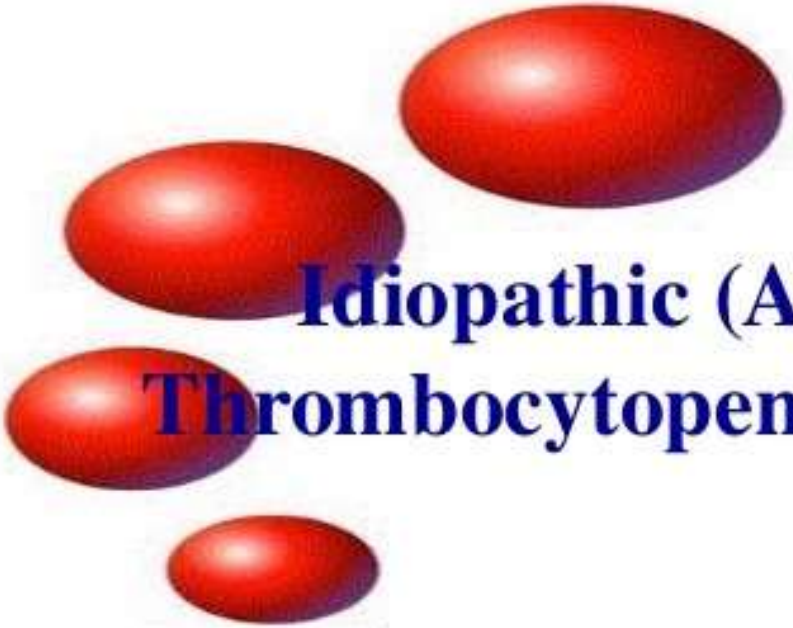
بسم الله الرحمن الرحيم

Presentation by

DR.AKRAMIPOUR

Ped.hematologist&oncologist



Idiopathic (Autoimmune) Thrombocytopenic Purpura(ITP)

Idiopathic (Autoimmune) Thrombocytopenic Purpura(ITP)

- The most common cause of acute onset of thrombocytopenia in an otherwise well child
- Estimated about 1 in 20,000 children
- A recent history of viral illness is described in 50-65% of cases of childhood ITP

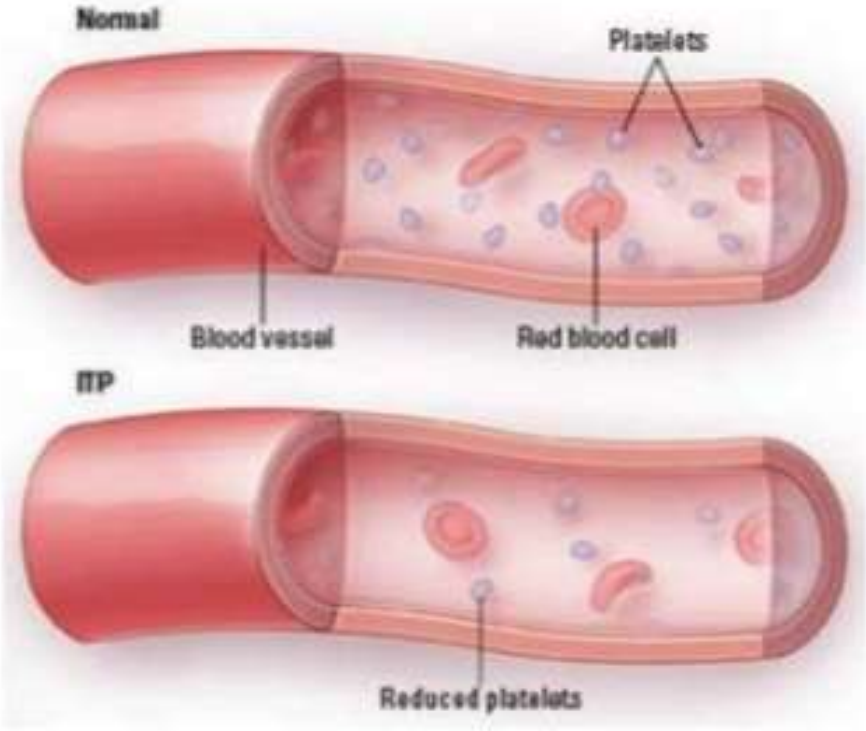
ITP (cont.)

