What did you call it?

Contents

Revised 2017

Introduction

This booklet was written to help answer some of the questions which arise with adult ITP. It is intended to help you cope with the trauma and the fear which accompanies the first few weeks after diagnosis - when you wonder just what lies ahead. You probably don't know if ITP is a serious illness - whether it is treatable or even life threatening. The questions you may be asking at such a time have been prepared in this booklet by those who have been through this very experience and know exactly what it is like. We hope it helps you to put ITP into a proper perspective and allow you to return to a normal life as soon as possible.

Medical science is changing rapidly, so it is important to remember that your consultant may have more up to date information about ITP and the techniques of treatment than what was available when this booklet was written. If you are in doubt on any matter, do not be afraid to ask your questions as no book or information pack can replace the essential face to face discussions which take place between consultant and patient.

What is ITP?

ITP describes a shortage of blood platelets caused by a disorder of the immune system. If skin or body tissue is cut or injured, normal blood contains a sufficient number of platelets to form the initial plug which helps stop the bleeding and allows the wound to heal. When ITP is diagnosed, it means that there are not enough platelets to do this job properly and the patient can suffer excessive bleeding and bruises.

So what's the difference between ITP and hæmophilia?

In normal blood there are 13 factors which act together to effect the full clotting mechanism, after any injury which causes bleeding. The platelets form a plug as the first part of the process, after which the other 12 factors complete the process and effectively stop the bleeding. A person with ITP is short of platelets, but a person with hæmophilia is deficient in one of the other twelve factors. Both are bleeding disorders, but whereas hæmophilia is inherited and permanent, ITP can develop at any age and can go into remission.

What does ITP stand for?

Formerly known as Idiopathic Thrombocytopenic Purpura. (Idiopathic = of unknown cause, purpura = bruising) it is now called Immune Thrombocytopenia meaning a shortage of blood platelets caused by a disorder of the immune system.

What are the symptoms?

Usually the first signs are unexpected bruises (often pain free), nose bleeds, black mouth blisters and tiny red spots (petechiae) on the skin where capillaries leak minute drops of blood. Women may have excessively heavy periods. Although not always recognised by doctors as a symptom, many people with ITP can feel fatigued.

Why does it happen?

The cause is not yet fully understood. However, it is known that the body's immune system, which deals with all physical illnesses from a minor cough or cold through to the very serious, mistakes the platelets in the blood for 'enemies' and destroys them.

How did I get it?

ITP is not contagious. You did not catch it from anyone and neither can you pass it to someone else. Although there is often no known cause, it is possible to develop ITP as a secondary condition whilst taking certain drugs, or following a viral infection or other illness.

How is it diagnosed?

If ITP is suspected a blood sample will be taken and the platelets counted (electronically). In one cubic millimetre of blood there should be between 150,000 and 400,000 platelets. In the UK this is referred to as a platelet count of 150 to 400. In ITP the red and white cell count is usually normal and only the platelet count is low, as determined by a blood test. A hæmatologist (blood specialist) may also request a bone marrow test, where a small sample is taken of the liquid part of the pelvic or chest bone. This is done to check that platelets are being made.

Should it be treated?

If your platelet count remains stable or at a level which allows you to remain symptom free, you may not need any treatment at all. If it is discovered that medicines you have been taking were the cause, then these will be stopped and alternatives investigated.

What if I do need treatment?

If drugs are prescribed for your condition, the two most frequently used first-line treatments are:-

Steroids: prednisolone is given in high doses for a short period of time to slow down the destruction of the platelets. Steroids may also stabilise the blood vessels, reducing the risk of bleeding.

Intravenous immunoglobulin: this is plasma processed from blood donations and is drip fed directly into the vein over a period of several days in hospital. It works by confusing the body's immune system.

Suppose treatment doesn't work?

There is no treatment yet which actually cures a person of ITP. Your doctor or consultants main concern at this stage will be to try to raise the platelet count to an acceptable level. Drugs which work well for one patient may not work as well for another, so it may take a while for the best course of treatment or dosage level to be determined by your consultant or doctor. If success with steroids or immunoglobulin is not forthcoming, your doctor may try other drugs such as rituximab, immunosuppressants, thrombopoietins, or even consider a splenectomy.

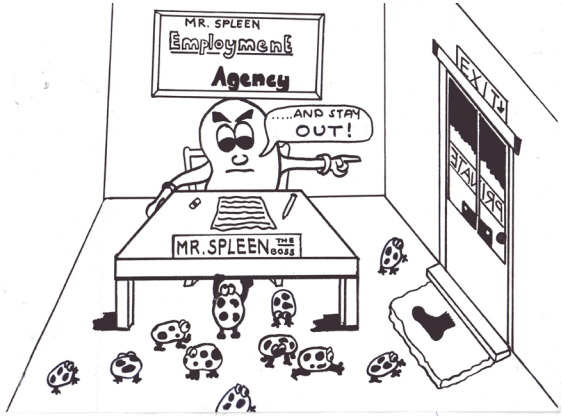
What are the risks from treatment?

