

# USH1G (N-15): sc-69070

## BACKGROUND

USH1G (Usher syndrome 1G), also known as ANKS4A or SANS, is a 461 amino acid protein that contains 3 ANK repeats and 1 SAM (sterile  $\alpha$  motif) domain. Expressed in the small intestine, as well as in tissue of the eye and inner ear, USH1G associates with harmonin and is thought to play a role in the development and maintenance of both auditory and visual systems, specifically by mediating the cohesion of hair bundles formed by inner ear sensory cells. Defects in the gene encoding USH1G are the cause of Usher syndrome type 1G (USH1G), a heterogeneous condition that is characterized by profound congenital sensorineural deafness, absent vestibular function and prepubertal onset of progressive retinitis pigmentosa, ultimately leading to blindness.

## REFERENCES

1. Kitamura, K., Kakoi, H., Yoshikawa, Y. and Ochikubo, F. 1992. Ultrastructural findings in the inner ear of Jackson shaker mice. *Acta Otolaryngol.* 112: 622-627.
2. Kikkawa, Y., Shitara, H., Wakana, S., Kohara, Y., Takada, T., Okamoto, M., Taya, C., Kamiya, K., Yoshikawa, Y., Tokano, H., Kitamura, K., Shimizu, K., Wakabayashi, Y., Shiroishi, T., Kominami, R. and Yonekawa, H. 2003. Mutations in a new scaffold protein SANS cause deafness in Jackson shaker mice. *Hum. Mol. Genet.* 12: 453-461.
3. Weil, D., El-Amraoui, A., Masmoudi, S., Mustapha, M., Kikkawa, Y., Lainé, S., Delmaghani, S., Adato, A., Nadifi, S., Zina, Z.B., Hamel, C., Gal, A., Ayadi, H., Yonekawa, H. and Petit, C. 2003. Usher syndrome type 1G (USH1G) is caused by mutations in the gene encoding SANS, a protein that associates with the USH1C protein, Harmonin. *Hum. Mol. Genet.* 12: 463-471.
4. Ouyang, X.M., Yan, D., Du, L.L., Hejtmancik, J.F., Jacobson, S.G., Nance, W.E., Li, A.R., Angeli, S., Kaiser, M., Newton, V., Brown, S.D., Balkany, T. and Liu, X.Z. 2005. Characterization of Usher syndrome type 1 gene mutations in an Usher syndrome patient population. *Hum. Genet.* 116: 292-299.
5. Adato, A., Michel, V., Kikkawa, Y., Reiners, J., Alagramam, K.N., Weil, D., Yonekawa, H., Wolfrum, U., El-Amraoui, A. and Petit, C. 2005. Interactions in the network of Usher syndrome type 1 proteins. *Hum. Mol. Genet.* 14(3): 347-356.
6. Aller, E., Jaijo, T., Beneyto, M., Nájera, C., Morera, C., Pérez-Garrigues, H., Ayuso, C. and Millán, J. 2007. Screening of the USH1G gene among Spanish patients with Usher syndrome. Lack of mutations and evidence of a minor role in the pathogenesis of the syndrome. *Ophthalmic Genet.* 28: 151-155.

## CHROMOSOMAL LOCATION

Genetic locus: USH1G (human) mapping to 17q25.1; Ush1g (mouse) mapping to 11 E2.

## SOURCE

USH1G (N-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of USH1G of human origin.

## PRODUCT

Each vial contains 200  $\mu$ g IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-69070 P, (100  $\mu$ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

USH1G (N-15) is recommended for detection of USH1G 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).

USH1G (D-20) is also recommended for detection of USH1G in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for USH1G siRNA (h): sc-63189, USH1G siRNA (m): sc-63190, USH1G shRNA Plasmid (h): sc-63189-SH, USH1G shRNA Plasmid (m): sc-63190-SH, USH1G shRNA (h) Lentiviral Particles: sc-63189-V and USH1G shRNA (m) Lentiviral Particles: sc-63190-V.

Molecular Weight of USH1G: 51 kDa.

## 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.

## 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](http://www.scbt.com) or our catalog for detailed protocols and support products.



Try **USH1G (D-10): sc-514418**, our highly recommended monoclonal alternative to USH1G (N-15).