

Perioperative Anesthetic Management of Elective Hernia Repair in a Patient With Hereditary Angioedema

Requiring Preoperative C1 Esterase Inhibitor Administration

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Introduction

- **Hereditary angioedema (HAE):** rare autosomal dominant disorder characterized by deficiency or dysfunction of C1 esterase inhibitor. Involves episodic, non–histamine-mediated angioedema that can involve the airway, gastrointestinal tract, and extremities.
- Perioperative stress, airway manipulation, mechanical pressure, and certain medications can precipitate life-threatening laryngeal edema.
- Optimal anesthetic care requires anticipatory planning, prophylactic therapy, medication availability, and advanced airway readiness, especially for patients **newly diagnosed** or **lacking prior exposure** to anesthesia.

Learning Objectives

1. Describe perioperative risk assessment and airway planning for patients with HAE.
2. Discuss indications and timing for prophylactic C1 esterase inhibitor administration.
3. Highlight multidisciplinary coordination required for safe anesthetic management in rare diseases.

Case Presentation

- **Patient:**
 - 51-year-old female
- **PMH:**
 - Hereditary angioedema (new diagnosis), GERD, Psoriasis, Lyme disease, Psoriatic arthritis, Rheumatoid arthritis, Chronic abdominal pain, Anxiety, Prediabetes
- **PSH:** Hysterectomy, breast augmentation, breast aspiration
- **Allergies:** Droperidol (anaphylaxis), Naproxen (anaphylaxis), Sulfasalazine (angioedema)
- **Presentation:**
 - Elective repair of a right inguinal hernia and incarcerated umbilical hernia.
 - Patient reported lifelong episodic abdominal pain and swelling, now attributed to HAE.
 - No prior history of airway edema.

References

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Our Anesthetic Plan

Preoperative Plan:

- 1500 units IV **Berinert** (C1 esterase inhibitor) administered prophylactically.
- Two units FFP immediately available if breakthrough angioedema occurred.
- Anaphylaxis and HAE emergency kits prepared.
- Difficult airway cart, video laryngoscope, and surgical airway tray at bedside.

Intraoperative Course:

- Monitored anesthesia care (MAC) with deep sedation.
- Avoided airway instrumentation.
- Procedure completed without hemodynamic or respiratory instability.
- No signs of edema, urticaria, or abdominal exacerbation.

Postoperative Course:

- Recovered uneventfully in PACU.
- No delayed swelling or respiratory compromise.
- Discharged home in stable condition.

Key Interventions

- Pharmacologic prophylaxis:** Berinert 1500 U IV (per current HAE guidelines)
- Trigger avoidance:**
 - No histamine-releasing drugs
 - No droperidol or NSAIDs due to prior anaphylaxis
 - Avoided airway manipulation via MAC
- Airway Preparedness:**
 - Surgical airway kit open
 - ENT on standby (if institutional protocol allows)
- Rescue Medications Available:**
 - C1-INH concentrate (extra dose)
 - Icatibant
 - Epinephrine
 - FFP
- Monitoring:**
 - Continuous capnography and respiratory pattern monitoring
 - Strict PONV control to avoid retching-induced swelling

Goals: Prevent airway edema, avoid triggers, ensure immediate rescue capability.

Discussion/Conclusion

- Hereditary angioedema creates unique anesthetic challenges because airway obstruction can develop rapidly and unpredictably.
- Unlike allergic angioedema, HAE does **not** respond to epinephrine, antihistamines, or steroids, making proper prophylaxis and equipment essential.

This case highlights several key features of perioperative care:

1. **Importance of Early Identification:**
Patients may present with vague abdominal symptoms for years before diagnosis, risking undetected perioperative flare potential.
2. **Prophylactic C1 Inhibitor is Evidence-Based:**
Current guidelines recommend pre-procedural C1-INH administration for surgeries involving mechanical stress, even without prior airway involvement.
3. **Economic and Institutional Challenges:**
Berinert is expensive and non-formulary at some hospitals. Advanced preparation and pharmacy coordination are critical for elective cases.
4. **Airway Strategy Matters:**
Although MAC was chosen to avoid instrumentation, full surgical airway readiness remained essential due to the potential for spontaneous laryngeal edema.
5. **Multidisciplinary Approach:**
Anesthesia, surgery, pharmacy, and nursing coordination contributed to safe outcomes.

Perioperative Management of Hereditary Angioedema

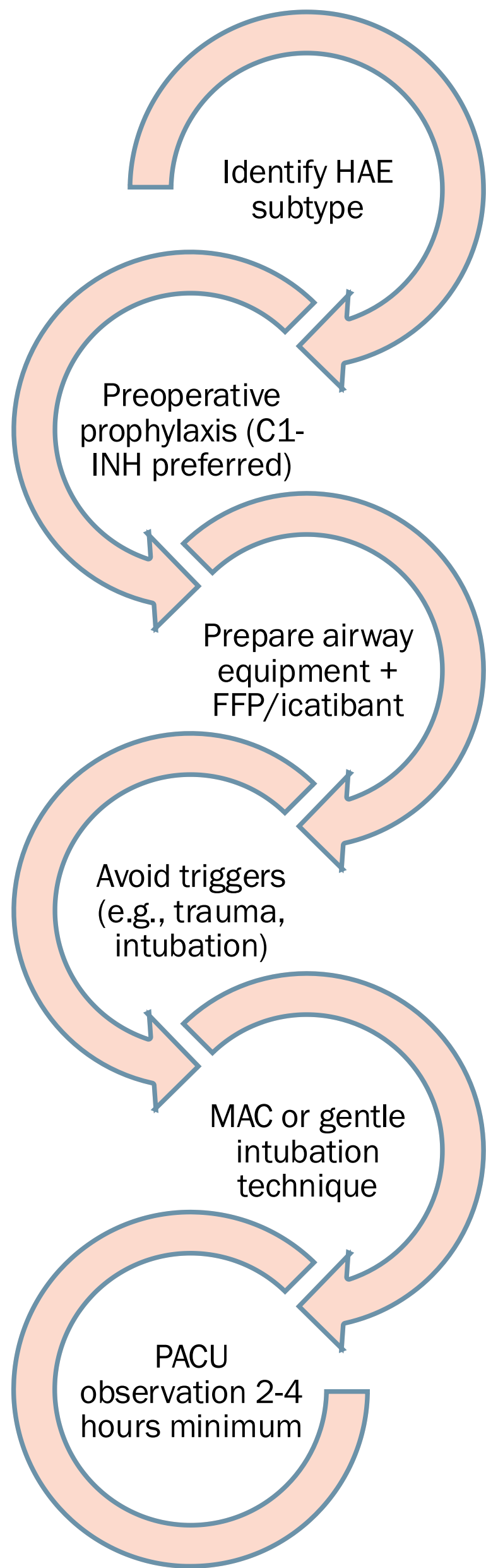


Table 1 – HAE Medications and Their Roles

Medication	Purpose	Notes
C1-INH (Berinert)	Prophylaxis and treatment	First-line for perioperative use
Icatibant	Acute bradykinin attack	Not prophylactic
FFP	Backup if C1-INH unavailable	Contains C1-INH but may worsen volume status