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

Measure: serum C4,
C1-INH antigenic protein,
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 C1-INH type I
      - Earlier age of onset and C1q normal
        - Consider acquired angioedema
    - Measure C1q and consider age of onset of symptoms
      - Later age of onset and/or low C1q
        - Consider other non-HAE causes of C4 consumption

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
    - Measure C1q and consider age of onset of symptoms
      - Earlier age of onset and C1q normal
        - Consider acquired angioedema
    - Later age of onset and/or low C1q
      - Consider other non-HAE causes of C4 consumption
  - C1-INH function decreased
    - HAE C1-INH Type II

C4, C1-INH protein normal
- Confirm C4, C1-INH normal during attack
  - Consider angioedema types other than HAE C1-INH types I and II
    - Angioedema from medications (eg, ACE inhibitors)
      - HAE type III
        - HAE-FXII
        - HAE-unknown