Perioperative Management of a Pregnant Patient with Hereditary Spherocytosis

- **Hereditary spherocytosis:** inherited RBC membrane defect → spherical, fragile RBCs → chronic hemolytic anemia
- **Pregnancy effects:** ↑ hemolysis, ↓ physiologic reserve, ↑ bleeding and thrombotic risk
- **Splenectomy:** improves anemia but increases thrombotic risk
- **Anesthetic concerns:** oxygen delivery, anemia tolerance, hemorrhage vs thrombosis balance
- **Key principle:** complex hematologic disease in pregnancy requires multidisciplinary planning

SCHOOL OF MEDICINE
ANESTHESIOLOGY & CRITICAL CARE MEDICINE

Case Presentation

23-year-old G9P0, 24 weeks gestation

- **Major Comorbidities**
  - Hereditary spherocytosis (status post splenectomy)
  - Non-alcoholic cirrhosis with portal hypertension
  - Esophageal & gastric varices
  - Portal vein thrombosis
  - Recurrent thromboembolic disease
- **Pregnancy Course**
  - Gastric variceal bleeding
  - Interstitial pancreatitis
  - Ascites
  - Progressive fetal growth restriction
- **Anesthetic & Delivery Plan**
  - Multidisciplinary planning with OB, anesthesia, hematology, neonatology
  - Urgent cesarean delivery under general anesthesia
  - Pre-induction central venous and arterial access
  - Rapid-sequence induction
  - Delivery achieved within 5 minutes
- **Analgesia Strategy**
  - Regional pain block prior to emergence
  - Multimodal analgesia pre- and post-emergence

Teaching Points

- Anesthetic challenges of pregnancy complicated by hereditary spherocytosis and associated co-morbidities
- Differentiating hereditary spherocytosis and its distinct perioperative risks
- Successful management requires multidisciplinary coordination, careful balancing of hemorrhagic and thrombotic risk, and individualized analgesic strategies
- Awareness of rare hematologic conditions and their interaction with pregnancy physiology is critical