- One - 4 wk after exposure to a common viral infection
- The peak age is 1-4 yr.
- ITP seems to occur more often in late winter and spring after the peak season of viral respiratory illness.

ITP (Pathophysiology)

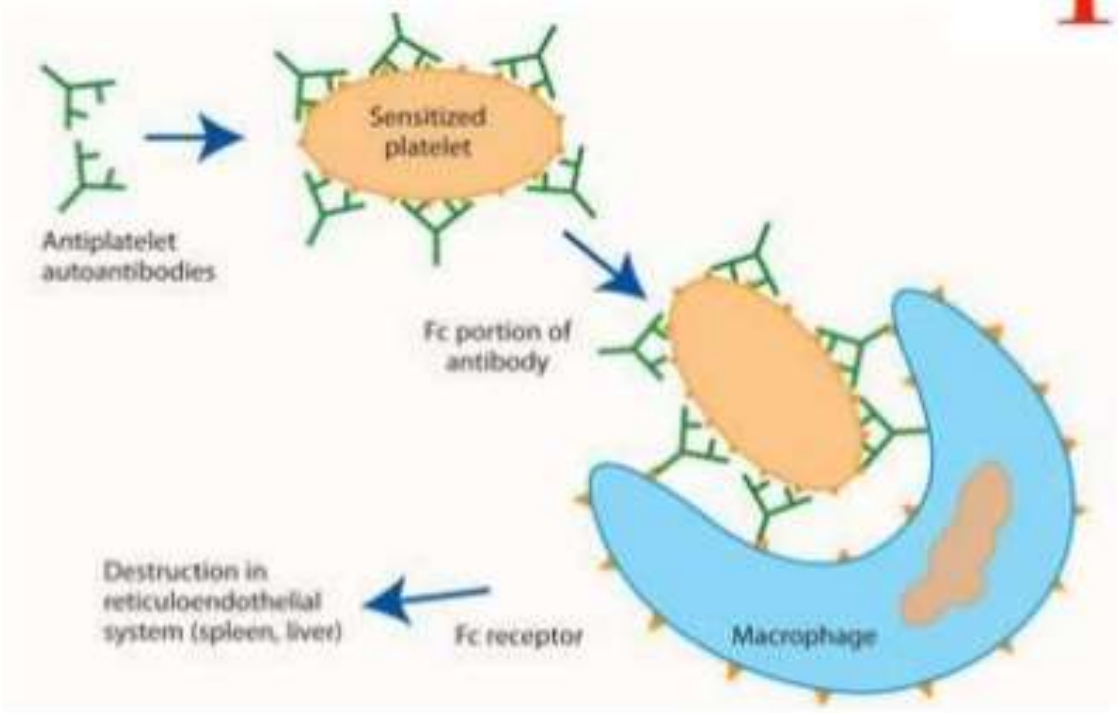
- An autoantibody directed against the platelet surface develops with resultant sudden onset of thrombocytopenia
- After binding of the antibody to the platelet surface, circulating antibody-coated platelets are **recognized** by the **Fc receptor on splenic macrophages**, **ingested**, and **destroyed**

ITP (Pathophysiology)(cont.)



