

Childhood Immune Thrombocytopenia (ITP)

Immune Thrombocytopenia (ITP) (previously called “idiopathic thrombocytopenia”) is a condition where the *platelets*, which are small blood cells that are important to help the blood clot, are reduced because they are attacked and removed by the body’s immune system. This leads to a tendency to bruise and bleed. ITP affects 1000 children each year in Gujarat.

What causes ITP?

ITP results from the body making antibodies against platelets that attack and remove them from the bloodstream, the main site of destruction being the spleen. In some children this may follow a viral illness (for example, EBV) but most often the cause is unknown. ITP is neither inherited, nor can it spread from one child to another in the family, or amongst playmates.

What are the symptoms of ITP?

ITP often presents dramatically with bruising that covers the body, and tiny red dots on the skin (*petechiae*). Sometimes nosebleeds, bleeding from the mouth and bowels can occur. Very rarely (around 1:1000 patients) life-threatening bleeding, such as bleeding into the brain occurs.

How is ITP diagnosed?

Usually these are healthy children without fevers, spleen enlargement, or bony aches and pains. The sudden onset of bruising/bleeding in an otherwise well-looking child is suspicious for ITP, and the blood tests listed below are often enough to make a diagnosis.

- **Complete Blood Count (CBC)** is a blood test that measures all the circulating blood cells, and should show decreased platelets with a normal red and white cell count. A normal platelet count is usually 150,000 to 400,000/mm³, but in ITP the counts can be 5000/mm³ or less which the CBC machine may struggle to count accurately
- **Peripheral blood smear** is a test where a trained pathologists looks at the blood under a microscope. This is extremely helpful as new platelets, being produced as the older ones are destroyed by antibodies, are large enough for the CBC machine to mistake as red cells but can be easily counted under the microscope. It is important to exclude other diseases that can cause low platelets and review of the blood smear can pick up any other blood cell abnormality

In children who have any abnormalities in their blood count or smear, or whose history or exam is atypical, your doctor may need to do a test called a bone marrow aspirate to exclude other blood diseases.

What is the outcome in ITP patients?

The majority of patients in childhood recover within the first few months without any treatment.

- **Newly Diagnosed ITP (60-70% children)** is temporary and lasts for less than 3 months. It is the most common form of ITP in children.
- **Persistent ITP (10-20% children)** disappears 3-12 months after diagnosis
- **Chronic ITP (10-20% children)** lasts over 12 months and is more common in adults.

Since children with ITP are rapidly producing platelets, which are quickly destroyed, young large functional platelets are present in the bloodstream and bleeding is rare. Since most children will recover without intervention the modern approach recommended by the US and UK Hematology societies is “watchful waiting”. This can be anxiety-provoking for the family, so the child is carefully followed by the hematologist. A CBC is checked every few weeks to look for platelet count recovery, but **we treat the bleeding, not the platelet count**. Fortunately, once ITP resolves it typically does not come back, although in 1-4% children this may happen.

How is ITP treated?

Although the majority of children will not need intervention, treatment is sometimes given to prevent or treat active bleeding (e.g. severe nosebleed), and medications fall into two categories:

Those that suppress destruction of platelets:

- **Corticosteroids (e.g. Prednisone)** suppresses the immune system and production of antibodies that are destroying platelets. Common side effects include irritability, weight gain, high BP, acne and upset stomach.
- **Immune globulin (IVIG)** blocks sites in the spleen where the platelets are being destroyed. Common side effects include fever, chills, headache, stiff neck, flushing, itching, and low BP.
- **Anti-D (WinRho)** is only an option for patients with Rh-positive blood type, and acts by destroying red cells that then interferes with destruction of platelets in the spleen. Common side effects include headache, chills and fever.
- **Rituximab (Rituxan)** is reserved for patients with chronic disease and destroys the white cells making antibodies against the platelets. Common side effects include fever, chills, weakness, nausea and headaches; the immune suppressive effect lasts over a year.
- **Splenectomy** is reserved for patients with chronic disease and is surgery to remove the spleen, which takes away the site where platelets are being destroyed. It increases the likelihood of severe infections in children, so hence rarely done.

Those that stimulate production of platelets:

- **Platelet stimulating drugs** (e.g. Eltrombopag, Romiplostim) mimic a hormone called “thrombopoietin” that helps the bone marrow make platelets more quickly. Side-effects vary by drug and your doctor will provide you with more information as these are not usually prescribed as a first-line treatment

What precautions should I take if my child has ITP?

- No specific dietary restrictions or supplements are helpful.
- Avoid contact sports, rough play, bike riding or activities which make head injury likely.
- Avoid medications containing aspirin or ibuprofen (NSAID), and check with your doctor if uncertain as many cough/cold or pain medications may have these drugs.
- Notify your hematologist if there is bleeding that will not stop within ten minutes, severe headache, vomiting of blood, or blood in the urine or stool.