Anti-CHRNE Antibody [5F11G8]

EM1711-01

Product Type:	Mouse monoclonal IgG1, primary antibodies
Species reactivity:	Human, Rat
Applications:	WB, FC
Molecular Wt:	54.7kDa
Clone number:	5F11G8

Description: Acetylcholine receptor subunit epsilon is a protein that in humans is encoded by the CHRNE gene. 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 achetylcholine 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. Congenital myasthenic syndrome (CMS) is associated with genetic defects that affect proteins of the neuromuscular junction. Postsynaptic defects are the most frequent cause of CMS and often result in abnormalities in the acetylcholine receptor (AChR). The majority of mutations causing CMS are found in the AChR subunits genes. Out of all mutations associated with CMS, more than half are mutations in one of the four genes encoding the adult AChR subunits. Mutations of the AChR often result in endplate deficiency. The most common AChR gene mutation that underlies CMS is the mutation of the CHRNE gene. The CHRNE gene codes for the epsilon subunit of the AChR. Most mutations are autosomal recessive loss-of-function mutations and as a result there is endplate AChR deficiency. CHRNE is associated with changing the kinetic properties of the AChR.

Immunogen: Purified recombinant fragment of human CHRNE (AA: extra 21-239) expressed in E. Coli.

Positive control: C6 cell SK-N-SH cells 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.

Subcellular location: Cell junction, synapse, postsynaptic cell membrane. Cell membrane.

Database links: SwissProt: Q04844 Human

Recommended Dilutions:

WB	1:500-1:2,000
FC	1:100-1:200

Storage Buffer: Purified antibody in PBS with 0.05% sodium azide.

Storage Instruction: 4℃; -20℃ for long term storage.

Purity: Protein G affinity purified.

Images

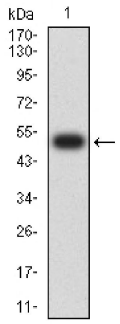

Fig1: Western blot analysis of CHRNE against human CHRNE (AA: extra 21-239) recombinant protein. Proteins were transferred to a PVDF membrane and blocked with 5% BSA in PBS for 1 hour at room temperature. The primary antibody (EM1711-01, 1/500) was used in 5% BSA at room temperature for 2 hours. Goat Anti-Mouse IgG - HRP Secondary Antibody at 1:5,000 dilution was used for 1 hour at room temperature.

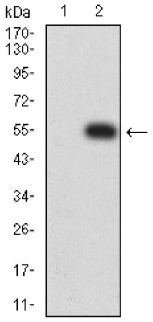

Fig2: Western blot analysis of EM1711-01 against HEK293 (1) and CHRNE (AA: extra 21-239)-hIgGFc transfected HEK293 (2) cell lysate.Proteins were transferred to a PVDF membrane and blocked with 5% BSA in PBS for 1 hour at room temperature. The primary antibody (EM1711-01, 1/500) was used in 5% BSA at room temperature for 2 hours. Goat Anti-Mouse IgG - HRP Secondary Antibody at 1:5,000 dilution was used for 1 hour at room temperature.

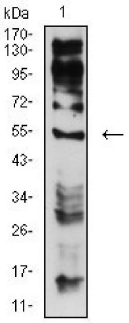

Fig3: Western blot analysis of EM1711-01 against C6 (1) cell lysate.Proteins were transferred to a PVDF membrane and blocked with 5% BSA in PBS for 1 hour at room temperature. The primary antibody (EM1711-01, 1/500) was used in 5% BSA at room temperature for 2 hours. Goat Anti-Mouse IgG - HRP Secondary Antibody at 1:5,000 dilution was used for 1 hour at room temperature.

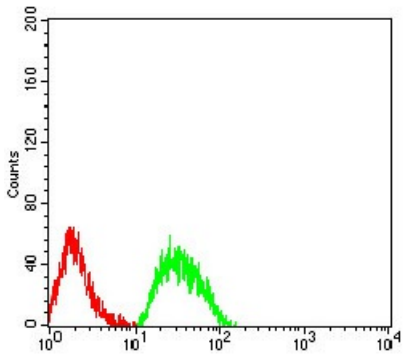

Fig4: Flow cytometric analysis of CHRNE was done on SK-N-SH cells. The cells were fixed, permeabilized and stained with the primary antibody (EM1711-01, 1/100) (green). After incubation of the primary antibody at room temperature for an hour, the cells were stained with a Alexa Fluor 488-conjugated goat anti-Mouse IgG Secondary antibody at 1/500 dilution for 30 minutes. Unlabelled sample was used as a control (cells without incubation with primary antibody; red).

Background References

1. J Neuroophthalmol. 2011 Mar;31(1):42-7.
2. Neurology. 2008 Dec 9;71(24):1967-72.