



Perioperative Considerations in a Patient with Hereditary Hemorrhagic Telangiectasia Undergoing Oocyte Retrieval



Ellie Cheng BA, Suzanne Karan M.D., Heather Lander M.D.
Center for Perioperative Medicine, University of Rochester

Introduction

- Hereditary Hemorrhagic Telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is a rare autosomal dominant disorder causing abnormal blood vessel formations (AVMs), often leading to bleeding and vascular complications.
- Common symptoms include epistaxis due to telangiectasias in the nasal mucosa, with potential AVMs affecting multiple organs (lungs, brain, liver) and risk of complications such as hemorrhage and stroke.
- Consider airway challenges, bleeding risks, and cardiovascular concerns, especially with AVMs and right-to-left shunting potential.

Case Description

A 38-year-old female with HHT presented for preoperative anesthesia evaluation before oocyte retrieval.

- **Medical History:** HHT with cutaneous telangiectasias, epistaxis, NSAID-induced gastritis, iron deficiency anemia, and migraines.
- **Diagnosis:** Initially diagnosed due to family history. Recent echocardiogram showed normal heart function with a small PFO, no intrapulmonary shunt or AVM.
- **Symptoms:** Three recent episodes of epistaxis and mild palpitations, self-limited, with in-clinic EKG showing sinus rhythm. Slight reduction in exercise tolerance.
- **ENT Evaluation:** No active bleeding sites or upper airway anomalies.

Preoperative Evaluation

ENT Clearance: Flexible laryngoscopy confirmed absence of upper airway telangiectasias. ENT advised substituting Flonase with a gentler nasal spray.

Cardiac & Pulmonary Screening: Echocardiogram revealed a small PFO without significant shunt or pulmonary AVMs. Palpitations evaluated as benign.

Epistaxis Consideration: Additional nasal care was advised to reduce dryness and prevent bleeding.

Anesthetic Plan

MAC vs. GA: Given the minimally invasive nature of oocyte retrieval, Monitored Anesthesia Care (MAC) was the initial choice. General Anesthesia (GA) prepared as a backup for complications.

Airway Management: No telangiectasias detected in the upper airway, but contingency plans developed for potential intubation complications/bleeding.

Perioperative Considerations

Bleeding Control: Local vasoconstrictors and nasal packing for potential epistaxis

Pulmonary AVM Screening: Re-screening advised if pregnancy is achieved due to the increased vascular load during pregnancy.

Pregnancy Risks: Should pregnancy occur post-retrieval → increased risks of bleeding and cardiovascular complications (e.g., heart failure, pulmonary hemorrhage, stroke), especially later trimesters.

Discussion

- **Anesthetic Management:** HHT presents unique perioperative challenges (e.g. bleeding risks and AVMs). Multidisciplinary team involved ENT, cardiology, and anesthesiology for thorough risk assessment.
- **Risk Mitigation:** To manage potential epistaxis, nasal care adjustments were made. A dual anesthetic approach (MAC with GA backup) was planned.
- **Pregnancy Considerations:** HHT elevates pregnancy risks, necessitating proactive AVM screening and cardiovascular assessment due to increased blood volume and vascular strain.

Conclusion

This case illustrates the value of comprehensive, interdisciplinary preoperative planning for HHT patients. Effective risk management allowed for safe oocyte retrieval, with ongoing monitoring recommended if pregnancy is achieved due to increased bleeding and cardiovascular risks.

References

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