

Pediatric Idiopathic Thrombocytopenic Purpura (ITP, Immune Thrombocytopenic Purpura) — Symptoms and Treatment

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ITP is a disorder that causes bleeding as a result of the lowered platelet count. The acute type is more common in children and the cause is unknown. It has more chances of spontaneous settlement without any specific pharmacological therapy. The chronic form continues for more than 6 months, it is linked to the defective immunological processes. Diagnosis is mostly clinical and some laboratory tests are performed. Treatment consists of general measures and specific medicines. Splenectomy is reserved for the resistant cases of ITP.



Discussion of Pediatric Idiopathic

Thrombocytopenic Purpura

It is a bleeding disorder that can occur in the pediatric population. ITP is an abbreviation of Idiopathic Thrombocytopenic Purpura. The bleeding occurs as the result of low platelets count, which is responsible for the formation of clots. The **deficiency of platelets results in impaired clotting mechanism and thus frequent bleeding.**

As the name suggests, the exact etiology is still not known, but many causative factors have been studied in association with ITP. ITP is also called primary immune thrombocytopenic purpura. The bone marrow and the production of platelets are mostly normal. The **number of platelets gets reduced as a result of destruction.**

The following **criteria** are set as the **definition** of ITP:

- Thrombocytopenia (It is defined as the decreased platelet count, <150,000 cubic mm)
- Bone marrow is normal
- Purpuric rash
- An absence of any other identifiable causes

Types of Pediatric Idiopathic Thrombocytopenic Purpura

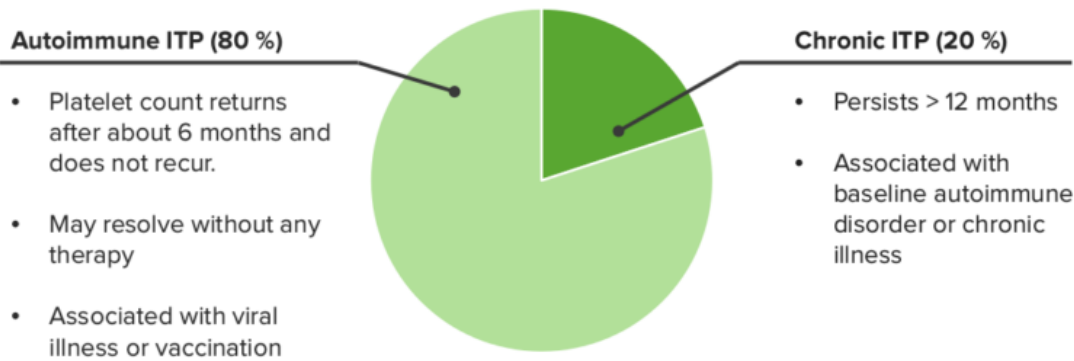
Acute form

It starts after an acute infection and resolves spontaneously within two months. It is the more common form of ITP that occurs in children. It follows viral infections such as rubella, measles, varicella and Epstein-Barr virus.

Chronic form

It persists more than 6 months without having any identified underlying cause. It results from the immune clearance of the platelets. The **platelets having the IgG or IgM antibodies are cleared from the circulation by the spleen.** The spleen performs both the function of the formation of antibodies and sequestering the platelets. It is mostly associated with the following factors:

1. **Female sex**
2. **The presence of other autoimmune diseases**
3. **The age of presentation is more than 10 years**



Epidemiology

In the US, there are about 66 cases of ITP out of 1,000,000 per year. In children, the average number of cases per year is 50 out of 1,000,000. About 40 % of ITP patients are less than 10 years of age, the condition is most prevalent in children 2 to 4 years old. In adults, the condition is most prevalent in people in the 20 to 50 years range.

In a study by the International Childhood ITP Registry (ICIS), 2031 children with the condition were studied. The research involved 136 centers across 38 countries. According to the study, ITP occurred most frequently in the spring and least frequently in the autumn.

Their relation between chronic ITP in children and *Helicobacter pylori* has not been proven. One study found *H. pylori*'s occurrence in children with the condition to be similar to the general population. Moreover, eliminating *H. pylori* did not have any effect on the recovery of the children.

