# Pediatric Immune Thrombocytopenia: ITP? IDK!

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# Objectives

- At the conclusion of this session, participants should be able to:
  - Define immune thrombocytopenia (ITP) based upon the patient's complete blood count
  - Identify pertinent past medical history details that may suggest ITP
  - Identify pertinent physical exam findings in a child with ITP

# Immune Thrombocytopenia

“Idiopathic thrombocytopenia”

“Idiopathic thrombocytopenic purpura”

“Autoimmune thrombocytopenic purpura”

# Blood Clotting

**Primary Hemostasis** (Platelet Plug)

- Initiated by platelets
- Thrombocytopenia or platelet dysfunction →
  - Bleeding into skin and mucous membranes → **petechiae**, ecchymosis, purpura

**Secondary Hemostasis** (Fibrin Clot)

- Initiated by coagulation factors
- Coagulation factor deficiency →
  - Bleeding into soft tissue, muscle, **joints**

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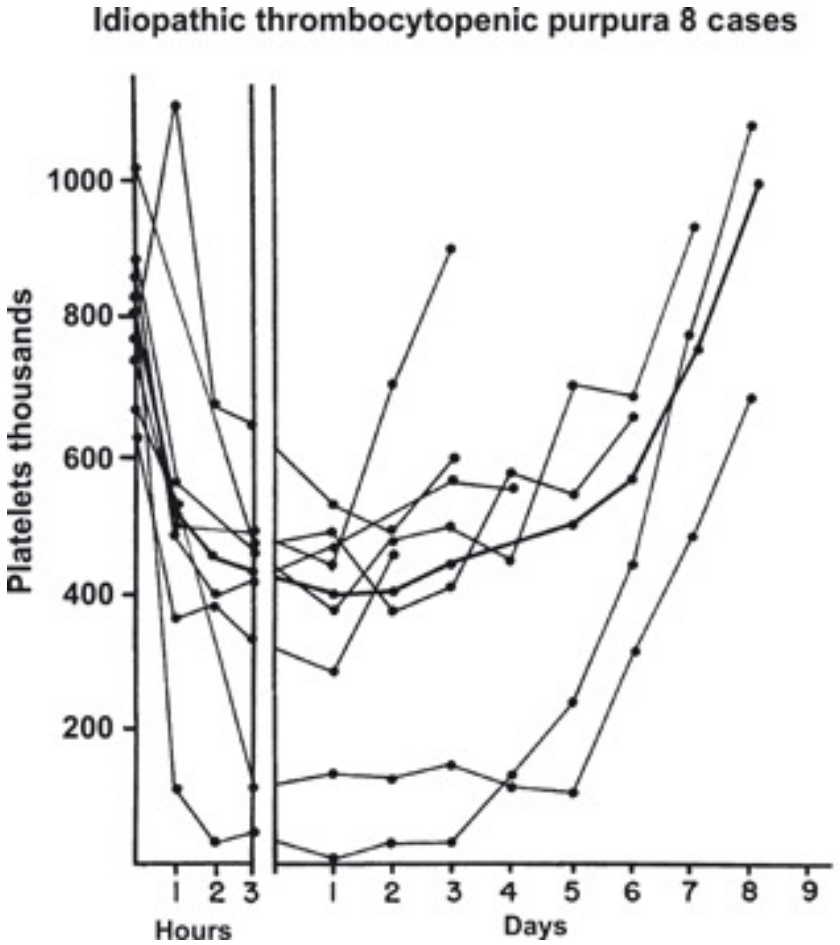
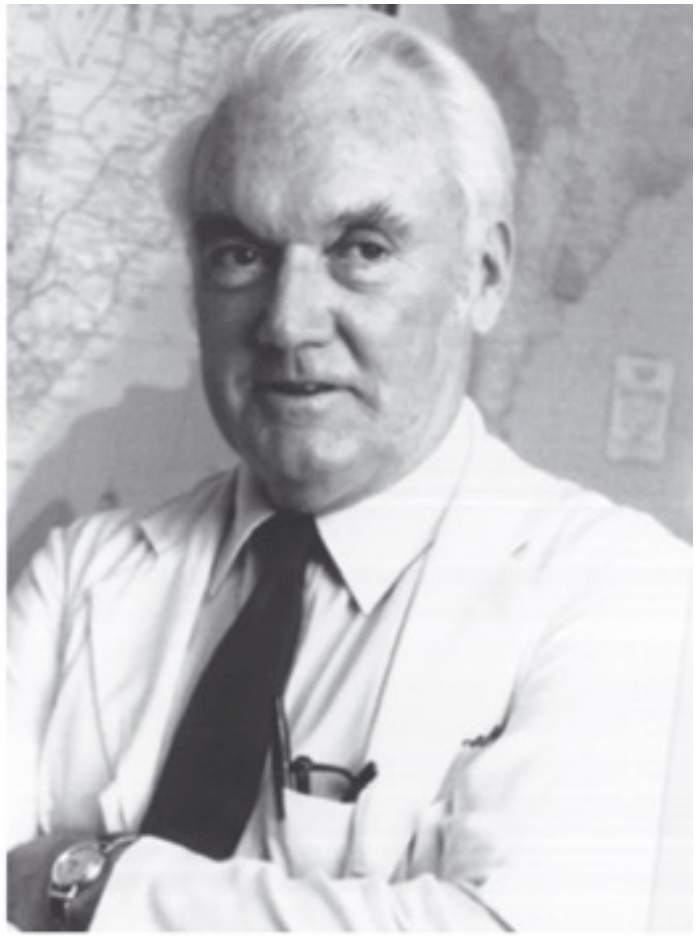


