

## Hereditary Angioedema (HAE)

Analyze C<sub>4</sub> (001834)  
Reference Interval:  
9 mg/dL–36 mg/dL

Analyze C<sub>1</sub> Esterase Inhibitor (004648)  
Reference Interval:  
21 mg/dL–39 mg/dL

Is C<sub>1</sub> Esterase Inhibitor  
less than 21 mg/dL?

No

Yes

Analyze C<sub>1</sub> Esterase Inhibitor, Functional (120220)  
Reference Interval:  
Normal: > 67%  
Equivocal: 41%–67%  
Abnormal: < 41%

Stop and Report C<sub>4</sub> and  
C<sub>1</sub> Esterase Inhibitor

**HAE Interpretation:**  
Results are consistent with  
hereditary angioedema, type 1

If C<sub>1</sub> Esterase Inhibitor, Functional  
is greater than 67%

If C<sub>1</sub> Esterase Inhibitor, Functional is  
41% to 67%

If C<sub>1</sub> Esterase Inhibitor, Functional is  
less than 41%

Stop and report C<sub>4</sub>;  
C<sub>1</sub> Esterase Inhibitor;  
and C<sub>1</sub> Esterase Inhibitor, Functional

Stop and report C<sub>4</sub>;  
C<sub>1</sub> Esterase Inhibitor;  
and C<sub>1</sub> Esterase Inhibitor, Functional

Stop and report C<sub>4</sub>;  
C<sub>1</sub> Esterase Inhibitor;  
and C<sub>1</sub> Esterase Inhibitor, Functional

**HAE Interpretation:** Results are not consistent with hereditary angioedema (type I or type II). Consider other types of angioedema. Please note that patients taking anabolic steroids may have falsely elevated C<sub>4</sub> values. Consider retesting after discontinuation of anabolic steroids or during an acute attack.

**HAE Interpretation:** Results are equivocal for hereditary angioedema type II. Interpret in conjunction with other clinical and laboratory data. Retest C<sub>1</sub> esterase inhibitor, functional during an acute attack if indicated.

**HAE Interpretation:** Results are consistent with hereditary angioedema type II.