abcam

Product datasheet

Anti-CHRND antibody ab26095

1 References 2 Images

Overview

Product name	Anti-CHRND antibody
Description	Rabbit polyclonal to CHRND
Tested applications	**Suitable for:** ELISA, WB, IHC-P
Species reactivity	**Reacts with:** Human **Predicted to work with:** Rat, Cow, Dog
Immunogen	Synthetic peptide derived from a region between residues 6-55 of human Acetylcholine Receptor Protein delta Precursor. (the amino acid sequence is considered to be commercially sensitive)
Positive control	Jurkat cell lysate (WB), Human Kidney (IHC)

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: None Constituents: 2% Sucrose, PBS
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab26095** 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
ELISA		1/62500.

Application	Abreviews	Notes
WB		Use a concentration of 1 µg/ml. Predicted molecular weight: 59 kDa. Good results were obtained when blocked with 5% non-fat dry milk in 0.05% PBS-T.
IHC-P		Use at an assay dependent concentration.

Target

Function

After binding acetylcholine, the AChR responds by an extensive change in conformation that affects all subunits and leads to opening of an ion-conducting channel across the plasma membrane.

Involvement in disease

Defects in CHRND are a cause of multiple pterygium syndrome lethal type (MUPSL) [MIM:253290]. Multiple pterygia are found infrequently in children with arthrogryposis and in fetuses with fetal akinesia syndrome. In lethal multiple pterygium syndrome there is intrauterine growth retardation, multiple pterygia, and flexion contractures causing severe arthrogryposis and fetal akinesia. Subcutaneous edema can be severe, causing fetal hydrops with cystic hygroma and lung hypoplasia. Oligohydramnios and facial anomalies are frequent.
Defects in CHRND are a cause of congenital myasthenic syndrome slow-channel type (SCCMS) [MIM:601462]. SCCMS is the most common congenital myasthenic syndrome. Congenital myasthenic syndromes are characterized by muscle weakness affecting the axial and limb muscles (with hypotonia in early-onset forms), the ocular muscles (leading to ptosis and ophthalmoplegia), and the facial and bulbar musculature (affecting sucking and swallowing, and leading to dysphonia). The symptoms fluctuate and worsen with physical effort. SCCMS is caused by kinetic abnormalities of the AChR, resulting in prolonged endplate currents and prolonged AChR channel opening episodes.
Defects in CHRND are a cause of congenital myasthenic syndrome fast-channel type (FCCMS) [MIM:608930]. FCCMS is a congenital myasthenic syndrome characterized by kinetic abnormalities of the AChR. In most cases, FCCMS is due to mutations that decrease activity of the AChR by slowing the rate of opening of the receptor channel, speeding the rate of closure of the channel, or decreasing the number of openings of the channel during ACh occupancy. The result is failure to achieve threshold depolarization of the endplate and consequent failure to fire an action potential.

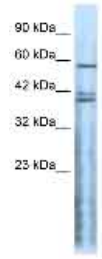
Sequence similarities

Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1) subfamily. Delta/CHRND sub-subfamily.

Cellular localization

Cell junction > synapse > postsynaptic cell membrane. Cell membrane.

Images



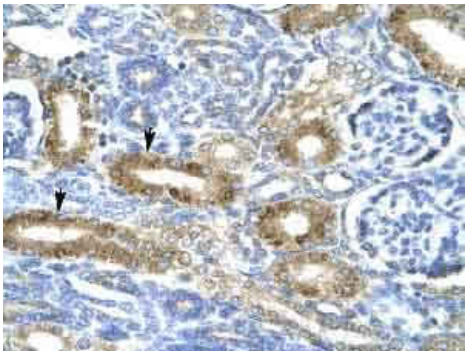
Western blot - Acetylcholine Receptor Protein delta Precursor antibody (ab26095)

Anti-CHRND antibody (ab26095) (at 1 ug/ml in 5% skim milk / PBS buffer) + Jurkat cell lysate 10ug.

Secondary

HRP conjugated anti Rabbit IgG diluted in 1/50,000 - 1/100,000.

Predicted band size : 59 kDa



Immunohistochemistry (Paraffin-embedded sections) - Acetylcholine Receptor Protein delta Precursor antibody (ab26095)

Ab26095 at a concentration of 4-8ug/ml staining paraffin embedded human kidney tissue by Immunohistochemistry. The arrows indicate the epithelial cells of renal tubule. Magnification: x400.

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