

Platelet Storage Pool Disorder in Pregnancy: An under recognised condition increasing risk of post-partum haemorrhage

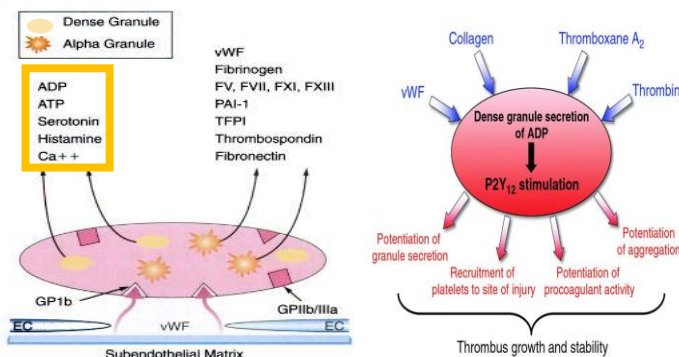
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INTRODUCTION

Platelet Storage Pool Disorder (PSPD) refers to an underrecognized group of bleeding disorders characterised by normal platelet counts but with impaired functioning due to deficiency of dense granules. Women with PSPD may have abnormal uterine bleeding and are at risk of Post-partum haemorrhage (PPH) during childbirth. According to reports from the U.S. Centres for Disease Control and Prevention, in 2021, 3528 patients were living with PSPD, 67% of whom were women. There are only a few reported cases in pregnancy and therefore the management remains unclear. Routine use of DDAVP (Desmopressin acetate) to prevent excessive bleeding has been suggested in some case reports.



CASE REPORT

A 26 year old Primigravida at 40 weeks gestation was admitted for elective induction of labour. She denied history of medical comorbidities during antenatal clinic visits. On admission to birth suite, she disclosed diagnosis of PSPD in childhood when she was investigated (platelet aggregation test) by Paediatric Haematologist for the cause of her easy bruising. Her other symptom included menorrhagia. She denied history of excessive bleeding during previous surgery in 2021. Platelet count and Coagulation studies were normal on admission. Due to high risk of PPH associated with PSPD and limited data available on its management, consultation was sought from Haematologist & Anaesthetist. Labour was induced, Epidural analgesia was avoided and platelets & DDAVP were kept readily available. Emergency Caesarean Section under Spinal Anesthesia was performed due to fetal distress.

INVESTIGATIONS

PLATELET AGGREGATION TEST

AGONIST	CONCENTRATION	COMMENT CODE
ADP	5.0 UMOLH.....
ADP	2.5 UMOLH.....
RISTOCETIN	1.5 ug/ml2.....
COLLAGEN	4.0 ug/mlH.....
COLLAGEN	2.0 ug/mlH.....
ARACHIDONIC ACID	1.0 mmolH.....
FINAL COMMENT/ CODE : ADP 10.0 umol	H.....

Comment codes :
 N Normal aggregation
 A No response
 I Impaired response

PO1 Comment : Platelet aggregation studies showed normal response to all stimuli used.
 PO2 Comment : The above aggregation studies are suggestive of a release defect, such as that due to aspirin or N.S.A.I.D ingestion.

PLATELET FUNCTION TEST

Against	Concentration	Result
	2.50 uM/mL	Reduced response.
	5.00 uM/mL	Reduced response.
	4.0 ug/mL	Normal response
	1.5 ug/mL	Normal response
Arachidonic Acid	1.0 mM/mL	Reduced response.

Comment: Collagen 2.0 ug/ml = Reduced response. Reduced response to ADP and low dose Collagen would suggest a PLATELET STORAGE POOL DEFECT. repeat study with family studies may be appropriate depending on clinical circumstances. Reviewed by Dr P Monagle, Staff Haematopathologist.

INVESTIGATION	RESULT	
	ON ADMISSION	POST-OP DAY 1
Platelet Count	190	172
PT/INR	11.5/1.0	-
aPTT	26	-

RESULT

There was 1100 ml PPH from brisk bleeding from the lower uterine segment. DDAVP was not administered as haemostasis was quickly achieved with 2 layer uterine closure, hemostatic sutures and oxytocin administration. Prophylactic intravenous tranexamic acid was given for 24 hours. She was discharged on 3rd post-operative day without any episodes of secondary PPH.

CONCLUSION

Women with PSPD have increased risk of PPH. There are only few reported cases, each with different management approaches. Our case highlights the importance of multi-disciplinary team approach and that DDAVP may not be required in all patients. Further studies need to be carried out to draw definitive conclusions on its management.

References

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- 2) Melissa JEWELL, Everett F. MAGANN, Andrew BARR and Ross BAKER; Management of platelet storage pool deficiency during pregnancy; *Australian and New Zealand Journal of Obstetrics and Gynaecology* 2003; 43: 171-172
- 3) Sadia Khakwani, Claire Winton, Nosheen Aslam, Suzanne Taylor; Platelet storage pool disorder: multidisciplinary planning in pregnancy; *BMJ Case Rep* 2021;14:e239321. doi:10.1136/bcr-2020-239321