Complications of ITP
A critical complication of ITP is bleeding. Typically, the risk for spontaneous bleeding is increased when the platelet count is below 20,000 cells/mm³ and usually below 10,000 cells/mm³, or when medications that interfere with platelet function are also utilized by the patient. Therefore, it is important to monitor platelet counts closely throughout the course of treatment.

Complete responses are generally defined as sustained platelet counts over 100,000 cells/mm³. In pediatric patients, one third resolve by six weeks after diagnosis, another third in six months and a further third become chronic. Patients diagnosed in infancy or in their adolescent years are at higher risk to develop a chronic course. One third of adult patients remain in remission 5 years from initial diagnosis, while two thirds are at increased risk for chronic ITP.

The most worrisome complication of ITP is bleeding. Typically, the risk for spontaneous bleeding is increased when the platelet count is below 20,000 cells/mm³ and usually below 10,000 cells/mm³, or when medications that interfere with platelet function are also utilized by the patient. Therefore, it is important to monitor platelet counts closely throughout the course of treatment.

Other frequent observed complications result from the ITP flare from the therapies utilized for treatment. Long-term steroid use may result in conditions such as hyperglycemia, diabetes mellitus, osteoporosis, weight gain, infections, and delay in the ability to become pregnant. When steroids are utilized for therapy, the shortest efficacious course should be employed to decrease the risk of complications. As with all medications, the goal of treatment should always be weighed against potential treatment related risks.

What is the pathophysiology of ITP?
INTRODUCTION:
What is ITP?
ITP, or immune thrombocytopenic purpura, is an acquired bleeding disorder in which the immune system destroys platelets. Blood cells that are normally produced in a primary event of fetal life, develop with a temporary (transient) primary immune thrombocytopenia (ITP) which develops with a chronic course. Most cases arise unexpectedly, and prompt diagnosis and treatment are essential to reduce the chances of death and disability caused by hemorrhage.

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Cytomegalovirus
Hepatitis C
HIV
Helicobacter pylori
CMV
HCV
HIV
H. pylori
Other bacteria
Shigella
Salmonella
Staphylococcus
Streptococcus
Other viruses
Echovirus
Parvovirus
Mumps
Measles
Epstein-Barr virus
Hodgkin’s disease
Non-Hodgkin’s lymphoma
Leukemia
Sickle cell disease
Sideroblastic anemia
Myelodysplasia
Other myeloproliferative disorders
What are the causes of ITP?
ITP can arise due to a variety of causes. They may be acquired, infectious, or idiopathic, and cannot be classified as autoimmune. ITP may be caused by a variety of factors, including but not limited to injury to the bone marrow, infection, or idiopathic. ITP may also occur as a result of medications or other substances that may cause a decrease in platelet production or an increase in platelet destruction.

The classic triad of chronic ITP consists of purpura (easy bruising), and petechiae (extravasation of blood under the skin) and oral gum bleeding. In addition, the spleen may be enlarged in some cases and the patient may experience paleness, fatigue, and dizziness.

What is the pathophysiology of ITP?
PATHOPHYSIOLOGY:
Immunologic mechanisms:
Immunologic mechanisms play a critical role in the pathophysiology of ITP. The immune system is responsible for removing cells and other foreign substances from the body. However, in ITP, the immune system mistakenly targets and destroys platelets, leading to their reduced number in the blood.

Infectious triggers:
The immune system plays a crucial role in fighting off infections. However, in some cases, the immune system mistakes normal blood cells for foreign substances and destroys them. This can lead to a decrease in the number of platelets, causing symptoms of ITP.

Maternal-fetal transfusion:
Maternal-fetal transfusion may occur during pregnancy when the mother’s immune system mistakes the fetal platelets for foreign substances. This can lead to a decrease in the number of platelets, resulting in symptoms of ITP.

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What is the epidemiology of ITP?

ITP is considered a rare hematologic condition with a prevalence of 100 per million in the general population. The incidence based on large registry studies and estimated to be 100 per million. Overall: 85% acute, 15% chronic. Overall incidence in childhood is based on large registry studies with an estimate of 1,000 per million. What is the age distribution of ITP?

ITP is more common in children than adults, with the majority of cases occurring in the first 2 years of life. In children, the most common age of occurrence is 3 to 12 months. In adults, the most common age of occurrence is over age 75 and less than 18 years. The predominance is primarily on patients who develop clinical symptoms. In adults, males are more commonly affected than females, with a male to female ratio of 1.2:1.0. There is some suggestion that ITP has seasonal variation, this finding has not been confirmed, however, it is known that ITP is more common in the cooler months of the year. The overall incidence in adulthood is based on large registry studies with an estimate of 100 per million. The incidence in adult males and females is approximately equal, however, a greater proportion of women older age subgroups in females exceed that of males. To date, there are no known genetic or ethnic subgroups in which ITP is more prevalent. What is chronic ITP?

