10320 Camino Santa Fe, Suite G
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ABCA4 (Rim Protein) Antibody
Mouse monoclonal antibody
Catalog # AN1167

Specification

ABCA4 (Rim Protein) Antibody - Product Information

Application	**IHC**
Primary Accession	O35600
Reactivity	**Bovine, Human, Mouse, Xenopus**
Host	**Mouse**
Clonality	**monoclonal**
Isotype	**IgG**
Calculated MW	**220 KDa**

ABCA4 (Rim Protein) Antibody - Additional Information

Gene ID	**11304**
Gene Name	**ABCA4**

Other Names
Retinal-specific ATP-binding cassette transporter, ATP-binding cassette sub-family A member 4, RIM ABC transporter, RIM protein, RmP, Abca4, Abcr

Target/Specificity
Partially purified bovine 220-kDa disc rim protein.

Dilution
IHC~~ 1:100

Format
Protein G purified culture supernatant

Antibody Specificity
Specific for ABCA4

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions
ABCA4 (Rim Protein) Antibody is for research use only and not for use in diagnostic or therapeutic procedures.

Shipping
Blue Ice

ABCA4 (Rim Protein) Antibody - Protocols

Provided below are standard protocols that you may find useful for product applications.

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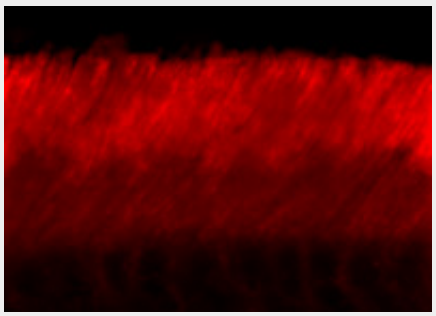
- Western Blot
- Blocking Peptides
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- Cell Culture

ABCA4 (Rim Protein) Antibody - Images



Immunohistochemical staining of adult mouse retina showingspecific immunolabeling of the ABCA4 protein. Photo courtesyof Mary Raven, University of California, Santa Barbara, CA.

ABCA4 (Rim Protein) Antibody - Background

ABCA4 (ATP-binding cassette, sub-family A (ABC1), member 4, Rim Protein) is a member of the superfamily of ATP-binding cassette (ABC) transporters (Illing et al., 1997). ABC proteins transport various molecules across extra- and intracellular membranes. This protein is a retina-specific ABC transporter with N-retinylidene-PE as a substrate. It is expressed exclusively in retina photoreceptor cells, indicating the gene product mediates transport of an essential molecule across the photoreceptor cell membrane. Mutations in this gene are found in patients diagnosed with Stargardt disease and are associated with retinitis pigmentosa-19 and age-related macular degeneration (Wiszniewski et al., 2003). Defects in ABCA4 are the cause of Stargardt disease type 1 (STGD1) (Molday et al., 2000). STGD is one of the most frequent causes of macular degeneration in childhood. Defects in ABCA4 are also known to cause fundus flavimaculatus (FFM), age-related macular degeneration type 2 (ARMD2) and cone-rod dystrophy type 3 (CORD3) (Klevering et al., 2005).

ABCA4 (Rim Protein) Antibody - References

Michelle Illing, Laurie L. Molday and Robert S. Molday. The 220-kDa Rim Protein of Retinal Rod Outer Segments Is a Member of the ABC Transporter Superfamily. J. Biol. Chem., (1997) Vol 272 (15) I April 11. 10303-10310.
Klevering BJ, Deutman AF, Maugeri A, Cremers FP, Hoyng CB (2005) The spectrum of retinal phenotypes casued by mutations in the ABCA4 gene. Graefes Arch Clin Exp Ophthalmol. 243(2):90-100.
Molday, L. et al., ABCR expression in foveal cone photoreceptors and its role in Stargardt macular dystrophy. Nature Genetics (2000) 25, 257 - 258.
Wiszniewski, W. et al., ABCA4 mutations causing mislocalization are found frequently in patients with severe retinal dystrophies. Human Molecular Genetics (2005) 14(19):2769-2778.