abcam

Product datasheet

Anti-Nicotinic Acetylcholine Receptor alpha 1/CHRNA1 antibody [mAb 192] ab288434

Recombinant

1 References 3 Images

Overview

Product name Anti-Nicotinic Acetylcholine Receptor alpha 1/CHRNA1 antibody [mAb 192]

Description Rat monoclonal [mAb 192] to Nicotinic Acetylcholine Receptor alpha 1/CHRNA1

Host species Rat

Specificity The antibody binds to human muscle AChR with a Kd of 10 pM, to mouse muscle AChR with a Kd

of 50 pM, and to rat muscle AChR with a Kd of 646 nM

Tested applications Suitable for: ICC/IF, WB

Species reactivity Reacts with: Mouse, Human

Immunogen Recombinant full length protein.

Database link: P02708

Positive control WB: Mouse skeletal muscle lysate ICC/IF: HeLa cells

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. Avoid freeze / thaw cycle.

Storage buffer Preservative: 0.02% Proclin 300

Constituent: 99% PBS

Purity Protein A purified

ClonalityMonoclonalClone numbermAb 192IsotypeIgG2bLight chain typekappa

Applications

The Abpromise guarantee Our Abpromise guarantee covers the use of ab288434 in the following tested applications.

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

Application	Abreviews	Notes
ICC/IF		Use a concentration of 10 µg/ml.
WB		Use a concentration of 0.03 µg/ml. Predicted molecular weight: 54 kDa.

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.

Tissue specificity

Isoform 1 is only expressed in skeletal muscle. Isoform 2 is constitutively expressed in skeletal muscle, brain, heart, kidney, liver, lung and thymus.

Involvement in disease

Defects in CHRNA1 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.

Note=The alpha subunit is the main focus for antibody binding in myasthenia gravis. Myasthenia gravis is characterized by sporadic muscular fatigability and weakness, occurring chiefly in muscles innervated by cranial nerves, and characteristically improved by cholinesterase-inhibiting drugs.

Defects in CHRNA1 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 CHRNA1 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.

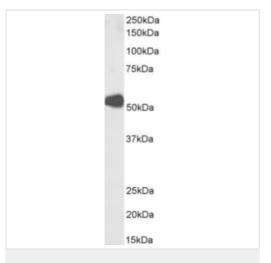
Sequence similarities

Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1) subfamily. Alpha-1/CHRNA1 sub-subfamily.

Cellular localization

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

Images



Western blot - Anti-Nicotinic Acetylcholine Receptor alpha 1/CHRNA1 antibody [mAb 192] (ab288434)

Anti-Nicotinic Acetylcholine Receptor alpha 1/CHRNA1 antibody [mAb 192] (ab288434) at 0.03 μ g/ml + Mouse Skeletal Muscle lysate at 35 μ g

Secondary

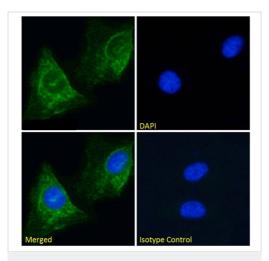
anti-mouse IgG1 secondary antibody

Predicted band size: 54 kDa **Observed band size:** 58.9 kDa

The image data was generated using the chimeric mouse version of the same antibody clone

Run on a 10% SDS PAGE gel.

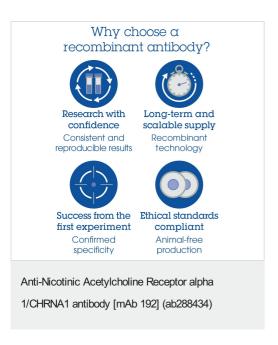
ab288434 successfully detected AChR in mouse skeletal muscle tissue lysate. Though this antibody is targetted to the human AChR it also reacts with mouse AChR.



Immunocytochemistry/ Immunofluorescence - Anti-Nicotinic Acetylcholine Receptor alpha 1/CHRNA1 antibody [mAb 192] (ab288434) The image data was generated using the chimeric mouse version of the same antibody clone

Immunofluorescence analysis of paraformaldehyde fixed HeLa cells, stained for muscle acetylcholine receptor using ab288434 at $10\mu g/ml$ for 1 hr. Followed by Alexa Fluor® 488 secondary antibody $(2 \mu g/ml)$, showing positive cytoplasmic staining.

The nuclear stain is DAPI (blue). Panels show from left-right, top-bottom ab288434, DAPI, merged channels and an isotype control. The isotype control was stained with an unknown specificity antibody (ab288434) followed by Alexa Fluor® 488 secondary antibody.



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