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
      - Consider Acquired Angioedema
  - **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
    - **HAE-C1INH-Type II**
    - Consider Acquired Angioedema
  - **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