

Product datasheet

Recombinant Human GGCX protein ab114658

画像数 1

製品の概要

製品名	Recombinant Human GGCX protein
タンパク質長	Protein fragment

製品の詳細

由来	Recombinant
由来	Wheat germ
アミノ酸配列	
アクセッション番号	P38435
生物種	Human
配列	ADFPGLHLENFVSEDLGNTSIQLLQGEVTVELVAEQKNQTLREGEKMLP AGEYHKVYTTSPSPSCYMYVYVNTTELALAEQDLAYLQELKEKVENG
分子量	36 kDa including tags
領域	533 to 629

特性

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.

アプリケーション	Western blot SDS-PAGE ELISA
製品の状態	Liquid
備考	Protein concentration is above or equal to 0.05 mg/ml. Best used within three months from the date of receipt.

前処理および保存

保存方法および安定性	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00
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Constituents: 0.3% Glutathione, 0.79% Tris HCl

関連情報

機能

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.

関連疾患

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.

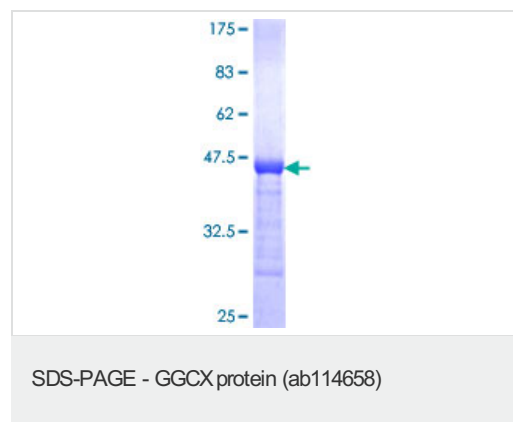
配列類似性

Belongs to the vitamin K-dependent gamma-carboxylase family.

細胞内局在

Endoplasmic reticulum membrane.

画像



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

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