Platelet Storage Pool Disorder in a Parturient

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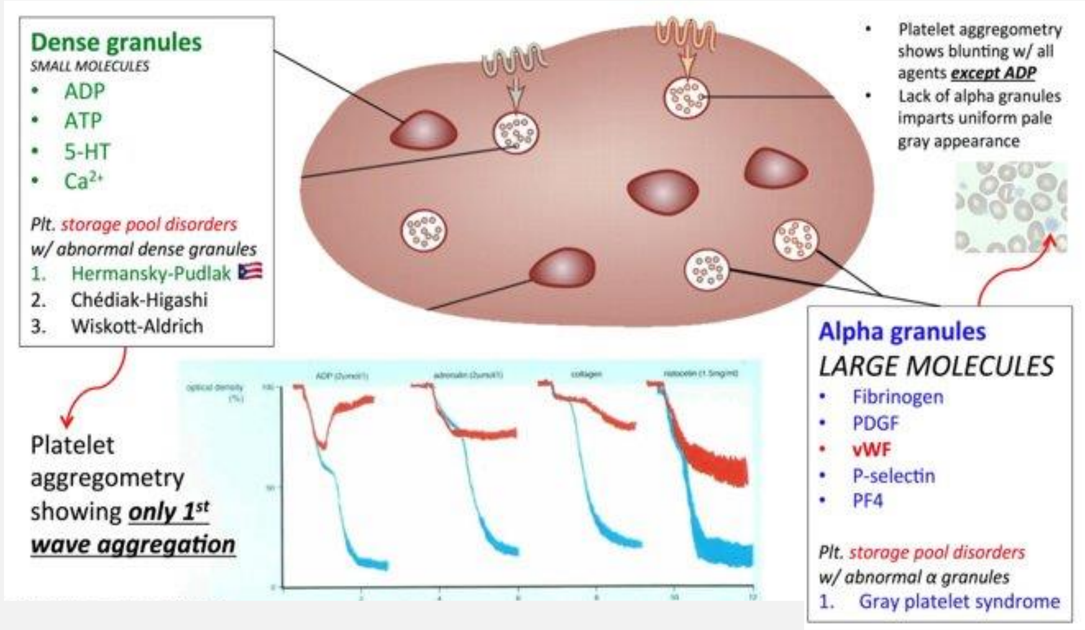
Introduction

- Platelet Storage Pool Disorder (PSPD) is a disease process characterized by defective alpha or dense granules in the structure of the platelet
- Leads to a **<u>qualitative</u>** defect in platelet function, despite normal platelet counts[1]
- Increases the risk for postpartum hemorrhage in mother
- Increases the risk for epidural hematoma with neuraxial anesthesia
- Increases the risk of intracranial hemorrhage in newborns with inherited PSPD (and in newborns with status unknown



