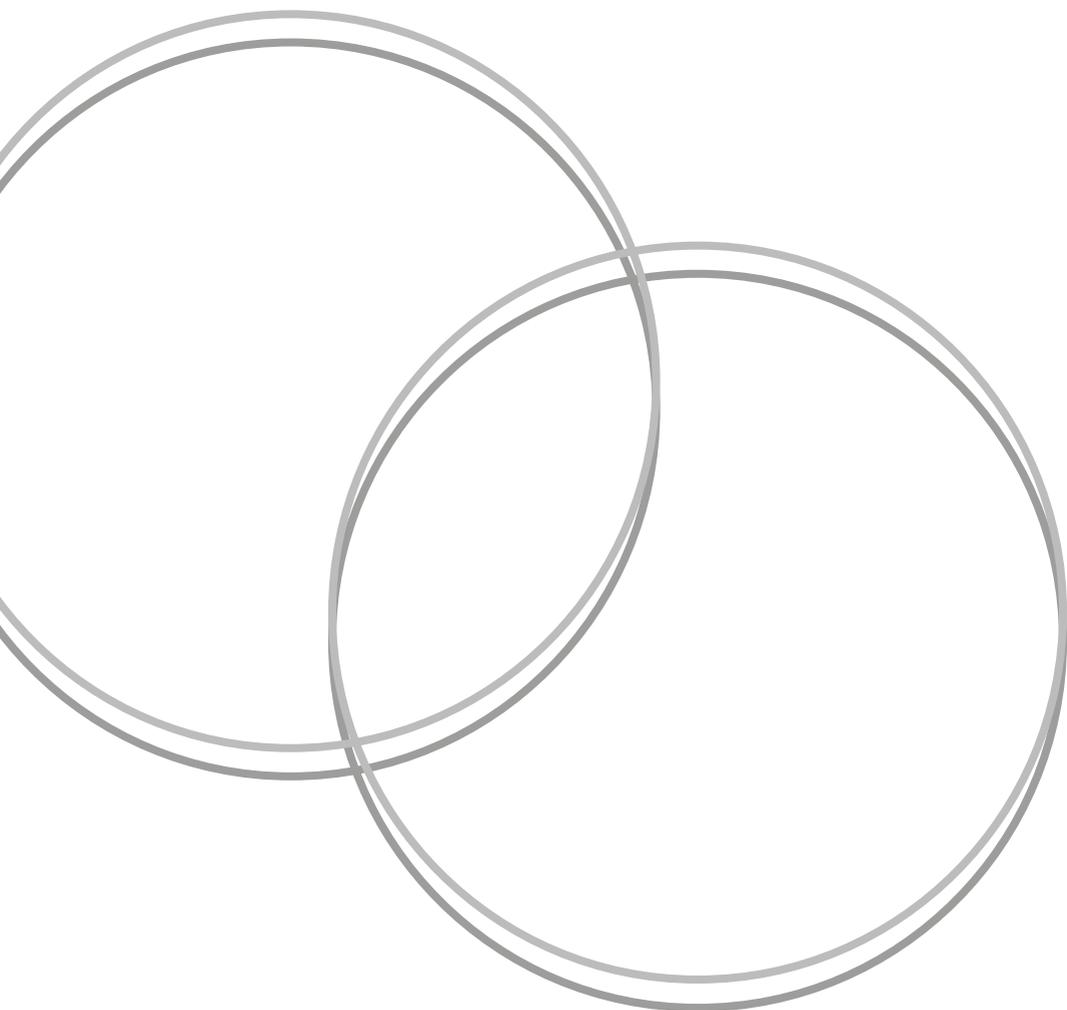




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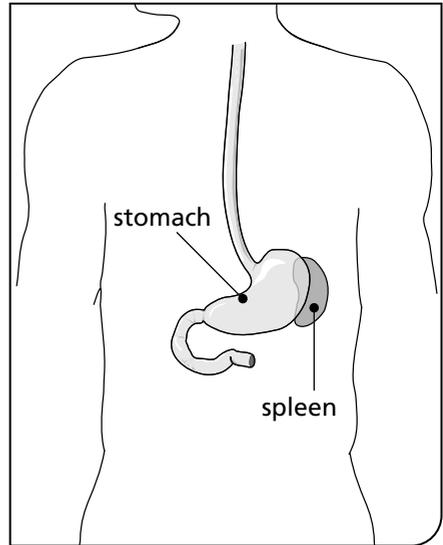
# Immune thrombocytopenic purpura (ITP)

Information for patients



## What is ITP?

Immune thrombocytopenic purpura (ITP) is a condition which causes the number of platelets in your blood to be reduced. Platelets are what makes blood clot and they are needed to help you stop bleeding and bruising after an injury. If you do not have enough platelets in your blood, you are likely to bruise very easily or may be unable to stop bleeding if you cut yourself.