Br J Haematol, Volume: 153, Issue: 4, Pages: 437-450, First published: 05 April 2011, DOI: (10.1111/j.1365-2141.2010.08562.x)

# Immune Thrombocytopenia (ITP)

**Pathophysiology**

- Antiplatelet IgG autoantibody → increased platelet destruction
- At least 50% of cases follow a viral infection

***<u>Frequently follows 4-6 weeks after a viral infection</u> or live virus vaccine (especially MMR)

# Immune Thrombocytopenia (ITP)

- Most common between ages 2-5 y/o
  - Can also occur in adults (usually more chronic form)

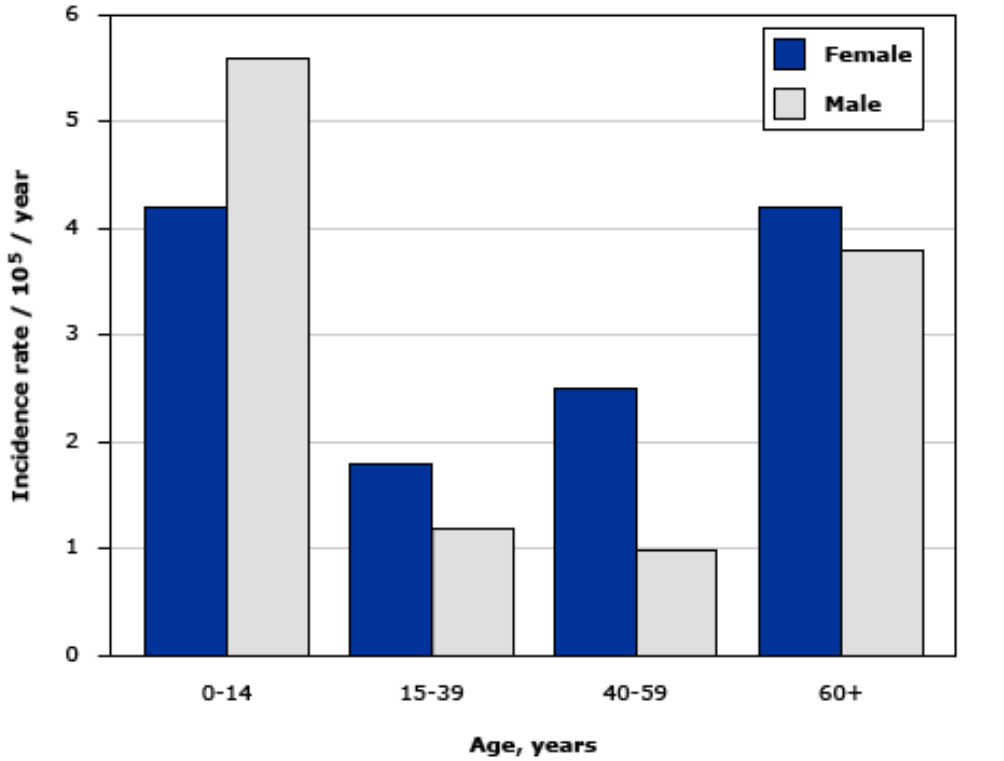
- Most common cause of **isolated thrombocytopenia** in children



