Hereditary Angioedema (HAE)

Analyze C₄ (001834)
Reference Interval: 14 mg/dL – 44 mg/dL

Analyze C₁ Esterase Inhibitor (004648)
Reference Interval: 21 mg/dL – 39 mg/dL

Is C₁ Esterase Inhibitor less than 21 mg/dL?

No

Analyze C₁ Esterase Inhibitor, Functional (120220)
Reference Interval:
Normal: > 67%
Equivocal: 41% – 67%
Abnormal: < 41%

Yes

Stop and Report C₄ and C₁ Esterase Inhibitor

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

If C₁ Esterase Inhibitor, Functional is greater than 67%

Stop and report C₄; C₁ Esterase Inhibitor; and C₁ 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 C₄ values. Consider retesting after discontinuation of anabolic steroids or during an acute attack.

If C₁ Esterase Inhibitor, Functional is 41% to 67%

Stop and report C₄; C₁ Esterase Inhibitor; and C₁ Esterase Inhibitor, Functional

HAE Interpretation: Results are equivocal for hereditary angioedema type II. Interpret in conjunction with other clinical and laboratory data. Retest C₁ esterase inhibitor, functional during an acute attack if indicated.

If C₁ Esterase Inhibitor, Functional is less than 41%

Stop and report C₄; C₁ Esterase Inhibitor; and C₁ Esterase Inhibitor, Functional

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