Paroxysmal Nocturnal Hemoglobinuria (PNH)

Overview

Rare, acquired hematologic disorder.

Complement-mediated hemolysis, thrombosis, bone marrow & renal failure.

Pregnancy Risks

Increased maternal morbidity & mortality.

Requires multidisciplinary management.

Therapies

Eculizumab (ECU): Well-studied in pregnancy.

Pegcetacoplan (PEG): Newer, improved hemoglobin stabilization.

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Case Report

24F G1 PNH

Initially on **PEG** ⟶ **ECU**
Reinstated **PEG** in 3rd trimester ⟶ stable control

Pre-op Course

Hb: 105 g/L
Plt: 126 x10^9/L
Last transfusion 8 wks prior to admission

Daily dalteparin for thromboprophylaxis

Anesthesia

Spinal anesthesia with intrathecal morphine, arterial line & 2 large-bore IVs

Outcome

Uncomplicated C/S, Discharge on POD3

Post-op **Hb**: 86-92 g/L
LDH peak: 378 U/L

Conclusions

- Neuraxial anesthesia is feasible in optimized PNH patients
- **Pegcetacoplan** may provide superior control during pregnancy
- Key considerations:
  - Continue thromboprophylaxis (e.g., dalteparin)
  - Monitor hemolysis & hematologic parameters
- Importance of **multidisciplinary planning** (Anesthesia, Hematology, Obstetrics)

1. Hillmen P, Szer J, Weitz I, Röth A, Höchsmann B, et al. Pegcetacoplan versus eculizumab in paroxysmal nocturnal hemoglobinuria. N Engl J Med. 2021 Mar 18;384(11):1028-1037.