Factor XIII Deficiency Testing

Factor XIII (FXIII) deficiency is a rare bleeding disorder that affects the final stage of blood coagulation and may lead to heavy bleeding. FXIII deficiency may be acquired or inherited. Diagnosis is difficult because many of the tests typically used to test coagulation (eg, prothrombin time [PT], activated partial thromboplastin time [aPTT], thrombin time, platelet count, bleeding time) return normal results in cases of FXIII deficiency. Specific testing for FXIII presence and activity can detect FXIII deficiency in individuals with a bleeding disorder who have normal screening test results.[1,2]

Tests to Consider

Factor XIII Activity 2006182

Method: Chromogenic Assay

- Preferred first-line test to diagnose inherited or acquired FXIII deficiency
- Appropriate for evaluation of individuals with a bleeding disorder who present with normal PT, PTT, and platelet count test results
- Monitor therapy in individuals being treated for FXIII deficiency
- Confirm abnormalities identified in the qualitative FXIII assay (clot solubility test)

Factor XIII, Qualitative, with Reflex to Factor XIII 1:1 Mix 2002819

Method: Qualitative Solubility

- Most useful if severe FXIII deficiency is suspected (<1% of normal activity)
- Distinguish between FXIII deficiency and a FXIII inhibitor
- Abnormal results should be confirmed with quantitative testing

Test Description

Factor XIII Activity

- Chromogenic assay
- Measures consumption of NADPH spectrophotometrically
  - Decrease in absorbance is directly proportional to FXIII activity
- Can quantitate factor activity as low as 5% of normal

Factor XIII, Qualitative, with Reflex to Factor XIII 1:1 Mix

- Qualitative solubility test
- If clot lysis occurs in the initial testing, factor XIII 1:1 mix added

Disease Overview

Incidence

~1/1-3 million (inherited)[1]

Inheritance

Autosomal recessive (when inherited and not acquired)[1]

Visit the Uncommon Factor Deficiencies Consult topic for additional information about characteristics of acquired and inherited FXIII deficiency.

Test Interpretation

Results

Factor XIII Activity

- Reference interval: FXIII activity, 69-143%
- Severe bleeding usually does not occur until FXIII level <1-3%
- Mild or moderate deficiencies may be associated with increased bleeding risk in some cases

- Deficient: clot lysis was observed in the qualitative factor XIII screening test but not when the test was repeated using a 1:1 mix of patient plasma and pooled normal plasma
- Clot lysis only occurs in samples with <1% of normal activity

Limitations

Factor XIII Activity

- A low FXIII level does not distinguish deficiency from a low value due to FXIII autoantibodies
- False-negative (artificially increased) results may be caused by lipemic plasma
- False-positive (artificially decreased) results may be caused by icteric plasma

Factor XIII, Qualitative, with Reflex to Factor XIII 1:1 Mix

- Normal results do not exclude deficiency
- Does not identify:
  - Mild or moderate deficiency
  - Heterozygous carriers for FXIII deficiency
  - Treated, yet deficient, individuals
  - Individuals with weak FXIII inhibitors that do not decrease FXIII activity to <1% of normal
- False-positive results may be caused by:
  - Heparin
  - Decreased or abnormal fibrinogen
  - Increased fibrinolysis
  - Fibrinolytic drugs

References

1. Karimi M, Peyvandi F, Naderi M, et al. Factor XIII deficiency diagnosis: challenges and tools. Int J Lab Hematol. 2018;40(1):3-11. PubMed

2. Kohler HP, Ichinose A, Seitz R, et al. Diagnosis and classification of factor XIII deficiencies. J Thromb Haemost. 2011;9(7):1404-1406. PubMed

Related Information

Uncommon Factor Deficiencies

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