

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**