

Junctophilin-3 (H-66): sc-292970

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. Holmes, S.E., et al. 2001. A repeat expansion in the gene encoding junctophilin-3 is associated with Huntington disease-like 2. *Nat. Genet.* 29: 377-378.
4. Nishi, M., et al. 2002. Motor discoordination in mutant mice lacking junctophilin type 3. *Biochem. Biophys. Res. Commun.* 292: 318-324.
5. Nishi, M., et al. 2003. Coexpression of Junctophilin type 3 and type 4 in brain. *Brain Res. Mol. Brain Res.* 118: 102-110.
6. Stevanin, G., et al. 2003. Huntington's disease-like phenotype due to tri-nucleotide repeat expansions in the TBP and JPH3 genes. *Brain* 126: 1599-1603.
7. Keckarevic, M., et al. 2005. JP-3 gene polymorphism in a healthy population of Serbia and Montenegro. *J. Genet.* 84: 69-71.
8. Keckarevic, M., et al. 2005. Yugoslav HD phenocopies analyzed on the presence of mutations in PrP, ferritin, and JP-3 genes. *Int. J. Neurosci.* 115: 299-301.
9. Costa Mdo, C., et al. 2006. Exclusion of mutations in the PRNP, JPH3, TBP, ATN1, CREBBP, POU3F2 and FTL genes as a cause of disease in Portuguese patients with a Huntington-like phenotype. *J. Hum. Genet.* 51: 645-651.

CHROMOSOMAL LOCATION

Genetic locus: JPH3 (human) mapping to 16q24.2; *Jph3* (mouse) mapping to 8 E1.

SOURCE

Junctophilin-3 (H-66) is a rabbit polyclonal antibody raised against amino acids 213-278 mapping within an internal region of Junctophilin-3 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

Junctophilin-3 (H-66) is recommended for detection of Junctophilin-3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, 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).

Junctophilin-3 (H-66) is also recommended for detection of Junctophilin-3 in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for Junctophilin-3 siRNA (h): sc-72009, Junctophilin-3 siRNA (m): sc-72010, Junctophilin-3 shRNA Plasmid (h): sc-72009-SH, Junctophilin-3 shRNA Plasmid (m): sc-72010-SH, Junctophilin-3 shRNA (h) Lentiviral Particles: sc-72009-V and Junctophilin-3 shRNA (m) Lentiviral Particles: sc-72010-V.

Molecular Weight of Junctophilin-3: 81 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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 or our catalog for detailed protocols and support products.