

Product name:	AChR β 1 Rabbit Polyclonal Antibody
Cat number:	ABN06501
Conjugate:	Unconjugated
Size:	100 μ L
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	The antiserum was produced against synthesized peptide derived from human CHRNB1. AA range:41-90
Reactivity:	Human,Mouse,Rat
Applications:	WB 1:500-1:2000,ELISA 1:5000-1:10000
Molecular Weight:	55kDa
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.

Background:

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],disease: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.,disease:Defects in CHRNA1 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.,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.,similarity:Belongs to the ligand-gated ionic channel (TC 1.A.9) family.,subunit:Pentamer of two alpha chains, and one each of the beta, delta, and gamma (in immature muscle) or epsilon (in mature muscle) chains.,