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GGCX Rabbit pAb

Catalog # AP98117

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P38435
Other Accession	P38435
Reactivity	Mouse
Predicted	Rat, Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	87561
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GGCX
Epitope Specificity	351-450/758
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the vitamin K-dependent gamma-carboxylase family.
SUBUNIT	Monomer. May interact with CALU.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes an enzyme which catalyzes the posttranslational modification of vitamin K-dependent protein. Many of these vitamin K-dependent proteins are involved in coagulation so the function of the encoded enzyme is essential for hemostasis. Mutations in this gene are associated with vitamin K-dependent coagulation defect and PXE-like disorder with multiple coagulation factor deficiency. Multiple transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, Nov 2008]

Additional Information

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Gene ID	
Other Names	VKCFD1; VKGC_HUMAN; GGCX; Gamma-glutamyl carboxylase; Peptidyl-glutamate 4-carboxylase; Vitamin K gamma glutamyl carboxylase; 4.1.1.90; GC; vitamin K-dependent gamma-carboxylase
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	GGCX
Synonyms	GC
Function	Mediates the vitamin K-dependent carboxylation of glutamate residues to calcium-binding gamma-carboxyglutamate (Gla) residues with the concomitant epoxidation of vitamin K hydroquinone to vitamin K epoxide (PubMed:17073445, PubMed:39880952, PubMed:39880037). Couples epoxidation and carboxylation in the same active site (PubMed:39880952, PubMed:39880037). Catalyzes gamma-carboxylation of blood coagulation factors (F2, F7, F9 and F10) and anticoagulants such as PROC, which are essential for thrombosis(PubMed:17073445, PubMed:39880037). Catalyzes gamma-carboxylation of osteocalcin/BGLAP, which is essential for bone metabolism (PubMed:17073445, PubMed:39880952). Catalyzes gamma-carboxylation of matrix Gla protein (MGP) and other transmembrane gamma-Gla proteins such as PRRG2, which are essential for calcium homeostasis and haemostasis (PubMed:17073445, PubMed:39880037).
Cellular Location	Endoplasmic reticulum membrane; Multi-pass membrane protein

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