

## Angioedema Panel

**Test Name:** Angioedema Panel  
**Test Code:** 401715  
**CPT Code:** 86160 x 3, 86161 x 1

**Description:**

Angioedema is characterized by a rapid onset of subcutaneous edema in various parts of the body, frequently involving the upper respiratory and gastrointestinal tract. Angioedema is a relatively common problem in differential diagnosis. Since the appropriate treatment will vary depending on the pathogenic mechanism, it is important to investigate the immunochemical basis of the angioedema symptoms. The hereditary form of angioedema should be differentiated from the acquired forms and from other related conditions such as allergic angioedema and idiopathic angioedema. The following panel of complement tests are often helpful in this diagnostic evaluation.

Tests	Method	CPT
C1 Inhibitor (Functional Activity)	EIA	86161
C1 Inhibitor (Protein Concentration)	RID	86160
C1q	RID	86160
C4	Neph	86160

**Specimen Requirement:** 3 mL of fresh serum divided into three separate 1 mL vials. Ship frozen via overnight courier.

**Units and Normal Reference Ranges:**

C1 Inhibitor (Functional Activity)	> 68% Ref
C1 INH Protein Concentration	19 - 34 mg/dL
C1q	11 - 19 mg/dL
C4	14 - 40 mg/dL

**Summary of Expected Results:**

	Hereditary (Variant)	Acquired	Other
C1Inhibitor Activity	↓	↓	N
C1INH Protein	↓	N	N or ↓
C1q	N	N	↓↓
C4	↓↓	↓↓	↓

N, Normal level of complement component.

Other, urticarias and other angioedemas

**Comments:**

Hereditary angioedema (HAE) is caused by depressed activity of the C1 Esterase Inhibitor (C1 INH), the regulator protein that binds to and dissociates activated C1. C1 INH is a protease inhibitor which regulates the enzymes of the complement, coagulation, fibrinolytic and kinin-forming enzyme systems. C1 INH inhibits Hageman factor and kallikrein, two enzyme systems involved in producing vasoactive peptides. Approximately 85% of the cases of HAE are due to diminished levels of the C1 INH protein and 15% of cases are variants resulting from the synthesis of a dysfunctional C1 INH protein. Acquired angioedemas are more often encountered clinically and may be associated with B cell lymphomas or one of the connective tissue diseases. In acquired angioedema the C1 INH levels are low because of increased consumption. In these cases the C1q levels are also often decreased. However, in HAE the C1q concentrations are usually within normal ranges.

**References:**

1. Kaplan, A.P. Heredity Angioedema. In: *Current Therapy in Allergy, Immunology and Rheumatology*. Edited by L. Lichtenstein and A. Fauci. Decker Publishers, 1988.
2. Fearon, D.T. (1983) *Journal of Allergy and Clinical Immunology*, 71: 520-529.
3. Figueroa, J. and Densen, P. (1991) *Clinical Microbiology Reviews*, 4: 359-395.

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