

Whirlin (h): 293T Lysate: sc-112679

BACKGROUND

Whirlin is a cytoplasmic PDZ domain-containing protein that plays a role in elongation and maintenance of stereocilia, mechanosensory organelles located in hair cells of the inner ear. Whirlin co-localizes with actin filaments and is primarily detected in cochlear hair cells. It is connected to the dynamic Usher protein interactome and has a pleiotropic function in both the retina and the inner ear. Myosin XVa is a motor protein that associates with the second and third PDZ domain of Whirlin through its C-terminal PDZ-ligand. Myosin XVa then delivers Whirlin to the tips of stereocilia, which are subsequently elongated. p55 also interacts with Whirlin, and mutations in DFNB31, the Whirlin gene, lead to an early ablation of p55 labeling of stereocilia, which may cause recessive hearing loss in rats and humans.

REFERENCES

1. Belyantseva, I.A., et al. 2003. Stereocilia: the long and the short of it. *Trends Mol. Med.* 9: 458-461.
2. Mburu, P., et al. 2003. Defects in Whirlin, a PDZ domain molecule involved in stereocilia elongation, cause deafness in the Whirler mouse and families with DFNB31. *Nat. Genet.* 34: 421-428.
3. Delprat, B., et al. 2005. Myosin XVa and Whirlin, two deafness gene products required for hair bundle growth, are located at the stereocilia tips and interact directly. *Hum. Mol. Genet.* 14: 401-410.
4. Adato, A., et al. 2005. Usherin, the defective protein in Usher syndrome type IIA, is likely to be a component of interstereocilia ankle links in the inner ear sensory cells. *Hum. Mol. Genet.* 14: 3921-3932.
5. Belyantseva, I.A., et al. 2005. Myosin XVa is required for tip localization of Whirlin and differential elongation of hair-cell stereocilia. *Nat. Cell Biol.* 7: 148-156.
6. Rzadzinska, A., et al. 2005. Balanced levels of Espin are critical for stereociliary growth and length maintenance. *Cell Motil. Cytoskeleton* 62: 157-165.
7. Tlili, A., et al. 2005. Identification of a novel frameshift mutation in the DFNB31/WHRN gene in a Tunisian consanguineous family with hereditary non-syndromic recessive hearing loss. *Hum. Mutat.* 25: 503.

CHROMOSOMAL LOCATION

Genetic locus: DFNB31 (human) mapping to 9q32.

PRODUCT

Whirlin (h): 293T Lysate represents a lysate of human Whirlin transfected 293T cells and is provided as 100 µg protein in 200 µl SDS-PAGE buffer.

STORAGE

Store at -20° C. Repeated freezing and thawing should be minimized. Sample vial should be boiled once prior to use. Non-hazardous. No MSDS required.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.

APPLICATIONS

Whirlin (h): 293T Lysate is suitable as a Western Blotting positive control for human reactive Whirlin antibodies. Recommended use: 10-20 µl per lane.

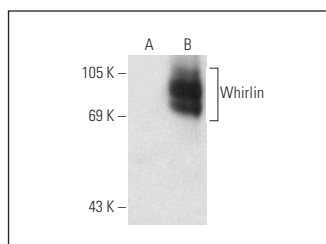
Control 293T Lysate: sc-117752 is available as a Western Blotting negative control lysate derived from non-transfected 293T cells.

Whirlin (A-12): sc-271508 is recommended as a positive control antibody for Western Blot analysis of enhanced human Whirlin expression in Whirlin transfected 293T cells (starting dilution 1:100, dilution range 1:100-1:1,000).

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended:
1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.

DATA



Whirlin (A-12): sc-271508. Western blot analysis of Whirlin expression in non-transfected: sc-117752 (A) and human Whirlin transfected: sc-112679 (B) 293T whole cell lysates.

RESEARCH USE

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