*Current* Ch. 14-04: Increased Platelet Destruction

Approximately 5 in 100,000 children annually

**Incidence of immune thrombocytopenia (ITP) by age and sex**




Data are presented for the incidence of ITP in females and males in different age groups. The incidence of ITP is highest in children, and may be greater in boys than in girls. In adults, the incidence increases with age. The incidence in women is greater than in men at younger ages, but in adults over age 60, the incidence of men and women is the same.

*Data from:*

*1. Data for children from: Zeller B, Rajantie J, Hedlund-Treutiger I, et al. Childhood idiopathic thrombocytopenic purpura in the Nordic countries: epidemiology and predictors of chronic disease. Acta Paediatrica 2005; 94:178.*

*2. Data for adults from: Frederiksen H, Schmidt K. The incidence of idiopathic thrombocytopenic purpura in adults increases with age. Blood 1999; 94:909.*

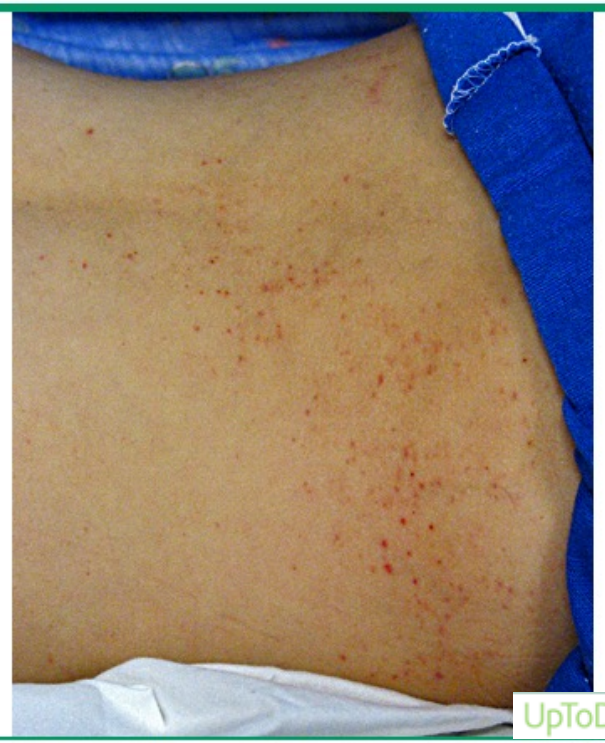
# Bleeding History

- Where?
  - Don’t forget occult bleeding in stool or microscopic hematuria

# Bleeding Terminology

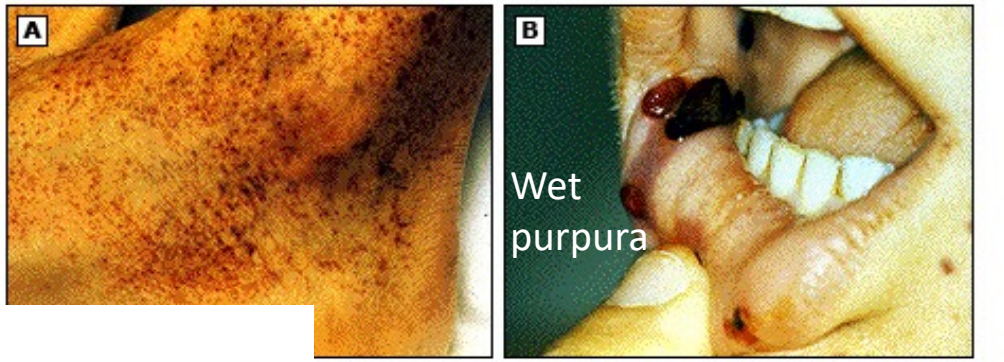
**"Purpura" = any bleeding under the surface of the skin (dry purpura) or mucous membrane (wet purpura)**

