

乙酰胆碱酯酶相关胶原蛋白多肽抗体

产品货号: mIR10932

英文名称: COLQ

中文名称: 乙酰胆碱酯酶相关胶原蛋白多肽抗体

别 名: Acetylcholinesterase-associated collagen; AChE Q subunit; asymmetric acetylcholinesterase; Collagen-like tail subunit (single strand of homotrimer) of asymmetric acetylcholinesterase; Colq; COLQ_HUMAN; EAD; OTTHUMP00000209566; OTTHUMP00000209567; single strand of homotrimeric collagen-like tail subunit of asymmetric acetylcholinesterase.

研究领域: 神经生物学 信号转导 细胞骨架

抗体来源: Rabbit

克隆类型: Polyclonal

交叉反应: Human, Mouse, Rat, Pig, Horse, Rabbit,

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: 45kDa

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 COLQ:301-400/455

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 $^{\circ}$ 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 $^{\circ}$ 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 $^{\circ}$ C.

PubMed: PubMed

产品介绍: This gene encodes the subunit of a collagen-like molecule associated with acetylcholinesterase in skeletal muscle. Each molecule is composed of three identical subunits. Each subunit contains a proline-rich attachment domain (PRAD) that binds an acetylcholinesterase tetramer to anchor the catalytic subunit of the enzyme to the basal lamina. Mutations in this gene are associated with endplate acetylcholinesterase deficiency. Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Jul 2008]

Function:

Anchors the catalytic subunits of asymmetric AChE to the synaptic basal lamina.

Subcellular Location:

Cell junction; synapse.

Tissue Specificity:

Found at the end plate of skeletal muscle.



Post-translational modifications:

The triple-helical tail is stabilized by disulfide bonds at each end.

DISEASE:

Defects in COLQ are the cause of congenital myasthenic syndrome Engel type (CMSE) [MIM:603034]; also known as end-plate acetylcholinesterase deficiency or congenital myasthenic syndrome type IC (CMS-IC). CMSE is a rare autosomal recessive congenital myasthenic syndrome characterized by onset during childhood, generalized weakness, abnormal fatigability on exertion, refrectoriness to acetylcholinesterase drugs, decremental electromyographic response and morphological abnormalities of the neuromuscular junctions.

Similarity:

Belongs to the COLQ family.

Contains 2 collagen-like domains.

SWISS:

Q9Y215

Gene ID:

8292

Important Note:

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