

# Immune Thrombocytopenic Purpura (ITP)

## What is Immune Thrombocytopenic Purpura?

Immune Thrombocytopenic Purpura (or ITP for short) is a medical term for a condition which affects the 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 main job is to help form clots and stop bleeding after an injury. Platelets, like red and white blood cells are formed in the bone marrow.

Platelets circulate in the blood for approximately eight to ten days. By taking a sample of blood it is possible to make an estimate on how many platelets are circulating in the bloodstream (this is called a platelet count). The normal platelet count is between 150-400  $\times 10^9/L$  of blood. In most cases of ITP the platelet count is less than 20  $\times 10^9/L$ . A low platelet count is called '*thrombocytopenia*'.

In ITP there is an increase in breaking down of the platelets and therefore there is bruising (purpura) because there are fewer platelets in blood than usual. ITP is not a cancer or malignancy. ITP is not contagious and you have done nothing to cause it.

About four in every 100,000 children develop ITP each year. There seems to be two groups who develop ITP: young children and young adults. Despite the excessive bruising, ITP is generally very safe with the risk <0.5% bleeding into the brain.

## What are the signs and symptoms of ITP?

The main symptom is that your child may develop serious bruising on any part of their body, or may bleed from their nose or gums. Your child will also likely have petechiae (pinprick blood spots under the skin). For menstruating females they may have increased and prolonged bleeding during this time. Except for these signs of bleeding your child may appear well.

## What causes ITP?

We are not certain what the cause of ITP is, but it usually follows childhood viral infections. We think that this infection/reaction will trigger the body to produce antibodies to destroy the virus but unfortunately sometimes these antibodies seem to attack the platelets too, destroying them very quickly.

## How is ITP diagnosed?

ITP is usually diagnosed using a blood test called a 'complete blood count - CBC'. Sometimes a small sample of bone marrow will need to be taken and examined to rule out other causes of a low platelet count.

## What is the outlook for children with ITP?

Just as we do not understand what 'triggers' ITP to develop, we do not understand why many children suddenly recover from it. The majority of children, particularly younger ones, suddenly improve within a few weeks/months whether or not treatment has been given.

## How is ITP classified?

- Acute ITP < 3 months
- Persistent ITP 3-12 months
- Chronic ITP > 12 months

## How is ITP treated?

Most children do not need any treatment unless they have severe bleeding/bruising. The type of treatment recommended depends more 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 with your doctor. Every child is different. The options for treating ITP include:

### Steroids

Steroids are sometimes given to children with ITP on a short-term basis in an attempt to increase their platelet count. Steroids should only be given for a short period of time as side effects such as weight gain and mood changes are common the longer a person stays on steroids. It is usually given with a stomach medicine as it can cause stomach upset.

### Intravenous Immunoglobulin (IVIg)

Immunoglobulins are antibodies which can reduce platelet destruction. They are a safe blood product produced from many donors. The risk of transmitting blood-borne infections is minimal. This solution is given by infusion into a vein over several hours. It works temporarily preventing platelet destruction in the spleen. The spleen is the body organ responsible for the rapid destruction of the platelets. The benefits can usually last from a few weeks to a month. Side effects such as headaches and fever are common.

### WinRho/Rhogam/Anti Rh D Immunoglobulin

These antibodies can only be used for blood group Rh (D) positive individuals (85% Caucasian). They target red cells and by doing so, distract the immune system to decrease immune platelet destruction. Side effects include a drop in the level of hemoglobin (red blood cells) in the blood and your child may appear paler and slightly yellower than normal.

### Splenectomy

If ITP does not respond to medical treatment, or the side effects are too severe, a splenectomy may be required. A splenectomy is the removal of the spleen by a surgical procedure. Following treatment, the platelet count should rise. Splenectomy is a major surgical procedure and carries long term risk of serious infection with a success rate of about 60-70%. It is usually considered a treatment of last resort.

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## Other treatments

If ITP is not responsive to steroids, IVIG or WinRho, “second line” treatment may be indicated. These are treatments with lower chance of response, and may have more side effects. These include Vincristine, Rituximab, or a combination of different treatments. Bone marrow platelet stimulants (Romiplostim, Eltrombopag) are recent additions to the armamentarium. These are reserved for the cases resistant to all other treatments. There are other treatments currently undergoing trials, the medical field is always advancing. Tried treatments should be reserved for children with serious long-term problems or complex disorders of their immune system.

## What else can I do?

Your child should also avoid drugs like aspirin and ibuprofen. This is because these drugs interfere with platelet function and may increase their risk of bleeding. Herbal medicines/Vitamins and Minerals like cod liver oil and Vitamin E should also be avoided. If you are unsure ask your hematologist or the pharmacist.

Finally, you should make sure that doctors and dentists know that your child has a low platelet count if they are due to have any procedure or operation.

## ITP and physical activity

Your child’s level of physical activity should be restricted until their platelet count is above  $50 \times 10^9/L$ . If you have any concerns regarding these restrictions, please contact your hematologist or the hematology nurse clinician. Limited physical activity will decrease the risk of bleeding from injury or trauma.

Contact sports like football, soccer and hockey should be avoided until your child’s platelet count has reached  $100 \times 10^9/L$ . Swimming is allowed, diving is not. If your child rides a bicycle, skateboards or roller skates, please ensure a helmet is worn at all times.

Seek medical attention right away if your child receives a head injury or bump to their head and develops headache, drowsiness, vomiting, or decreased level of consciousness.

## When to seek help

- A prolonged (over 30 minutes nose bleed) which does not stop despite pinching the nose
- Prolonged gum bleeding
- Blood in stool or urine
- Following a heavy blow to the head
- Persistent or severe headaches
- Vomiting and drowsiness