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Ryanodine Receptor Rabbit pAb

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Catalog # AP58415

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P21817
Reactivity	Mouse
Predicted	Rat, Pig, Dog, Human, Rabbit
Host	Rabbit
Clonality	Polyclonal
Calculated MW	565176
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Ryanodine Receptor
Epitope Specificity	4701-4800/5038
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Sarcoplasmic reticulum membrane; Multi-pass membrane protein (Probable). Membrane; Multi-pass membrane protein. Microsome membrane; Multi-pass membrane protein.
DISEASE	Malignant hyperthermia 1 (MHS1) [MIM:145600]: Autosomaldominant pharmacogenetic disorder of skeletal muscle and is one ofthe main causes of death due to anesthesia. In susceptible people,an MH episode can be triggered by all commonly used inhalationalanesthetics such as halothane and by depolarizing muscle relaxantssuch as succinylcholine. The clinical features of the myopathy arehyperthermia, accelerated muscle metabolism, contractures,metabolic acidosis, tachycardia and death, if not treated with thepostsynaptic muscle relaxant, dantrolene. Susceptibility to MH canbe determined with the 'in vitro' contracture test (IVCT):observing the magnitude of contractures induced in strips of muscletissue by caffeine alone and halothane alone. Patients with normalresponse are MH normal (MHN), those with abnormal response tocaffeine alone or halothane alone are MH equivocal (MHE(C) andMHE(H) respectively). Note=The disease is caused by mutationsaffecting the gene represented in this entry.Central core disease of muscle (CCD) [MIM:117000]:Autosomal dominant congenital myopathy, but a severe autosomalrecessive form also exists. Both clinical and histologicalvariability is observed. Affected individuals typically displayhypotonia and proximal muscle weakness in infancy, leading to thedelay of motor milestones. The clinical course of the disorder isusually slow or nonprogressive in adulthood, and the severity ofthe symptoms may vary from normal to significant muscle weakness.Microscopic examination of CCD-affected skeletal muscle reveals apredominance of type I fibers containing amorphous-looking areas(cores) that do not stain with oxidative and phosphorylasehistochemical techniques. Note=The disease is caused by mutationsaffecting the gene represented in this entry. Multiminicore disease with external ophthalmoplegia(MMDO) [MIM:255320]: Clinically heterogeneous neuromusculardisorder. General features include neonatal

	hypotonia, delayedmotor development, and generalized muscle weakness and amyotrophy,which may progress slowly or remain stable. Muscle biopsy showsmultiple, poorly circumscribed, short areas of sarcomeredisorganization and mitochondria depletion (areas termed minicores)in most muscle fibers. Typically, no dystrophic signs, such asmuscle fiber necrosis or regeneration or significant endomysialfibrosis, are present in multiminicore disease. Note=The disease iscaused by mutations affecting the gene represented in this entry. Congenital myopathy with fiber-type disproportion (CFTD)[MIM:255310]: Genetically heterogeneous disorder in which there isrelative hypotrophy of type 1 muscle fibers compared to type 2fibers on skeletal muscle biopsy. However, these findings are notspecific and can be found in many different myopathic andneuropathic conditions. Note=The disease is caused by mutationsaffecting the gene represented in this entry. Note=Defects in RYR1 may be a cause of Samaritanmyopathy, a congenital myopathy with benign course. Patientsdisplay severe hypotonia and respiratory distress at birth. Unlikeother congenital myopathies, the health status constantly improvesand patients are minimally affected at adulthood.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	6261
Other Names	Ryanodine receptor 1, RYR-1, RyR1, Skeletal muscle calcium release channel, Skeletal muscle ryanodine receptor, Skeletal muscle-type ryanodine receptor, Type 1 ryanodine receptor, RYR1 (HGNC:10483), RYDR
Dilution	IHC-P=1:100-500,IHC-F=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

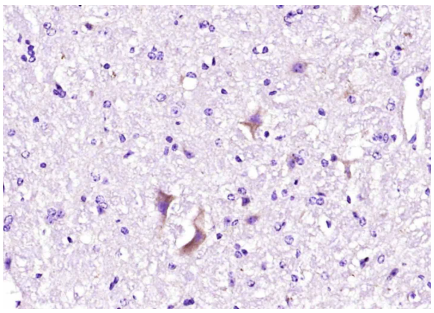
Name	RYR1 (HGNC:10483)
Synonyms	RYDR
Function	Cytosolic calcium-activated calcium channel that mediates the release of

	Ca(2+) from the sarcoplasmic reticulum into the cytosol and thereby plays a key role in triggering muscle contraction following depolarization of T-tubules (PubMed:11741831, PubMed:16163667, PubMed:18268335, PubMed:18650434, PubMed:26115329). Repeated very high- level exercise increases the open probability of the channel and leads to Ca(2+) leaking into the cytoplasm (PubMed:18268335). 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).
Cellular Location	Sarcoplasmic reticulum membrane; Multi-pass membrane protein. Note=The number of predicted transmembrane domains varies between orthologs. Both N-terminus and C-terminus are cytoplasmic. {ECO:0000250|UniProtKB:P11716}
Tissue Location	Skeletal muscle and brain (cerebellum and hippocampus).

Background

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Images



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 (AP58415) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.

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