



Rabbit Anti-Ryanodine Receptor antibody

SL6305R

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| Product Name: | Ryanodine Receptor |
| Chinese Name: | 心肌兰尼碱受体抗体(脑肌兰尼碱受体) |
| Alias: | Arrhythmogenic right ventricular dysplasia 2; ARVC 2; ARVC2; ARVD 2; ARVD2; Brain ryanodine receptor calcium release channel; Brain type ryanodine receptor; Cardiac muscle ryanodine receptor; Cardiac muscle ryanodine receptor calcium release channel; Cardiac muscle type ryanodine receptor; CCO; Central core disease of muscle; HBRR; hRYR 2; hRYR2; MHS; MHS1; Ryanodine receptor 1 (skeletal); Ryanodine receptor 1; Ryanodine receptor 2 (cardiac); Ryanodine receptor 2; Ryanodine receptor 3; Ryanodine receptor type1; RYDR; RYR 1; RYR 2; RYR 3; RYR; RYR1; RYR2; RYR3; Sarcoplasmic reticulum calcium release channel; Skeletal muscle calcium release channel; Skeletal muscle ryanodine receptor; Skeletal muscle type ryanodine receptor; SKRR; Type 1 like ryanodine receptor; VTSIP. |
| Organism Species: | Rabbit |
| Clonality: | Polyclonal |
| React Species: | Human,Mouse,Rat,Dog,Pig,Cow,Rabbit, |
| Applications: | ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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: | 566kDa |
| Cellular localization: | cytoplasmicThe cell membrane |
| Form: | Lyophilized or Liquid |
| Concentration: | 1mg/ml |
| immunogen: | KLH conjugated synthetic peptide derived from human Ryanodine Receptor:4701-4800/5038 |
| 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 |

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| | 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: | <p>The Ryanodine Receptor (RyR) is the channel responsible for calcium release from muscle cell Sarcoplasmic Reticulum (SR) and also plays a role in calcium regulation in non-muscle cells. The RyR exists as a homotetramer and is predicted to have a short cytoplasmic C-terminus and 4-10 transmembrane domains. The remainder of the protein, termed the "foot" region, is located in the cytoplasm between the transverse tubule and the SR. Mammalian RyR isoforms are the product of three different genes: RyR-1 is expressed predominantly in skeletal muscle and areas of the brain; RyR-2 is expressed predominantly in heart muscle but also found in the stomach, endothelial cells and diffuse areas of the brain; and RyR-3 is found in smooth muscle and the brain (striatum, thalamus and hippocampus). In non-mammalian vertebrates, the RyR isoforms are termed alpha, beta and cardiac which correlate loosely to the mammalian RyR-1, RyR-3 and RyR-2 isoforms respectively.</p> <p>Function: Calcium channel that mediates the release of Ca(2+) from the sarcoplasmic reticulum into the cytoplasm and thereby plays a key role in triggering muscle contraction following depolarization of T-tubules. Repeated very high-level exercise increases the open probability of the channel and leads to Ca(2+) leaking into the cytoplasm. Can also mediate the release of Ca(2+) from intracellular stores in neurons, and may thereby promote prolonged Ca(2+) signaling in the brain. Required for normal embryonic development of muscle fibers and skeletal muscle. Required for normal heart morphogenesis, skin development and ossification during embryogenesis (By similarity).</p> <p>Subunit: Homotetramer. Can also form heterotetramers with RYR2. Interacts with CALM; CALM with bound calcium inhibits the RYR1 channel activity. Interacts with S100A1. Interacts with FKBP1A; this stabilizes the closed conformation of the channel. Interacts with CACNA1S; interaction with CACNA1S is important for activation of the RYR1 channel. Interacts with CACNB1. Interacts with TRDN and ASPH; these interactions stimulate RYR1 channel activity (By similarity). Identified in a complex composed of RYR1, PDE4D, PKA, FKBP1A and protein phosphatase 1 (PP1). Repeated very high-level exercise decreases interaction with PDE4D and protein phosphatase 1 (PP1).</p> <p>Subcellular Location: Sarcoplasmic reticulum membrane; Multi-pass membrane protein (Probable). Membrane; Multi-pass membrane protein. Microsome membrane; Multi-pass membrane protein.</p> <p>Tissue Specificity: Brain, skeletal muscle, placenta and possibly liver and kidney. In brain, highest levels are found in the cerebellum, hippocampus, caudate nucleus and amygdala, with lower levels in the corpus callosum, substantia nigra and thalamus.</p> |

Post-translational modifications:

Channel activity is modulated by phosphorylation. Phosphorylation at Ser-2843 may increase channel activity. Repeated very high-level exercise increases phosphorylation at Ser-2843. [PTM] Activated by reversible S-nitrosylation. Repeated very high-level exercise increases S-nitrosylation.

DISEASE:

Malignant hyperthermia 1 (MHS1) [MIM:145600]: Autosomal dominant pharmacogenetic disorder of skeletal muscle and is one of the main causes of death due to anesthesia. In susceptible people, an MH episode can be triggered by all commonly used inhalational anesthetics such as halothane and by depolarizing muscle relaxants such as succinylcholine. The clinical features of the myopathy are hyperthermia, accelerated muscle metabolism, contractures, metabolic acidosis, tachycardia and death, if not treated with the postsynaptic muscle relaxant, dantrolene. Susceptibility to MH can be determined with the 'in vitro' contracture test (IVCT): observing the magnitude of contractures induced in strips of muscle tissue by caffeine alone and halothane alone. Patients with normal response are MH normal (MHN), those with abnormal response to caffeine alone or halothane alone are MH equivocal (MHE(C) and MHE(H) respectively). Note=The disease is caused by mutations affecting the gene represented in this entry.

