

SUCCESSFUL LABOR NEURAXIAL ANALGESIA IN PARTURIENT WITH SUSPECTED BERNARD-SOULIER SYNDROME

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Case Presentation

- 27-year-old G1P0 at 39w0d presenting for consultation in setting of rare sub-type thrombocytopenia
 - 1st trimester thrombocytopenia (**80-100** $10^9/L$)
 - **70** $10^9/L$ one week before induction
 - Bleeding history: post-tonsillectomy, mild gum bleeding, & easy bruising
- Hematology consult: c/f mild-AD Bernard-Soulier Syndrome
 - Genetic testing deferred until post-partum
- Hematology recommendations
 - Pre-neuraxial platelet (plt) transfusion
 - CBC q6 hours during labor
 - At delivery: 1 gm tranexamic acid and plt transfusion

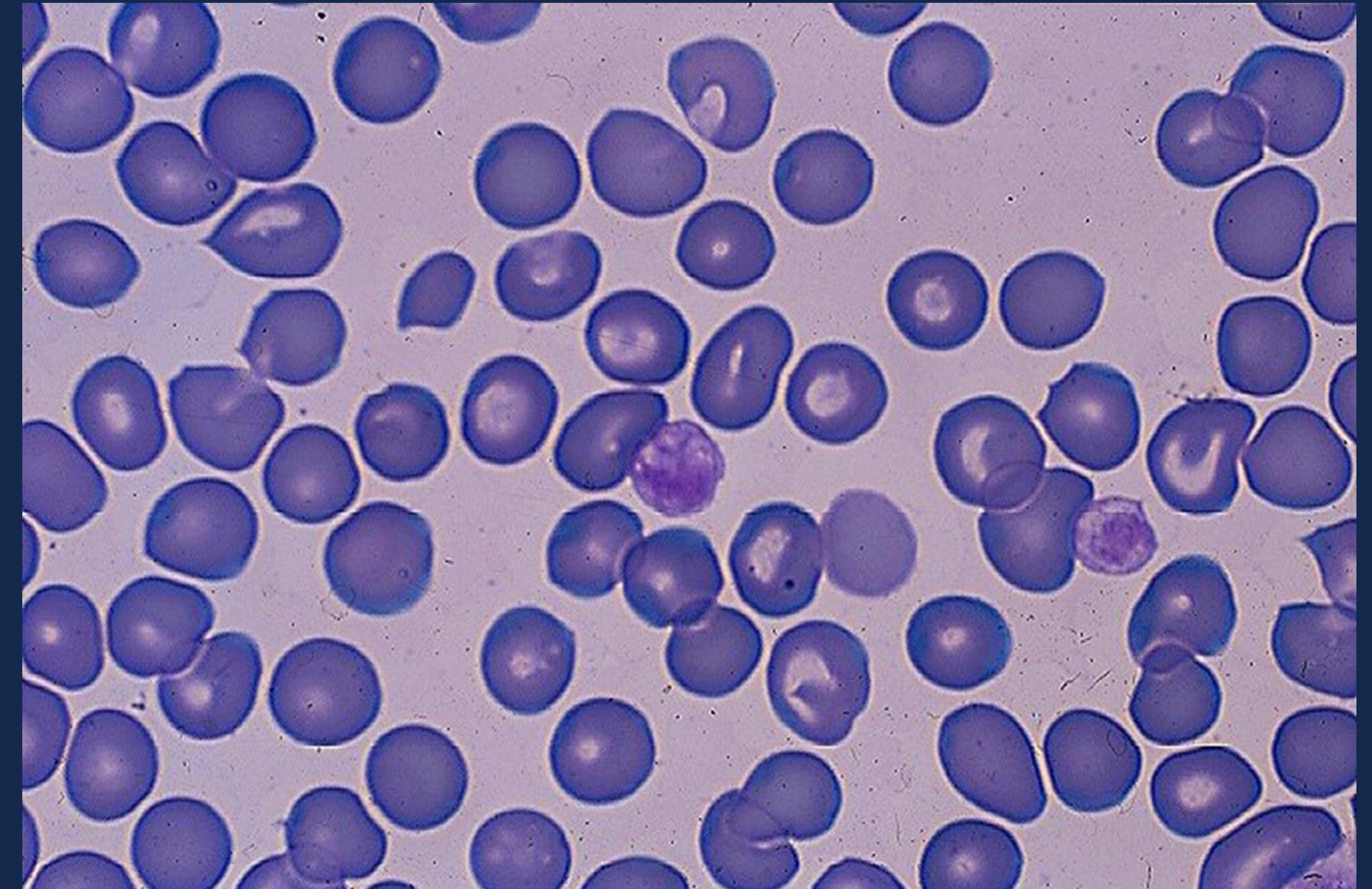


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Peripartum Course

- Admission platelets **97** $10^9/L$, transfused one unit of platelets with a rise to **124** $10^9/L$, & oral TXA started
- Labor epidural placed prior to Pitocin initiation
- Before delivery, the platelet count was **129** $10^9/L$ and at delivery she received a unit of platelets and one gram of intravenous TXA
- Delivery was complicated by a first-degree laceration and her quantitative blood loss was 306 ml
- Post-delivery, the platelet count was **176** $10^9/L$
- Serial lower extremity neuro checks were performed x24 hours post epidural removal, which occurred 2 hours post-partum