**Petechiae** < 2mm

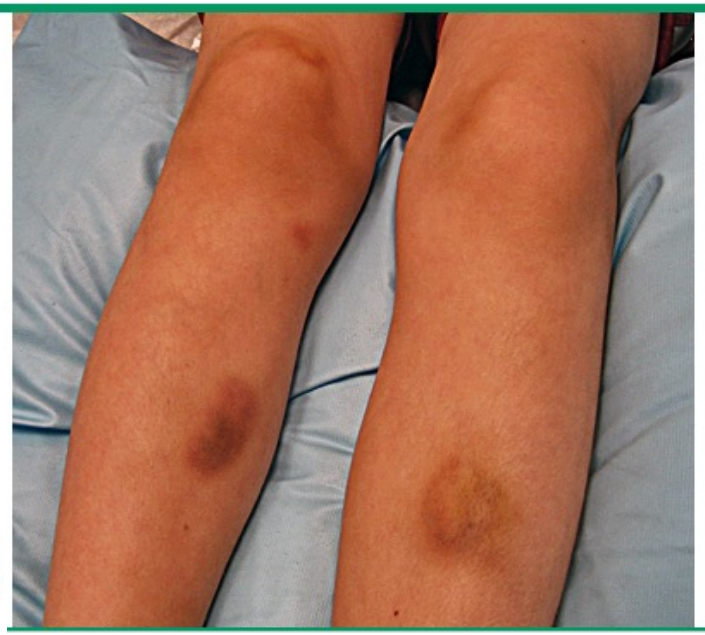


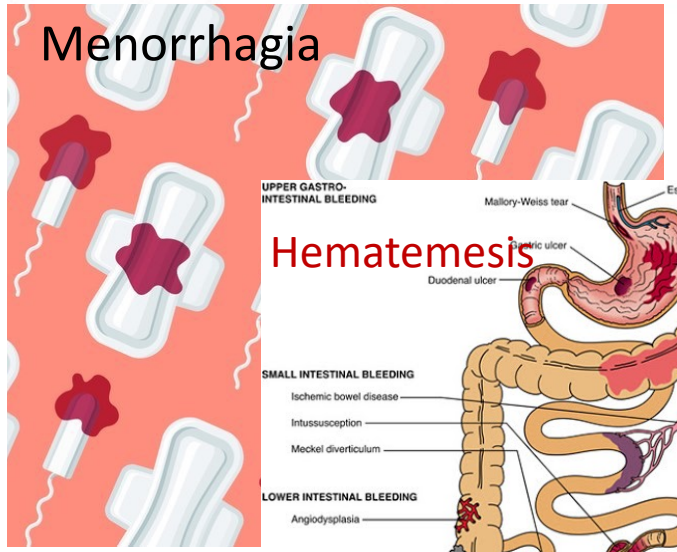


**Petechiae in immune thrombocytopenia (ITP)**

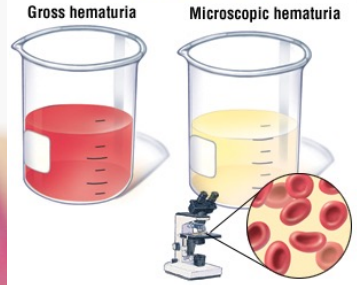



**Ecchymoses** "bruise" > 2mm








Hematuria

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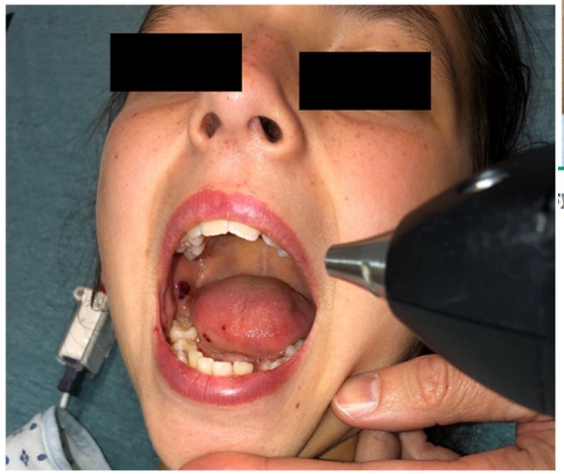
# Bleeding: History

