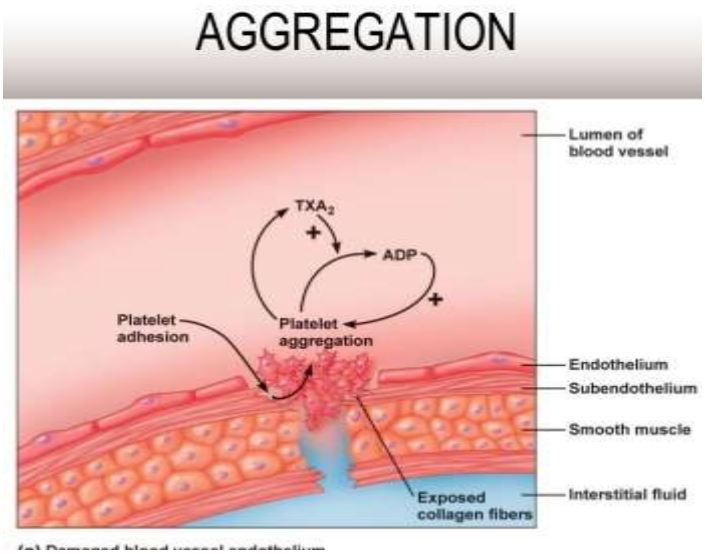
ITB

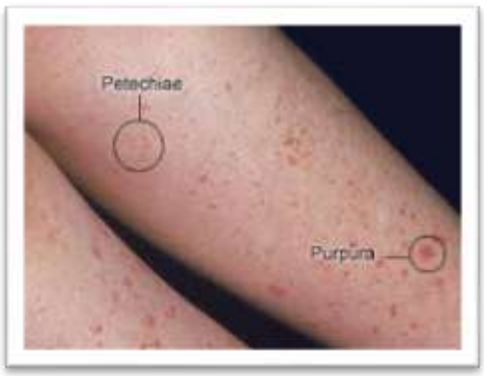
Primary hemostasis consists of:

- Vascular Phase: Vasoconstriction
- Platelets Phase:
  - Platelet plug formation (Depends on number + functioning platelets)
    - *Thrombasthenia (platelets paralysis) = Non-functioning platelets*
  - Von-Willebrand factor (glue factor)



Presentation:

History: Epistaxis



Examination of skin:

- Petechia: Small pin point lesion
- Purpura: Larger than petechia
- Ecchymosis: Larger than purpura

If purpura in skin → Dry purpura

- Next step after finding dry purpura → Check mucous membranes

If purpura in mucous membranes → **Wet purpura** (Very important)

- Significance of wet purpura:
  - More serious
  - Herald a developing CNS bleeding
- Next step after finding wet purpura → Fundoscopy examination because:
  - Retinal bleeding indicates high risk of CNS bleeding

So, Epistaxis + Dry purpura + Wet purpura = **<u>Mucocutaneous bleeding</u>** (Feature of primary homeostatic defect)

Mucocutaneous bleeding confirmed, **what are next steps?**

- Don't forget **fundoscopy** to check the risk of CNS Bleeding
- **Examination** from scalp to toe – All Negative (Nooothing):
    - No pallor!

    - No fever!

    - No Organomegaly! (Neither spleen nor liver)

    - No Lymphadenopathy!

- **Investigation**:
  <u>Complete blood count + Blood film</u>
  (*Nooothing, only …*):
    - Hb= Normal!
    - WBC = Normal!
    - RBC = Normal!

↓

    - Platelets = **Looooow**
        - i.e. = *Isolated thrombocytopenia*

- So, **diagnosis** is: ITP (immune/idiopathic thrombocytopenic purpura)
    - i.e. diagnosis is made by exclusion! If any of the 'nothings' above is present, it is not ITP, e.g.:
        - Epistaxis + fever + toxic ill →acute leukemia
        - Epistaxis + anemia → Aplastic anemia

ITP: is an auto-immune disorder, due to formation of antibodies causing:

- <u>Over destruction</u> of platelets → Rx: Immuno-suppressant
- <u>Or under-production</u> from bone morrow (due to destruction of megakaryocytes (platelets precursor cells)) → Rx: TRA (Thrombopoietin receptor agonist)

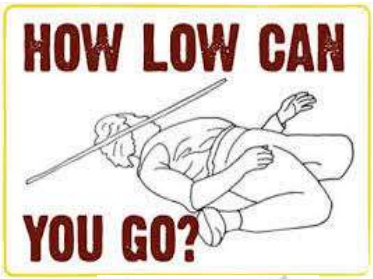
Etiology: Where those antibodies come from?

- Primary ITP (Idiopathic TP)
- Due to other causes that may be diagnosed initially as ITP, so they should be investigated:
    - Lupus (SLE), especially if female, Invx for lupus markers.

    - Virology (HIV, Hepatitis C, ± Hepatitis B)

Management:

- How low can you get with platelets level before starting treatment? (i.e. how many platelets are needed to maintain stable hemostatic function in normal situation?)



    - 20,000, only!

        - *(The last updated number is 7,200, but for now let's stick to the old 20K number)*
- Anyway, you should treat the patient, not the platelets number! *So, if patient is bleeding, treat him, otherwise not needed.*

Options of treatments:

- <u>First-line therapy: Steroid, IVIG and Anti-D:</u>
    - Quick (24–48 hours)
    - Efficacy (70–80%)
    - Transient
    - Palliative

- Regarding **steroids**:
  - Type: prednisone Vs dexamethasone
  - *Prednisone:*
    - *Dose 1-2mg /kg*
    - *Duration: for 3 weeks:*
      - *If improved ☺ → Taper*
      - *If not improved ☹ → Switch*
  - *Dexamethasone:*
    - 40 mg/day
    - 4 days
    - 4 cycles
    - 14 days interval
- <u>TPO receptor agonists (Romiplostim, eltrombopag)</u>
  - Effective
  - Non-immunosuppressant
  - Not steroid based
- <u>Splenectomy</u>
  - The problem with spleen is, it is the place where opsonized platelets are destroyed.

Bone morrow examination is *not a routine* in patient with ITP, it is indicated in:

- Failure to respond to, or relapse following, first-line therapy
- Age > 60 years
- Presence of atypical clinical or laboratory features
- Myelodysplastic syndromes (MDS), usually attack elderly patient, presented with isolating thrombocytopenia, mimicking ITP.
- Failure of splenectomy

Thank you ,,,