烟碱型乙酰胆碱受体ε抗体

产品货号： mlR19244

英文名称： CHRNE

中文名称： 烟碱型乙酰胆碱受体ε抗体

别　　名： Nicotinic Acetylcholine Receptor epsilon; Acetylcholine receptor subunit epsilon; ACHE_HUMAN; AchR epsilon subunit; ACHRE; Cholinergic receptor, nicotinic, epsilon polypeptide; Chrne; CMS1D; CMS1E; CMS2A; FCCMS; SCCMS.

研究领域： 细胞生物　神经生物学　细胞膜受体

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat,

产品应用： ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 ICC=1:100-500 IF=1:100-500 （石蜡切片需做抗原修复）
not yet tested in other applications.
optimal dilutions/concentrations should be determined by the end user.

分 子 量： 53kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human CHRNE:111-210/492 <Extracellular>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. 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.

PubMed： PubMed

产品介绍： 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]

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.

Subunit:

Pentamer of two alpha chains, and one each of the beta, delta, and gamma (in immature muscle) or epsilon (in mature muscle) chains.

Subcellular Location:

Cell junction; synapse; postsynaptic cell membrane. Cell membrane.

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.

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.

SWISS:

Q04844

Gene ID:

1145

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.