



Platelet and White Blood Cell Disorders.

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Platelet Disorders

Platelet function

Wound healing requirements

Platelet count methods.

Automated methods shortcomings

Platelet count regulation.

A microscopic image of platelets, showing a large, dark, irregularly shaped mass on the left side of the frame. The mass has a rough, textured surface with some lighter, granular areas. To the right of this mass, there are several smaller, dark, circular spots scattered across a lighter, textured background. The overall appearance is that of a biological specimen, likely platelets, under a microscope.

Platelet Structure

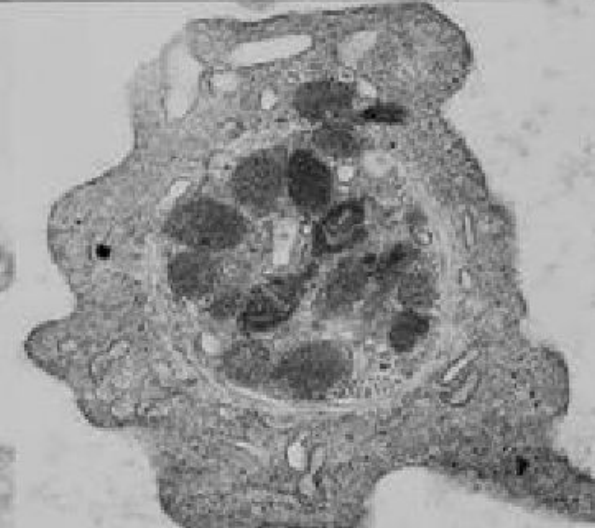
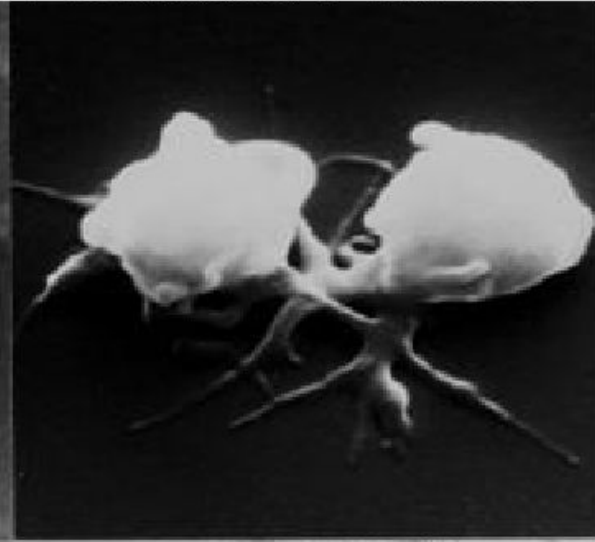
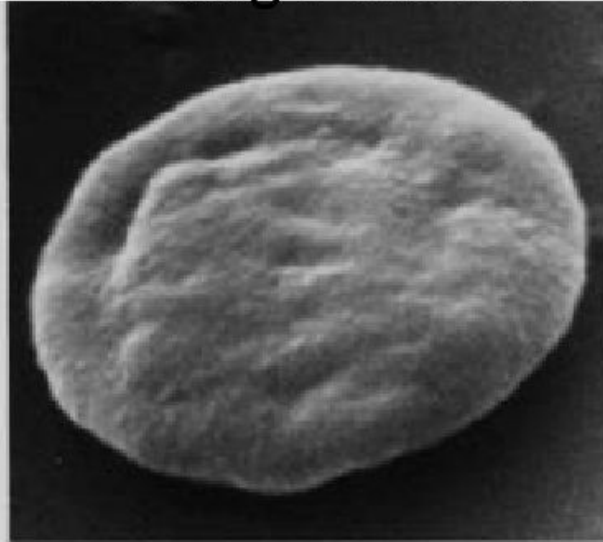
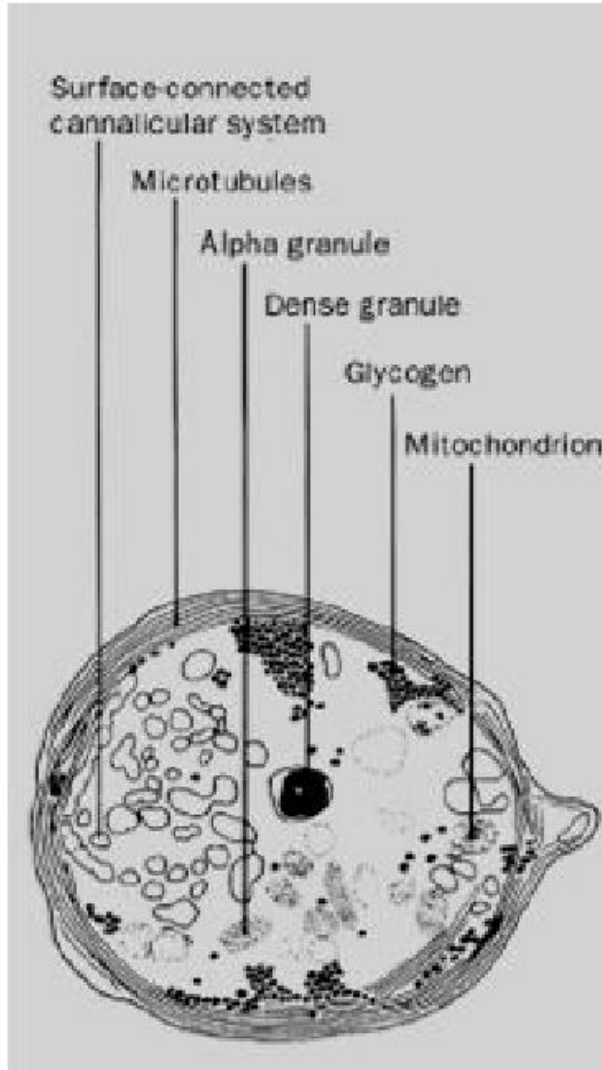
- Small disks.
- No nuclei as they don't need them.
- Mitochondria, lysosomes, peroxisomes, alpha granules (contain VWF, fibrinogen, PF4, PDGF) and dense granules (contain ADP, ATP, serotonin, calcium).