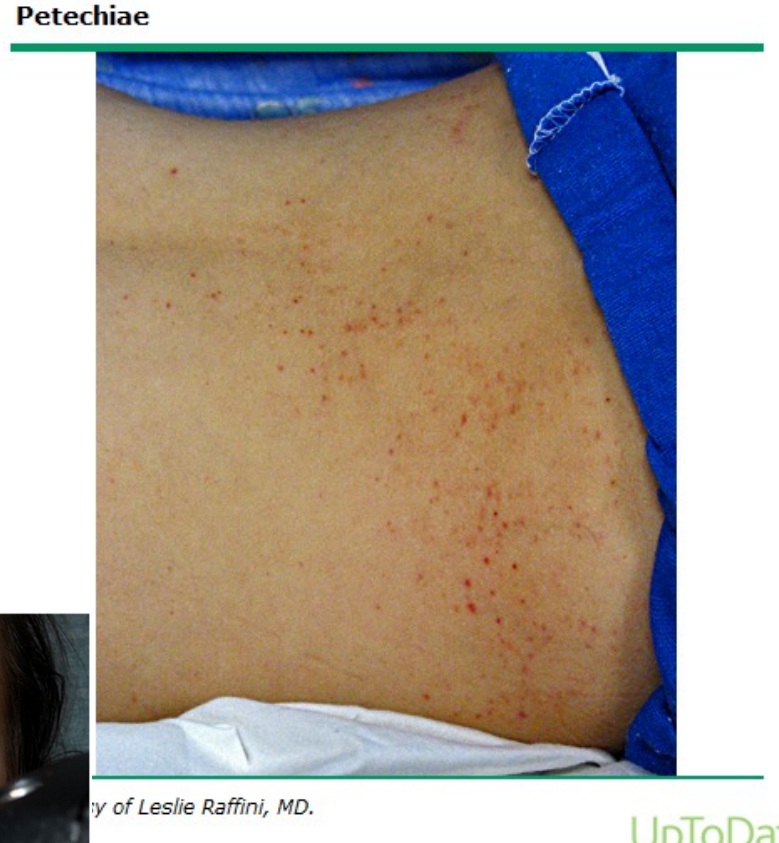
- Where?
  - Don’t forget occult bleeding in stool or microscopic hematuria
- When? Post-operative bleeding (i.e. after circumcision?), spontaneous, traumatic
- How long? Duration of bleeding?
- Who? Any family history of bleeding disorders (hemophilia, von Willebrand disease)?
- PMH
  - *Recent viral infection? Recent vaccinations?*
  - Etoh, liver disease
  - Hematologic disorders, malignancy, autoimmune disorder
- Any new medications?
  - Aspirin
  - NSAIDs
  - Anticoagulants

# Immune Thrombocytopenia (ITP)

- Symptoms: <u>Sudden onset</u> petechial rash and mucocutaneous bleeding
  - LACK of systemic red flag symptoms (fever, weight loss, night sweats, fatigue, bone pain)

- PE: *NORMAL, well-appearing child on exam*, except related to bleeding
  - Particularly small, ***<u>mucocutaneous bleeding—wet purpura</u>***
  - Look for red flags (lymphadenopathy, hepatosplenomegaly)



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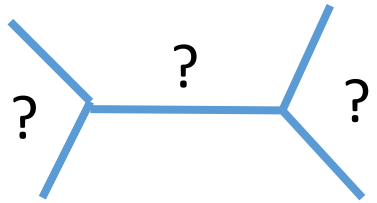
*Current* Ch. 14-04: Increased Platelet Destruction

# Bleeding: General Workup

- CBC with differential & peripheral smear
  - Which cell lines are affected?
  - Thrombocytopenia? HOW low?
  - Hgb → how MUCH are they bleeding?
  - Peripheral smear?
- Coags
  - PT/INR → extrinsic pathway
  - PTT → intrinsic pathway
  - Fibrinogen
  - D-dimer



# If you discover new thrombocytopenia, the first thing you should do is…

- REPEAT the CBC to confirm!

