

Junctophilin-1 (2E6): sc-517084

BACKGROUND

Junctophilins are components of the junctional complexes between plasma membranes and endoplasmic or sarcoplasmic reticulums present in all excitable cells. Junctophilins contain a cytoplasmic domain which binds to the plasma membrane, as well as an ER/SR membrane spanning hydrophobic C-terminal segment. The three subtypes in this family are Junctophilin-1, -2 and -3. Junctophilin-1 is predominantly expressed in skeletal muscle, but is also expressed at low levels in heart. Junctophilin-2 is expressed in heart and skeletal muscle. Mutant mice lacking the Jph2 gene exhibit embryonic lethality and possess cardiac myocytes that express abnormal calcium transients. Junctophilin-3 is expressed in brain. The JPH3 alternatively spliced exon 2A has been suggested as a site for CTG repeat expansion leading to a Huntington disease-like autosomal dominant disorder.

REFERENCES

1. Takeshima, H., et al. 2000. Junctophilins: a novel family of junctional membrane complex proteins. *Mol. Cell* 6: 11-22.
2. Margolis, R.L., et al. 2001. A disorder similar to Huntington's disease is associated with a novel CAG repeat expansion. *Ann. Neurol.* 50: 373-380.
3. Takeshima, H., et al. 2001. Junctophilins: molecular components contributing junctional membrane complexes between the cell-surface membrane and endoplasmic/sarcoplasmic reticulum. *Clin. Calcium* 11: 758-762.
4. Komazaki, S., et al. 2002. Deficiency of triad formation in developing skeletal muscle cells lacking junctophilin type 1. *FEBS Lett.* 524: 225-229.
5. Komazaki, S., et al. 2003. Abnormal junctional membrane structures in cardiac myocytes expressing ectopic junctophilin type 1. *FEBS Lett.* 542: 69-73.
6. Moriguchi, S., et al. 2006. Functional uncoupling between Ca^{2+} release and afterhyperpolarization in mutant hippocampal neurons lacking junctophilins. *Proc. Natl. Acad. Sci. USA* 103: 10811-10816.
7. Phimister, A.J., et al. 2007. Conformation-dependent stability of Junctophilin-1 (JP1) and ryanodine receptor type 1 (RyR1) channel complex is mediated by their hyper-reactive thiols. *J. Biol. Chem.* 282: 8667-8677.
8. Matsushita, Y., et al. 2007. Mutation of Junctophilin type 2 associated with hypertrophic cardiomyopathy. *J. Hum. Genet.* 52: 543-548.

CHROMOSOMAL LOCATION

Genetic locus: JPH1 (human) mapping to 8q21.11; Jph1 (mouse) mapping to 1 A3.

SOURCE

Junctophilin-1 (2E6) is a mouse monoclonal antibody raised against amino acids 501-579 representing partial length Junctophilin-1 of human origin.

PRODUCT

Each vial contains 100 μ g IgG_{2a} kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Junctophilin-1 (2E6) is recommended for detection of Junctophilin-1 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Junctophilin-1 siRNA (h): sc-72005, Junctophilin-1 siRNA (m): sc-72006, Junctophilin-1 shRNA Plasmid (h): sc-72005-SH, Junctophilin-1 shRNA Plasmid (m): sc-72006-SH, Junctophilin-1 shRNA (h) Lentiviral Particles: sc-72005-V and Junctophilin-1 shRNA (m) Lentiviral Particles: sc-72006-V.

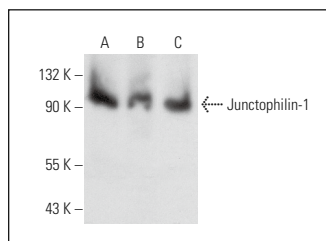
Molecular Weight of Junctophilin-1: 72 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, Jurkat whole cell lysate: sc-2204 or Sol8 cell lysate: sc-2249.

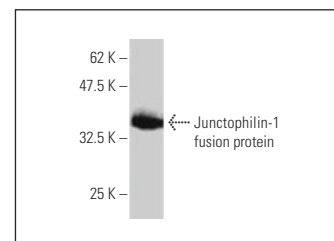
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).

DATA



Junctophilin-1 (2E6): sc-517084. Western blot analysis of Junctophilin-1 expression in HeLa (A), Jurkat (B) and Sol8 (C) whole cell lysates.



Junctophilin-1 (2E6): sc-517084. Western blot analysis of human recombinant Junctophilin-1 fusion protein.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.