Pathophysiology of Pediatric Idiopathic Thrombocytopenic Purpura

The peripheral destruction of platelets takes place in ITP. Many patients exhibit antibodies in their blood that attach to the glycoprotein on the platelets' membrane. The **spleen and other reticuloendothelial organs destroy the antibody-coated platelets**. In addition to this immunological destruction of the platelets, there is some evidence that the **defective platelet production can also be the cause**, especially in the chronic type of ITP.

Although the number of megakaryocytes is normal or slightly increased in the bone marrow, the plasma thrombopoietin levels which are responsible for maturation and proliferation of progenitor cells are reduced. This confirms the defective activity of megakaryocytes.

Clinical Features of Pediatric Idiopathic Thrombocytopenic Purpura

The **signs and symptoms** of ITP include:



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- Petechia, ecchymosis, bruising, and purpuric rash
- Bleeding from mucosal membranes, such as gum bleeding and nose bleeding
- Bleeding from the urinary tract, conjunctivae, and retina
- Generalized bruises
- No lymphadenopathy, hepatosplenomegaly, or other signs of leukemia
- In the chronic cases, the onset is slow and a patient can be completely asymptomatic



Image: "Trombocitopenia 01" by AfroBrazilian. License: [CC BY-SA 4.0](https://creativecommons.org/licenses/by-sa/4.0/)

Diagnosis of Pediatric Idiopathic Thrombocytopenic Purpura

The diagnosis is **mostly clinical**. The following investigations are performed to confirm the clinical diagnosis:

- **Complete blood count:** The platelet count is low, usually less than 50,000/cubic mm. In about 10 % of the children, there can be mildly reduced red blood cells count
- The **PT** (Prothrombin Time) and **APTT** (Activated Partial thromboplastin time) are normal.
- Bleeding time (**BT**) is prolonged
- **Bone marrow examination** may show increased megakaryocytes
- **Platelet-associated IgM** and **IgG** are demonstratable on platelets
- **A CT scan of the head** can be performed if there is any suspicion of intracranial hemorrhage

Management of Pediatric Idiopathic Thrombocytopenic Purpura

The management will be discussed separately for acute and chronic cases of ITP.

Treatment of Acute ITP

General approach

- Securing the airway
- Maintenance of fluids and electrolytes
- Preventing the trauma
- Avoid using medicines such as aspirin, chloramphenicol, carbamazepine, sulfonamides, valproic acid, heparin, and digoxin

Specific Therapy

- Most of the cases **do not require any further treatment**
- Platelets transfusion is started in the emergency cases if the count is significantly dropped
- **IV immunoglobins** are the mainstay of therapy. They can raise the count within 2-3 days
- **Corticosteroids** (Prednisolone): The dose is 2-4 mg/kg in the divided doses for one to four weeks. It is then slowly tapered off
- **Anti-Rh (D)**: It is only useful in Rh-positive patients. 80% of the children show a response to this treatment

Treatment of Chronic ITP

General approach

- Avoid medicines that suppress the production of platelets
- Restrict the activities that can increase the risks of trauma
- **Pharmacological therapy** includes the IVIG, steroids, anti-Rh D, androgens and immunosuppressive agents (Cyclophosphamide, Rituximab, and Azathioprine)

Specific therapy

Most of the chronic cases of ITP keep remitting; therefore, a more useful therapy is needed to maintain the normal platelet count to avoid the repeated bleeding.

Splenectomy

It is done in cases which are **resistant to medical therapy**. It is considered after one year of persistent thrombocytopenia.

- This procedure is associated with increased **risks of infections**, especially in very young children; therefore, it is **delayed until the child is at least 5 years old**
- The **vaccination cover** for Hemophilus influenza b, meningococcal, and pneumococcal is provided. Prophylactic penicillin is also given to the patient

Complications of Pediatric Idiopathic Thrombocytopenic Purpura

Intracranial bleeding is the most feared complication. The other complications are the side-effects of corticosteroids.

- Infections
- Increased blood sugar levels
- Thinning of skin
- Cataracts
- Osteoporosis

Prognosis of Pediatric Idiopathic Thrombocytopenic Purpura

The overall prognosis of Idiopathic Thrombocytopenic Purpura is **good**. 90 % of children will have a spontaneous recovery from the disease.

References

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