Evelina London

Guy's and St Thomas' NHS
NHS Foundation Trust

Primary Immune Thrombocytopenia (ITP)

Information for patients and their families

showing we care

What is ITP?

Primary immune thrombocytopenia (ITP) is a medical term for an immune condition causing a shortage of platelets (thrombocytopenia) and bruising (purpura).

ITP that arises suddenly is known as acute ITP, if the platelet count remains low after three months it will be called persistent ITP, and if the platelet count has not returned to normal after 12 months it will be called chronic ITP. For example, someone with chronic ITP will have a troublesome condition with a low platelet count for over a year.

What are platelets?

Platelets are one of the three types of blood cells, along with red and white blood cells. Platelets are small and sticky and their job is to help the blood clot after an injury so that the wound can heal. If the blood doesn't have enough platelets it is unable to clot successfully. The result is easy bruising and the tendency to bleed for a long time after a cut. Platelets, like red and white cells, are formed in the bone marrow, which is found on the inside of bones. The normal platelet count is between150-400.

.

How common is ITP?

About four in every 100,000 children develop ITP each year.

It can occur in children and young people of any age. In older children it is more common in girls than boys, but in younger children there is little difference.

One in four children that develop ITP will go on to develop chronic ITP. The majority of children with chronic ITP will still have recovery of platelets at a later date

What causes ITP?

The specific cause of ITP is unknown. However the cause is due to something going wrong with the immune system (the body's defence against infection). In ITP the immune system mistakes the platelets as foreign and produces antibodies against the platelets. The antibody coated platelets are then destroyed by the spleen. In most cases this may follow a viral infection or vaccination during which the immune system makes antibodies against the virus but also makes antibodies against the platelets by mistake. Most children that develop ITP are quite healthy and the ITP usually resolves itself without any treatment.

What are the symptoms of ITP?

Most children with ITP will have bruising and petechiae (pinprick blood spots under the skin). Bruising usually happens following minor knocks, but may occur without any history of trauma. Apart from bruising and bleeding children are otherwise well. Common sites of bleeding are the gums and the nose. Girls may have heavy periods.

Less common, but more serious are bleeds occurring from the gut (stomach), kidneys, blood in the urine or in the brain. The risk from serious bleeds is about three in 100 children with ITP and the risk of brain bleeds is about one in 300 children with ITP. These bleeds occur most often in the first week of ITP and are usually picked up at the time of diagnosis and are most frequent in children with platelet counts less than10.

How is ITP diagnosed?

ITP is diagnosed using a blood test called a “full blood count”. In addition, a sample of your child’s blood is examined under the microscope to look at all the blood cells closely. This is to rule out other conditions that affect the blood and may also cause low platelets. If the low platelet count improves quickly and no treatment is needed your child will not require further tests.

If the platelet count does not show signs of recovery or there are findings on physical examination or in the full blood count that may indicate another diagnosis, a small sample of bone marrow may need to be taken and examined under the microscope.

Additional blood tests may be obtained at this time to check for rare clotting or immune diseases that can look similar to ITP.

Is ITP contagious?

No. ITP is not contagious.

What is the outlook for children with ITP?

Most children with ITP will improve within six weeks whether or not treatment has been given. Most of those that do not completely recover will have a platelet count over 20 and will have fewer bleeding problems. Most of these children will improve over several years. About one in 20 children with a history of ITP will have a recurrence of ITP. This usually occurs at the time of an infection or immunisation. The ITP is usually milder and resolves faster.

How is ITP treated?

Most children with ITP do not need treatment unless they have severe bleeding, and most children improve whether or not treatment is given. The type of treatment recommended depends on the bleeding symptoms rather than the platelet count. All the forms of treatment can temporarily improve the platelet count and do not cure the condition. Children with ITP are followed up regularly in the haematologist's or paediatrician's office. The frequency of clinic visits and blood tests will depend upon the severity of bleeding symptoms and the therapy the child is receiving.

These treatment options will be discussed with you and your child in more detail at your appointment. When treatment options are considered it is important to think about the risks and benefits of these as opposed to having no treatment.

The options for treatment include:

1. **No treatment**
   The majority of children with ITP have a low platelet count but do not have dangerous bleeding and so will not require therapy. If severe bleeding is not present at the time of diagnosis it is uncommon for dangerous bleeding to develop later

2. **Tranexamic acid**
   Tranexamic acid does not increase the platelet count but does help the blood to produce clots. It is useful for gum bleeds, nose bleeds or heavy periods and may be used to reduce bleeding after a dental extraction. It is best taken as a liquid three times a day.

3. **Intravenous immunoglobulin (IVIG)**
   Immunoglobulin is a liquid concentrate of antibodies purified from healthy donors. IVIG is believed to work by overwhelming the spleen with antibody so it cannot recognise the antibody coated platelets. About three out of four children have a good response to this medication within 24-48 hours of it being given and its effect can last 3-6 weeks. Immunoglobulin is administered in the hospital, over several hours through an intravenous cannula.

IVIG is used when there are significant bleeding symptoms and a higher platelet count is desired. Side effects such as fever, nausea (feeling sick) and headache are common so it is usually given with paracetamol, anti-sickness medication and chlorphenamine (also known as Piriton). There is a very low risk of transmission of blood-borne infections.

