Introduction

This leaflet explains about immune thrombocytopenic purpura (ITP), which is a blood disorder affecting the platelets. It also explains what to expect when your child is diagnosed with the condition.

What are platelets?

Platelets are one of the three types of blood cell, along with red and white blood cells. Platelets are small and sticky and their job is to prevent bruising and stop bleeding after an injury. Platelets, like red and white blood cells, are formed in the bone marrow. A rough idea of how many platelets are circulating in the bloodstream (platelet count) can be made using a sample of blood. The normal platelet count is between is 150 to 400 x 10^9/l. In most cases of ITP the platelet count is less than 20 x 10^9/l. A low platelet count is called ‘thrombocytopenia’.

What is immune thrombocytopenic purpura?

Immune thrombocytopenic purpura is a medical term for a condition in which there is bruising (purpura) because there are fewer platelets in the blood than usual (thrombocytopenic) and is usually caused by something going wrong with the immune system (the body’s defence against infection) or an allergic reaction of some kind.

Chronic ITP is the term for ITP that has not gone away on its own after 6 months. Only 1 in 4 children with ITP will develop chronic ITP. The majority of children with “chronic” ITP will still have some recovery of the platelet count at a later date and the majority of younger children will still completely recover after a few years even if the ITP is still present at 6 months.

How common is ITP and who does it affect?

About four in every 100,000 children develop ITP each year. There seem to be two groups who develop ITP: young children and young adults. It is more common in girls than boys.

What are the symptoms of ITP?

Most children with a platelet count of under 20 x 10^9/l will have petechiae (pinprick blood spots under the skin) and limited bruising. Bruising most commonly follows minor knocks (“easy bruising”) but may also occur spontaneously without trauma. Apart from the bruising/bleeding the children are otherwise well. Common sites of spontaneous bleeding are the gums and nose. Girls may be troubled with heavy periods.

Less common and potentially serious are spontaneous bleeds occurring from the gut or brain. Data from international studies suggests that the risk of serious bleeds is about 3 in 100 children and the risk of brain bleeds is about 1 in 300 children. These bleeds most often occurred in the first week of ITP and were often caused by a rare pre-existing abnormality of the blood vessels in the head. The risk of serious bleeding is much lower when the platelet count recovers to over 20 x 10^9/l.
What causes ITP?

ITP commonly results due to the immune system mistaking platelets as being foreign and attacking the platelets. In many cases this may follow a viral infection or vaccination during which time the immune system attacks the virus but the immune system then goes on to think that the platelets are viral material and starts to attack the platelets.

How is ITP diagnosed?

ITP is usually diagnosed using a blood test called a ‘full blood count’. When a sample of your child’s blood is examined under a microscope, a haematologist can examine each blood cell type closely. This is to rule out other conditions that may cause similar symptoms to ITP. If the platelets, red blood cells and white blood cells all look normal, this rules out leukaemia. If the low platelet count improves quickly and no treatment is needed, your child will not need any further tests.

If the platelet count is not showing signs of recovery by 3 to 6 months then a small sample of bone marrow will need to be taken and examined under the microscope. Additional blood tests may be taken at this time to exclude rare clotting or immune diseases that can mimic ITP. If the bone marrow looks normal, with the usual or higher number of platelet parent cells (megakaryocytes) and other blood tests are normal then the doctor will diagnose chronic ITP.

What is the outlook for children with ITP?

Many children, particularly younger ones, suddenly improve within six weeks, whether or not treatment has been given. Three out of four children will have improved by 6 months after the start of ITP. Even those who fail to recover completely will reach a platelet count over 20 x $10^9/l$ and have fewer bleeding problems. After six months about 25% of children will fully recover over the following year and over half will recover over several years.

When ITP recovers about one in 20 children will have a further occurrence in the future.

How is ITP treated?

Most children do not need any treatment unless they have severe bleeding, and most children improve whether or not treatment is given. The type of treatment recommended depends on your child’s symptoms rather than their platelet count. All the various forms of treatment aim to temporarily improve the platelet count and do not cure the condition itself. When treatments are considered, you will have the chance to discuss the risks and benefits of these, as opposed to no treatment, with the doctor. The options for treating ITP include:

1) No treatment
The majority of children with ITP have a low platelet count but do not have dangerous bleeding. If severe bleeding is not present at the time of diagnosis then it is very rare for dangerous bleeding to develop later. Without treatment most children will have a platelet count > 20 x$10^9/l$ within 5 days and a normal platelet count by six months.

