Disorder	Defect	Platelet count	Blood Film	ADP [2µM]	ADP [5-10 µM]	Collagen [1 µg/mL]	AA [100 µM]	Adrenaline [10 µM]	Ristocetin [1.2 mg/mL]	Ristocetin [0.5 mg/mL	Thrombin [0.2 U/mL]	Platelet Nucleotides	Platelet Procoagulant Activity	Comments
Normal	None	Normal [150-400 x 10^9/L	No abnormality	Primary and secondary wave aggregation seen	Secondary wave aggregation only	Normal	Normal	Normal	Normal	Absent		Normal ATP:ADP ratio >2.0 but this can vary from lab to lab	Normal	None!
Glanzmann's Thrombasthenia [see comments]	Defect in GpIIb/IIIa receptor	Normal [150-400 x 10^9/L]	No abnormality MPV ↑	Absent	Absent	Absent	Absent	Absent	Normal [although maximal light transmission is decreased]	Absent	Absent	Normal	May be reduced	Flow cytometry shows decreased expression of the Gp IIb/IIIa receptor A similar picture will be seen in Afibrinogenaemia as Fibrinogen is essential for normal platelet aggregation via the GpIIb/IIIa receptor
Bernard Soulier Syndrome [see comments]	Defect in GpIb-V-IX receptor	Reduced	MPV ↑ Giant platelets Thrombocytopaenia	Normal	Normal	Normal	Normal	Normal	Absent	Absent	Normal	Normal	May be reduced	Important to check that VWF levels are normal. Flow cytometry shows decreased expression of the Gp Ib receptor and may show increased expression of the Gp IIb/IIIa receptor due to the presence of large platelets.
Velocardiofacial Syndrome	Chromosomal deletion of 22q11.2 including the *GpIb* beta gene	Reduced	MPV ↑ Giant platelets Thrombocytopaenia	Normal	Normal	Normal	Normal	Normal	Absent	Absent	Normal	Normal	May be reduced	The deleted chromosomal region in >90% of VCFS patients includes the GpIb beta gene, encoding for one subunit of the platelet GpIb-V-IX receptor.
Aspirin/NSAIDs	Inhibits cyclooxygenase either irreversibly [aspirin] or irreversibly [NSAIDs]	Normal [150-400 x 10^9/L]	No abnormality	Primary wave only [Absent second wave]	Primary wave only [Absent second wave]	Decreased or absent	Absent	Decreased	Normal	Absent	No effect	Normal	Normal	
Cyclooxygenase Deficiency & defects in Thromboxane synthesis [Similar to Aspirin]	COX-1 deficiency due either to a complete absence of the enzyme or a functional abnormality. Leads to defective TxA2 generation	Normal	No abnormality	Primary wave only [Absent second wave]	Primary wave only [Absent second wave]	Decreased or absent	Absent	Decreased	Normal	Absent	No effect		Normal	
Gp1a/IIa & GpVI deficiency [Collagen receptor deficiency]	Defect in the platelet collagen receptor	Normal	Normal	Normal	Normal	Absent	Normal	Normal	Normal	Absent	Normal	Normal	Normal	GpIa and GpVI are the major collagen receptors on platelets.
Gray Platelet Syndrome [α-granule deficiency]	Deficiency of α-granules	Often decreased	Platelets appear gray in colour - sometimes referred to as platelet 'ghosts' MPV ↑	Normal or decreased	Normal or decreased	Variable but usually decreased	Normal	Normal	Normal	Absent	Decreased/Absent			Decreased α-granules on EM Association with myelofibrosis
2B VWD	Dysfunctional VWF protein	Decreased	Thrombocytopaenia	Normal	Normal	Normal	Normal	Normal	Normal	Increased	Normal	Normal	Normal	Type 2B VWD and Platelet-Type VWD can be distinguished from each other by: - Mixing studies - Genetic Tests
Platelet-type VWD	Mutation in the GpIb receptor - usually within the hinge region of the protein	Decreased	Thrombocytopaenia	Normal	Normal	Normal	Normal	Normal	Normal	Increased	Normal			
Dense Granule Deficiency [δ-granule deficiency]	Defect in dense granules	Normal	Usually normal	Normal	Absent	Decreased	Normal	Absent secondary wave	?? Primary Wave Only	Absent	Normal or decreased	Increased ATP:ADP ratio	Decreased or normal	Decreased dense granules on EM Decreased ATP release by lumiaggregometry Albinism in patients with Hermansky Pudlak disorder or Chediak Higashi syndrome
Secretion Defect	Dense granules are present but not released	Normal	Usually normal	Normal	Absent	Decreased	Normal	Absent secondary wave	?? Primary Wave Only	Absent	Normal or decreased	Normal	Normal	
Scott Syndrome	Defect in platelet procoagulant activity characterised by impaired transmembrane migration of procoagulant phosphatidylserine Other disorders can also be associated with defective PCA	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Normal	Absent	Normal	Normal	Decreased	In some cases appears due to a mutation within the ABCtransporter A1 [ABCA1] gene or to a *trans*-acting regulatory gene sequence that controls its expression Decreased PF3a
ADP antagonists e.g. Clopidogrel ADP receptor [$P2Y_{12}$] abnormality	Inhibition of the ADP receptor	Normal	Normal	Absent	Absent	Normal	Normal	Normal	Normal	Absent	Normal	Normal		Important to check that VWF levels are normal. Flow cytometry shows decreased expression of the Gp Ib receptor and may show increased expression of the Gp IIb/IIIa receptor due to the presence of large platelets.
Signal Transduction Disorders	Multiple - poorly defined	Normal	Normal	Primary wave only [Absent second wave]	Primary wave only [Absent second wave]	Decreased			Normal	Absent		May be reduced	??? Normal	Normal Granule contents but there may be a defect in ADP release
Myeloproliferative Disorders		Decreased or increased		Inconsistent findings	Inconsistent findings	Inconsistent findings	Inconsistent findings	Inconsistent findings	Inconsistent findings	Absent	Inconsistent findings	May have an acquired SPD		Membrane GpIIb/IIIa expression may be reduced
Uraemia	Acquired SPD	May be decreased		Normal	Decreased	Normal or decreased	Normal or decreased	Decreased	Normal or decreased	Absent	Normal or decreased	Normal or decreased	Decreased	GPIb and IIb/IIIa expression may be reduced
Liver Disease	Multiple	May be decreased	May show Thrombocytopaenia	Primary wave only [Absent second wave]	Primary wave only [Absent second wave]	Decreased	???	Primary wave only [Absent second wave]	Normal or decreased	Absent	Decreased	May have an acquired SPD		
DIC	Multiple	May be decreased	May show Thrombocytopaenia	Decreased	Decreased	Decreased	Decreased	Decreased	Decreased	Absent	Decreased	Acquired SPD		
Cardio-Pulmonary Bypass	Acquired SPD	May be decreased	May show Thrombocytopaenia	May be decreased	May be decreased	May be decreased	May be decreased	May be decreased	Decreased	Absent	May be decreased	Deficiency of α and δ granules		Most ECMO patients also demonstrate decreased platelet counts and impaired platelet function. The platelet secretion defects are similar to storage pool deficient in inherited Platelet Disorders