abcam

Product datasheet

Anti-Rapsyn antibody ab118491

1 Image

Overview

Product name	Anti-Rapsyn antibody
Description	Rabbit polyclonal to Rapsyn
Tested applications	**Suitable for:** WB, ELISA, IHC-P
Species reactivity	**Reacts with:** Human **Predicted to work with:** Mouse, Rat
Immunogen	Synthetic peptide corresponding to Human Rapsyn aa 399-412 (C terminal) conjugated to Keyhole Limpet Haemocyanin (KLH).
Positive control	Human brain (cortex) tissue.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term.
Storage buffer	Preservative: 0.01% Sodium azide Constituents: 50% Glycerol, 49% PBS
Purity	Protein A purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab118491** 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 1 µg/ml. Predicted molecular weight: 46 kDa.
ELISA		Use at an assay dependent concentration.

Application	Abreviews	Notes
IHC-P		Use a concentration of 10 - 20 µg/ml. Perform heat mediated antigen retrieval with citrate buffer pH 6 before commencing with IHC staining protocol.

Target

Function

Thought to play some role in anchoring or stabilizing the nicotinic acetylcholine receptor at synaptic sites. It may link the receptor to the underlying postsynaptic cytoskeleton, possibly by direct association with actin or spectrin.

Involvement in disease

Defects in RAPSN are a cause of congenital myasthenic syndrome with acetylcholine receptor deficiency (ACHRDCMS) [MIM:608931]. ACHRDCMS is a post-synaptic congenital myasthenic syndrome. Congenital myasthenic syndromes (CMS) are inherited disorders of neuromuscular transmission that stem from mutations in presynaptic, synaptic, or postsynaptic proteins. Postsynaptic disorders result from mutations in proteins forming the subunits of the muscle acetylcholine receptor (AChR). The kinetic abnormalities of AChR result in either prolonged ion channel activations that underlie 'slow-channel myasthenic syndromes' (SCCMS) or abbreviated channel activations that underlie the abnormally rapid decay of endplate currents in 'fast-channel syndromes' (FCCMS). ACHRDCMS is the third disorder associated with postsynaptic CMS which could result from mutations in the proteins forming the muscle AChR. Mutations underlying AChR deficiency cause a 'loss of function' and show recessive inheritance.
Defects in RAPSN are the cause of fetal akinesia deformation sequence (FADS) [MIM:208150]; also known as Pena-Shokeir syndrome type 1 or fetal akinesia sequence or arthrogryposis multiplex congenita with pulmonary hypoplasia. FADS is a rare condition characterized by decreased intrauterine fetal movement, congenital limb contractures, pulmonary hypoplasia, polyhydramnios and craniofacial abnormalities.

Sequence similarities

Belongs to the RAPsyn family.
Contains 1 RING-type zinc finger.
Contains 7 TPR repeats.

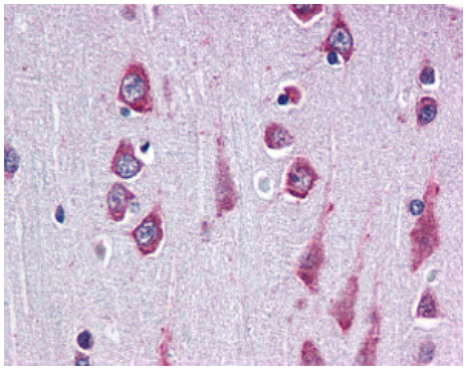
Domain

A cysteine-rich region homologous to part of the regulatory domain of protein kinase C may be important in interactions of this protein with the lipid bilayer.

Cellular localization

Cell membrane. Cell junction > synapse > postsynaptic cell membrane. Cytoplasm > cytoskeleton. Cytoplasmic surface of postsynaptic membranes.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Rapsyn antibody (ab118491)

ab118491 at 20µg/ml staining Rapsyn in Formalin-fixed, Paraffin-embedded Human brain (cortex) tissue by Immunohistochemistry, followed by biotinylated secondary antibody, alkaline phosphatase-streptavidin and chromogen.

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