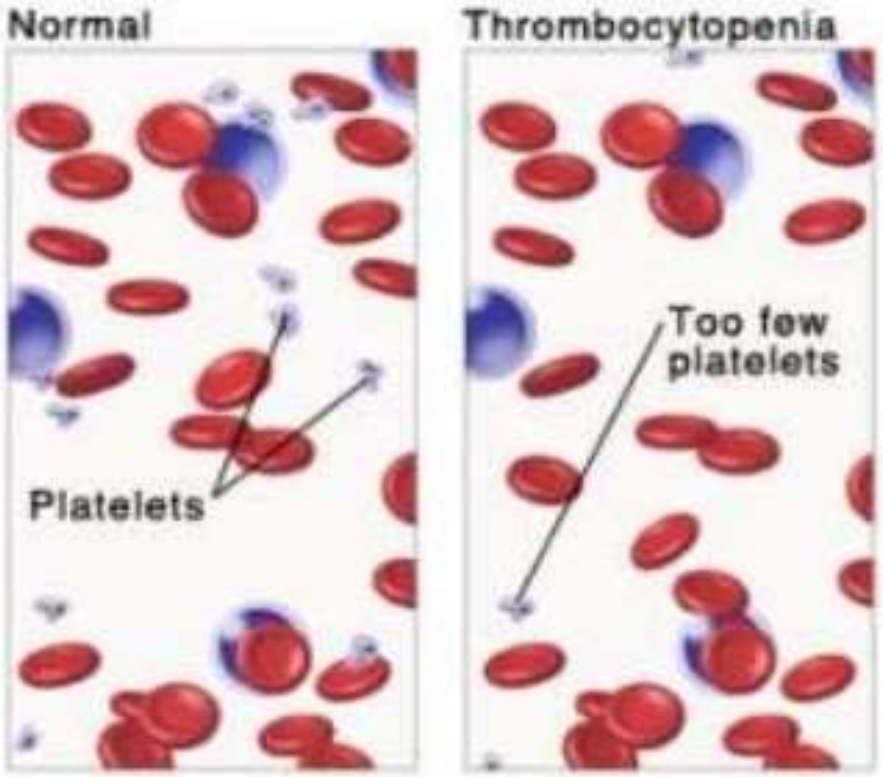
weishendopublications.com

ITP (Pathophysiology)(cont.)



clinicalstudiestoday.blogspot.com

ITP (Pathophysiology)(cont.)



skperdon.hubpages.com

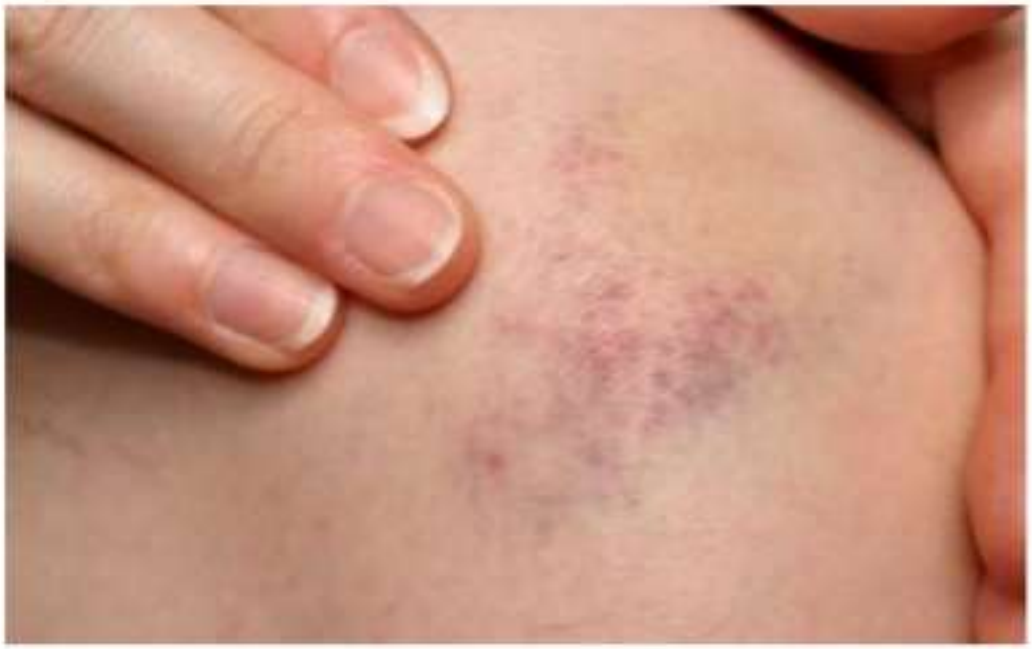
ITP (Pathophysiology)(cont.)

- Most common viruses have been described in association with ITP, including Epstein-Barr virus
- In some patients ITP appears to arise in children infected with Helicobacter pylori or rarely following the measles, mumps, rubella vaccine

Clinical Manifestations (Cont.)

