abcam

Product datasheet

Recombinant Human GGCX protein ab114658

1 Image

Description

Product name	Recombinant Human GGCX protein
Expression system	Wheat germ
Accession	P38435
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	ADFPGLHLENFVSEDLGNTSIQLLQGEVTVELVAEQKNQT LREGEKMQLP AGEYHKVYTTSPSPSCYMYVYVNTTELALEQDLAYLQELK EKVENGS
Predicted molecular weight	36 kDa including tags
Amino acids	533 to 629

Specifications

Our Abpromise guarantee covers the use of **ab114658** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Western blot SDS-PAGE ELISA
Form	Liquid

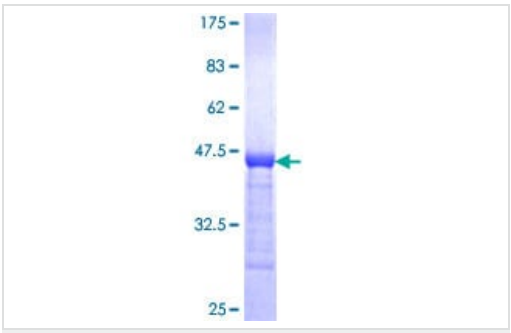
Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl

General Info

Function	Mediates the vitamin K-dependent carboxylation of glutamate residues to calcium-binding gamma-carboxyglutamate (Gla) residues with the concomitant conversion of the reduced hydroquinone form of vitamin K to vitamin K epoxide.
Involvement in disease	Defects in GGCX are a cause of combined deficiency of vitamin K-dependent clotting factors type 1 (VKCFD1) [MIM:277450]; also known as multiple coagulation factor deficiency III (MCFD3). VKCFD leads to a bleeding tendency that is usually reversed by oral administration of vitamin K. Defects in GGCX are the cause of pseudoxanthoma elasticum-like disorder with multiple coagulation factor deficiency (PXEL-MCFD) [MIM:610842]. This syndrome is characterized by hyperlaxity of the skin involving the entire body. Important phenotypic differences with classical PXE include much more severe skin laxity with spreading toward the trunk and limbs with thick, leathery skin folds rather than confinement to flexural areas, and no decrease in visual acuity. Moreover, detailed electron microscopic analyzes revealed that alterations of elastic fibers as well as their mineralization are slightly different from those in classic PXE.
Sequence similarities	Belongs to the vitamin K-dependent gamma-carboxylase family.
Cellular localization	Endoplasmic reticulum membrane.

Images



SDS-PAGE - Recombinant Human GGCX protein (ab114658)

SDS-PAGE analysis of ab114658 on a 12.5% gel stained with Coomassie Blue.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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