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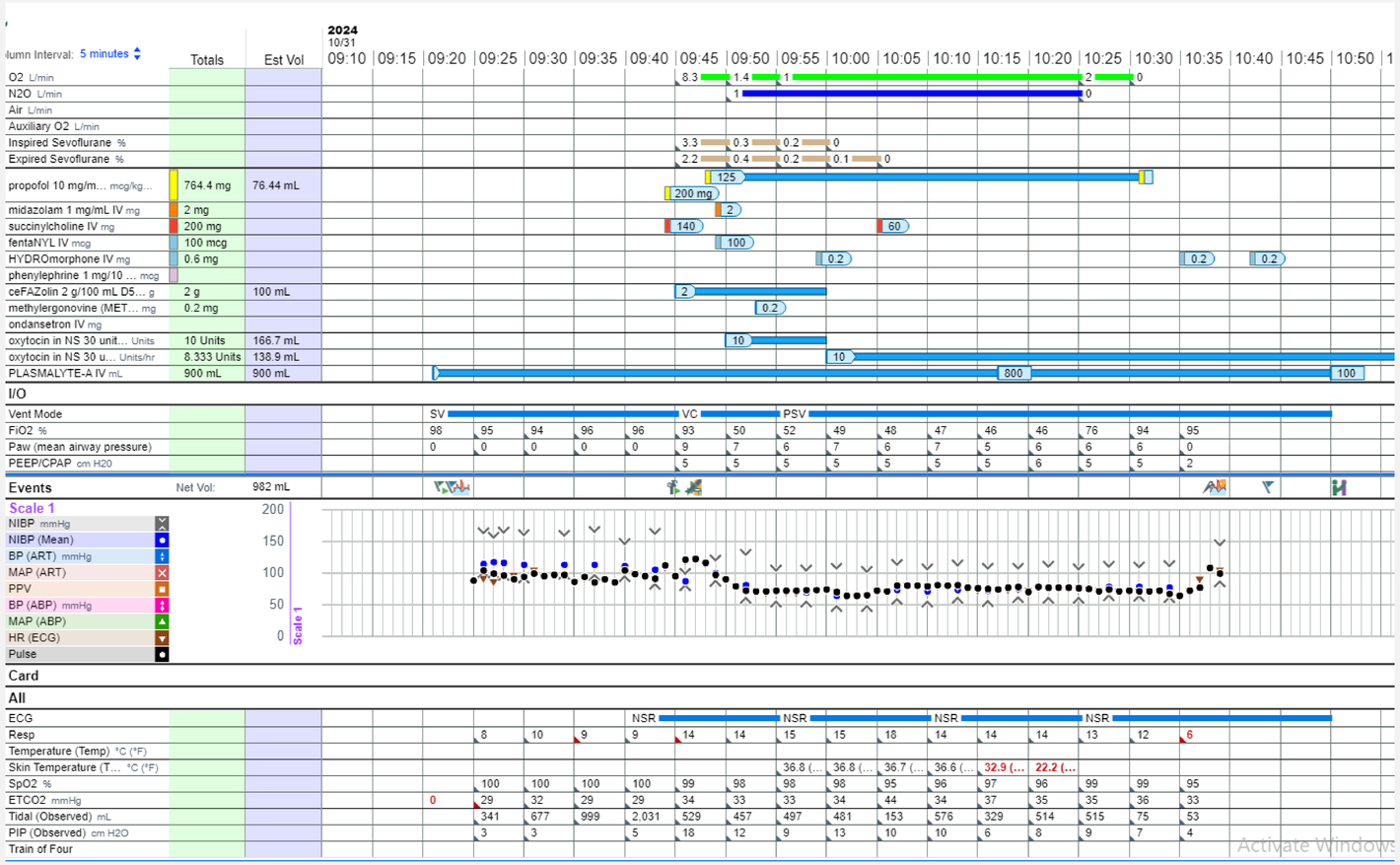
Case Presentation

- 28-year-old G1P0 F w/ family history of PSPD, personal history of easy bruising and hematoma formation following laparoscopic appendectomy presented to anesthesia preoperative clinic to discuss delivery options and anesthetic considerations
- Multi-disciplinary discussion between MFM, Anesthesia, and Hematology teams
- Risks Considered: Maternal bleeding with C-section vs neonatal intracranial hemorrhage with vaginal delivery vs epidural hematoma with neuraxial anesthesia
- Shared decision made to move forward with scheduled c-section under general anesthesia
- Preoperative labs: Hgb 11.5g/dL, PLT 248 K/uL, collagen aggregation and collagen ATP release slightly decreased
- Thromboelastography was unavailable to assess clot integrity for a possible neuraxial anesthetic
- Hematology recommended platelet transfusion, recombinant coagulation factor VIIa, tranexamic acid, and DDAVP as needed for intraoperative bleeding.
- GA proceeded uneventfully. TIVA implemented after delivery to decrease the risk of uterine atony. Moderate uterine atony was treated with oxytocin and IM methergine with good response.
- The patient emerged from anesthesia and was extubated without complications. QBL was ~1270mL w/o need for transfusion of products
- Both mother and baby were discharged without any long-term complications or morbidity three days later.



