SANTA CRUZ BIOTECHNOLOGY, INC.

Harmonin (D-18): sc-26286

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

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. Sen-sory 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 stereocilia bundle.

REFERENCES

1. Scanlan, M.J., et al. 1999. Isoforms of the human PDZ-73 protein exhibit differential tissue expression. Biochim. Biophys. Acta 1445: 39-52.
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-Glindzicz, 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 IC are allelic mutations of USHIC. 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.

CHROMOSOMAL LOCATION

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

SOURCE

Harmonin (D-18) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the C-terminus of Harmonin 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.

Blocking peptide available for competition studies, sc-26286 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

Harmonin (D-18) is recommended for detection of Harmonin isoforms 1 and 2 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), immunohistochemistry (including paraffin-embedded sections) (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 (D-18) is also recommended for detection of Harmonin isoforms 1 and 2 in additional species, including canine, bovine and porcine.

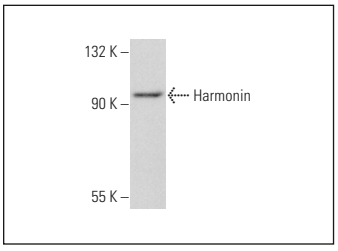
Suitable for use as control antibody for Harmonin siRNA (h): sc-40648, Harmonin siRNA (m): sc-40649, Harmonin shRNA Plasmid (h): sc-40648-SH, Harmonin shRNA Plasmid (m): sc-40649-SH, Harmonin shRNA (h) Lentiviral Particles: sc-40648-V and Harmonin shRNA (m) Lentiviral Particles: sc-40649-V.

Positive Controls: mouse kidney extract: sc-2255.

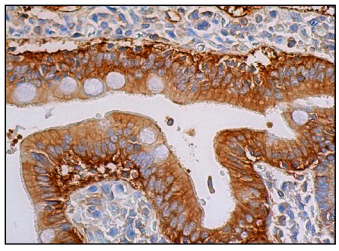
RECOMMENDED SECONDARY REAGENTS

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) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) 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. 4) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

DATA



Harmonin (D-18): sc-26286. Western blot analysis of Harmonin expression in mouse kidney tissue extract.



Harmonin (D-18): sc-26286. Immunoperoxidase staining of formalin fixed, paraffin-embedded human small intestine tissue showing cytoplasmic and membrane staining of glandular cells.

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.