

# Retinoschisin (H-65): sc-48755

## 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 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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- 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.
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- 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 Xp22.13; Rs1 (mouse) mapping to X F4.

## SOURCE

Retinoschisin (H-65) is a rabbit polyclonal antibody raised against amino acids 126-190 mapping near the C-terminus of Retinoschisin 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.

## APPLICATIONS

Retinoschisin (H-65) 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), 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) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Retinoschisin (H-65) is also recommended for detection of Retinoschisin in additional species, including equine, canine, bovine and avian.

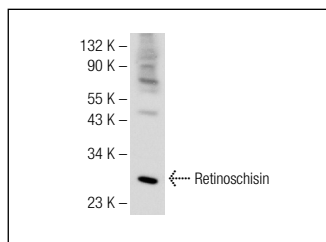
Suitable for use as control antibody for Retinoschisin siRNA (h): sc-44771, Retinoschisin siRNA (m): sc-44772, Retinoschisin shRNA Plasmid (h): sc-44771-SH, Retinoschisin shRNA Plasmid (m): sc-44772-SH, Retinoschisin shRNA (h) Lentiviral Particles: sc-44771-V and Retinoschisin shRNA (m) Lentiviral Particles: sc-44772-V.

Molecular Weight of Retinoschisin: 24 kDa.

## RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

## DATA



Retinoschisin (H-65): sc-48755. Western blot analysis of Retinoschisin expression in 293T whole cell lysate.

## 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.