Hereditary Angioedema (HAE) Laboratory Testing and Codes

Recommended lab testing for HAE[1,2]

- —Serum C4 levels
- —C1 INH antigenic level concentration
- —C1 INH antigenic function

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% Equivocal: 41–67% Abnormal: <41%
Complement C1q, Quantitative	016824	86160		Male: 11.8–23.8 mg/dL Female: 11.8–24.4 mg/dL

National Jewish (ADx)[b] 1-303-270-2541

Test Name	Laboratory Code	CPT Code	ICD-10-CM Code	Normal Range
C4 Level	C4	86160	D84.1	11–61 mg/dL (depending on age)
Ratio of C4d to C4	C4RAT	86160 (x2)		Male/Female: C4: 0.112–0.441 mg/mL C4d: 0.52–7.88 mcg/mL Ratio: <25
C1-Esterase Inhibitor Level (C1-INH)	CEIQ	86160		20–37 mg/dL
C1-Inhibitor (C1-INH) Function, Chromogenic Assay	CEICHR	86161		
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		11–26 mg/dL
C1 Inhibitor, Functional	297	86161		Normal: ≥68% Equivocal: 41–67% Abnormal: ≤40%
Complement Component C1q	981	86160		5.0–8.6 mg/dL

Current as of March 2017
[a]Laboratory Corporation of America® Holdings
[b]Advanced Diagnostic Laboratories, National Jewish Health—Affiliated with the University of Colorado, Denver
[c]Quest Diagnostics Incorporated

When to Suspect Hereditary Angioedema (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 abdominal pain attacks
- Failure to respond to treatment with antihistamines, glucocorticoids, or epinephrine
- Presence of prodromes

HAE Is Frequently Misdiagnosed as Another Condition[1]

Up to 65% of patients with HAE may have been misdiagnosed with another condition prior to receiving their HAE diagnosis. Additionally, 19 to 24% of patients undergo unnecessary procedures as a result of misdiagnosis, including removal of appendix.[5,a]

Most common misdiagnoses[5,a]:

- 38% allergic reaction
- 17% appendicitis

[a]In a survey of 313 patients.

References: 1. Banerji A. The burden of illness in patients with hereditary angioedema. *Ann Allergy Asthma Immunol*. 2013;111(5):329-336. **2.** Craig T, Aygören-Pürsün E, Bork K, et al. WAO guideline for the management of hereditary angioedema. *World Allergy Organ J*. 2012;5(12):182-199. **3.** Zuraw BL. 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.** Lunn ML, Santos CB, Craig TJ. Is there a need for clinical guidelines in the United States for the diagnosis of hereditary angioedema and the screening of family members of affected patients? *Ann Allergy Asthma Immunol*. 2010;104(3):211-214.