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Discussion

- PSPD is a platelet aggregation abnormality that can result in a bleeding diathesis due to defective platelet granules[1]
- Patient's may not receive a diagnosis even with bleeding history, due to normal platelet counts
- Diagnostic tests for PSPD include platelet aggregation, electron microscopy, and thromboelastography (TEG)[1]
- Patients with known PSPD or familial history of PSPD benefit from multidisciplinary discussion between obstetrics, anesthesia, pediatrics and hematology in guiding decision making
- Considerations for mother: increased risk of postpartum hemorrhage and/or epidural hematoma formation with neuraxial technique
- Considerations for newborn: Increased risk of intracranial hemorrhage, as has been seen in infants with inherited hemophilia[2]
- Highlights the importance of TEG which can allow physicians to determine which specific blood products can be given to slow down postpartum hemorrhage.

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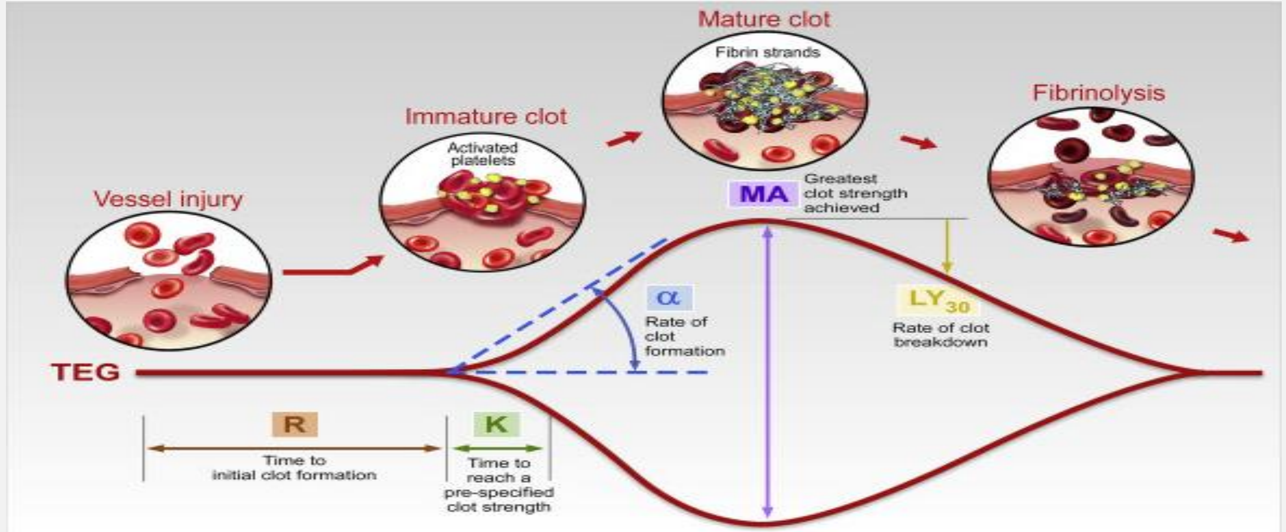

	R-time (R)	K-time (K)	Alpha angle (α)	Maximum Amplitude (MA)	Lysis % at 30 mins. (LY30)
Definition	Time to first deviation from baseline	Time for tracing to reach 20 mm amplitude	Angle between baseline and tangent line that intersects initial deviation	Maximum deviation of tracing from baseline	Decrease in curve amplitude (relative to MA) at 30 minutes
Controlling Pathways	Coagulation cascade	Fibrinogen cleavage Fibrin polymerization	Fibrinogen cleavage Fibrin polymerization	Fibrinogen activity Platelet count / quality	Fibrinolysis
Interpretation	↑ = hypocoagulable ↓ = hypercoagulable			↓ = hypocoagulable ↑ = hypercoagulable	
Therapeutic Implications	↑ = administer fresh frozen plasma	If K time is ↑ or alpha angle is ↓, then: administer cryoprecipitate or fibrinogen concentrate		↓ = administer platelets	↓ = administer tranexamic acid

References

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2.Lavee O, Kidson-Gerber G. Update on inherited disorders of haemostasis and pregnancy. Obstet Med. 2016 Jun;9(2):64-72. doi: 10.1177/1753495X15624307. Epub 2016 Jan 20. PMID: 27512496; PMCID: PMC4950409
3.New Uses for Thromboelastography and Other Forms of Viscoelastic Monitoring in the Emergency Department: A Narrative Review. Tyler, Patrick D. et al.. Annals of Emergency Medicine, Volume 77, Issue 3, 357 - 366