Most strong substances taken internally have side effects and medical drugs are no different. Speak to your consultant if your side effects are serious. Steroids can cause mood changes ranging from euphoria to irritability; they can cause weight gain, roundness in the face and neck, and irritation of the stomach. There is also an increased risk of infections and, if taken long term, loss of calcium from the bones, kidney stones, increased blood pressure and pancreatitis. The dosage prescribed by your doctor or consultant will be carefully assessed to try to avoid such problems. Patients receiving

immunoglobulin could experience temporary headaches, nausea, light headedness, shakes and shivers, or even a slight fever. If symptoms become severe anti-histamine and cortisone can be given before the next treatment. Rarely, you could be allergic to immunoglobulin and treatment would be discontinued.

What about a splenectomy?

A splenectomy is the surgical removal of the spleen. It is no longer done routinely for ITP, and is only undertaken in chronic cases (i.e. those with ITP for six months or more), for people with severe symptoms or where drug related methods have been unsuccessful or insufficient. Splenectomy is successful in about 60-70% of cases, but even in the remainder may bring some improvement to the platelet count.



Why remove the spleen?

The spleen is an organ which filters out 'foreign' or waste materials from the blood. Healthy platelets normally pass through the spleen unmolested, but with ITP patients the platelets become coated with 'antibodies'. These fool the spleen into believing that they are foreign to the body and need to be removed. It is thought that, to some extent, the spleen might be responsible for coating the platelets with antibodies in the first place.

What are the risks of a splenectomy?

Patients who undergo a splenectomy will need to be vaccinated against pneumococcus and hæmophilus influenzae before the operation, and are usually prescribed lifelong antibiotics. Afterwards they will have an increased risk of infections or fevers which will require prompt medical attention.

Will ITP affect my periods?

Women with a platelet count of 20 or below may have prolonged periods with heavier menstrual bleeding. If it becomes necessary, this can usually be controlled with a hormonal preparation, such as the contraceptive pill or mirena coil.

What happens if I become pregnant?

Many pregnant women with ITP will not need to be treated, but this depends on the platelet count and their previous history. A care plan will be decided upon between the obstetrician and hæmatologist with experience of the condition.

Will ITP affect my life span?

Although ITP can affect a person's quality of life because of activity restrictions to avoid the danger of bleeding, it is extremely rare for someone with ITP to suffer a severe hæmorrhage.

Who should I tell?

It is important to inform your dentist and any other medical practitioner (including, chiropodist, osteopath, acupuncturist or chiropractor. It may also be helpful to discuss your condition with your boss, personnel manager and those you work with so that in the unlikely event of an emergency they would know what to do. You can obtain a leaflet from the Support Association for employers, giving basic information about ITP.
If you see a doctor for any other medical condition remind them that you have ITP, especially if you are prescribed drugs as they may affect your platelet count or interact with any treatment you have for ITP.

How should I stop a nose bleed?

Pinch the sides of the nose just below the bone - above the nostrils, or use a swimming nose clip which gives gentle, even pressure. Sit quietly for five minutes and then remove hand or nose clip. Keep still for a further five minutes. Afterwards, gently wash the dried blood away to help reduce nasal irritation. Ice packs on the nose are also useful to stem the blood flow.

How can I cope with the range of emotions I experience?

You're very likely to feel a wide range of emotions.

- Fear - because you don't know what lies ahead
- Relief - that it wasn't a more serious illness
- Anger - that it has happened to you
- Guilt - that you might have brought it upon yourself
- Helplessness - because you cannot make yourself well
- Frustration -platelet counts can rise or fall unpredictably
- Anxiety - wondering how to cope with work and family

The ITP Association has three contact volunteers with ITP who provide a 'listening ear' and are very willing to hear your problems and offer their understanding and advice.

What about my family?

Other members of your family are likely to be affected emotionally and worry about you. Keep them informed about your ITP and its management and/or treatment, so that you can support each other. Share their feelings and fears and make sure they understand that ITP is not infectious or hereditary.

And what if it goes on?

If the condition persists, you may feel that you're on a sort of roller coaster – going through the ups and downs of symptoms and platelet counts. With a reasonably stable condition, however, you should be able to continue to live a fairly normal life. In any case it is important to 'fold' ITP into your life style as much as possible. Of course there will be stressful times, but as you understand

more about ITP you should feel that you are more on top of the situation than you were at first.

How do I find an ITP specialist?

The Association website has a map of ITP Clinical Centres in the UK, which are recognised specialist ITP centres, or you can ask to see one of the Association's medical advisors (on the NHS). Whoever you see you will need a referral from your consultant or GP so the new doctor has access to your full medical notes showing your ITP history, symptoms and tests which have been carried out. Before you visit your doctor or consultant, you may find it useful to write down a brief memo of any questions or problems you wish to discuss as it is easy to forget something important when you are in the consulting room. It can be helpful to take a relative or friend to your appointments to prompt you with questions if necessary and to help remember the answers or advice given. Don't be intimidated by the medical jargon and if there is something you don't understand ask for a simpler explanation.