Chronic ITP refers to the development of isolated thrombocytopenia with an a platelet count below the normal range (less than 150,000 cells/mm³) that resolves most often in less than 6 months is termed acute. “acute” refers not to the onset of the disorder, but rather its duration. Acute ITP refers to the development of isolated thrombocytopenia with a platelet count below the normal range (less than 150,000 cells/mm³) that resolves most often in less than 6 months is termed acute. “acute” refers not to the onset of the disorder, but rather its duration. Chronic ITP that resolves most often in less than 6 months is termed acute. “acute” refers not to the onset of the disorder, but rather its duration. Oski’s Hematology of Infancy and Childhood.

Table 2. Characteristics of Childhood versus Adult ITP

<table>
<thead>
<tr>
<th>Age</th>
<th>Course</th>
<th>Predominance</th>
<th>Age</th>
<th>Course</th>
<th>Predominance</th>
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<tbody>
<tr>
<td>Primarily peripheral destruction of platelets</td>
<td>Male to female ratio 1.2 : 1.0</td>
<td>3 to 12 months</td>
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What is the pathophysiology of ITP?

ITP is a clinicopathologic diagnosis. A detailed history, including the onset and pattern of bleeding, is important in the diagnosis of ITP in conjunction with appropriate laboratory testing. The typical finding is a bone marrow biopsy of a patient with ITP is an increase in megakaryocytes without other characteristic abnormalities. Some marrow aspirates and biopsies are performed based on the clinical context and evidence indicated in the patient's medical history.

FIGURE 1 & 2. Case presentation

2. Bone marrow evaluation. If the clinical presentation and review of the blood smear is typical of ITP, then bone marrow aspiration and biopsy is not indicated. However, if the clinical presentation and review of the blood smear is atypical, bone marrow aspiration and biopsy are performed based on the clinical context and evidence indicated in the patient's medical history.

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4. Viral studies: platelet clumping in EDTA anticoagulant and cold agglutinins. Testing for platelet antibodies is only performed in specialized laboratories. The absence of platelet antibodies does not rule out ITP, therefore, platelet antibody testing is not performed on a routine basis. Many hematologists perform a bone marrow aspirate and biopsy as part of the diagnostic workup. The most common cause of thrombocytopenia is autoimmune destruction of platelets.

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What is acute ITP?

ITP is considered acute ITP by most hematologists if it has persisted for 3 to 12 months. It should be noted that children of any age can be affected with ITP. Between ages 2 - 5 years, followed by adolescence.

What is the epidemiology of ITP?

The annual incidence of ITP is about 3 to 8 cases per 100,000 children with a peak in the two to five year age group. It should be noted that since platelets play a pivotal role in primary hemostasis, quantitation of platelet-associated biochemical and mechanical processes is important in the diagnosis of ITP.

Since platelets play a pivotal role in primary hemostasis, quantitation of platelet-associated biochemical and mechanical processes is important in the diagnosis of ITP.

Uncommonly, patients may be asymptomatic and ITP is incidentally detected during laboratory testing performed for an unrelated issue.

Overview of Adult versus Pediatric ITP

Chronic ITP is usually defined in children as a platelet count below the normal range (less than 150,000 cells/mm³) that remains below this level for 3 to 12 months without treatment. Chronic ITP is considered chronic ITP by most hematologists if it has persisted for more than 12 months. Chronic ITP is usually defined in adults as a platelet count below the normal range (less than 150,000 cells/mm³) that remains below this level for 12 months or more without treatment. Chronic ITP is usually defined in adults as a platelet count below the normal range (less than 150,000 cells/mm³) that remains below this level for 12 months or more without treatment.
The disease is acute by definition, with a mean duration of 3 to 12 months, although this varies from patient to patient. The majority of children recover within 1 year. The incidence of ITP is higher in adolescents, with more chronic cases seen in this age group. In general, ITP is considered chronic if it persists beyond 12 months. The overall incidence of ITP is 1.5 to 10.0 cases per 100,000 children per year, with a female predominance of 1.2:1.

**Epidemiology:**

### Incidence of Pediatric vs. Adult ITP

- **Pediatric ITP:**
  - Approximately 3 to 10 cases per 100,000 children are affected, with a peak incidence between the ages of 2 to 5 years.
  - The disease is more common in children than in adults, with a prevalence of 1.5 to 10.0 cases per 100,000 children per year.
  - A female predominance of 1.2:1 is observed.

- **Adult ITP:**
  - The incidence is lower, with approximately 30 cases per 100,000 adults per year.
  - The disease is more common in adults than in children, with a prevalence of 0.5 to 2.0 cases per 100,000 adults per year.
  - A female predominance of 2:1 is observed.