# Thrombocytopenia

- Normal platelet count: 150-450K
  - Mild thrombocytopenia: 70-150K
  - Moderate thrombocytopenia: 20-70K
  - Severe thrombocytopenia: <20K

- Impaired hemostasis with platelets <75K
- Risk of spontaneous intracranial hemorrhage at <20K

- Platelet life span: 8-10 days

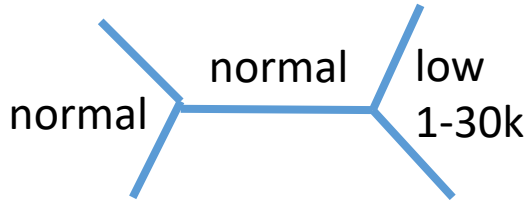


Williams Hematology Ch. 3

# Immune Thrombocytopenia (ITP) Diagnosis

- **Isolated, severe thrombocytopenia with platelet count <30k**
  - CBC otherwise normal unless significant bleeding!
- No role for antiplatelet antibody testing (low yield)
- Bone marrow aspiration/biopsy usually not indicated

- *ITP is a diagnosis of exclusion!*



# Differential Diagnosis of Thrombocytopenia

- Decreased production — Are they making platelets? Is the bone marrow working?
  - Marrow replacement/infiltration (*leukemia*, myelodysplastic syndrome, metastasis)
  - Marrow is empty (aplastic anemia, bone marrow failure syndromes)
  - Marrow injury (chemotherapy, radiation)
  - Marrow suppression (*drug-induced*, *viral suppression* [parvovirus, varicella, HCV, COVID-19])
  - Nutrient deficiency (B12, folate, alcoholism)
- Decreased platelet lifespan — Are their platelets being destroyed?
  - Autoimmune destruction (ITP, SLE, *sepsis*)
  - Increased consumption (DIC, HUS, TTP, HIT)
- Abnormal platelet distribution — Are their platelets getting stuck?
  - Hypersplenism/Splenomegaly/Splenic sequestration



Williams Hematology Ch. 3

# Immune Thrombocytopenia (ITP) Treatment

- Treatment:
  - Consult/refer to hematology!
  - In children: *75-80% resolve spontaneously – watchful waiting may be indicated!*
  - Treatment indicated for significant bleeding
  - Treatment options:
    - Steroids
    - Intravenous immunoglobulin (IVIG)
    - Thrombopoitein receptor antagonists
    - Splenectomy
  - **What about platelet transfusion???**
- Chronic ITP → chronic therapy



UpToDate
*Current* Ch. 14-04: Increased Platelet Destruction

# Immune Thrombocytopenia (ITP) Treatment

| | |
|---|---|
| **Platelet count <20k with no or mild bleeding** | Outpatient management |
| | Observation (rather than steroids, IVIG, rituximab) |
| | Follow up with a hematologist within 24 to 72 hours |
| | |
| **Non-life-threatening mucosal bleeding** | Prednisone 2-4mg/kg/day, maximum 120mg daily, for 5-7 days |
| | |
| **Second-line therapy** | Thrombopoietin receptor antagonists |
| | |



American Society of Hematology 2019 Guidelines

# Supportive Care

- Avoid contact sports (boxing, rugby, football, martial arts)
- Avoid OTC platelet-inhibiting drugs
  - Aspirin
  - NSAIDS
- Menstrual suppression

## Control of benign epistaxis

The child on the right is showing the correct way to stop a nosebleed. The nasal alae should be pressed together closing off the nasal airway. The incorrect way to stop a nosebleed is demonstrated by the child on the left.

*Courtesy of Anna H Messner, MD.*

# References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Thank you!