ITP is an autoimmune condition. This means that your body's immune system is acting against your own cells. ITP causes the white blood cells in your body's immune system to destroy your platelets. This happens mainly in the spleen, an organ in your abdomen.

ITP in adults is more common in women than men. It is very different to ITP in children. Children usually get ITP after a viral infection and it almost always gets better on its own without any treatment. ITP in adults usually needs treatment.

Some people with ITP have other autoimmune conditions, such as rheumatoid arthritis, or infections such as hepatitis or HIV. If you have any of these medical issues, your ITP may be treated slightly differently.

## How is ITP diagnosed?

ITP is diagnosed from a blood test. Sometimes it is found if someone experiences bleeding. More often, it is found when a blood test is taken for another reason.

## **What causes ITP?**

The cause of ITP is usually unknown.

In rare cases, ITP happens after a viral infection or, very rarely, in response to a vaccination.

## **What is a 'normal' platelet count?**

A normal platelet count is between 150 and 400 thousand million platelets per litre of blood. This is usually referred to by doctors just using the first three numbers, such as '150' or '400'. You are unlikely to get bleeding symptoms unless your platelet count is below 20 or even 10. If you needed to have an operation this would be safe as long as your platelet count is more than 50.

Your doctor will not recommend treatment for ITP unless you have bleeding symptoms, or if your platelet count is very low, usually below 20.

## **What is the treatment for ITP?**

If your ITP needs treating, we usually prescribe steroids. These reduce the level of antibodies in your bloodstream and stop your immune system from destroying your platelets. Steroids are a good treatment for ITP and you will usually only need a short course of treatment.

We will ask for your permission – or 'consent' – before we carry out treatment. Staff will explain the risks, benefits and alternatives if there are any before they ask for your consent. Remember that it is your choice whether you give your consent or not.

Sometimes it is not possible to ask for consent if you are admitted in an emergency. We will discuss your treatment with you at the earliest opportunity.

If you have any doubts or would like more information about your treatment please ask us. We will do everything we can to help you.

## **Are there any side effects of steroids?**

Over a short period of time, steroids usually cause no problems. However, steroids can have side effects, especially if you need repeated courses of treatment or have to take them for a long time. Some side effects are related to stopping the white cells working properly; this can increase your chance of getting infections, as the white cells will also be less able to fight off bacteria and viruses. Steroids also have other side effects, such as thinning of the bones (osteoporosis), stomach ulcers and high blood sugar levels (diabetes). They can change your facial appearance and cause thinning and bruising of the skin. People feel they want to eat more while they are on steroids and often put on weight. These side effects usually reverse when the steroids are stopped.

## **Are there any alternatives to steroids?**

Your doctor may recommend that you try another treatment for your ITP. This could be because:

- you didn't respond to steroids at all
- you responded but you 'relapsed' (your platelet count fell again) when the dose of steroids was cut down
- you have responded to steroids in the past, but your ITP has come back and because of the side effects you do not want to have them again.

## **Should I stop taking steroids?**

No. If you are taking steroids it is very important that you do not stop them without advice from your doctor, as your body starts to rely on them. They may need to be cut down slowly so that your body has time to adjust, otherwise you might 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.

## **Why don't steroids cure everyone?**

There are many reasons why you might not respond to steroids. If this happens, your doctor will want to make sure there is no other reason for your low platelet count; this may involve some extra blood tests (including checking for infections if this hasn't already been done) or a bone marrow test. There is no test available that is completely accurate in diagnosing ITP.

## **Will I need other treatments?**

7 out of 10 people who are treated with steroids will have another drop in their platelet count in future.

This does not always need treatment. Many people with ITP have a low platelet count without experiencing bleeding, and do not need further treatment. This is called being 'in remission' from ITP.

However, we will discuss with you other treatment options that you may need in future if your platelets fall further, if you have symptoms of bleeding, or if you need an operation.

