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
- Confirm C4 and C1-INH are normal during attack
- Consider angioedema types other than HAE types I and II:
  - HAE type IV (familial)
  - Medication related (e.g., due to ACE inhibitors)
  - Estrogen related
  - Factor XII related
  - Other

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

C4 and C1-INH protein quantities decreased
- 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