East Kent Hospitals University NHS Foundation Trust

Immune Thrombocytopenia Purpura (ITP)

Information for patients and carers from the Haematology Department

What is ITP?

Immune thrombocytopenic purpura (ITP) is a condition which causes the number of platelets in your blood to be reduced. Platelets are cells that help blood to clot and they help to prevent bleeding and bruising 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 if you cut yourself.

In ITP, your body's immune system destroys your own platelets. White blood cells in your blood and your spleen (an organ in your abdomen) are part of your immune system. One of their actions is to produce antibodies which help your body to fight infections. If you develop ITP, your immune system becomes overactive and produces antibodies that cause your platelets to be destroyed in the spleen; this results in a low platelet count. ITP is a type of autoimmune condition (which means your immune system is acting against your body rather than for it).

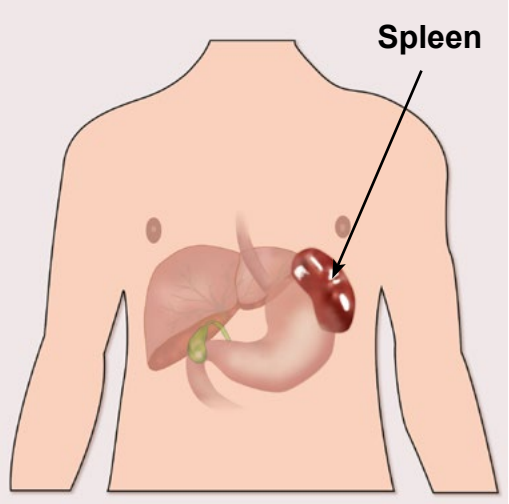

Diagram showing the position of the spleen

ITP in adults is more common in women than men. It is very different from ITP in children, who usually get ITP after a viral infection but who recover without any treatment. ITP in adults normally 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.

We care

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

What is the treatment for ITP?

If your ITP needs treating, the aim is to get your platelet count to a safe level; this is called **first line treatment**. There are a number of medicines which can be used alone or in combination, particularly if your platelet count is very low or you are experiencing bleeding problems. These medications are also used for **rescue treatment** if, at any time in the future, your platelet count drops (a relapse in your ITP). The three main options are:

- steroids
- Intravenous Immunoglobulin (IVIG), and
- Anti-D.

In some cases, you may have already been given one or more of these treatments before you come to the Haematology Outpatients' Clinic.

- **Steroids**
  These work by preventing your immune system from destroying your platelets, by reducing the level of antibodies in your blood stream. Steroids are a good treatment for ITP and you will usually only need a short course of this medication to be successful.

  Are there any side effects to using steroids?
  Over a short period of time, steroids usually cause no problems. However, they can have side effects, especially if you need repeated courses of treatment or have to take them for a long period of time.

  Some side effects are related to stopping the white blood 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 can 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. Patients can feel they want to eat more whilst they are on steroids and, as such, they might put on weight. These side effects usually reverse when the steroids are stopped.

  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 over time so that your body can readjust, otherwise you might experience weakness and fatigue (tiredness). If you are worried about any possible side effects of steroids, please discuss your treatment with your doctor before making any changes to your medication.

  Please see the **Prednisolone** and **Dexamethasone** patient information sheets for more information. Ask a member of staff for copies or go to the ITP Patient Information web page www.ekhuft.nhs.uk/patient-information-itp/

- **Intravenous Immunoglobulin (IVIG)**
  This medicine contains antibodies (immunoglobulin) which are 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 a human blood product, which means that the antibodies have been collected from blood donors. Nobody understands exactly how IVIG works to treat ITP, but it is thought that the extra antibodies stop your white blood cells from destroying your platelets.

  What are the advantages of IVIG?
  IVIG works quickly, usually within a few days. Unfortunately, the effect does not 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 an allergic reaction (such as a fast heart rate or breathlessness) whilst the IVIG is being given, so you will be monitored closely by a nurse at the time. There is also a small risk of developing a rare complication called aseptic meningitis, which causes a headache, neck stiffness, and a dislike of bright lights. Aseptic meningitis is not to be confused with other types of meningitis, and usually gets better on its own.

  If you develop any of these symptoms after treatment with IVIG you must see either your GP or go to your nearest Emergency Department immediately.

