

Recombinant Human GGCX protein ab114658

1 图像

描述		
产品名称	重组人GGCX蛋白	
表达系统	Wheat germ	
Accession	<b>P38435</b>	
蛋白长度	Protein fragment	
无动物成分	No	
性质	Recombinant	
种属	Human	
序列	ADFPGLHLENFVSEDLGNTSIQLLQGEVTVELVAEQKNQTLR EGEKMQLP AGEYHKVYTTSPSPSCYMYVYVNTTELALQDLAYLQELKEK VENG	
预测分子量	36 kDa including tags	
氨基酸	533 to 629	

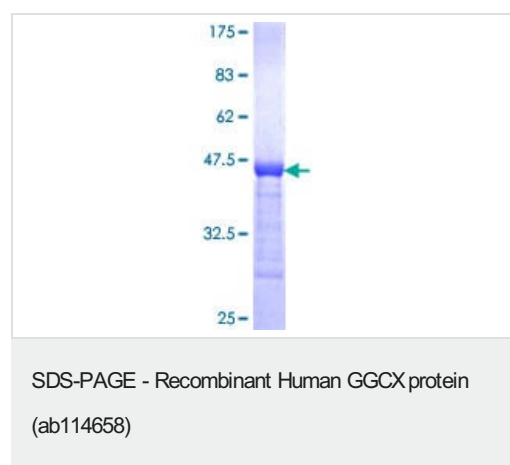
技术指标		
Our <b>Abpromise guarantee</b> covers the use of <b>ab114658</b> in the following tested applications.		
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.		
应用	Western blot  SDS-PAGE  ELISA	
形式	Liquid	

制备和贮存		
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.  pH: 8.00 Constituents: 0.3% Glutathione, 0.79% Tris HCl	

常规信息		
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功能	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.
疾病相关	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.
序列相似性	Belongs to the vitamin K-dependent gamma-carboxylase family.
细胞定位	Endoplasmic reticulum membrane.

## 图片



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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