

# Childhood ITP

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## Abstract

Idiopathic (or immune) thrombocytopenic purpura (ITP) in children is generally an acute, benign, and self-limited autoimmune disease. It is characterized by the sudden onset of an isolated thrombocytopenia in association with petechiae and bruises in an otherwise well child. Affected children may experience epistaxis, gum bleeding, hematuria or gastrointestinal hemorrhage. Although serious or life-threatening bleeding is infrequent, less than 1% of children with ITP may suffer from intracranial hemorrhage. There is often antecedent history of a viral illness or immunization. In approximately 80% to 90% of children, the thrombocytopenia is resolved within 6 months after the onset of disease regardless of treatment with complete remission in ~60% of children within the first month. However, approximately 10-20% of children will have a chronic form of ITP defined as persistent thrombocytopenia for longer than 6 months (some defined it as 12 months). In these children with chronic ITP, the probability of spontaneous remission is much less, however, they may attain complete remission as long as 10 years after the diagnosis. The peak age for acute ITP is 2-4 years and males and females are equally affected. In contrast, the chronic form of ITP is usually seen in children older than 10 years of age and is quite similar to adult-onset ITP in that the onset of thrombocytopenia may be insidious and more females than males are affected. Physical examination is entirely normal other than mucocutaneous bleeding manifestations, most commonly diffuse petechiae and ecchymoses. If affected individuals have hepatosplenomegaly, lymphadenopathy, or skeletal anomaly, other acquired or inherited disorders that could cause thrombocytopenia must be ruled out.

As etiology of ITP in children, autoantibodies are considered to be responsible for immune destruction of platelets. For example, antiviral antibodies following viral infection may be cross-reactive with platelet antigens. The specificity of autoantigens identified includes GP IIb/IIIa, Ib/IX, V, Ia/IIa, IV, etc. Limited studies in adult patients with chronic ITP showed that there was no clear immune response linkage to HLA loci. Other investigators suggest that the activated platelet itself may play a role in autoantibody production and immune dysregulation associated with ITP. Diagnosis of ITP is established by excluding other causes of thrombocytopenia, some of which include viral infections, such as HIV, parvovirus, Epstein-Barr virus, hepatitis C virus, immunodeficiency disorders, drug ingestion, HUS, TTP, SLE, congenital/hereditary form of thrombocytopenia, etc. For confirmation of diagnosis, bone marrow examination is not always necessary. However, it is appropriate to perform bone marrow aspiration prior to corticosteroid therapy or when ITP persists for longer than 6 months. Treatment of childhood ITP depends on largely the platelet count at diagnosis. However, other factors must be considered, e.g., the age and activity of the child or type and severity of bleeding. Although the platelet count per se or mucocutaneous bleeding manifestations are not predictors of the risk of life-threatening bleeding, it is appropriate to treat children with extensive mucosal bleeding, i.e., wet purpura, or retinal petechiae. The goal of treatment should be aimed at preventing serious or life-threatening hemorrhage rather than achieving normal platelet counts. However, therapies have not been shown to alter the duration of the disease course. There are four options as initial treatment: 1) no treatment, 2) oral steroids, 3) intravenous anti-D, or 4) intravenous immune globulin (IVIg).

The majority of children with chronic ITP have a benign clinical course. However, a subset of children with chronic ITP experience varying degrees of bleeding and are refractory to standard therapies and thus require other measures. Although splenectomy results in complete remission in about 75% of children with ITP, the risk of overwhelming postsplenectomy infection is a serious consideration. To avoid splenectomy or in the case of failure of response to splenectomy, immunosuppressive or other experimental therapies have been tried or are under investigation. However, their efficacy has not been fully evaluated in children and risks may be greater than benefits. Pros and cons of various treatment regimens including new treatments and management of life-threatening bleeding, especially in children with ITP, which is refractory to conventional treatment, will be discussed.

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