

# USH3A (C-20): sc-69073

## BACKGROUND

USHSA (Usher syndrome type 3), also known as CLRN1 (clarin-1) or USH3, is a 232 amino acid multi-pass membrane protein that exists as multiple alternatively spliced isoforms and belongs to the clarin family. Expressed in a variety of tissues, including retina, USH3A is thought to be involved in the maintenance of the inner ear and retina, specifically playing a role in excitatory ribbon synapse junctions between hair cells and cochlear ganglion cells, as well as in analogous synapses within the retina. Defects in the gene encoding USH3A are the cause of Usher syndrome type 3 (USH3), a genetically heterogeneous condition characterized by postlingual progressive deafness and onset of retinitis pigmentosa in the second decade of life.

## REFERENCES

1. Sankila, E.M., et al. 1995. Assignment of an Usher syndrome type III (USH3) gene to chromosome 3q. *Hum. Mol. Genet.* 4: 93-98.
2. Joensuu, T., et al. 1996. Refined mapping of the Usher syndrome type III locus on chromosome 3, exclusion of candidate genes, and identification of the putative mouse homologous region. *Genomics* 38: 255-263.
3. Adato, A., et al. 1999. Possible interaction between USH1B and USH3 gene products as implied by apparent digenic deafness inheritance. *Am. J. Hum. Genet.* 65: 261-265.
4. Joensuu, T., et al. 2000. A sequence-ready map of the Usher syndrome type III critical region on chromosome 3q. *Genomics* 63: 409-416.
5. Joensuu, T., et al. 2001. Mutations in a novel gene with transmembrane domains underlie Usher syndrome type 3. *Am. J. Hum. Genet.* 69: 673-684.
6. Fields, R.R., et al. 2002. Usher syndrome type III: revised genomic structure of the USH3 gene and identification of novel mutations. *Am. J. Hum. Genet.* 71: 607-617.
7. Adato, A., et al. 2002. USH3A transcripts encode Clarin-1, a four-transmembrane-domain protein with a possible role in sensory synapses. *Eur. J. Hum. Genet.* 10: 339-350.

## CHROMOSOMAL LOCATION

Genetic locus: CLRN1 (human) mapping to 3q25.1; Clrn1 (mouse) mapping to 3 D.

## SOURCE

USH3A (C-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of USH3A 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-69073 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## RESEARCH USE

For research use only, not for use in diagnostic procedures.

## APPLICATIONS

USH3A (C-20) is recommended for detection of USH3A 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), 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).

USH3A (C-20) is also recommended for detection of USH3A in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for USH3A siRNA (h): sc-63191, USH3A siRNA (m): sc-63192, USH3A shRNA Plasmid (h): sc-63191-SH, USH3A shRNA Plasmid (m): sc-63192-SH, USH3A shRNA (h) Lentiviral Particles: sc-63191-V and USH3A shRNA (m) Lentiviral Particles: sc-63192-V.

Molecular Weight of USH3A: 26 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. 3) Immunohistochemistry: use ImmunoCruz™: sc-2053 or ABC: sc-2023 goat IgG Staining Systems.

## DATA



USH3A (C-20): sc-69073. Immunoperoxidase staining of formalin fixed, paraffin-embedded human kidney tissue showing cytoplasmic staining of cells in tubules.

## STORAGE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.