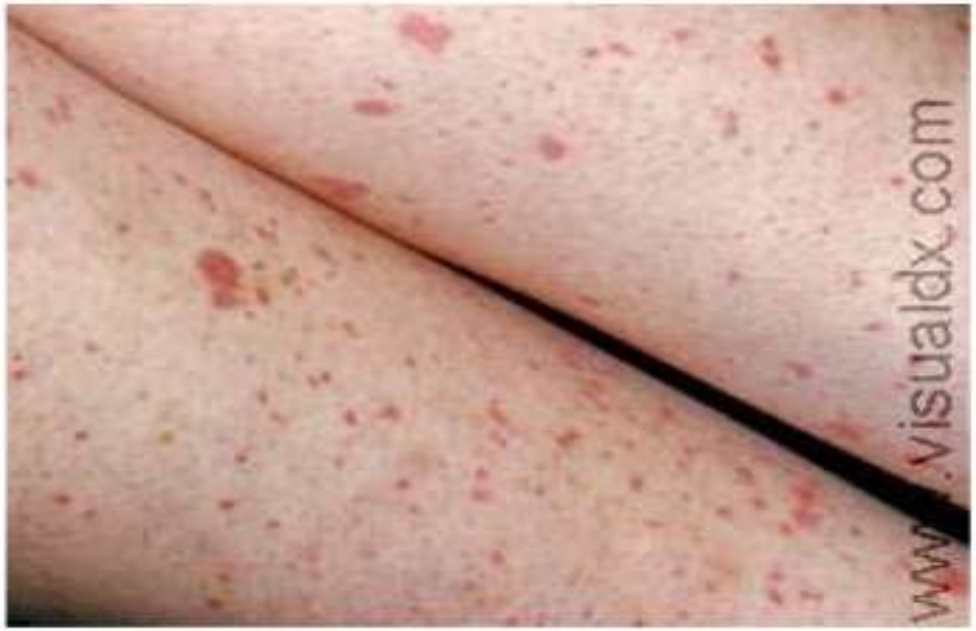
- The classic presentation of ITP is a previously healthy 1-4 yr old child who has sudden onset of generalized petechiae and purpura
- Often there is bleeding from the gums and mucous membranes, particularly with profound thrombocytopenia (platelet count <10 × 109/L).

Clinical Manifestations(cont.)



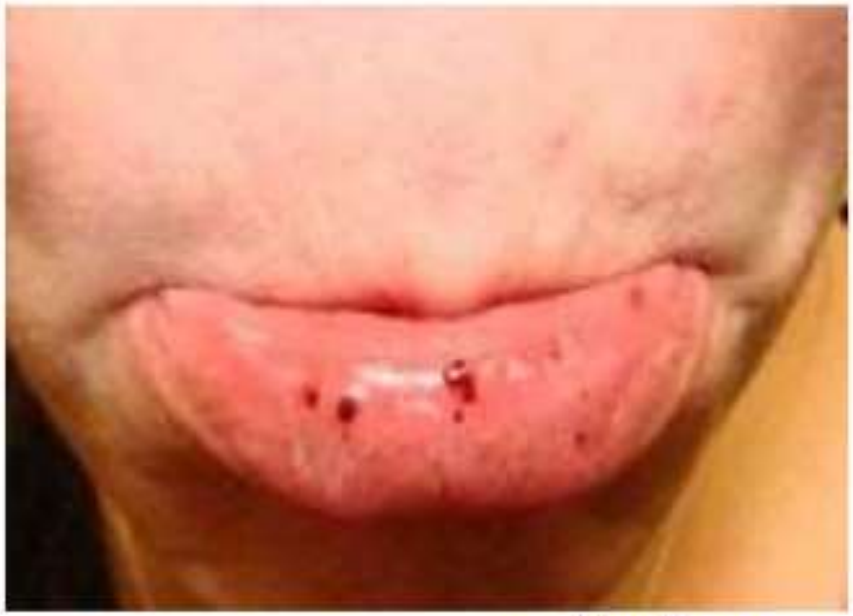
www.itriagehealth.com

Clinical Manifestations (cont.)



