C1-INH Deficiency Testing Algorithm

INDICATIONS FOR TESTING
- Recurrent angioedema (without urticaria)
- Recurrent episodes of abdominal pain and vomiting
- Laryngeal edema
- Family history of angioedema

ORDER
C4 and C1-INH testing, including quantity and function

C4 and C1-INH protein quantities normal

C4 quantity low, but C1-INH protein normal or elevated

C4 and C1-INH protein quantities decreased

Determine C1-INH function

Confirm C4 and C1-INH are normal during attack

C1-INH function normal

C1-INH function decreased

Consider angioedema types other than HAE types I and II:
- HAE type IV (familial)
- Medication related (eg, due to ACE inhibitors)
- Estrogen related
- Factor XII related
- Other

Angioedema due to C1-INH deficiency

Confirm by second measurement of C4 and C1-INH quantity and function

Family history of angioedema

No family history of angioedema

Measure C1q

Earlier age of onset and/or C1q normal

Later age of onset and/or low C1q

Hereditary angioedema

Acquired angioedema

Abbreviations

ACE	Angiotensin-converting enzyme
C1-INH	Complement component 1 esterase inhibitor
C1q	Complement component 1q
C4	Complement component 4
HAE	Hereditary angioedema