Hereditary Angioedema (HAE) Laboratory Testing and Codes

1/Banerji 2013 p330, col2 ¶2, ln 1-12

2/Craig 2012 p187 col 2 Rec 1; p188 col 1 ¶1, lines 6-8

3/Kaplan 2005 p383 col 1 ¶3, lines 1-3; col 2 Table VII

Recommended lab testing for HAE[1-3]

—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		9–36 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		8.0–19.5 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		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

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

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2/Craig 2012 p187 col 2 ¶3 lines 1-3

4/Frank 2006 p655 ¶1 lines 1-3

2/Craig 2012 p187 col 2 ¶3 lines 3-8

5/Kaplan 2010 p193 col1 ¶1 lines 4-6 (calculation: 100%-25%=75%)

2/Craig 2012 p187 col 2 ¶3 lines 3-8

2/Craig 2012 p187 col 2 ¶3 lines 3-8

2/Craig 2012 p187 col 2 ¶3 lines 3-8

2/Craig 2012 p187 col 2 ¶3 lines 3-8

2/Craig 2012 p187 col 2 ¶3 lines 3-8

1/Banerji 2013 p330 col 1 ¶4 lines 1-2

6/Lunn 2010 p211 col 2 ¶2 lines 8-9

1/Banerji 2013 p330 col 1 ¶4 lines 3-5

6/Lunn 2010 p212 col 2 ¶2 lines 11-15

1/Banjeri 2013 p330 col 1 ¶4 lines 3-4

6/Lunn 2010 p212 col 2 ¶2 ln 11-15

1/Banjeri 2013 p330 col 1 ¶4 ln 3-4

4/Frank 2006 p653 ¶2 lines 8-9

7/Nzeako 2001 p2418 col 3 ¶2 lines 34-38

When to Suspect Hereditary Angioedema (HAE)

HAE should be suspected in patients with:

- ❑ History of recurrent angioedema without urticaria[2]
- ❑ Unilateral angioedema, often locally occurring attacks without symmetry[4]
- ❑ Positive family history[2]
  —Present in approximately 75% of patient population[5]
- ❑ Onset of symptoms in childhood/adolescence[2]
- ❑ Recurrent abdominal pain attacks[2]
- ❑ Occurrence of laryngeal episode[2]
- ❑ Failure to respond to treatment with antihistamines, glucocorticoids, or epinephrine[2]
- ❑ Prodromal signs or symptoms before swellings[2]

HAE Is Frequently Misdiagnosed[1,6]

Common misdiagnoses include:

- ❑ Allergic reaction[1,6]
- ❑ Appendicitis[1,6]
- ❑ Irritable bowel syndrome[1]
- ❑ Episodic abdominal pain[4]
- ❑ Psychosomatic illness[7]

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.** Kaplan AP, Greaves MW. Angioedema. *J Am Acad Dermatol.* 2005;53:373-388. **4.** Frank MM. Hereditary angioedema: the clinical syndrome and its management in the United States. *Immunol Allergy Clin North Am.* 2006;26(4):653-668. **5.** Kaplan AP, Joseph K. The bradykinin-forming cascade and its role in hereditary angioedema. *Ann Allergy Asthma Immunol.* 2010;104(3):193-204. **6.** 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. **7.** Nzeako UC, Frigas E, Tremaine WJ. Hereditary angioedema: a broad review for clinicians. *Arch Intern Med.* 2001;161:2417-2429.

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