### Acute vs. Chronic ITP

- **Acute ITP:** A diagnosis of acute ITP is made when the platelet count is below 50,000/mm³ and there are no signs of active bleeding.
  - The disease is self-limited and typically resolves within 12 months.
  - The signs and symptoms are usually mild, with a resolution rate of 85% acute, 15% chronic.
  - The causes include: infections, vaccinations, and drugs.

- **Chronic ITP:**
  - A diagnosis of chronic ITP is made when the disease persists beyond 12 months.
  - The disease is characterized by recurrent episodes of thrombocytopenia and a high risk of bleeding.
  - The causes include: autoimmune disorders, infections, and malignancies.

### Diagnosis of ITP

**Common Presentations & Symptoms**

**What are the signs and symptoms of ITP?**

- Petechiae, purpura, ecchymoses, subcutaneous hemorrhage, mucosal bleeding, bleeding from open wounds, gingival bleeding, epistaxis, conjunctival bleeding, menorrhagia, and hematochezia.

**DISEASE DURATION**

- Acquired ~ 3 - 8 cases per 100,000 children per year.
- This range is dependent on the development of bleeding symptoms.

**Overall: 85% acute, 15% chronic**

**What is acute ITP?**

Acute ITP refers to the development of isolated thrombocytopenia with bleeding symptoms. The signs and symptoms are usually mild, with a resolution rate of 85% acute, 15% chronic.

**What is the epidemiology of ITP?**

- ITP is considered acute if the platelet count is below 50,000/mm³ and there are no signs of active bleeding.
- The disease is self-limited and typically resolves within 12 months.
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**Acute ITP**

- The disease is acute and self-limited, with a mean duration of 3 to 12 months.
- The incidence of ITP is higher in adolescents, with more chronic cases seen in this age group.
- In general, ITP is considered chronic if it persists beyond 12 months.
- The overall incidence of ITP is 1.5 to 10.0 cases per 100,000 children per year, with a female predominance of 1.2:1.

**Chronic ITP**

- The disease is chronic and may persist for years, with a female predominance of 2:1.
- The signs and symptoms are usually mild, with a resolution rate of 85% acute, 15% chronic.
- The causes include: autoimmune disorders, infections, and malignancies.

**What is ITP?**

ITP, or idiopathic thrombocytopenic purpura, is a disease of unexplained thrombocytopenia (i.e., low platelet count) in association with the clinical features of spontaneous thrombocytopenia and bleeding manifestations. ITP is characterized by the absence of any known cause or identifiable trigger for the thrombocytopenia. The disease is more common in children than in adults, with a prevalence of 1.5 to 10.0 cases per 100,000 children per year.

**INCIDENCE**

- Incidence: 1.5 to 10.0 cases per 100,000 children per year.
- Female predominance: 1.2:1.

**Prevalence**

- Prevalence: 0.5 to 2.0 cases per 100,000 adults per year.
- Female predominance: 2:1.

**Pathophysiology**

- **Pediatric ITP:**
  - Characterized by the destruction of platelets in the spleen and liver, leading to a decrease in the number of circulating platelets.
  - The spleen is often enlarged.

- **Adult ITP:**
  - Characterized by the destruction of platelets in the bone marrow, leading to a decrease in the number of circulating platelets.
  - The spleen is often enlarged.

**Common Presentations & Symptoms**

- Petechiae, purpura, ecchymoses, subcutaneous hemorrhage, mucosal bleeding, bleeding from open wounds, gingival bleeding, epistaxis, conjunctival bleeding, menorrhagia, and hematochezia.

**Common Presenting Symptoms in Patients with ITP**

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Platelet Disorder Support Association (PDSA): www.pdsa.org
The ITP Support Association: www.itpsupport.org.uk

What is ITP?
Primary immune thrombocytopenia (ITP) is an acquired bleeding disorder in which the immune system destroys platelets. Blood cells that naturally circulate for a primary reason to be removed. Patients with ITP develop thrombocytopenia with a platelet count below the normal range, generally defined as less than 100,000 cells/mm³. Thrombocytopenia commonly manifests as a bleeding tendency, including epistaxis (nosebleeds), petechiae (small purpura), and purpura (large purpura). Blood from capillaries into skin and mucous membranes.

Introduction
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What is the pathophysiology of ITP? The underlying pathologic process resulting in ITP is the generation of autoantibodies that react with the platelet surface antigens. Once bound to the platelets, these autoantibodies cause platelets to be removed from circulation through phagocytosis by the reticuloendothelial system, primarily by the spleen. The resulting shortened platelet half-life results in thrombocytopenia, the level of thrombocytopenia observed is based upon each affected individual's burden to platelet production. The unreacted, or pool of mononuclear reactive platelets, can also be found reactive platelets.

