

Rabbit Anti-GGCX antibody

SL10530R

Product Name:	GGCX
Chinese Name:	γ-谷氨酰羧化酶抗体
Alias:	Gamma glutamyl carboxylase; Gamma-glutamyl carboxylase; GC antibody GGCX; Peptidyl glutamate 4 carboxylase; Peptidyl-glutamate 4-carboxylase; Vitamin K dependent gamma carboxylase; Vitamin K gamma glutamyl carboxylase; Vitamin K- dependent gamma-carboxylase; VKCFD 1; VKCFD1; VKGC HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100- 500 (Paraffin sections need antigen repair)
	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	87kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human GGCX:351-450/758
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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]

Function:

Mediates the vitamin K-dependent carboxylation of glutamate residues to calciumbinding gamma-carboxyglutamate (Gla) residues with the concomitant conversion of the reduced hydroquinone form of vitamin K to vitamin K epoxide.

Subunit: Monomer. May interact with CALU.

Subcellular Location: Endoplasmic reticulum membrane.

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.

Similarity:

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

SWISS:

P38435

Gene ID: 2677

Database links:

Entrez Gene: 2677 Human

<u>Omim: 137167</u> Human

SwissProt: P38435 Human

	Unigene: 77719 Human
	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	
	Paraformaldehyde-fixed, paraffin embedded (mouse brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (GGCX) Polyclonal Antibody, Unconjugated (SL10530R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.