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Rabbit Anti-CHRND antibody

SL12114R

Product Name:	CHRND
Chinese Name:	烟碱型乙酰胆碱受体δ/AChRδ抗体
Alias:	Acetylcholine receptor delta subunit; Acetylcholine receptor subunit delta; ACHD_HUMAN; ACHRD; Cholinergic receptor, nicotinic, delta polypeptide; CHRND; CMS2A; FCCMS; Nicotinic acetylcholine receptor delta polypeptide precursor; SCCMS.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	57kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human CHRND:145-190/517<Extracellular>
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	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
Product Detail:	Members of the ligand-gated ion channel receptor family are characterized by their fast transmitting response to neurotransmitters. Two important members of this family are the nicotinic acetylcholine and glutamate receptors, both of which are composed of five

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	homologous subunits forming a transmembrane aqueous pore. These transmembrane receptors change conformation in response to their cognate neurotransmitter. Nicotinic acetylcholine receptors (AChRs) are found at the postsynaptic membrane of the neuromuscular junction and bind acetylcholine molecules, allowing ions to move through the pore. Glutamate receptors are found in the postsynaptic membrane of cells in the central nervous system. The activity that is generated at the synapse by the binding of acetylcholine is terminated by acetylcholinesterase, an enzyme that rapidly hydrolyzes acetylcholine. AChR delta, also known as CMS2A, FCCMS, SCCMS or CHRND, is a 517 amino acid multi-pass membrane protein that is associated with lethal type multiple pterygium syndrome, congenital myasthenic syndrome slow-channel type (SCCMS) and congenital myasthenic syndrome fast-channel type (FCCMS). **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; Multi-pass membrane protein. Cell membrane; Multi-pass membrane protein. **DISEASE:** Defects in CHRND are a cause of multiple pterygium syndrome lethal type (MUPSL) [MIM:253290]. Multiple pterygia are found infrequently in children with arthrogryposis and in fetuses with fetal akinesia syndrome. In lethal multiple pterygium syndrome there is intrauterine growth retardation, multiple pterygia, and flexion contractures causing severe arthrogryposis and fetal akinesia. Subcutaneous edema can be severe, causing fetal hydrops with cystic hygroma and lung hypoplasia. Oligohydramnios and facial anomalies are frequent. Defects in CHRND 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 CHRND 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,

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	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. **Similarity:** Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1) subfamily. Delta/CHRND sub-subfamily. **SWISS:** Q07001 **Gene ID:** 1144 **Database links:** Entrez Gene: 281687Cow Entrez Gene: 1144Human Entrez Gene: 54240Rat SwissProt: Q07001Human SwissProt: P25110Rat Unigene: 156289Human Unigene: 41469Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

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