



Rabbit Anti-Junctophilin-2 antibody

SL0702R

Product Name:	Junctophilin-2
Chinese Name:	亲联蛋白2抗体
Alias:	FLJ40969; JP-2; JP2; JPH2; JPH2_HUMAN; Junctophilin 2; Junctophilin type 2; Junctophilin-2; OTTHUMP00000031651; OTTHUMP00000031652.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Cow,Rabbit,
Applications:	WB=1:500-2000ELISA=1:500-1000 not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	74kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Junctophilin-2:351-450/696<Cytoplasmic>
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:	Junctional complexes between the plasma membrane and endoplasmic/sarcoplasmic reticulum are a common feature of all excitable cell types and mediate cross talk between cell surface and intracellular ion channels. The protein encoded by this gene is a component of junctional complexes and is composed of a C-terminal hydrophobic segment spanning the endoplasmic/sarcoplasmic reticulum membrane and a remaining cytoplasmic domain that shows specific affinity for the plasma membrane. This gene is

a member of the junctophilin gene family. Alternative splicing has been observed at this locus and two variants encoding distinct isoforms are described. [provided by RefSeq, Jul 2008].

Function:

Junctophilins contribute to the formation of junctional membrane complexes (JMCs) which link the plasma membrane with the endoplasmic or sarcoplasmic reticulum in excitable cells. Provides a structural foundation for functional cross-talk between the cell surface and intracellular calcium release channels. JPH2 is necessary for proper intracellular Ca(2+) signaling in cardiac myocytes via its involvement in ryanodine receptor-mediated calcium ion release. Contributes to the construction of skeletal muscle triad junctions.

Subcellular Location:

Cell membrane; Peripheral membrane protein. Endoplasmic reticulum membrane; Single-pass type IV membrane protein. Sarcoplasmic reticulum membrane; Single-pass type IV membrane protein. Note=Localized predominantly on the plasma membrane. The transmembrane domain is anchored in endoplasmic/sarcoplasmic reticulum membrane, while the N-terminal part associates with the plasma membrane. In heart cells, it predominantly associates along Z lines within myocytes. In skeletal muscle, it is specifically localized at the junction of A and I bands.

Tissue Specificity:

Specifically expressed in skeletal muscle and heart.

Post-translational modifications:

Phosphorylation on Ser-165, probably by PKC, affects RYR1-mediated calcium ion release, interaction with TRPC3, and skeletal muscle myotubule development.

DISEASE:

Defects in JPH2 are the cause of familial hypertrophic cardiomyopathy type 17 (CMH17) [MIM:613873]. CMH17 is a hereditary heart disorder characterized by ventricular hypertrophy, which is usually asymmetric and often involves the interventricular septum. The symptoms include dyspnea, syncope, collapse, palpitations, and chest pain. They can be readily provoked by exercise. The disorder has inter- and intrafamilial variability ranging from benign to malignant forms with high risk of cardiac failure and sudden cardiac death.

Similarity:

Belongs to the junctophilin family.
Contains 8 MORN repeats.

SWISS:

Q9BR39

Gene ID:

57158

Database links:

[Entrez Gene: 57158](#) Human

[Entrez Gene: 59091](#) Mouse

[Entrez Gene: 296345](#) Rat

[Omim: 605267](#) Human

[SwissProt: Q9BR39](#) Human

[SwissProt: Q9ET78](#) Mouse

[SwissProt: Q2PS20](#) Rat

[Unigene: 441737](#) Human

[Unigene: 34459](#) Mouse

[Unigene: 6206](#) Rat

Important Note:

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