10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

# CHRNG Rabbit pAb

Catalog # AP97953

## Product Information

| | |
|---|---|
| **Application** | WB, IHC-P, IHC-F, IF |
| **Primary Accession** | P07510 |
| **Other Accession** | P07510 |
| **Predicted** | Rat, Pig, Human, Mouse, Rabbit, Dog, Horse |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Calculated MW** | 57883 |
| **Physical State** | Liquid |
| **Immunogen** | KLH conjugated synthetic peptide derived from human CHRNG |
| **Epitope Specificity** | 151-250/517 |
| **Isotype** | IgG |
| **Purity** | affinity purified by Protein A |
| **Buffer** | 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol. |
| **SIMILARITY** | Belongs to the ligand-gated ion channel (TC 1.A.9) family. Acetylcholine receptor (TC 1.A.9.1) subfamily. Gamma/CHRNG sub-subfamily. |
| **DISEASE** | Defects in CHRNG 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 CHRNG are a cause of multiple pterygium syndrome Escobar variant (MUPSE) [MIM:265000]; also known as nonlethal type multiple pterygium syndrome. Escobar syndrome is a non-lethal form of arthrogryposis multiplex congenita. It is an autosomal recessive condition characterized by excessive webbing (pterygia), congenital contractures (arthrogryposis), and scoliosis. Variable other features include intrauterine death, congenital respiratory distress, short stature, faciocranial dysmorphism, ptosis, low-set ears, arachnodactyly and cryptorchism in males. Congenital contractures are common and may be caused by reduced fetal movements at sensitive times of development. Possible causes of decreased fetal mobility include space constraints such as oligohydramnion, drugs, metabolic conditions or neuromuscular disorders including myasthenia gravis. is a. |
| **Important Note** | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Background Descriptions** | The mammalian muscle-type acetylcholine receptor is a transmembrane pentameric glycoprotein with two alpha subunits, one beta, one delta, and one epsilon (in adult skeletal muscle) or gamma (in fetal and denervated muscle) subunit. This gene, which encodes the gamma subunit, is expressed prior to the thirty-third week of gestation in humans. The gamma subunit of the acetylcholine receptor plays a role in neuromuscular organogenesis and ligand binding and disruption of gamma subunit expression prevents the correct localization of the receptor in cell membranes. Mutations in this gene |

cause Escobar syndrome and a lethal form of multiple pterygium syndrome. Muscle-type acetylcholine receptor is the major antigen in the autoimmune disease myasthenia gravis.[provided by RefSeq, Sep 2009]

## Additional Information

| | |
|---|---|
| **Gene ID** | 1146 |
| **Other Names** | ACHRG; ACHG_HUMAN; CHRNG; cholinergic receptor nicotinic gamma subunit; cholinergic receptor, nicotinic, gamma; cholinergic receptor, nicotinic, gamma (muscle); cholinergic receptor, nicotinic gamma; acetylcholine receptor, nicotinic, gamma (muscle) |
| **Dilution** | WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500 |
| **Storage** | Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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. |

## Protein Information

| | |
|---|---|
| **Name** | CHRNG ( HGNC:1967) |
| **Synonyms** | ACHRG |
| **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. |
| **Cellular Location** | Postsynaptic cell membrane; Multi-pass membrane protein. Cell membrane; Multi-pass membrane protein |

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.