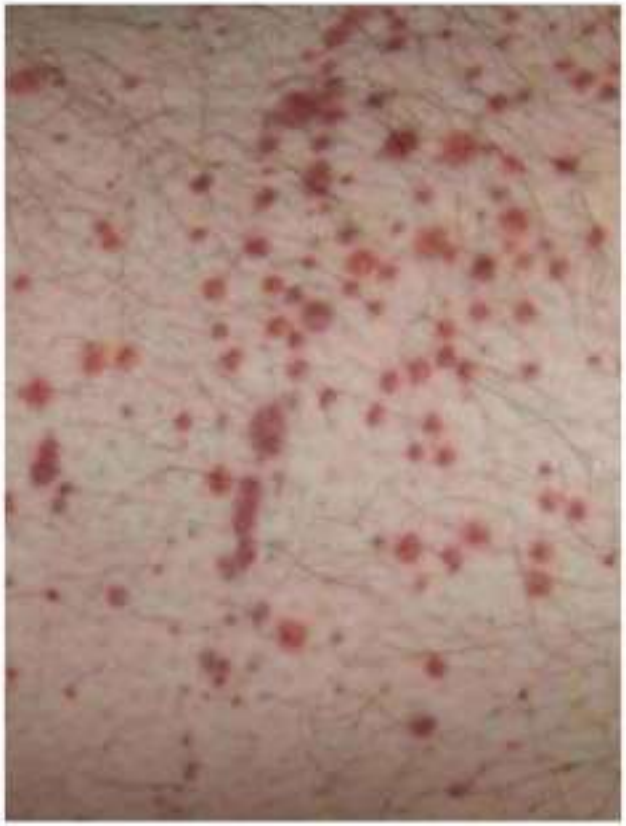
www.lookfordiagnosis.com

Clinical Manifestations (cont.)



en.wikipedia.org

Clinical Manifestations (cont.)



clinicalstudies.com.au

Clinical Manifestations (Cont.)

- There is a history of a preceding viral infection 1-4 wk before the onset of thrombocytopenia
- Findings on physical examination are normal, other than the finding of petechiae and purpura

Classification system

Depending on the basis of symptoms and signs, but not platelet count; ITP is classified as:

Class 1: No symptomes

Class 2. Mild symptoms:

–Bruising and petechiae

–Occasional minor epistaxis

–Very little interference with daily living

Classification system(cont.)

Class 3. Moderate:

- More severe skin and mucosal lesions
- More troublesome epistaxis and menorrhagia

Classification system(cont.)

Class 4. Severe:

– Bleeding episodes—*menorrhagia*, *epistaxis*, *melena*—requiring transfusion or hospitalization

- Symptoms interfering seriously with the quality of life

Clinical Manifestations (Cont.)

- The presence of abnormal findings such as hepatosplenomegaly, bone or joint pain, or remarkable lymphadenopathy suggests other diagnoses

Prognosis

- Severe bleeding is rare (<3% of cases)
- In 70-80% of children who present with acute ITP, spontaneous resolution occurs within 6 mo
- Fewer than 1% of patients develop an intracranial hemorrhage.
- Approximately 20% of children who present with acute ITP go on to have chronic ITP

Prognosis(cont.)

- The outcome/prognosis may be related more to age, as:
  - ITP in younger children is more likely to resolve
  - The development of chronic ITP in adolescents approaches 50%.

Laboratory Findings

- Severe thrombocytopenia (platelet count <20 × 109/L) is common, and platelet size is normal or increased, reflective of increased platelet turnover
- In acute ITP, the hemoglobin value, white blood cell (WBC) count, and differential count should be normal.

Laboratory Findings(cont.)

- Bone marrow examination shows normal granulocytic and erythrocytic series, with characteristically normal or increased numbers of megakaryocytes

Laboratory Findings(cont.)

- Indications for bone marrow aspiration/biopsy include:
  1. An abnormal WBC count or differential
  2. Unexplained anemia
  3. Findings on history and physical examination suggestive of a bone marrow failure syndrome or malignancy.
- Other laboratory tests should be performed as indicated by the history and physical examination

Laboratory Findings(cont.)

