

Harmonin (N-17): sc-26285

BACKGROUND

The gene encoding human Harmonin (NY-CO-38/PDZ-73) maps to chromosome 11p15.1. Mutations in the Harmonin gene cause Usher syndrome type I subtype C and non-syndromic deafness. Alternative splicing generates three Harmonin isoforms. Harmonin isoform 1 contains three PDZ protein-protein interaction domains. Renal and colon cancer patients frequently develop autoantibodies to Harmonin, which is present in kidney, brain, small intestine and colon. Sensory hair cells (stereocilia) in the inner ear also express Harmonin. The first PDZ domain of Harmonin binds mutated in colon cancer 2 (MCC2) at the carboxy terminal. Harmonin also interacts with cadherin 23 and myosin VIIA in growing stereocilia of the inner ear. The harmonin/cadherin 23/myosin VIIA complex influences the shaping of a functional stereo-cilia bundle.

REFERENCES

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2. Verpy, E., et al. 2000. A defect in Harmonin, a PDZ domain-containing protein expressed in the inner ear sensory hair cells, underlies Usher syndrome type 1C. *Nat. Genet.* 26: 51-55.
3. Bitner-Glindzic, M., et al. 2000. A recessive contiguous gene deletion causing infantile hyperinsulinism, enteropathy and deafness identifies the Usher type 1C gene. *Nat. Genet.* 26: 56-60.
4. Ishikawa, S., et al. 2001. Interaction of MCC2, a novel homologue of MCC tumor suppressor, with PDZ-domain Protein AIE-75. *Gene* 267: 101-110.
5. Ahmed, Z.M., et al. 2002. Nonsyndromic recessive deafness DFNB18 and Usher syndrome type 1C are allelic mutations of USH1C. *Hum. Genet.* 110: 527-531.
6. Ouyang, X.M., et al. 2002. Mutations in the alternatively spliced exons of USH1C cause non-syndromic recessive deafness. *Hum. Genet.* 111: 26-30.
7. Siemens, J., et al. 2002. The Usher syndrome proteins cadherin 23 and Harmonin form a complex by means of PDZ-domain interactions. *Proc. Natl. Acad. Sci. USA* 99: 14946-14951.
8. Boeda, B., et al. 2002. Myosin VIIa, Harmonin and cadherin 23, three Usher I gene products that cooperate to shape the sensory hair cell bundle. *EMBO J.* 21: 6689-6699.

CHROMOSOMAL LOCATION

Genetic locus: USH1C (human) mapping to 11p15.1; Ush1c (mouse) mapping to 7 B4.

SOURCE

Harmonin (N-17) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of Harmonin of human origin.

STORAGE

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

PRODUCT

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

Blocking peptide available for competition studies, sc-26285 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Harmonin (N-17) is recommended for detection of Harmonin isoforms 1, 2 and 3 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

Harmonin (N-17) is also recommended for detection of Harmonin isoforms 1, 2 and 3 in additional species, including canine, bovine and porcine.

Suitable for use as control antibody for Harmonin siRNA (h): sc-40648, Harmonin siRNA (m): sc-40649, Harmonin shRNA Plasmid (h): sc-40648-SH, Harmonin shRNA Plasmid (m): sc-40649-SH, Harmonin shRNA (h) Lentiviral Particles: sc-40648-V and Harmonin shRNA (m) Lentiviral Particles: sc-40649-V.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.