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CHRNB Rabbit pAb

Catalog # AP97951

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P11230
Other Accession	P11230
Predicted	Rat, Pig, Human, Mouse, Rabbit, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	56698
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human CHRNB
Epitope Specificity	1-100/501
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. Beta-1/CHRNB1 sub-subfamily.
DISEASE	Defects in CHRNB1 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 CHRNB1 are a cause of congenital myasthenic syndrome with acetylcholine receptor deficiency (ACHRDCMS) [MIM:608931]. ACHRDCMS is a post-synaptic 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	The muscle acetylcholine receptor is composed of five subunits: two alpha subunits and one beta, one gamma, and one delta subunit. This gene encodes the beta subunit of the acetylcholine receptor. The acetylcholine receptor changes conformation upon acetylcholine binding leading to the opening of an ion-conducting channel across the plasma membrane. Mutations in this gene are associated with slow-channel congenital myasthenic syndrome. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	1140

Other Names	ACHRB; CHRNB; CMS1D; CMS2A; CMS2C; SCCMS; Achr-2; Acrb; RNACRB1; nAChR; ACHB_BOVIN; CHRNB1; ACHB_HUMAN; ACHB_MOUSE; ACHB_RAT; cholinergic receptor nicotinic beta 1 subunit; cholinergic receptor, nicotinic, beta polypeptide 1 (muscle); cholinergic receptor, nicotinic, beta 1 (muscle); cholinergic receptor, nicotinic beta 1; acetylcholine receptor, nicotinic, beta 1 (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	CHRNB1 (HGNC:1961)
Synonyms	ACHRB, CHRNB
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

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.