- A direct antiglobulin test (Coombs) should be done
  1. If there is unexplained anemia to rule out Evans syndrome (autoimmune hemolytic anemia and thrombocytopenia)
  2. Before instituting therapy with IV anti-D.

Diagnosis/
Differential Diagnosis

- Autoimmune thrombocytopenia may be an initial manifestation of :
    1. SLE
    2. HIV infection
    3. Common variable immunodeficiency
    4. Lymphoma(rarely)

Treatment

- Platelet transfusion in ITP is usually contraindicated unless life-threatening bleeding is present (Antiplatelet antibodies bind to transfused platelets as well as they do to autologous platelets)

Treatment (cont.)

- Initial approaches to the management of ITP include the following:

1. No therapy other than education and counseling of the family and patient for patients with minimal, mild, and moderate symptoms, as defined earlier.

- This approach is:
  - Far less costly
  - Side effects are minimal

Treatment(cont.)

2.Intravenous immunoglobulin (IVIG).

- IVIG at a dose of 0.8- 1.0 g/kg/day for 1-2 days induces a rapid rise in platelet count (usually >20 × 109/L) in 95% of patients within 48 hr.
- IVIG appears to induce a response by downregulating Fc-mediated phagocytosis of antibody-coated platelets.

Treatment(cont.)

2.Intravenous immunoglobulin (IVIG).(cont.)

IVIG therapy is :

- Expensive
- Time-consuming to administer
- After infusion, there is a high frequency of headaches and vomiting, suggestive of IVIG-induced aseptic meningitis.

Treatment(cont.)

3.Intravenous anti-D therapy.

For Rh positive patients:

IV anti-D at a dose of 50-75 μg/kg causes a rise in platelet count to >20 × 10^9/L in 80-90% of patients within48-72 hr.

Treatment(cont.)

4. Prednisone.

- Doses of prednisone of 1-4 mg/kg/24 hr
- Corticosteroid therapy is usually continued for 2-3 wk or until a rise in platelet count to >20 × 10_9/L has been achieved, with a rapid taper
- long-term side effects of corticosteroid therapy:
    1. Growth failure
    2. Diabetes mellitus
    3. Osteoporosis

Treatment(Cont.)

Intracranial hemorrhage

Multiple modalities should be used, including:

1. Platelet transfusion
2. IVIG
3. High-dose corticosteroids
4. Prompt consultation by neurosurgery and surgery.

Treatment(Cont.)

- The role of splenectomy in ITP should be reserved for 1 of 2 circumstances.
  1. The older child (≥4 yr) with severe ITP that has lasted >1 yr (chronic ITP)
  2. Whose symptoms are not easily controlled with therapy
  3. Life-threatening hemorrhage (intracranial hemorrhage) complicates acute ITP
  4. Platelet count cannot be corrected rapidly with transfusion of platelets and administration of IVIG and corticosteroids

Treatment(Cont.)

- The role of splenectomy in ITP should be reserved for 1 of 2 circumstances.
  1. The older child (≥4 yr) with severe ITP that has lasted >1 yr (chronic ITP)
  2. Whose symptoms are not easily controlled with therapy
  3. Life-threatening hemorrhage (intracranial hemorrhage) complicates acute ITP
  4. Platelet count cannot be corrected rapidly with transfusion of platelets and administration of IVIG and corticosteroids

References

- Nachman RL, Rafii S: Platelets, petechiae, and preservation of the vascular wall, N Engl J Med 359:1261–1270, 2008.
- Donato H, Picón A, Martinez M, et al. Demographic data, natural history, and prognostic factors of idiopathic thrombocytopenic purpura in children: a multicentered study from Argentina. Pediatr Blood Cancer 2009; 52:491.
- Newman PK, Newman DK: Platelets and the vessel wall. In Orkin SH, Nathan DG, Ginsberg D, et al, editors: *Nathan and Oski's hematology of infancy and childhood*, ed 7, Philadelphia, 2009, Saunders Elsevier, pp.1399–1379
- www.uk-itp.org
- clinicalstudies.com.au

www.my-thank-you-site.com