



**PhosphoSolutions®**  
Antibodies that work™

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## Anti-ABCA4 (Rim Protein)

**Catalog Number:** 115-ABCA4      **Size:** 100 µl      **Clone:** 3F4      **Isotype:** IgG

**Product Description:** Mouse monoclonal antibody

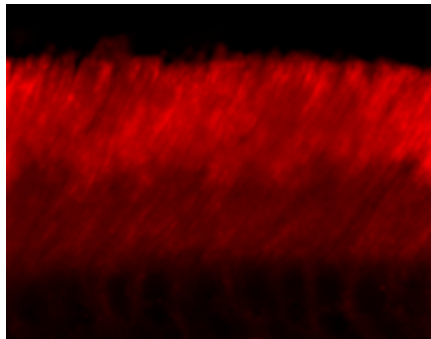
**Applications:**    **WB:** 1:1000    **IHC:** 1:100

**Antigen:** Partially purified bovine 220-kDa disc rim protein.

**Species reactivity:** The antibody has been directly tested for reactivity in bovine, human, mouse and *Xenopus* tissues.

**Biological Significance:** 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).

### Anti-ABCA4



**Immunohistochemical** staining of adult mouse retina showing specific immunolabeling of the ABCA4 protein. Photo courtesy of Mary Raven, University of California, Santa Barbara, CA.

**WB** = Western Blot    **IF** = Immunofluorescence    **IHC** = Immunohistochemistry    **IP** = Immunoprecipitation

**Packaging:** 100 µl in 10 mM HEPES (pH 7.5), 150 mM NaCl, 100 µg per ml BSA and 50% glycerol. Adequate amount of material to conduct 10-mini Western Blots.

**Storage and Stability:** Store at -20°C in undiluted aliquots; stable for up to one year after date of receipt. Avoid freeze/thaw cycles.

**Shipment:** Domestic - Blue Ice; International - Blue Ice or Dry Ice.

**For Research Use Only.**

**Purification Method:** Protein G purified culture supernatant

**Antibody Specificity:** Specific for ABCA4

**Quality Control Tests:** Western blots performed on each lot.

**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) | April 11. 10303-10310.

Klevering BJ, Deutman AF, Maugeri A, Cremers FP, Hoyng CB (2005) The spectrum of retinal phenotypes caused 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.

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