abcam

Product datasheet

Anti-GGCX antibody ab106924

1 Image

Overview

Product name	Anti-GGCX antibody
Description	Goat polyclonal to GGCX
Host species	Goat
Specificity	This antibody is expected to recognize both isoforms (NP_000812.2; NP_001135741.1).
Tested applications	**Suitable for:** WB
Species reactivity	**Reacts with:** Human **Predicted to work with:** Dog, Orangutan
Immunogen	Synthetic peptide: C-PPESNPDPVHSE , corresponding to C terminal amino acids 746-757 of Human GGCX isoform 1 (NP_000812.2) or amino acids 689-700 of isoform 2 (NP_001135741.1). Run BLAST with Run BLAST with
Positive control	Human Muscle lysates.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
Storage buffer	Preservative: 0.02% Sodium Azide Constituents: 0.5% BSA, Tris buffered saline, pH 7.3
Purity	Immunogen affinity purified
Purification notes	Purified from goat serum by ammonium sulphate precipitation followed by antigen affinity chromatography using the immunizing peptide.
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab106924** in the following tested applications.

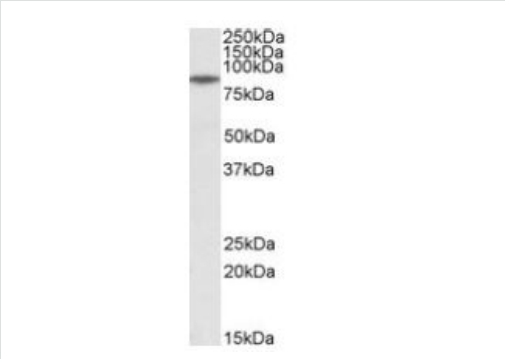
The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
WB		Use a concentration of 0.3 - 1 µg/ml. Predicted molecular weight: 88 kDa.

Target

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.
Involvement in 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.
Sequence similarities	Belongs to the vitamin K-dependent gamma-carboxylase family.
Cellular localization	Endoplasmic reticulum membrane.

Images



Western blot - Anti-GGCX antibody (ab106924)

Anti-GGCX antibody (ab106924) at 0.3 µg/ml (for 1 hour) + Human Muscle lysate in RIPA buffer at 35 µg

Developed using the ECL technique.

Predicted band size: 88 kDa

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