F13B gene

coagulation factor XIII B chain

Normal Function

The *F13B* gene provides instructions for making one part, the B subunit, of a protein called factor XIII. This protein is part of a group of related proteins called coagulation factors that are essential for normal blood clotting. They work together as part of the coagulation cascade, which is a series of chemical reactions that forms blood clots in response to injury. After an injury, clots seal off blood vessels to stop bleeding and trigger blood vessel repair. Factor XIII acts at the end of the cascade to strengthen and stabilize newly formed clots, preventing further blood loss.

Factor XIII in the bloodstream is made of two A subunits (produced from the *F13A1* gene) and two B subunits (produced from the *F13B* gene). The role of the B subunits is to carry and stabilize the A subunits, protecting them from being broken down. When a new blood clot forms, the A and B subunits separate from one another, and the A subunits are cut (cleaved) to produce the active form of factor XIII (factor XIIIa). The active protein links together molecules of fibrin, the material that forms the clot, which strengthens the clot and keeps other molecules from breaking it down.

Studies suggest that factor XIII has additional functions, although these are less well understood than its role in blood clotting. Specifically, factor XIII is likely involved in other aspects of wound healing, immune system function, maintaining pregnancy, bone formation, and the growth of new blood vessels (angiogenesis).

Health Conditions Related to Genetic Changes

Factor XIII deficiency

At least 17 mutations in the *F13B* gene have been found to cause inherited factor XIII deficiency, a rare bleeding disorder. Without treatment, affected individuals have a greatly increased risk of abnormal bleeding episodes, including life-threatening bleeding inside the skull (intracranial hemorrhage). *F13B* gene mutations severely reduce the amount or disrupt the function of the B subunit of factor XIII, preventing it from stabilizing and protecting the A subunit. The resulting loss of factor XIII activity weakens new blood clots and prevents them from stopping blood loss effectively.

Age-related macular degeneration

MedlinePlus Genetics provides information about Age-related macular degeneration

Other Names for This Gene

- coagulation factor XIII B chain precursor
- coagulation factor XIII, B polypeptide
- fibrin-stabilizing factor B subunit
- FXIIIB
- protein-glutamine gamma-glutamyltransferase B chain
- TGase
- transglutaminase B chain

Additional Information & Resources

Tests Listed in the Genetic Testing Registry

- Tests of F13B (https://www.ncbi.nlm.nih.gov/gtr/all/tests/?term=2165[geneid])

Scientific Articles on PubMed

- PubMed (https://pubmed.ncbi.nlm.nih.gov/?term=%28F13B%5BTIAB%5D%29+OR+%28FXIIIB%5BTIAB%5D%29+OR+%28%28coagulation+factor+XIII%5BTIAB%5D%29+AND+%28beta%5BTIAB%5D%29%29+OR+%28%28coagulation+factor+XIII%5BTIAB%5D%29+AND+%28B%5BTIAB%5D%29%29+AND+english%5Bla%5D+AND+human%5Bmh%5D+AND+%22last+1800+days%22%5Bdp%5D)

Catalog of Genes and Diseases from OMIM

- FACTOR XIII, B SUBUNIT; F13B (https://omim.org/entry/134580)

Gene and Variant Databases

- NCBI Gene (https://www.ncbi.nlm.nih.gov/gene/2165)
- ClinVar (https://www.ncbi.nlm.nih.gov/clinvar?term=F13B[gene])

References

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Genomic Location

The *F13B* gene is found on chromosome 1 (https://medlineplus.gov/genetics/chromosome/1/).

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