## **What other options are there?**

There are different treatments available and it is important you understand a bit about them, so that you and your doctor can decide which would be most appropriate. The other possible treatments are:

- Intravenous immunoglobulin (IVIG)
- Splenectomy
- Rituximab
- Romiplostim or Eltrombopag
- Immunosuppressants
- Dapsone
- Anti-D

## **Intravenous immunoglobulin (IVIG)**

This is a medicine containing antibodies (immunoglobulin) which is given into a vein, usually in your arm, through a drip (intravenously). Antibodies are produced by white blood cells, to fight infections. It is a human blood product, which means that the antibodies have been collected from numerous blood donors. This means that **if you receive IVIG you will never be able to donate blood** in the UK, even when you recover from ITP.

Nobody understands exactly how IVIG works to treat ITP, but it is thought that the extra antibodies stop your own white blood cells from destroying your platelets.

IVIG is given as an infusion (liquid mixture) through a drip over a few hours.

### **What are the advantages of IVIG?**

IVIG works quite quickly, usually within a few days. Unfortunately, the effect doesn't last long (a few weeks at most) and so it will not cure your ITP. It is generally given before surgery, or if you have significant bleeding symptoms and your platelet count needs to be increased urgently.

### **What are the risks of IVIG?**

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 also a small risk of developing a rare complication called aseptic meningitis. This causes a headache, neck stiffness and dislike of bright lights. This condition usually gets better on its own, but if you develop any of these symptoms after treatment with IVIG you must seek medical attention immediately.

IVIG can very rarely cause kidney damage and there is also an extremely small risk (less than one in many millions) of infections such as hepatitis and HIV (as it is made from donated blood).

## **Splenectomy (removal of the spleen)**

As your platelets are mainly being destroyed when they are in your spleen, removing your spleen can cure the condition. This will be carried out during an operation under general anaesthetic (where you are made to be asleep). The operation can usually be done laparoscopically (using very small cuts to carry out keyhole surgery), which means you should recover more quickly. You will usually be in hospital for a couple of days, although it can take six weeks to fully recover from the operation.

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.

### **What are the advantages of splenectomy?**

Splenectomy has been used to treat ITP for decades and offers the best chance of curing ITP. For every three people who have the operation, two will not need further treatment. Unfortunately it is difficult to predict whether you will be cured by this operation. Your doctor may ask you to have a special scan to look at your spleen, which can help them to decide whether a splenectomy will work for you. The test can sometimes show whether your spleen is the main place that your platelets are being destroyed, but it cannot completely predict whether you will be cured by the surgery.

### **What are the risks of splenectomy?**

Any surgical operation has risks. 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.

You may be at more at risk of complications from surgery if you have other medical conditions, are very overweight, or if it is not possible to increase your platelet count before the operation. Your doctor will discuss your own situation and specific risks with you.

To reduce the risk of long term infection you will need to have vaccinations (immunisations or 'jabs') before having surgery. After the surgery you may need to take long-term low dose antibiotics to help prevent infection, or you may be given a packet of antibiotics to keep at home in case you become unwell. This is because some of the white cells which would normally help your body to fight infection would have been made in your spleen.

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.

There is an information leaflet produced by the ITP Support Association called 'Splenectomy in ITP', which you may find helpful. You can order it from their website at [www.itpsupport.org.uk/](http://www.itpsupport.org.uk/)

## Rituximab

Rituximab is a drug which was first used to treat cancer, but has also been used for nearly 20 years to treat ITP. Like steroids, it stops the immune system destroying platelets, but it has fewer side effects than steroids. It is a manufactured antibody (developed by a medicines company) which affects the 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. Soon we may be able to give it as a slow-release injection under the skin.

It usually takes a few weeks for rituximab to work, although some people respond many months after treatment. If rituximab works well for you, the treatment can be repeated months or years later, if needed.

### **What are the advantages of rituximab?**

Around two out of three people given rituximab will have some increase in their platelet count. Most people experience further episodes of ITP (their platelet count will drop again) but usually the platelet count stays at a good level for at least a year.

### **What are the risks of rituximab?**

Most people who are treated with rituximab for ITP 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.

Although rituximab works by stopping the white cells from making antibodies, you aren't likely to have any problems with infections. There is an extremely rare viral infection which can affect the nervous system, which a few people treated with rituximab have had. The condition is even rarer among patients with ITP (there is only 1 report of this happening from all the people who have received rituximab for ITP over nearly 20 years).

