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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.
Research Area	Cardiovascular Cell biology
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Mouse, (predicted: Human, Rat,)
Applications	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 （Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	87kDa
Cellular localization	cytoplasmic The cell membrane
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human GGCX: 351-450/758
Lsotype	IgG
Purification	affinity purified by Protein A
Buffer Solution	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Shipped at 4℃. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Attention	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

PubMed PubMed

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]

Product Detail

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.

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:

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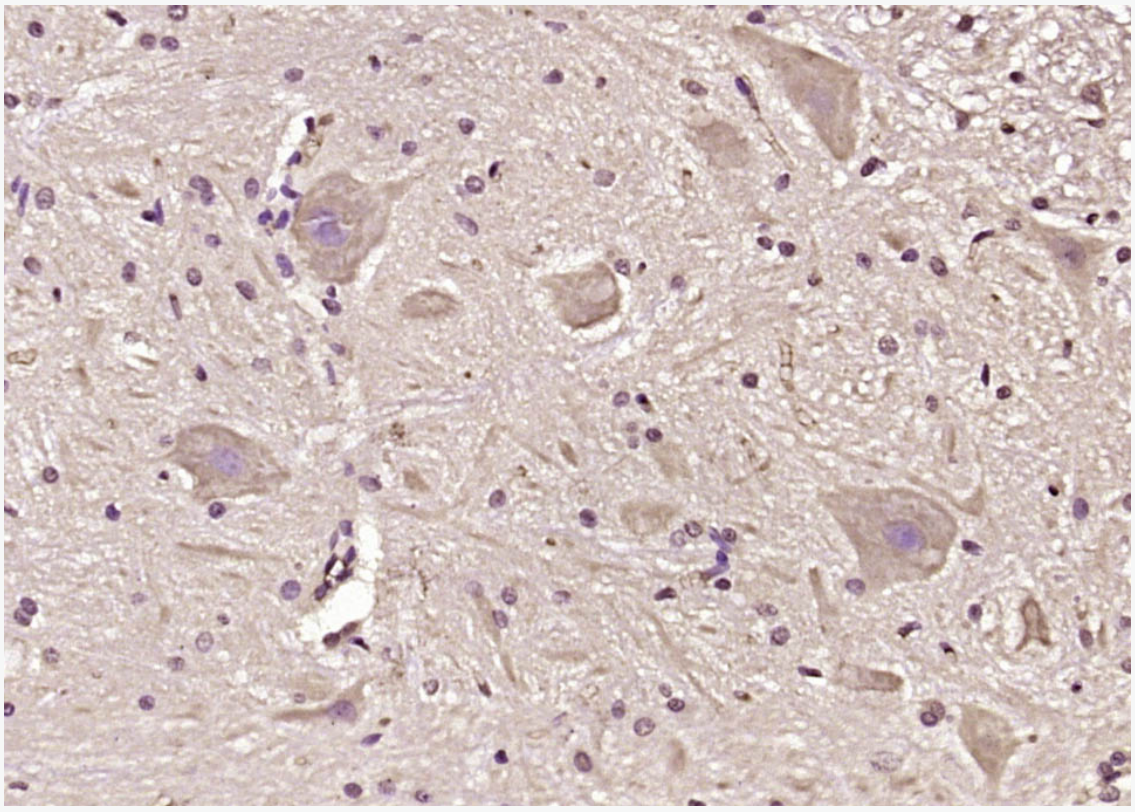
Entrez Gene: 2677 Human

Omim: 137167 Human

SwissProt: P38435 Human

Unigene: 77719 Human

Product 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.