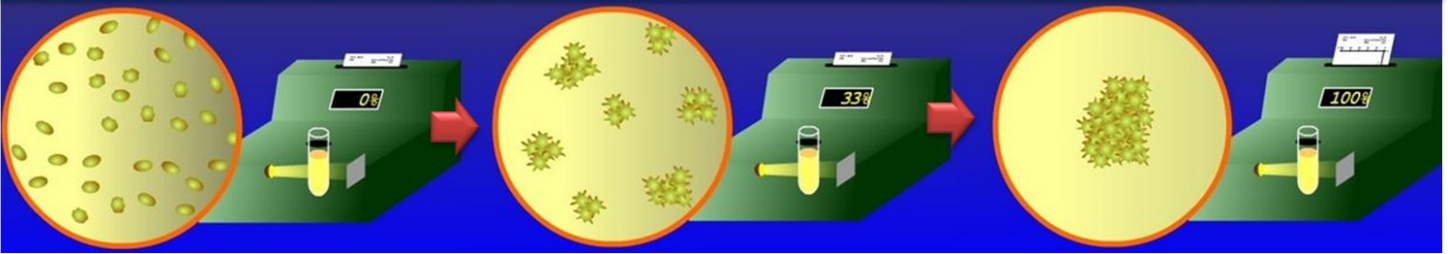


Light Transmission Aggregometry

A light beam passing through platelet rich plasma is measured. As platelets aggregate, more light passes



Dx	GPVI deficiency	GPIIb/IIIa deficiency	Fibrinogen deficiency	Dense Granule Def. or Secretion defect ⁴	Thromboxane synth def or Aspirin	ADP receptor mutation or ADP receptor blocker (e.g. plavix)
PT/PTT	Normal	Normal	Prolonged	Normal	Normal	Normal
PLT	Normal	Normal	Normal	Normal	Normal	Normal
Bleeding Time	Prolonged	Prolonged	Prolonged	Prolonged	Prolonged	Prolonged
Ristocetin Co Activity	Normal	Normal ²	Normal	Normal	Normal	Normal
Aggregation-Collagen	Absent	Absent	Absent	Decreased	Decreased	Normal
Aggregation-ADP	Normal	Absent	Absent	1° wave present 2° wave absent	1° wave present 2° wave absent	Absent
Aggregation-Epinephrine	Normal	Absent	Absent	1° wave present 2° wave absent	Decreased	Normal ⁶
Aggregation-Arachidonic Acid ¹	Normal	Absent	Absent	Normal ³	Absent	Normal

Notes:

1) Arachidonic Acid is converted to TxA2 if COX-1 and Thromboxane synthase are present causing a strong platelet activation.

2) Ristocetin causes a conformation change in VWF which increases its affinity for GP1b/IX/V. This causes a non-physiologic "agglutination" of platelets where they are connected by Gp1b and VWF rather than GPIIb/IIIa and fibrinogen (the predominant linker of platelets in vivo).

3) AA gets converted into supra-physiologic amounts of TxA2 leading to strong platelet activation independent of the dense granule related 2nd wave.

4) Dense Granule Deficiency AKA Storage Pool Disease can be an isolated syndrome or part of other genetic diseases such as Hermansky-Pudlak, Chediak-Higashi, or Wiskott-Aldrich

5) Distinguish a deficiency in dense granules from a disorder of secreting dense granules by electron microscopy to see whether dense granules are present

6) Epinephrine causes a 2nd wave despite the absence of ADP signaling since other dense granule contents are still present (ATP and serotonin)