2) Tranexamic acid
Tranexamic acid does not increase the platelet count but does help the blood to produce clots. It is particularly useful for gum bleeds, nose bleeds or heavy periods and helps the blood to form clots without altering the platelet count. It is best taken as a liquid (“swish and swallow”) three times per day. It most not be used if there is any blood in the urine.

3) Steroid treatment
Steroids are sometimes given to children with ITP on a short-term basis in an attempt to increase their platelet count. However, when the steroid dose is reduced, the platelet count will drop again after a few days. Steroids should only be given for a
short period of between 4 to 7 days. Side effects such as weight gain and mood changes are common. Longer courses of steroids may dampen the immune system, weaken bones, cause diabetes or obesity and stunt growth.

4) Intravenous immunoglobulin
Immunoglobulins are antibodies which can reduce platelet destruction. They are a blood product produced from many donors and have a theoretical but very low risk of transmitting blood-borne infections. One course of treatment with immunoglobulin takes two to five days as an in-patient in the hospital and the benefit will usually last about a month. Side effects such as fever and headaches are common.

5) Anti – D (WinRho)
WinRho can be used in Rhesus positive children (about 85% of children). WinRho is similar to immunoglobulin in producing antibodies which the immune system targets rather than the platelets. Anti-D is also a blood product but produced from a small number of donors. A small drop in the haemoglobin is common, rarely (1 in 40000 recipients) a severe and dangerous drop in the haemoglobin is seen. Anti-D can be given as a day case over about ten minutes and the benefit may last for several weeks.

6) Splenectomy
In ITP the majority of platelets are destroyed in the spleen. Removing the spleen (splenectomy) is often effective in preventing early destruction of the platelets and allows the count to rise. In children however this is rarely necessary unless the ITP persists and the child has recurrent severe bleeds. Splenectomy is a major surgical procedure and carries a long term risk of severe infection.

What about school, sport and holidays?

Most severe bleeds tend to occur in the first week and in children with a platelet count under 20 x10^9/l. In those children with a count over 20 x10^9/l they can return to school immediately after the head teacher has been informed about the ITP. In children with a lower platelet count school can resume after the first week and when the school have been informed. For primary school aged children it may be best if they take breaks inside if these can not be supervised. The ITP Support Association produces a document for schools, clubs and playgroups.

If your child is on steroids and has not had chicken pox then school will need to inform you if anyone in your child’s class/nursery comes down with chicken pox.

At home it is best to take sensible precautions which all children should follow such only cycling with a helmet and if swimming no diving into the shallow end! It is sensible to avoid sports where there is a risk of head injury whilst the platelet count is below 50 x10^9/l. Make sure any sports teachers are aware. With a platelet count between 50 and 100 x10^9/l there will still be more bruising so encourage the use of shin pads etc. For further details discuss with your consultant.

It is best not to take any holidays abroad in the first three months of ITP as it may be difficult to get insurance. After this time most cases of ITP will have resolved. If the ITP does persist you will need to discuss further with your doctor and you will need specialist medical insurance. A list of recommended insurance companies can be obtained from ITP Support Association (details below)
What else can I do?

Your child should also avoid drugs like aspirin, ibuprofen or herbal medication which can increase the risk of bruising and bleeding. Finally, you should make sure that doctors and dentists know that your child has a low platelet count if they are due to have an operation.

When to seek help?

When your child is sent home you will be given a clinic appointment for review at the hospital and an emergency number (usually the phone number to the children’s ward). You should contact the hospital in the following circumstances:

- A prolonged (over 30 minutes) nosebleed which will not stop despite pinching the nose
- Prolonged gum bleeding
- Blood in the poo or urine
- Following a heavy blow to the head, particularly if the child is stunned or sickly
- Persistent or severe headache
- Vomiting or drowsiness
- Children on steroids are at a greater risk of a severe form of chickenpox. If your child has not had chicken pox then contact the hospital if your child is in direct contact with someone who has chicken pox or who develops chickenpox within 7 days of being with your child.

Is there a support group?

The ITP Support Association
‘Synehurste’
Kimbolton Road
Bolnhurst
Bedford MK44 2EW
Tel 0870 777 0559  Website: www.itpsupport.org.uk

Is there a UK registry?

To maintain accurate numbers of cases of childhood ITP and investigate possible markers for risk of severe bleeding a UK registry has been established (www.uk-itp.org) Families may be routinely asked to consent for anonymous data to be stored on the registry.