# Hereditary angioedema (HAE) laboratory testing and codes

## Recommended lab testing for HAE[1,2]

- Serum levels of C4
- Serum levels of antigenic and functional C1-INH

| LabCorp[a] 1-800-845-6167 | | | | |
|---|---|---|---|---|
| Test Name | Laboratory Code | CPT Code | ICD-10-CM Code | Normal Range |
| **Hereditary Angioedema (HAE) (Panel includes all tests below)** | **123020** | **86160 (x2)** | D84.1 | **See below** |
| Complement C4, Serum | 001834 | 86160 | | 13–44 mg/dL |
| Complement C1 Esterase Inhibitor, Serum | 004648 | 86160 | | 21–39 mg/dL |
| Complement C1 Esterase Inhibitor, Functional | 120220 | 86161 | | Normal: >67%<br>Equivocal: 41%–67%<br>Abnormal: <41% |
| Complement C1q, Quantitative | 016824 | 86160 | | Male: 11.8–23.8 mg/dL<br>Female: 11.8–24.4 mg/dL |

| National Jewish Health (ADx)[b] 1-800-550-6227 | | | | |
|---|---|---|---|---|
| Test Name | Laboratory Code | CPT Code | ICD-10-CM Code | Normal Range |
| C4 Level | C4 | 86160 | D84.1 | 13–52 mg/dL<br>(depending on age) |
| Ratio of C4d to C4 | C4RAT | 86160 (x2) | | Male/Female:<br>C4: 0.112–0.441 mg/mL<br>C4d: 0.52–7.88 mcg/mL<br>Ratio: <25 |
| C1-Esterase Inhibitor Level (C1-INH) | CEIQ | 86160 | | 20–37 mg/dL |
| C1-Inhibitor (C1-INH) Function, Chromogenic Assay | CEICHR | 86161 | | Units for CEICHR 74%–147% of Normal |
| C1q Level | C1Q | 86160 | | 83–125 mcg/mL |

| Quest Diagnostics[c] 1-800-222-0446 | | | | |
|---|---|---|---|---|
| Test Name | Laboratory Code | CPT Code | ICD-10-CM Code | Normal Range |
| **Angioedema Panel, Hereditary, Comprehensive (Panel includes all tests below)** | **17706** | **86160 (x2), 86161** | D84.1 | **See below** |
| Complement Component C4c | 353 | 86160 | | Adults: 16–47 mg/dL |
| C1 Esterase Inhibitor, Protein | 298 | 86160 | | 21–39 mg/dL |
| C1 Inhibitor, Functional | 297 | 86161 | | Normal: ≥68%<br>Equivocal: 41%–67%<br>Abnormal: ≤40% |
| Complement Component C1q | 981 | 86160 | | 5.0–8.6 mg/dL |

| Exsera BioLabs[d] 1-303-724-7592 | | | | |
|---|---|---|---|---|
| Test Name | Laboratory Code | CPT Code | ICD-10-CM Code | Normal Range |
| C1-INH Chromogenic Function | C1INHF | 86161 | D84.1 | 70%–142% of normal |
| Intact C4[e] | C4LMX | 86160 | | 130–342 mcg/mL |
| Total C4 | C4IM | 86160 | | 16.9–55.1 mg/dL |
| C1q Level | C1QLMX | 86160 | | 59–148 mcg/mL |

Current as of April 2019
[a]Laboratory Corporation of America® Holdings.
[b]Advanced Diagnostic Laboratories, National Jewish Health—Affiliated with the University of Colorado, Denver.
[c]Quest Diagnostics Incorporated.
[d]Exsera BioLabs—Affiliated with the University of Colorado, Aurora.
[e]Intact C4 is a novel assay that measures only uncleaved C4. Useful for patients with unexpectedly normal C4 levels.

# When to suspect HAE

**HAE should be suspected in patients with[2,3]:**

- History of recurrent angioedema without urticaria
- Positive family history
  - Present in approximately 75% of patient population[4]
- Onset of symptoms in childhood/adolescence
- Recurrent painful abdominal attacks
- Failure to respond to treatment with antihistamines, glucocorticoids, or epinephrine
- Presence of prodromes

# There is a need to improve diagnostic delays in HAE[5]

**8.4**

In a 2017 survey of 445 patients with HAE, people with HAE on average experienced a delay of 8.4 years in getting an accurate diagnosis.[5]

**47.1%**

In a 2015 survey of 106 patients with HAE Type I or II, one-fourth were diagnosed within 1 year of onset of HAE symptoms, but almost half (47.1%) experienced a delay of ≥10 years between their initial onset of symptoms and their diagnosis.[6]

Misdiagnoses may contribute to diagnostic delays and result in unnecessary treatments, potentially increasing the risk of death.[7]

**References: 1.** Banerji A. The burden of illness in patients with hereditary angioedema. *Ann Allergy Asthma Immunol.* 2013;111(5):329-336. **2.** Busse PJ, Christiansen SC, Riedl MA, et al. US HAEA Medical Advisory Board 2020 guidelines for the management of hereditary angioedema. *J Allergy Clin Immunol Pract.* 2021;9(1):132-150. doi: 10.1016/j.jaip.2020.08.046. **3.** Zuraw BL. Clinical practice. Hereditary angioedema. *N Engl J Med.* 2008;359(10):1027-1036. **4.** Altman KA, Naimi DR. Hereditary angioedema: a brief review of new developments. *Curr Med Res Opin.* 2014;30(5):923-930. **5.** Banerji A, Davis KH, Brown TM, et al. Patient-reported burden of hereditary angioedema: findings from a patient survey in the United States. *Ann Allergy Asthma Immunol.* 2020;124(6):600-607. **6.** Banerji A, Li Y, Busse P, et al. Hereditary angioedema from the patient's perspective: a follow-up patient survey. *Allergy Asthma Proc.* 2018;39(3):212-223. doi:10.2500/aap.2018.39.4123. **7.** Longhurst HJ, Bork K. Hereditary angioedema: an update on causes, manifestations and treatment. *Br J Hosp Med (Lond).* 2019;80(7):391-398. doi:10.12968/hmed.2019.80.7.391.

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