Are there any special precautions?

Do not take aspirin or non-steroidal like ibuprofen (also sold as Nurofen), which make platelets work less effectively, but paracetamol can be used instead. Remember to tell any doctor prescribing you drugs that you have ITP, as some affect platelet function. A list of these is available from the Support Association. You would be advised to carry an ITP Support Association Emergency Card or jewellery at all times to alert others to your condition, especially if you have had a splenectomy.

Should I restrict my life style?

Try to live as normal a life as possible, but remember that activities which involve energetic body contact, aggressive sports, or a fall from any height should be avoided. Sports protective clothing, such as knee pads, should be worn where appropriate, and if you ride a bicycle, you are strongly advised to wear a crash helmet.

Some people with ITP suffer from overwhelming fatigue when their platelet count is very low, and at such times you may need to pace yourself carefully. Don't be afraid to ask family and friends for help!

There is no reason why you can't go on holiday but you may need to plan more carefully before you go, and if your ITP is severe you may wish to restrict you holiday destination to a country where you can easily obtain medical attention if necessary

What about holiday arrangements?

Before you go:-

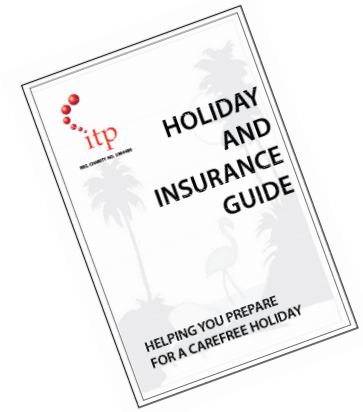
• Discuss any required vaccinations with your doctor, bearing in mind that they are not effective within three months of taking steroids or receiving an infusion of immunoglobulin. You should not have vaccinations if you are receiving rituximab and immunosuppressant drugs.

• Order any medication well in advance and make sure that you have enough to last the entire holiday.

• Tell your holiday insurance company about your ITP, or you may find you are not covered in the event of a claim (they may require a doctor's letter). The ITP Support Association has a list or recommended insurance companies in its Holiday and Insurance Guide.

• Find out what ITP is called in the country you are going to in case of an emergency. You should be able to obtain this from the country's embassy.

• Find out where the nearest doctor and hospital are situated to where you are staying.



The ITP Support Association publishes a Holiday and Insurance Guide which includes essential information about ITP, holiday insurance, flying, and vaccinations, and also has ITP factsheets in various languages with an English translation should you need to explain ITP in a foreign country.

What research is being carried out into ITP?

The Association has been funding much needed ITP clinical research since 2000. In addition to projects looking to increase understanding of all aspects of ITP and why it arises, we have also funded research into the cause of fatigue in ITP and funded a study investigating the relationship of food intolerance and ITP, and possible links with gut bacteria.

In order to increase knowledge about ITP, to try to discover the possible causes of ITP, to predict who is at risk of fatal bleeding and requires aggressive treatment, to find out why symptoms vary so much between patients and to identify its most effective treatments we support the collection of data by funding two registries. The UK Childhood Registry (under the auspices of Dr John Grainger at Manchester Children's Hospital) and the UK Adult ITP Registry (initiated by Dr Drew Provan at The Royal London Hospital) collect anonymous data on ITP patients in a long term follow-up.

We desperately need more funds to support research proposals in the pipeline. There is no state funding for ITP research.

Is ITP managed the same way in Europe and the USA?

UK ITP specialists liaise with other experts around the world, and they all share their findings at meetings, when lecturing and through publications in medical journals. The available treatments for ITP patients in Europe and the USA are the same (sometimes with different drug names) but the emphasis may vary a little from one country to another, or even one doctor to another.

Splenectomy is used more often in the USA and France than in the UK, and some of the newer drugs may have restrictions on use in some countries because of funding problems.

The 'watch and wait' option, used in particular for children in the UK but also for adults who wish to avoid drug side effects, has been emulated in other countries, particularly Canada and the USA.

We hope you found this booklet useful. If you have any questions e-mail us at info@itpsupport.org.uk and we will do our best to answer them.

The ITP Support Association is always looking for ways to promote ITP awareness and raise funds for ITP research. If you can help us by persuading your company to do a dress-down day or by taking part in a fundraising or sponsored event in aid of our charity we would be delighted to hear from you. Anyone who raises money for our cause receives a certificate and if they send a photo it will appear in our quarterly journal The Platelet.

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The Platelet Mission, Kimbolton Road,
Bolnhurst, Beds, MK44 2EL
Tel: 01234 376559
Email: info@itpsupport.org.uk
www.itpsupport.org.uk