

Bernard-Soulier Syndrome in Pregnancy: A Case Report

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INTRODUCTION

Bernard-Soulier Syndrome (BSS) is a rare inherited bleeding disorder caused by a defect or deficiency in the glycoprotein Ib-IX-V complex on platelets. Increases the risk of excessive bleeding. Managing pregnancy in women with BSS requires a multidisciplinary approach to address bleeding risks, transfusion challenges, medication, and planned analgesia.

Bleeding Risk

- **Antepartum:** Women with BSS are at risk for spontaneous bleeding, including mucosal bleeding and ecchymoses. Monitoring for signs of bleeding during pregnancy is essential.
- **Intrapartum:** Delivery is a high-risk period due to the potential for severe postpartum hemorrhage (PPH). Vaginal and cesarean deliveries carry significant bleeding risks.
- **Postpartum:** Risk of delayed bleeding can persist in the postpartum period, requiring close monitoring.

CASE REPORT

A 29-year-old woman at 36 weeks of pregnancy (G3, P0+2) with BSS and HLA antibodies presented in early labor.

- HLA antibodies, which impair platelet survival after transfusion.
- ICU transfer for labor augmentation.

Past Obstetric History

- Marked by PPH

Management

- Intravenous immunoglobulin
- Recombinant factor VIIa
- HLA-matched platelets
- Packed red blood cells

Investigations at presentation:

- Hemoglobin: 9.2g/dL
- Platelets: $42 \times 10^9/L$

PATIENT CUMULATIVE REPORT				
Test Name	02/10/2024 15:50:09 2462018141	02/10/2024 10:18:55 2462017766	01/10/2024 21:09:08 2462013254	01/10/2024 17:57:56 2462015118
# PT	8.00 L	12.30		
# PT INR	0.698 L	1.070		
APTT Patient	25.63	26.90		
APTT Ratio	0.86	0.90		
WBC	8.41	10.40 H	9.46	8.50
RBC	3.15 L	3.46 L	3.91 L	3.85 L
Hemoglobin	74.00 L	82.00 L	92.00 L	91.00 L
Hematocrit	0.282 L	0.275 L	0.309 L	0.303 L
MCV	79.00	79.40	79.10	78.70
MCH	23.50 L	23.60 L	23.70 L	23.60 L
MCHC	294.00 L	298.00 L	298.00 L	300.00 L
RDW	17.90 H	18.00 H	17.70 H	18.00 H
Platelet Count	29.00 L	39.00 L	42.00 L	41.00 L
MPV	10.00	11.70 H	10.00	11.00
Retic %	2.34			
Retic #	0.02			
Neutrophils %	83.10 H	88.90 H	79.30 H	76.10 H
Lymphocytes %	10.80 L	6.30 L	15.30 L	17.00 L
Monocytes %	4.10	3.30 L	3.90 L	4.40
Eosinophils %	0.40 L	0.40 L	0.70 L	0.80 L
Basophils %	0.50	0.30	0.30	0.40
PLC%	1	1	0	1
Neutrophils #	7.00 H	9.25 H	7.40 H	6.55 H
Lymphocytes #	0.91 L	0.66 L	1.45	1.46
Monocytes #	0.34	0.34	0.37	0.38
Eosinophils #	0.03 L	0.04 L	0.07 L	0.07 L
Basophils #	0.04	0.03	0.03	0.03
HGB#	0	0	0	0
Thrombin_C	4.20			
2-Dimer_Femostil	1026.00			

DISCUSSION

- Due to the increased risk of bleeding in these situations, prophylactic measures against bleeding may include the use of tranexamic acid, desmopressin (DDAVP), recombinant factor VIIa (rFVIIa), and platelet transfusions.
- It is preferable to refrain from platelet transfusion due to the potential risk of alloantibody formation.
- The fetus typically carries a heterozygous form of the disease, resulting in its platelets potentially containing antigens not present in the mother's platelets.
- In cases of cesarean sections or bleeding during vaginal deliveries, the first line of treatment involves the infusion of HLA-compatible platelets along with tranexamic acid.

CONCLUSION

The condition is highly variable and somewhat unpredictable. Close monitoring of the mother's hematocrit and platelet count is essential to promptly identify and address any bleeding during the peripartum period. Managing pregnancy multidisciplinary approach and in such cases remains a subject of debate, necessitating a personalized medical care.

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