Platelet utilization

- Platelet adhesion: attaching to subendothelial vWF/ collagen via platelet GP1b-IX-V and GPIIb/IIIa.
- Activation: shape changes to expose GPIIb/IIIa receptors.
- Aggregation: cross linking of platelet activated GPIIb/IIIa by fibrinogen/ vWF
- Propagation of coagulation: coagulation factor complexes/ enzymes attach to the to the activated platelet surface.

Resting Platelet

Activated Platelet



A microscopic image of platelets, showing several dark, irregularly shaped cells with granular internal structure. The cells are clustered together, with some appearing to be in the process of aggregation. The background is light and shows some smaller, less distinct particles.

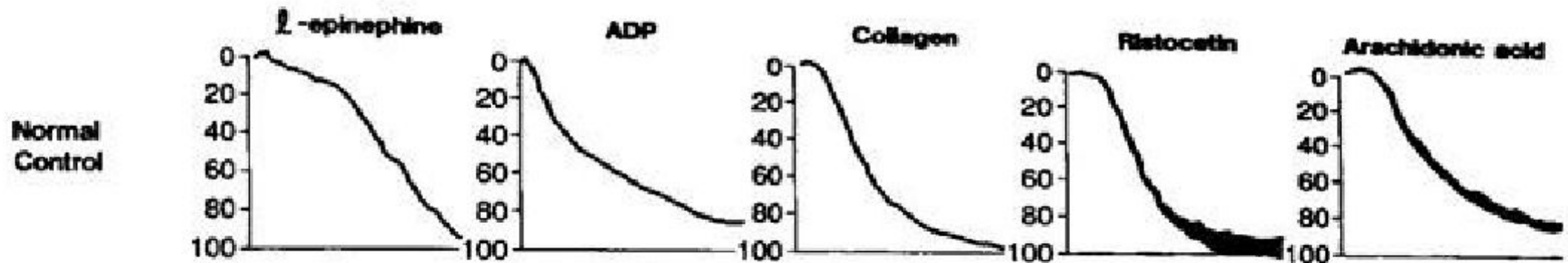
Platelet function testing

- Bleeding time: not used anymore
- PFA-100
- Aggregation studies.

PFA-100 Results

Diagnosis	Col/Epi result	Col/ADP result
Normal Patient	Normal (<187 sec)	Not performed
Aspirin effect	Prolonged	Normal (<114 sec)
Possible platelet defect or Von Willebrand Disease	Prolonged	Prolonged

Platelet Aggregation: Normal Tracing



Platelet related bleeding



Tends to happen immediately after the trauma.



Petechiae are more indicative of platelet problems than are bigger bleeds



Family history can be very helpful as many of the defects may be genetic.

Quantitative platelet disorders

Pseudothrombocytopenia.

Primary bone marrow problem.

Loss of external drive/ inhibition.

Pooling.

Increased utilization.

Increased destruction/ shortened life span.



Shortened
platelet life span

Non-immune:

- DIC.
- TTP.
- HUS.
- Kasabach-Merritt syndrome.

Shortened platelet life span

Immune:

ITP Mechanism, presentation, natural history, treatment and outcome.

Heparin- induced thrombocytopenia/
drug induced

Immune thrombocytopenia in neonates:
Autoimmune vs alloimmune.





