

UPDATE IN IMMUNE THROMBOCYTOPENIC PURPURA.

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Abstract: Primary immune thrombocytopenia (ITP) is an acquired immune-mediated disorder characterized by isolated thrombocytopenia, defined as a peripheral blood platelet count less than $100 \times 10^9/L$, and the absence of any initiating or underlying cause of the thrombocytopenia.

Once regarded as idiopathic, immune thrombocytopenia (ITP) is now known to have a complex pathogenesis: presence of antibodies against multiple platelet antigens leading to reduced platelet survival as well as impaired platelet production.

The incidence in adults is approximately equal for the sexes, except for the middle age where female gender is more prevalent. Categories of ITP have also been established in order to facilitate management decisions, as follows: newly diagnosed (under 3 months of evolution), persistent (3-12 months' duration), chronic (≥ 12 months' duration) and severe ITP.

Whereas ITP in adults typically has an insidious onset followed by a chronic course, ITP in children is usually transitional, with at least 2/3 recovering spontaneously within 6 months. Signs and symptoms vary widely: some have no symptoms or minimal bruising, whereas others experience serious bleeding. The severity of thrombocytopenia correlates to some extent but not entirely with the bleeding risk, which is also influenced by additional individual factors as: age, lifestyle factors, comorbidities, etc.

Multiple therapies with different mechanisms of action are available to treat ITP, and the treatment should be individualized according to clinical course, ITP onset, hemorrhagic risk and/or individual features (age, pregnancy, diabetes, etc).

While the first line therapy is based on corticosteroids and immunoglobulins, the guidelines for managing ITP in adults has changed with the advent of new agents (thrombopoietin receptor agonists and rituximab) as options for second-line therapy. Splenectomy continues to provide the highest cure rate (60%-70% at 5+ years) with no significant complications. Each approach has advantages and disadvantages, therefore treatment needs to be individualized.