

Junctophilin-2 (B-6): sc-398125

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. Walker, R.H., et al. 2003. Huntington's disease—like 2 can present as chorea-acanthocytosis. *Neurology* 61: 1002-1004.
4. Margolis, R.L., et al. 2004. Huntington's disease-like 2 (HDL2) in North America and Japan. *Ann. Neurol.* 56: 670-674.
5. Minamisawa, S., et al. 2004. Junctophilin type 2 is associated with caveolin-3 and is downregulated in the hypertrophic and dilated cardiomyopathies. *Biochem. Biophys. Res. Commun.* 325: 852-856.
6. Margolis, R.L., et al. 2005. Huntington's disease-like 2: review and update. *Acta Neurol. Taiwan.* 14: 1-8.
7. Matsushita, Y., et al. 2007. Mutation of Junctophilin type 2 associated with hypertrophic cardiomyopathy. *J. Hum. Genet.* 52: 543-548.
8. Rudnicki, D.D., et al. 2007. Huntington's disease—like 2 is associated with CUG repeat-containing RNA foci. *Ann. Neurol.* 61: 272-282.

CHROMOSOMAL LOCATION

Genetic locus: JPH2 (human) mapping to 20q13.12; Jph2 (mouse) mapping to 2 H3.

SOURCE

Junctophilin-2 (B-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 552-579 near the C-terminus of Junctophilin-2 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-398125 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

Junctophilin-2 (B-6) is recommended for detection of Junctophilin-2 isoform 1 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Junctophilin-2 siRNA (h): sc-72007, Junctophilin-2 siRNA (m): sc-72008, Junctophilin-2 siRNA (r): sc-270010, Junctophilin-2 shRNA Plasmid (h): sc-72007-SH, Junctophilin-2 shRNA Plasmid (m): sc-72008-SH, Junctophilin-2 shRNA Plasmid (r): sc-270010-SH, Junctophilin-2 shRNA (h) Lentiviral Particles: sc-72007-V, Junctophilin-2 shRNA (m) Lentiviral Particles: sc-72008-V and Junctophilin-2 shRNA (r) Lentiviral Particles: sc-270010-V.

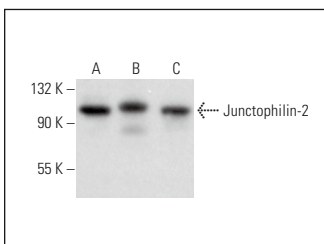
Molecular Weight of Junctophilin-2: 100 kDa.

Positive Controls: human heart extract: sc-363763, human skeletal muscle extract: sc-363776 or Sol8 cell lysate: sc-2249.

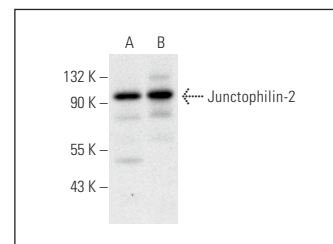
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ 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). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



Junctophilin-2 (B-6): sc-398125. Western blot analysis of Junctophilin-2 expression in Sol8 (A), C2C12 (B) and A-10 (C) whole cell lysates.



Junctophilin-2 (B-6): sc-398125. Western blot analysis of Junctophilin-2 expression in human heart (A) and human skeletal muscle (B) tissue extracts.

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.