SANTA CRUZ BIOTECHNOLOGY, INC.

Retinoschisin (K-12): sc-46478

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The Power to Question

BACKGROUND

X-linked juvenile retinoschisis (XLRS), the most common form of early onset macular degeneration in males, is characterized by delamination of the inner retinal layers and severe loss of vision. XLRS is caused by over 125 different mutations in the RS1 gene, which encodes the 24 kDa discoidin domain-containing protein Retinoschisin. Retinoschisin functions as a cell adhesion protein that maintains the cellular organization and synaptic structure of the retina. It is secreted from retinal tissues, specifically photoreceptor and bipolar cells, as an octamer, the subunits of which are joined together by Cys 59-Cys 223 intermolecular disulfide bonds. The interaction of cysteine residues in the Retinoschisin protein are critical for proper folding and subunit assembly. Misfolding of the discoidin domain, defective disulfide-linked subunit assembly and inability of Retinoschisin to insert into the endoplasmic reticulum membrane are responsible for the loss of function of Retinoschisin and the pathogenesis of XLRS.

REFERENCES

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2. Molday, L.L., et al. 2001. Expression of X-linked retinoschisis protein RS1 in photoreceptor and bipolar cells. Invest. Ophthalmol. Vis. Sci. 42: 816-825.
3. Wu, W.W., et al. 2003. Defective discoidin domain structure, subunit assembly and endoplasmic reticulum processing of Retinoschisin are primary mechanisms responsible for X-linked retinoschisis. J. Biol. Chem. 278: 28139-28146.
4. Reid, S.N., et al. 2003. Retinoschisin, a photoreceptor-secreted protein, and its interaction with bipolar and muller cells. J. Neurosci. 23: 6030-6040.
5. Takada, Y., et al. 2004. A retinal neuronal developmental wave of Retinoschisin expression begins in ganglion cells during layer formation. Invest. Ophthalmol. Vis. Sci. 45: 3302-3312.
6. Tantri, A., et al. 2004. X-linked retinoschisis: a clinical and molecular genetic review. Surv. Ophthalmol. 49: 214-230.
7. Wu, W.W., et al. 2005. RS1, a discoidin domain-containing retinal cell adhesion protein associated with X-linked retinoschisis, exists as a novel disulfide-linked octamer. J. Biol. Chem. 280: 10721-10730.

CHROMOSOMAL LOCATION

Genetic locus: RS1 (human) mapping to Xp22.2-p22.1; Rs1h (mouse) mapping to X F4.

SOURCE

Retinoschisin (K-12) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of Retinoschisin 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-46478 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

Retinoschisin (K-12) is recommended for detection of Retinoschisin 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).

Suitable for use as control antibody for Retinoschisin siRNA (h): sc-44771 and Retinoschisin siRNA (m): sc-44772.

Molecular Weight of Retinoschisin: 24 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.

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.