

Consider Hereditary Angioedema (HAE):

- Recurrent angioedema (without urticaria)
- Recurrent episodes of abdominal pain and vomiting
- Laryngeal edema
- Positive family history of angioedema

Measure: serum complement factor 4 (C4),  
C1 inhibitor (C1-INH) antigenic protein  
C1 inhibitor (C1-INH) functional level if available

C4 and C1-INH protein  
quantities decreased

Confirm decreased  
C4 and C1-INH protein  
by second measurement

Family history  
of angioedema

**HAE-  
C1INH-  
Type I**

Earlier age of  
onset  
and C1q  
normal

No family  
history of  
angioedema

Measure C1q and  
consider age of  
onset of symptoms

Later Age of  
onset  
and/or low C1q

Consider  
**Acquired  
Angioedema**

C4 quantity low but  
C1-INH protein normal  
or elevated

Determine C1-INH  
function and repeat C4  
and C1-INH protein levels

C1-INH  
function  
normal

Consider  
other  
non-HAE  
causes of C4  
consumption

C1-INH  
function  
decreased

**HAE-  
C1INH-  
Type II**

C4, C1-INH protein  
normal

Confirm C4, C1-INH  
normal during attack

Consider angioedema (AE)  
types other than HAE-C1-  
INH types I and II

**AE from Medications**  
Eg. ACE inhibitors

**HAE Type III**  
- **HAE-FXII**  
- **HAE-unknown**