## Idiopathic Thrombocytopenic Purpura (ITP)

<table>
<thead>
<tr>
<th>Feature</th>
<th>Disease Facts</th>
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<td><strong>Synonyms</strong></td>
<td>ITP, primary immune thrombocytopenic purpura, autoimmune thrombocytopenic purpura.</td>
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<td><strong>Epidemiology</strong></td>
<td>True incidence is unknown, since individuals with mild disease may be asymptomatic and undiagnosed. In the United States, symptomatic disease occurs in about 70 adults/1,000,000 and 50 children/1,000,000.</td>
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<td><strong>Etiology &amp; Pathogenesis</strong></td>
<td>Autoimmune disease, with anti-platelet antibody formation (usually specific for the platelet GPIIbIIIa receptor) and peripheral platelet destruction. Acute ITP usually occurs in children, follows an acute viral infection or recent live virus immunization, and usually undergoes a spontaneous remission within two months. Chronic ITP is more common in adults, shows an insidious presentation, and lasts &gt; 6 months. The pathogenesis is complex and multifactorial, including a failure of self-antigen recognition and tolerance, altered cytokine secretion, impaired megakaryopoiesis, and impaired cell-mediated cytotoxicity.</td>
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<td><strong>Clinical Presentation</strong></td>
<td>Symptomatic individuals present with “platelet-type” bleeding problems. These include non-palpable petechiae, purpura, epistaxis, gingival bleeding, menorrhagia, and easy bruising. GI bleeding, hematuria, retinal hemorrhage, and even intracranial hemorrhage can occur in severe cases. Splenomegaly is unusual.</td>
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<td><strong>Differential Diagnosis</strong></td>
<td>All causes of thrombocytopenia must be excluded. The major considerations include pseudothrombocytopenia, DIC, viral infections (especially HIV and EBV), TTP, and drug-induced thrombocytopenia (heparin, alcohol, sulfonamides, quinine, quinidine). Other disease in the differential include pregnancy-associated thrombocytopenia, autoimmune disease (especially lupus erythematosus), other infections, liver disease, hematologic and other malignancies, transfusion reaction.</td>
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<td><strong>Laboratory Features</strong></td>
<td>Laboratory studies are more valuable in the exclusion of other diseases than in providing definitive evidence of ITP. Variable thrombocytopenia with a normal white blood count and erythrocytic indices (in the absence of severe hemorrhage). Peripheral blood smear examination reveals thrombocytopenia with normal leukocytic and erythrocytic morphology. Coagulation assays are within normal limits. Bone marrow evaluation reveals normal to increased megakaryocytes with normal morphology. Assays for platelet-associated immunoglobulin by ELISA or flow cytometry may be positive, but these assays are presently considered unreliable. Pseudothrombocytopenia must be excluded by peripheral smear examination for platelet clumping. The exclusion of other causes of thrombocytopenia should include other laboratory assays, especially a CBC, D-dimer and FDP assay, LDH, viral serology (especially for HIV), anti-nuclear antibody, rheumatoid factor studies, and ESR, liver function studies, and pregnancy test.</td>
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### Treatment

Supportive care, including platelet transfusions, must be provided until remission occurs or treatment is effective. No specific treatment is usually provided for children with platelet counts >30,000/µL or adults with >50,000 platelets/µL. Intravenous immunoglobulin (IVIG, IV Rh immune globulin, IV anti-Rh(D)) and corticosteroids are the first line of therapy for patients with severe thrombocytopenia. Rituximab is often used in patients with refractory ITP.

### References