  In rare cases, IVIG can cause kidney damage. As the IVIG is made from donated blood, there is also an extremely small risk (less than one in many millions) that you may acquire a blood-borne virus, such as hepatitis and/or HIV. There are ways in which these risks are minimised by the drug company. The first is that donors are carefully screened to make sure they do not carry these viruses. Secondly, testing for signs of viruses at each donation, and finally the IVIG is heat treated to inactivate or remove any possible viruses.

- **Anti-D**
  Like IVIG, Anti-D is a collection of antibodies made from donor blood. 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 (a small tube or cannula into a vein in your arm) over a few minutes, as a one-off dose. If you have Anti-D you could still be a blood donor (but only if you no longer have ITP).

  If 10 people are given Anti-D, seven will respond with an increase in their platelets but, in half of those seven people, the response lasts less than three weeks. However when it works, it is an effective treatment and the dose can be repeated at a later date, for ongoing treatment.

  With Anti-D, there is a small risk that there will be a breakdown of red blood cells, which can lead to kidney damage and severe anaemia. As with IVIG there is an extremely small risk of getting a blood-borne virus but the same steps taken for IVIG are also taken for Anti-D.

What happens if my platelet count is still low or drops again after steroid treatment?

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

- you have been given steroids but have not responded to them
- you responded to steroids but you 'relapsed' (your platelet count fell again) when the dose was cut down, or
- you have responded to steroids in the past, but your ITP has come back and your doctor does not want to give you more steroids because of the side effects (or you may not want to take them again because of the side affects you had before).

Why do steroids alone not cure everyone with ITP?

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

For every 10 adults with ITP who are treated with steroids, only three (at most) will not need further treatment. In the other seven the platelet count will drop again and more treatment may be needed.

As spontaneous bleeding (bleeding without injury) only happens when your platelet count is very low, your doctor may not recommend any further treatment for the time being. Many people with ITP have a platelet count that is below normal, but is kept at a safe level without any treatment. However, it is important that we discuss with you what treatment may be given if your platelet count falls further, you have bleeding symptoms or need an operation or dental treatment.

What other options are there?

Unfortunately all treatments for ITP have possible side effects, which is why your doctor will not recommend treatment unless you have bleeding symptoms or your platelet count is very low (usually below 20).

There are several other treatments available for ITP and it is important you understand something about them, so that you and your doctor can decide which would be the most appropriate for you.

- Steroid sparing immunosuppressants
- Rituximab
- Thrombopoietin (TPO) Receptor Agonists
- Splenectomy (surgical removal of your spleen)
- Helicobacter Pylori treatment.

- **Steroid sparing immunosuppressants**
  These are useful treatments for some people with ITP and do not usually cause many side effects. However, they work in a similar way to steroids but their benefit is that they do not have the long-term side effects of steroids. They can take a while to start working, in some cases up to 12 weeks, during which time we may have to give you rescue treatment with either short courses of steroids, IVIG, or Anti-D. Steroid sparing immunosuppressants can also increase the risk of anaemia and low white blood cell counts, which can increase the risk of infection.

  Some of these drugs can also affect your kidneys, so you will be closely monitored while you are taking them. Examples of steroid sparing immunosuppressant drugs include:

  - Mycophenolate mofetil (MMF)
  - Azathioprine
  - Cyclophosphamide
  - Vincristine
  - Cyclosporin.

  All these medications are given as a tablet, except Vincristine, which is given as an injection into a vein. If your doctor recommends one of these drugs more information is available.

- **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. It is a manufactured antibody which affects the white blood cells and 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 each time.

  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 every three people given Rituximab will have some increase in their platelet count.

  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 fast heart rate or breathlessness), but you will be monitored closely whilst it is given.

  Although Rituximab works by stopping the white blood cells from making antibodies, you are not 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 developed. The condition is even rarer amongst patients with ITP given Rituximab and there is only one report of this happening from all the people who have received Rituximab for ITP over almost 20 years.

  Please see the **Rituximab** patient information sheets for more information. Ask a member of staff for copies or go to the ITP Patient Information web page www.ekhuft.nhs.uk/patient-information-itp/

- **Thrombopoietin (TPO) Receptor Agonists: Romiplostim (NPlate™), Eltrombopag (Revolade™)**
  These two drugs have become available in the last few years. 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. These drugs can currently 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 for any reason.