The most common mechanisms involved in the development of ITP include: autoantibodies, viral infections, and idiopathic. Autoantibodies are most often directed against platelet glycoproteins. Viral infections can result in ITP due to the immune response to the virus. Autoantibody formation can also result in ITP. In this case, the autoantibodies are directed against the platelet surface antigens.

What are the future directions in the management of ITP? The future directions in the management of ITP include: more effective therapies, better understanding of the disease, and improved quality of life. More effective therapies may include the use of new drugs, such as eltrombopag, which is approved for the treatment of ITP. Improved understanding of the disease may include the discovery of new mechanisms underlying the disease. Improved quality of life may include the development of new treatments that reduce the symptoms of ITP.

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What are the future directions in the management of ITP? The future directions in the management of ITP include: more effective therapies, better understanding of the disease, and improved quality of life. More effective therapies may include the use of new drugs, such as eltrombopag, which is approved for the treatment of ITP. Improved understanding of the disease may include the discovery of new mechanisms underlying the disease. Improved quality of life may include the development of new treatments that reduce the symptoms of ITP.

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One of the most worrisome complications of ITP is bleeding. Typically, the risk for spontaneous bleeding is increased when the platelet count is below 20,000 cells/mm³ and usually below 10,000 cells/mm³, or when medications that interfere with platelet function are also utilized by the patient. Patients diagnosed in infancy or in their adolescent years are at higher risk to develop a chronic course. One third of adult patients remain in remission 5 years from initial diagnosis, while two thirds are at risk for relapse. Patients diagnosed in infancy or in their adolescent years are at higher risk to develop a chronic course. One third of adult patients remain in remission 5 years from initial diagnosis, while two thirds are at risk for relapse.

CLINICAL HISTORY OF BLEEDING

First Tier Testing
- Thrombocytopenia platelet count < 50,000
- Platelet function testing
- History of bleeding
- Personal and family history
- Physical examination

Second Tier Testing
- Computing platelet life span
- Plasma exchange
- Fibrinogen assessment
- Additional tests for storage pool disorders

Third Tier Testing
- Bone marrow aspiration and biopsy
- Platelet electron microscopy
- Flow cytometry
- Flow cytometric analysis of granulocytes

CLINICAL HISTORY OF BLEEDING

PATHOPHYSIOLOGY:

- Immunologic mechanisms:
  - Development of autoantibodies against platelet membrane antigens
  - Removal of platelet precursors from the bone marrow
  - Increased platelet destruction in the spleen
- Cell mediated mechanisms:
  - Lysis of platelets by activated T lymphocytes
- Hematopoietic mechanisms:
  - Reduction in the rate of platelet production
- Other mechanisms:
  - Reduced production of platelets in the bone marrow
  - Increased destruction of platelets in the spleen

COMPLICATIONS OF ITP

- Hemorrhagic shock
- Severe anemia
- Splenic rupture
- Pneumothorax
- Intracranial hemorrhage
- Other complications

What are the future directions in the management of ITP?

- Development of novel therapeutic strategies
- Improved understanding of the underlying pathogenesis
- Personalized medicine
- Gene therapy

IMMUNE THROMBOCYTOPENIC PURPURA (ITP):

- A New Look at an Old Disorder

INTRODUCTION:

- What is ITP?
- Immune thrombocytopenic purpura (ITP) is an acquired bleeding disorder in which the immune system destroys platelets. Blood cells that circulate in the body play a primary role in the body’s defense system. Patients with ITP develop thrombocytopenia with a platelet count below the normal range usually defined as less than 150,000 cells/mm³. Thrombocytopenia commonly manifests as a bleeding tendency, including spontaneous bruising and petechiae (small blood spots from capillaries into skin and mucous membranes).

PHYSIOPATHOLOGY:

- The underlying pathophysiological process in ITP is the generation of autoantibodies that react with platelet antigens. Once bound to the platelet, the autoantibodies cause platelet to be removed from circulation through phagocytosis by the reticuloendothelial system, primarily the spleen. The resulting shortened survival of platelet in the peripheral circulation is the level of thrombocytopenia observed based on such affected individual’s balance between production and natural destruction. The platelet survival time in ITP is normal, and no advisories of compensatory platelet production in ITP.

Three other frequent complications result not from the ITP but from the therapies utilized for treatment. Long-term steroid use may alter the HPA axis, and manifest as growth failure and delayed sexual maturation. Corticosteroids can also cause immunosupression, a state where the body’s immune system will not respond to infections. Other long-term complications include structural changes in the skeletal system, such as osteoporosis, and other immunosupression, where the immune system response to infection is reduced.

The most common mechanisms involved in ITP development is the generation of autoantibodies. These are most frequently directed against platelet glycoprotein IIb/IIIa (GPIIb/IIIa), a cell surface antigen that facilitates the attachment of platelets. Other mechanisms include genetic defects affecting platelet surface antigens, such as loss of expression of specific antigens, which may be involved in the generation of ITP.

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