4. Steroids

Steroids are sometimes given to children with ITP on a short term basis in an attempt to increase the platelet count. However, when the steroid dose is decreased the platelet count may decrease again after a few days. Steroids should only be given for a short period of time 4-14 days. Side effects such as mood changes, stomach upset, and increased appetite are common. Longer courses or repeated short courses of steroids may dampen the immune system, weaken bones, cause weight gain, puffy cheeks and acne and ultimately may lead to diabetes. If your child is on treatment with steroids for a long time, they should avoid exposure to anyone with chicken pox, if they have not already had chicken pox.

5. Platelet transfusions

Platelet transfusions may temporarily (24 hours) increase the platelet count in children with ITP and are only given to children with severe or life-threatening bleeding. Platelet transfusions are a blood product and are obtained from single donors so there is a very low risk of transmitting blood borne infections.

6. **Splenectomy and other treatments.**
   In ITP the majority of platelets are destroyed in the spleen. Splenectomy (removing the spleen) is often effective in preventing destruction of the platelets. This is rarely necessary in children, unless the ITP persists longer than a year and the child has recurrent severe bleeding. Splenectomy is a major surgical procedure and carries a long term risk of severe infection.

Other treatments to suppress the immune system may be considered prior to a splenectomy. These may include medicines such as rituximab, or ciclosporin.

There are newer medicines that stimulate platelet production (eltrombopag, romiplostim), which can be used if other treatments have failed.

Other treatments include medicines such as rituximab, ciclosporin, eltrombopag or romiplostim. These medicines will be explained in more detail if necessary.

What will happen when my daughter starts menstruating/

Girls with ITP may experience heavy bleeding and prolonged menstruation with their first or all of their periods. If this becomes a problem oral contraceptives can be used to decrease the severity of menstruation. If this does not work, a hormone injection can be given every three months to temporarily stop menstruation.

What about school, sport and holidays?

Children who have a platelet count greater than 20 may attend the school immediately. The teacher/ school nurse should be informed about the ITP. In children with a lower platelet count school can resume after the first week. If your child is on steroids and has not had chicken pox, the school will need to inform you if anyone in your child's class or nursery comes down with chicken pox.

At home it is best to take sensible precautions which all children should follow such as cycling with a helmet and no diving into the shallow end of a swimming pool. It is sensible to avoid contact sports, where there is a risk of head injury such as rugby or boxing. There is a suggestion below on what to tell sport teachers and what action they need to take if an injury occurs.

It is best not to take any holidays abroad in the first three months of ITP, as it may be difficult to get insurance. By this time most cases of ITP will have resolved. If the ITP persists you will need to discuss this with your consultant and obtain appropriate insurance.

What else can I do for my child?

Your child should avoid medicines that affect how your platelets work such as aspirin, ibuprofen or some herbal medications. You should make sure your doctors and dentists know that your child has a low platelet count.

Keep your child active. Just because they can't play rugby doesn't mean they can't play tennis, swim or run.

Concentrate on what they can do, not what they can't.

What should I tell people?

What you tell people about your child's ITP will depend on their role in your child's life and how much your family wants other people to know.

For acquaintances you may want to say:
"*ITP is a blood clotting disorder. She/he bruises easily but it is not contagious*".

For teachers, nursery, sports staff and coaches you may want to say:
"*ITP puts him/her at risk of increased bleeding with an injury. If they bleed this is how to reach me. If trauma occurs and there is loss of consciousness, or if my child has a massive bleed, call an ambulance immediately and then call me"*.

If your child is on steroid medication you may want to add
"*the medicine makes her/him hungry, tired, irritable*".

When to seek help

Your child will be sent home with a follow up clinic appointment for review in the hospital.

You should bring your child to the hospital **immediately** in the following circumstances:

- A prolonged (over 30 minutes) nosebleed which will not stop despite pinching the nose
- prolonged gum bleeding
- blood in the faeces or urine
- a heavy blow to the head resulting in loss of consciousness – in this case call 999
- persistent or severe headache with loss of vision
- Vomiting or drowsiness.
- Your daughter has a very heavy period, lasting longer than usual (7 days) or she experiences extreme tiredness (fatigue) and dizziness

Contact details

Paediatric Haematologist - 0207 188 7774

UK ITP support group - www.itpsupport.org.uk

For more information leaflets on conditions, procedures, treatments and services offered at our hospitals, please visit www.evelinalondon.nhs.uk/leaflets

Contact us

Evelina London Medicines Helpline
If you have any questions or concerns about your child's medicines, please speak to the staff caring for them or contact our helpline.
t: 020 7188 3003 10am to 5pm, Monday to Friday
e: letstalkmedicines@gstt.nhs.uk

Your comments and concerns
For advice, support or to raise a concern, contact our Patient Advice and Liaison Service (PALS). To make a complaint, contact the complaints department.
t: 020 7188 8801 (PALS) **e:** pals@gstt.nhs.uk
t: 020 7188 3514 (complaints) **e:** complaints2@gstt.nhs.uk

Language and accessible support services
If you need an interpreter or information about your care in a different language or format, please get in touch:
t: 020 7188 8815 **e:** languagesupport@gstt.nhs.uk

NHS Choices
Provides online information and guidance on all aspects of health and healthcare, to help you make choices about your health.
w: www.nhs.uk

Leaflet number: 4356/VER1
Date published: March 2017
Review date: March 2020