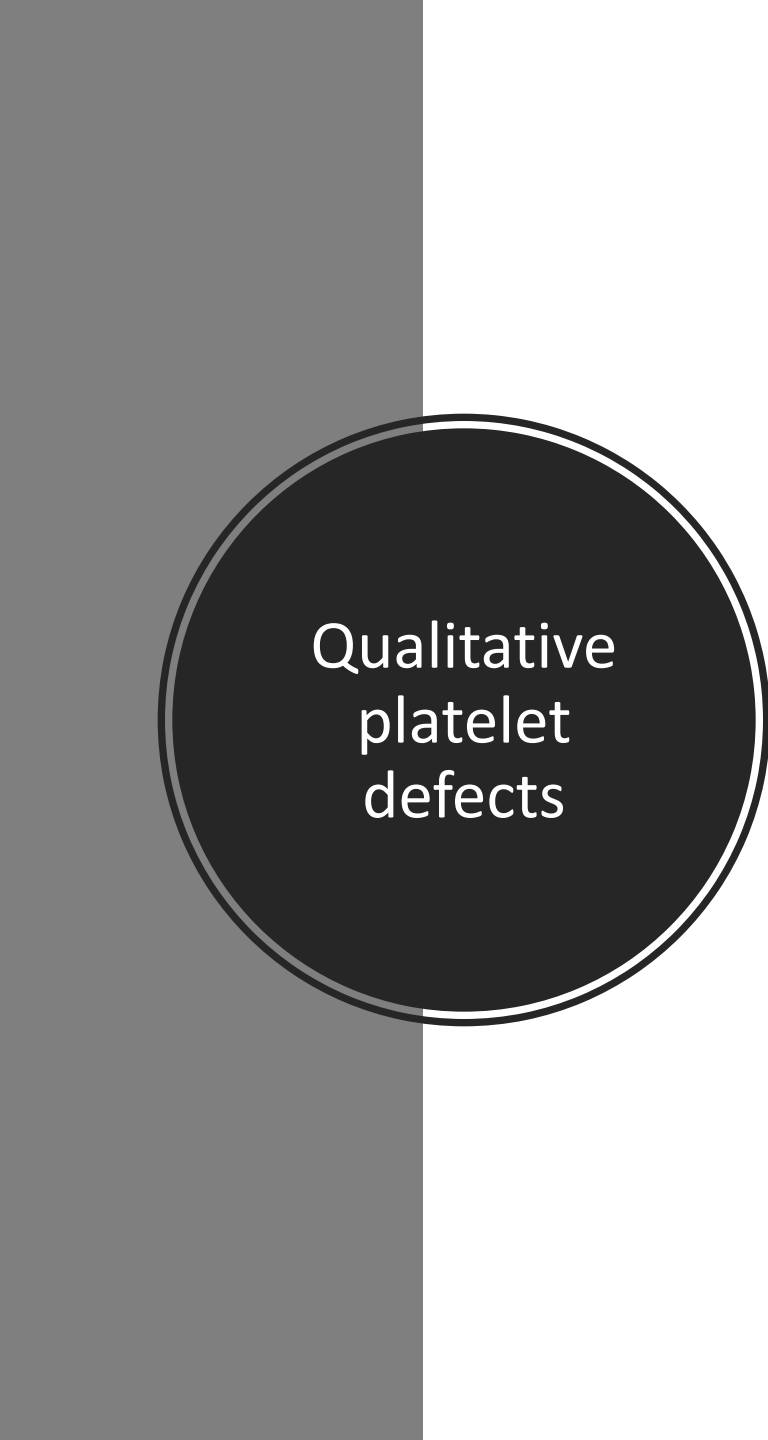
Congenital causes.

Wiskott-Aldrich syndrome.

X-linked thrombocytopenia.

Thrombocytopenia
Absent Radii (TAR)
syndrome.

Congenital
Amegakaryocytic
thrombocytopenia.



Qualitative
platelet
defects

Glanzmann's Thrombasthenia: platelet membrane fibrinogen receptor (GPIIb/IIIa).

Bernard-Soulier syndrome: absent or abnormal platelet surface receptor for vWF (GP Ib/IX).

Gray platelet syndrome.

Hermansky-Pudlak syndrome: pulmonary fibrosis, OC albinism and absent dense granules on EM



**Disorders of
white blood
cells**

Neutropenia secondary to infection.

Drug induced neutropenia.

Autoimmune neutropenia .

Chronic benign neutropenia of childhood.

Alloimmune neutropenia.

Chronic granulomatous disease



**Disorders of
white blood
cells**

Kostmann's syndrome.

Cyclic neutropenia.

Schwachman-Diamond syndrome.

Chronic idiopathic neutropenia.

Leukocyte adhesion deficiency.

Chediak-Higashi syndrome