Central core disease of muscle (CCD) [MIM:117000]: Autosomal dominant congenital myopathy, but a severe autosomal recessive form also exists. Both clinical and histological variability is observed. Affected individuals typically display hypotonia and proximal muscle weakness in infancy, leading to the delay of motor milestones. The clinical course of the disorder is usually slow or nonprogressive in adulthood, and the severity of the symptoms may vary from normal to significant muscle weakness. Microscopic examination of CCD-affected skeletal muscle reveals a predominance of type I fibers containing amorphous-looking areas (cores) that do not stain with oxidative and phosphorylase histochemical techniques. Note=The disease is caused by mutations affecting the gene represented in this entry.

Multiminicore disease with external ophthalmoplegia (MMDO) [MIM:255320]: Clinically heterogeneous neuromuscular disorder. General features include neonatal hypotonia, delayed motor development, and generalized muscle weakness and amyotrophy, which may progress slowly or remain stable. Muscle biopsy shows multiple, poorly circumscribed, short areas of sarcomere disorganization and mitochondria depletion (areas termed minicores) in most muscle fibers. Typically, no dystrophic signs, such as muscle fiber necrosis or regeneration or significant endomysial fibrosis, are present in multiminicore disease. Note=The disease is caused by mutations affecting the gene represented in this entry.

Congenital myopathy with fiber-type disproportion (CFTD) [MIM:255310]: Genetically heterogeneous disorder in which there is relative hypotrophy of type 1 muscle fibers compared to type 2 fibers on skeletal muscle biopsy. However, these findings are not specific and can be found in many different myopathic and neuropathic conditions. Note=The disease is caused by mutations affecting the gene represented in this entry. Note=Defects in RYR1 may be a cause of Samaritan myopathy, a congenital myopathy with benign course. Patients display severe hypotonia and respiratory distress at birth.

Unlike other congenital myopathies, the health status constantly improves and patients are minimally affected at adulthood.

Similarity:

Belongs to the ryanodine receptor (TC 1.A.3.1) family. RYR3 subfamily.

Contains 3 B30.2/SPRY domains.

Contains 5 MIR domains.

SWISS:

P21817, Q92736, Q15413

Gene ID:

6261

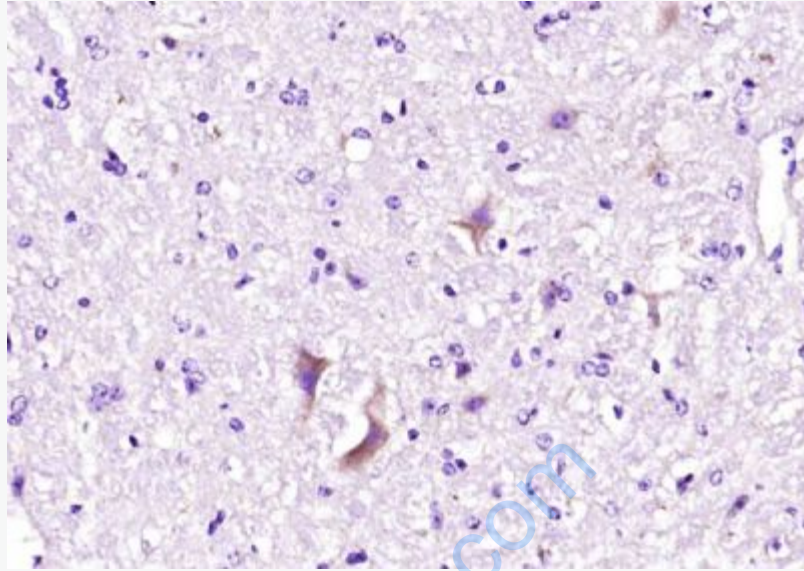
Database links:

UniProtKB/Swiss-Prot: P21817.3

Important Note:

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

Defects in the RYR2 gene are the cause of several heart diseases: 1) familial arrhythmogenic right ventricular dysplasia 2 (ARVD2), also known as arrhythmogenic right ventricular cardiomyopathy 2 (ARVC2), 2) an autosomal dominant form of stress-induced polymorphic ventricular tachycardia (VTSIP), also known as catecholaminergic polymorphic ventricular tachycardia (CPVT) and 3) familial polymorphic ventricular tachycardia (FPVT). Ryanodine Receptor 2 contains several phosphorylatable sites. Specifically, Ser-2030 and Ser-2809 (or at Ser-2808 depending on the species) can be phosphorylated by protein kinase A (PKA) and Ser-2815 (or at Ser-2814 depending on the species) can be phosphorylated by CaMKII (Ca²⁺/calmodulin-dependent protein kinase II).



Picture:

Paraformaldehyde-fixed, paraffin embedded (mouse cerebellum tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Ryanodine Receptor) Polyclonal Antibody, Unconjugated (SL6305R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.