Background

- Coagulopathy in pregnancy increases maternal and fetal morbidity and mortality due to higher risk for postpartum hemorrhage and transfusion.
- Bernard–Soulier Syndrome (BSS) is a **rare autosomal recessive platelet disorder** with Prevalence: **<1 per 1,000,000.**
- Defect in **GP Ib-IX-V receptor**, impairing platelet adhesion to vWF, thrombin binding. Leads to large dysfunctional platelets, prolonged bleeding time, variable bleeding phenotype.
- Presents unique peripartum challenges due to limited evidence guiding anesthetic management.

The Case

- A 31-year-old primigravida with suspected Bernard–Soulier–variant platelet disorder presented at 39 weeks and 3 days for delivery planning.
- A platelet abnormality had been identified years earlier before lumbar surgery, but the diagnosis was incompletely characterized.
- Hematology studies demonstrated reduced ristocetin-induced platelet aggregation and decreased CD42a/CD42b expression on flow cytometry, consistent with a Bernard–Soulier–like phenotype.
- Patient reported heavy menstrual bleeding, and IVF-related preconception testing ruled out common BSS mutations.

- Care coordination including maternal–fetal medicine, obstetrics, anesthesiology, and hematology

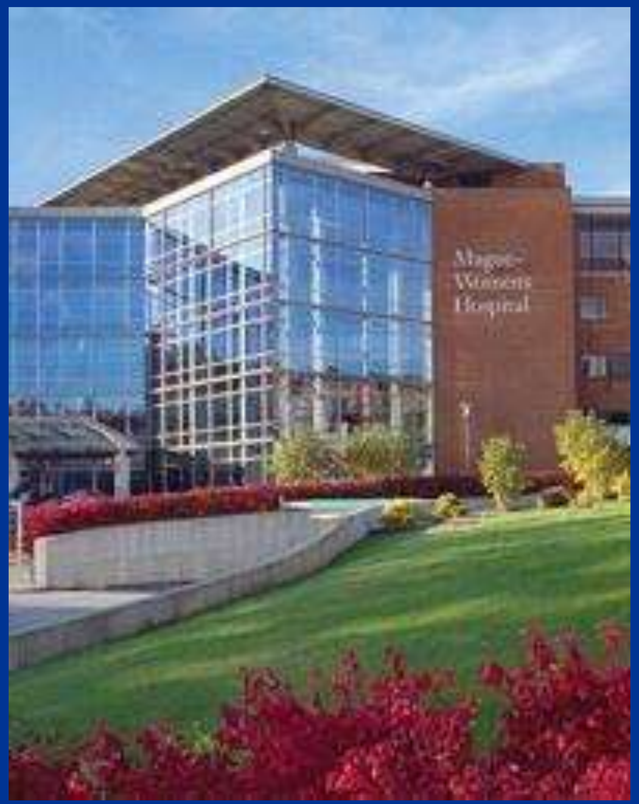
- Peripartum plan included prophylactic platelet transfusion and perioperative tranexamic acid, with postoperative duration guided by clinical course

- Neuraxial anesthesia was contraindicated; labor analgesia included nitrous oxide and remifentanil patient-controlled analgesia

- Labor arrest of descent necessitated cesarean under general anesthesia

Teaching Points

- Inherited platelet disorders challenge obstetric anesthesia because functional defects may not correlate with platelet count. Phenotype varies widely; bleeding severity cannot be reliably predicted.
- Most published BSS obstetric cases **avoid neuraxial anesthesia** due to risk of spinal/epidural hematoma.
- Neuraxial considered only after **platelet transfusion**, documented functional improvement, and **hematology agreement**.
- Even with prophylactic platelets and TXA, **significant hemorrhage** is frequently reported. Establishing a management algorithm is essential.
- Diagnostic insights and multidisciplinary planning can guide anesthetic planning and optimize outcomes in high-risk parturients.



University of Pittsburgh

Yaghoubi M, Sabeti N, Hajmollarezaei E. Successful vaginal delivery in a pregnant woman with Bernard–Soulier syndrome: a case report. *Clin Case Rep.* 2026;14(2):e71706.

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