## **Romiplostim (Nplate™), eltrombopag (Revolade™)**

Thrombopoietin (TPO) receptors are on the surface of the cells that make platelets in the bone marrow. These drugs use these receptors to tell the cell to make more platelets. The 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.

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 four hours before or 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.

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.

### **What are the advantages of romiplostim and eltrombopag?**

If we gave 10 people with ITP (that has already been treated with steroids and another treatment) one of these drugs, we would expect 8 or 9 of them to have some response (their platelet count may increase or they may have less bleeding). These effects will continue in the long term in about 5 of these people.

## **What are the risks of romiplostim and eltrombopag?**

Most people have no side effects with these drugs. Some people get headaches and a few people taking romiplostim have developed scarring of the bone marrow. The scarring of the bone marrow doesn't appear to stop the bone marrow from working properly.

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 frequent blood tests and clinic visits when you start to take this medicine.

## **Immunosuppressants**

These other medications can also be used to treat ITP. They are used less often now, as they are thought to have more side effects than the newer treatments. They may be tried if you have no response to the newer drugs.

Immunosuppressants are often used in cancer treatment, but the doses used in ITP are much lower than for cancer. They are useful treatments for some people with ITP and don't usually cause many side effects. However, they can stop bone marrow from working properly, which can lead to anaemia and low white blood cell counts. This increases the risk of infection.

Some of these drugs can also affect the kidneys, so you will be closely monitored while you are taking them. 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.

## Dapsone

Dapsone is an antibiotic, but it can also be used to treat ITP. It is not really known how it works, but it appears to dampen down the autoimmune process, which can stop your body from attacking your platelets. It is taken as a tablet once a day and has few side effects. About 50% of people (50 in 100) who have already had steroids and rituximab will respond to dapsone. However less than half of these people will still have a good platelet count after 6 months.

Before you are given dapsone, your doctor will check that you don't have a condition called G6PD deficiency, which is a rare condition that affects your 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 as a one-off dose. If you have anti-D you can still give blood if you want to (but only once you no longer have ITP).

If 10 people are given anti-D, 7 will respond with an increase in their platelets, but in half of these people the response lasts less than 3 weeks. However, if it is effective, the dose can be repeated at a later date. Anti-D is not used much in the UK because some people treated with it experienced a breakdown of red blood cells, which has led to kidney damage or death in a small number of people. However, anti-D may be an option if other treatments have not worked for you.

## 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 about tranexamic acid?

Tranexamic acid is a medication which helps blood clots to last longer once they have formed. The clots are more stable than usual and more resistant to being broken down. Tranexamic acid does not treat ITP, but can be useful if you have bleeding while your platelet count is low. It is a tablet that is taken 3 or 4 times a day. It should not be taken if you have blood in your urine. It can sometimes cause indigestion, which may get better if you take a lower dose.

## 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 Secretaries:

Tel: **01865 235 185**

(9.00am to 5.00pm, Monday to Friday)

Email: **[orh-tr.clinicalhaematology@nhs.net](mailto:orh-tr.clinicalhaematology@nhs.net)**

If you would like any more information, there is a list of places you can find it at the end of this leaflet.

## Contact details

If you have bleeding symptoms please contact:

### **Haematology Triage**

Tel: **01865 572 192**

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. **If you have excessive bleeding or bruising, you must go to the Emergency Department at your nearest hospital.**

## 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/](http://www.itpsupport.org.uk/)**

Email: **[itpsupport.org.uk](mailto:itpsupport.org.uk)**

NICE is the organisation that advises doctors which treatments they can prescribe. They have produced guidance on the TPOR agonists, which is available on the following websites:

Romiplostim: **<http://www.nice.org.uk/guidance/ta221>**

Eltrombopag: **<http://www.nice.org.uk/guidance/ta293>**

## Further information

If you would like an interpreter, please speak to the department where you are being seen.

Please also tell them if you would like this information in another format, such as:

- Easy Read
- large print
- braille
- audio
- electronic
- another language.

We have tried to make the information in this leaflet meet your needs. If it does not meet your individual needs or situation, please speak to your healthcare team. They are happy to help.

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[www.ouh.nhs.uk/information](http://www.ouh.nhs.uk/information)



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