ASRA PAIN MEDICINE 51st Annual Regional Anesthesiology and Acute Pain Medicine Meeting
April 16-18, 2026 | Phoenix, Arizona

Spinal Anesthesia in a Patient with Hereditary Angioedema Due to C1 Esterase Deficiency

INTEGRATED ANESTHESIA ASSOCIATES
DEPARTMENT OF ANESTHESIA RESEARCH & DEVELOPMENT

Kodilichi V. Baldino, MD[1,2], Adam J. House, DO[1,2]; Patrick Bergamo, MD[1,2]; Dakota Balkir[2]; Kevin Finkel, MD[1,2]

[1] IAA Fellowship in Regional Anesthesia at Bone and Joint institute, Hartford CT, [2]Integrated Anesthesia Associates, Hartford, CT

Introduction

- Hereditary angioedema (HAE) is a genetic disorder characterized by recurrent episodes of edema without urticaria or pruritus[1.]
- HAE most often affects the mucosa of the respiratory/GI tracts, as well as the skin, with the most deleterious result leading to fatal obstruction due to airway/laryngeal involvement.
- Episodes may be triggered by multiple stressors, including emotional stress, physical exertion, weather, menstruation, eating surgery/procedures, and endotracheal intubation[2].
- While in most cases, the swelling from HAE is self-limited, the risk of potential airway edema leads to the suggestion that a regional anesthetic and minimal airway manipulation is likely the safest surgical option for these patients. However, the role of C1INH in regulating clotting and fibrinolytic pathways raises additional questions as to the safety of a regional anesthetic in these patients.
- This case report presents the outcome of spinal anesthesia use with minimal airway manipulation in a patient with C1INH deficiency.

Methods

This case was reviewed retrospectively, and utilizes de-identified patient health information, therefore, it is exempt from IRB review requirements as per Hartford Hospital policy. Patient provided signed consent for publication at the time of encounter

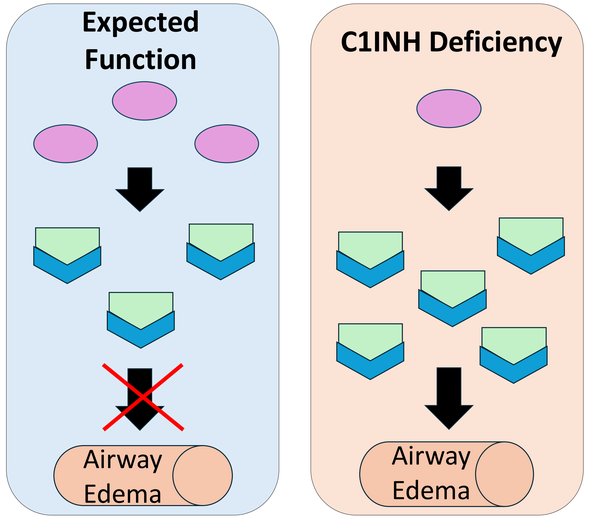

Figure 1. Expected results of proper C1INH inhibition of bradykinin levels (left) compared to uninhibited, bradykinin production, resulting in airway edema resulting from C1NH deficiency (right).

Case Report

- A 74-year-old female presenting with a femoral neck fracture, had a medical history significant for HAE due to C1 inhibitor (C1INH) deficiency. Her other relevant medical history includes bipolar disorder and depression.
- The location of surgery was determined by Cinryze (C1 esterase inhibitor) availability, with 20 U/kg of the drug being administered one hour prior to surgery.
- She received spinal anesthesia with MAC sedation; airway manipulation was minimized during surgery.
- Recovery from anesthesia and subsequent postoperative care were unremarkable.
- The patient was discharged on post-operative day 3 to a skilled nursing facility.

Discussion

The most studied and well-understood form of HAE relates to C1INH deficiency/dysfunction:

- C1INH limits bradykinin production and affects steps in intrinsic clotting and fibrinolytic pathways[1].
- Unchecked bradykinin results in the edema of HAE[1].
- Inhibition of multiple proteins is also common, including coagulation factor XII, coagulation factor XI, thrombin, plasma kallikrein, plasmin, and tissue plasminogen activator[1].

Regional anesthesia remains a safe option for patients with HAE caused by C1INH deficiency:

- C1INH promotes a thrombotic state despite coagulation and fibrinolysis effects, leaving patients more apt to venous thromboembolism than bleeding diathesis[3].
- By minimizing airway manipulation and potentially fatal airway edema/obstruction.

Preventative options exist for patients exposed to known triggers:

- Prophylaxis with C1INH supplementation (IV, SQ, and oral options)[4]. These medications are often expensive with high disposal rates, creating a need for efficient methods of acquisition.

References

1. Zuraw, B. L., & Farkas, H. (2022). Hereditary angioedema (due to C1 inhibitor deficiency): Pathogenesis and diagnosis. UpToDate. Retrieved December 22, 2025, fromhttps://www.uptodate.com/contents/hereditary-angioedema-due-to-c1-inhibito r-deficiency-pathogenesis-and-diagnosis
2. Zotter, Z., Csuka, D., Szabó, E., Czaller, I., Nébenführer, Z., Temesszentandrási, G., Fust, G., Varga, L., & Farkas, H. (2014). The influence of trigger factors on hereditary angioedema due to C1-inhibitor deficiency. Orphanet journal of rare diseases, 9, 44. https://doi.org/10.1186/1750-1172-9-44
3. Sundler Björkman, L., Pirouzifard, M. N., Grover, S. P., Egesten, A., Sundquist, J., Sundquist, K., & Zöller, B. (2024). Increased risk of venous thromboembolism in young and middle-aged individuals with hereditary angioedema: A family study. Blood, 144(4), 435–444. https://doi.org/10.1182/blood.2023022996
4. Maurer, M., Magerl, M., Betschel, S., Aberer, W., Ansotegui, I. J., Aygören-Pürsün, E., Banerji, A., Bara, N.-A., Boccon-Gibod, I., Bork, K., Bouillet, L., Boysen, H. B., Brodszki, N., Busse, P. J., Bygum, A., Caballero, T., … Craig, T. (2022). The international WAO/EAACI guideline for the management of hereditary angioedema — The 2021 revision and update. Allergy, 77(7), 1961–1990. https://doi.org/10.1111/all.15214