

Catalog Number:	NB300-164SS
Background:	Best macular dystrophy (BMD) or vitelliform macular dystrophy (VMD2), is an autosomal form of macular degeneration, characterized by a depressed light peak in the electrooculogram (EOG). It is inherited and has an early onset. Bestrophin is a 68 kDa basolateral plasma membrane protein encoded by the VMD2 gene. Bestrophin's function is still unknown, but data suggests that it is a chloride channel that plays a role in generating the altered EOG in Best disease patients. In addition, Bestrophin is a useful biochemical and histological marker of RPE (retinal pigment epithelial cells) cells.
Research Areas:	317,336,0
Immunogen:	Synthetic peptide conjugated to KLH [KDHMDPYWALENRDEAHS].
Clone:	E6-6
Isotype:	IgG1 kappa
Species Reactivity:	NB 300-164 recognizes human, porcine, and monkey. This antibody does not work in rat. Other species have not been tested.
Uses:	By Western blot, this antibody recognizes a band at ~68 kDa representing Bestrophin.** It is also useful for immunofluorescence. **Please see protocol for treatment of cell extract. * Other applications have not been tested.
Dilutions:	Suggested working dilutions * immunofluorescence Assay dependent, Western Blot 1:1,000 * Investigator should determine optimal working dilutions.
Positive Controls:	Pig RPE (Retinal Pigment Epithelium) whole cell extract
Packaging:	0.025 ml Ascites Mouse ascites.
Preservative:	0.1% sodium azide
Storage:	Aliquot and store at -20C or -80C. Avoid freeze-thaw cycles.
Product Specific References:	1. Marmorstein, A.D., et al. Bestrophin, the product of the Best vitelliform macular dystrophy gene (VMD2), localizes to the basolateral plasma membrane of the retinal pigment epithelium. PNAS. 97(23): 12758-12763 (2000)
Novus Specific References:	1. Klimanskaya, I., Hipp, J., Rezai, K., et al. Derivation and Comparative Assessment of Retinal Pigment Epithelium from Human Embryonic Stem Cells Using Transcriptomics. Cloning and Stem Cells. Sept. 2004: 217-245.
General References:	2. Petrushkin, K., et al. Identification of the gene responsible for Best macular dystrophy. Nature Genetics. 19: 241-247 (1998) 3. Caldwell, G.M., et al. Bestrophin gene mutations in patients with Best Vitelliform



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Macular Dystrophy. Genomics. 58: 98-100 (1999)

4. Marmorstein, L.Y., et al. Bestrophin physically and functionally interacts with protein phosphatase 2A. JBC. 277(34): 30591-30597 (2002)

Gene Id: 7439

Image(s)