  Once you have started these treatments you will need to continue taking them for as long as your ITP continues, which may be many years.

  Romiplostim (Nplate™) is given by an injection under your skin usually once a week. You can be taught to give this injection yourself or a family member can be shown how to do it.

  Eltrombopag (Revolade™) is a tablet which is taken once a day. It cannot be absorbed by the gut if there is calcium nearby, so you must not eat foods high in calcium for four hours before and two hours after you take the tablet. Most people find it easier to take the medication either 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 when taking this medication.

  What are the advantages of the TPO Receptor Agonists?
  If we give 10 people with ITP (that had already been treated with steroids and another treatment) one of these drugs, we would expect eight or nine of them to have some response (such as their platelet count may increase or they may have less bleeding). These effects will continue in the long term in about five of these people.

  What are the risks of the TPO Receptor Agonists?
  Most people have no side effects with these drugs. Some people get headaches and a few people taking Romiplostim have developed scarring of their bone marrow. This scarring does not 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 these medications so this can be risk-managed. As such, you will have frequent blood tests and clinic visits when you start to take this medication.

- **Splenectomy (surgical 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 operation is carried out under a general anaesthetic, you will be asleep throughout the procedure. The operation can usually be done laparoscopically, which means there are only small cuts made to carry out the keyhole surgery. With this type of operation you should recover more quickly, and you will usually only be in hospital for a couple of days. It can take six weeks to recover fully from the operation.

  Sometimes the splenectomy needs to be done using open surgery (a larger cut), but your surgeon will discuss this with you if they think you are likely to need this type of operation.

  You may also need some additional treatment to increase your platelet count before you have the operation.

- **What are the advantages of splenectomy?**
  Splenectomy has been used to treat ITP for decades. For every three people who have the operation, two will not need any further treatment. Unfortunately it is very 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 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 having a splenectomy?
  There is a risk with any surgery, but that risk is very small, and keyhole surgery is generally thought to be far safer than open surgery. These risks will be discussed with you before any surgery to remove your spleen.

  Risks from surgery include:
  - a 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, and
  - infection after surgery.

  You may be at more 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 your operation. Your doctor will discuss your own situation and specific risks with you.

  To reduce your risk of long term infection you will need to have vaccinations before surgery. After your surgery you may need to take long term low dose antibiotics to help prevent infection, or you may be given antibiotics to keep at home in case you become unwell. This is because some of the white blood cells which would normally help your body to fight infection would have been made in your spleen, which has now been removed.

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

  For more information, please read the ITP Support Association leaflet **Splenectomy in ITP**. You can order a copy from their website www.itpsupport.org.uk/

- **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 cure or improve the ITP. Helicobacter Pylori is diagnosed using a breath test or a stool sample. Improvements to the 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.

  Why can I not 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 receive other people's platelets (given by transfusion) they would only last minutes to 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 last longer once they have been formed. The clots are more stable than usual and more resistant to being broken down.

Tranexamic Acid does not treat ITP, but it can be useful if you have bleeding while your platelet count is very low. This treatment is taken as a tablet, three times a day usually just for a few days whilst your platelet count is very low. 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.

Further information

- The **ITP Support Association** has a very good website. You can also request leaflets from them on specific topics (such as splenectomy).

  Website: www.itpsupport.org.uk/
  Email: itpsupport.org.uk

- **NICE** (National Institute for Health and Care Excellence) is the organisation that advises doctors which treatments they can prescribe. They have produced guidance on the TPO Receptor Agonists, which is available on the following websites.

  Romiplostim: www.nice.org.uk/guidance/ta221
  Eltrombopag: www.nice.org.uk/guidance/ta293

This leaflet has been produced with and for patients

If you would like this information in **another language, audio, Braille, Easy Read, or large print** please ask a member of staff. You can ask someone to contact us on your behalf.

Any complaints, comments, concerns, or compliments please speak to your doctor or nurse, or contact the Patient Advice and Liaison Service (PALS) on 01227 78 31 45, or email ekh-tr.pals@nhs.net

Patients should not bring in large sums of money or valuables into hospital. Please note that East Kent Hospitals accepts no responsibility for the loss or damage to personal property, unless the property had been handed in to Trust staff for safe-keeping.

Further patient leaflets are available via the East Kent Hospitals web site www.ekhuft.nhs.uk/patientinformation

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