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CHRNE Rabbit pAb

Catalog # AP97952

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	Q04844
Other Accession	Q04844
Predicted	Rat, Human, Mouse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	54697
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CHRNE
Epitope Specificity	111-210/492
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1) subfamily. Epsilon/CHRNE sub-subfamily.
SUBUNIT	Pentamer of two alpha chains, and one each of the beta, delta, and gamma (in immature muscle) or epsilon (in mature muscle) chains.
DISEASE	Note=The muscle AChR is the major target antigen in the autoimmune disease 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 CHRNE 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 CHRNE 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. Defects in CHRNE are a cause of congenital myasthenic syndrome with acetylcholine receptor deficiency (CMS-ACHRD) [MIM:608931]. CMS-ACHRD is a postsynaptic congenital myasthenic syndrome. Mutations underlying AChR deficiency cause a 'loss of function' and show recessive inheritance.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions	Acetylcholine receptors at mature mammalian neuromuscular junctions are pentameric protein complexes composed of four subunits in the ratio of two alpha subunits to one beta, one epsilon, and one delta subunit. The acetylcholine receptor changes subunit composition shortly after birth when the epsilon subunit replaces the gamma subunit seen in embryonic receptors. Mutations in the epsilon subunit are associated with congenital myasthenic syndrome. [provided by RefSeq, Sep 2009]

Additional Information

Gene ID	1145
Other Names	ACHRE; CMS1A1; CMS1D; CMS1E; CMS2A; CMS4A; CMS4B; CMS4C; FCCMS; FIM1; FIMG; FIMG1; MGI; SCCMS; AChrepsilon; Acre; nAChRE; nAChR; EGK_08058; ACHE_BOVIN; CHRNE; ACHE_HUMAN; ACHE_MOUSE; ACHE_RAT; cholinergic receptor nicotinic epsilon subunit; cholinergic receptor, nicotinic, epsilon; cholinergic receptor, nicotinic, epsilon (muscle); cholinergic receptor, nicotinic epsilon; acetylcholine receptor, nicotinic, epsilon (muscle)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

Protein Information

Name	CHRNE (HGNC:1966)
Synonyms	ACHRE
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.
Cellular Location	Postsynaptic cell membrane; Multi-pass membrane protein. Cell membrane; Multi-pass membrane protein

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