

Peripartum Management of Hypodysfibrinogenemia

Rachel Brock, MD and Jaime Daly, MD

- Rare congenital quantitative and qualitative fibrinogen disorder
 - May manifest as bleeding, thrombosis, or both
- Genetics:
 - Caused by mutations in the fibrinogen-encoding genes (FGA, FGB, FGG), most commonly heterozygous missense mutations, leading to incomplete segregation of mutations with clinical phenotypes
- Pregnancy:
 - Increased risk of miscarriage (~30%), placental abruption (~8%), and postpartum hemorrhage (~20%)
- Neuraxial:
 - Historically, neuraxial has often been avoided due to concern for hematoma formation



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Case:

- Presentation: 31-year-old G1P0000 at 37 weeks and 4 days' gestation with the hypercoagulable subtype of familial hypodysfibrinogenemia presents for planned IOL.
 - Strong family hx of VTE, maintained on prophylactic enoxaparin
- Admission: Fibrinogen level is 61 mg/dL; receives fibrinogen concentrate (70 mg/kg), increasing level to 179 mg/dL
 - CSE Placed w/o complication, removed after delivery
- Postpartum: PPD1, fibrinogen is 147 mg/dL; she receives an additional 1 g fibrinogen concentrate
 - Resumes therapeutic enoxaparin that evening



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Safe Neuraxial Requires:

- Laboratory-guided replacement
- Risk-balanced anticoagulation
- Multidisciplinary collaboration

Recommendations:

- Minimum fibrinogen level of 150 mg/dL
- Fibrinogen concentrate is preferred
- Avoid antifibrinolytics
- Utilize TEG, especially in PPH



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