Discussion

- Bernard-Soulier Syndrome (BSS)
 - Rare inherited platelet disorder with an estimated prevalence of 1/1,000,000
 - Typical autosomal recessive inheritance pattern, but more mild forms can demonstrate an autosomal dominant inheritance
- Characterized by impaired platelet aggregation caused by a defect within the membrane glycoprotein Ib-IX-V complex
- Systematic reviews and case reports in around 30 pregnancies
 - Modes of delivery split 50/50 between cesarean and vaginal
 - Primary post-partum hemorrhage occurred in a third of all cases with two leading to hysterectomies
 - Neuraxial anesthesia was not utilized in any of the cases, but platelet counts were typically below **80** $10^9/L$
- Currently, no guidelines exist regarding optimal mode of delivery and treatment is focused on supportive measures including platelet transfusions, TXA administration, and consideration for recombinant factor VIIa

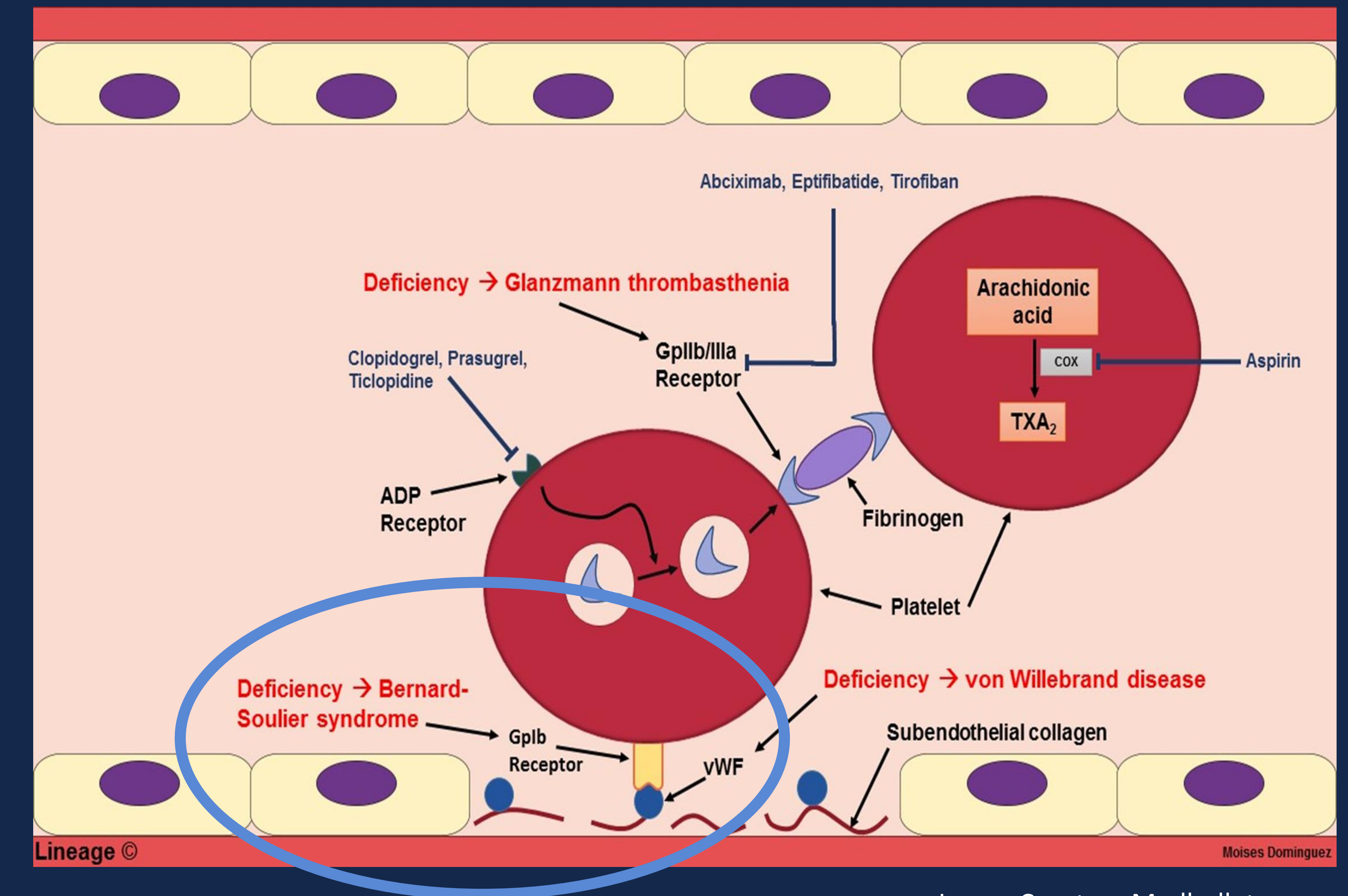


Image Courtesy Medbullets.com

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