

Polyclonal Antibody to Bestrophin-4 (C-term amidated) - Aff - Purified

Alternate names:	BEST4, VMD2L2, Vitelliform macular dystrophy 2-like protein 2
Catalog No.:	AP09915PU-N
Quantity:	0.1 mg
Concentration:	0.75 - 1.2 mg/ml (lot specific)
Background:	In the recent past at least 3 different family for "chloride channel" have been characterized: CLC family, ligand gated channels for the GABA and glycine receptor family and the cystic fibrosis membrane conductance regulator. These chloride channels play important role in mainting resting potentials, ion refluxes, acidification of internal organnles such as lysosomes in both excitatory and non-excitatory nerve and muscle cells. The bestrophins are a newly described family of "anion channels" unrelated in primary sequence to any previously characterized channel proteins. Bestrophins were originally defined as a family of over 20 related sequences of the C. elegans. The first mammalian bestrophin was identified as the vitelliform macular dystrophy (VMD), 1 also known as Best disease (1). Three more members of the bestrophin family members were cloned andindenfied recently, Bestrophin 2, 3 and 4. The bestrophin family members are membrane protein with 2-TMD and have a conserved 350-400 amino domain including the invariant peptide motif RFP. Each of the Bestrophin proteins has a unique C-terminus that lack similarity to other proteins or motifs. Bestrophin 1 gene is localised to chromosome 19p13.2-p13.12, Bestrophin 2 to 1p32.3-p33 and Bestrophin 3 to 12q14.2-q15. RT-PCR analyses revealed tissue-restricted expression of the three genes with both Bestrophin 1 and Bestrophin 2 are abundantly transcribed in colon. Bestrophin 1 is present in the retinal pigment epithelium while Bestrophin 3 shows predominant expression in skeletal muscle (2, 3). Functionally the bestrophines oligomerise to form tetramers and pentamers in order to act as calcium sensitive chloride channels. It has been shown that Bestrophin interacts with beta catalytic subunit of protein phosphatase 2A (PP2Ac). Such Protein-protein interaction between bestrophin and PP2Ac and the structural subunit of PP2A, PR65, was confirmed by reciprocal immunoprecipitation. The interaction between PP2Ac and the Bestrophin takes place near the Carboxy-terminal end of the protein. Okadic acid induce the phosphorylation of Bestrophin in vitro. Bestrophin also serves in the signal transduction pathway that modulates the light peak of the EOG, that is regulated by phosphorylation of the Bestrophin that in turn is regulated by protein phosphatase 2A (PP2A).
Uniprot ID:	Q8NFUQ
NCBI:	NP_695006
GeneID:	266675
Host:	Rabbit

Immunogen:	Synthetic peptide for Bestrophin 4 close to C-terminal corresponding to amino acids 457-473 AA Sequence: ari eee sae sgd eal ep Remarks: The peptide was post-synthetically modified to achieve desired immunogenicity. The carboxy peptide were amidated before conjugation.
Format:	State: Liquid Ig fraction Purification: Affinity chromatography Buffer System: Stabilization buffer
Applications:	ELISA. Western blot > 1:500. Immunoprecipitation and i.p pull-down assays: > 1:200. Other applications not tested. Optimal dilutions are dependent on conditions and should be determined by the user.
Specificity:	This antibody detects Bestrophin-4 (C-term).
Species Reactivity:	Tested: Mouse, rat
Storage:	Store (in aliquots) at -20 °C. Avoid repeated freezing and thawing. Shelf life: one year from despatch.
General Readings:	1. Marmorstein LY, McLaughlin PJ, Stanton JB, Yan L, Crabb JW, Marmorstein AD. J. Biol. Chemistry 2002; June 10, Electronic publication. 2. Stohr H, Marquardt A, et. al., Eur J Hum Genet. 2002 Apr;10(4):281-4. 3. Takashi Tsunenari , Hui Sun, John Williams, Hugh Cahill , Philip Smallwood, King-Wai Yau ** and Jeremy Nathans. J. Biol. Chem., Vol. 278, Issue 42, 41114-41125, October 17, 2003.
Pictures:	Western blot of anti Bestrophin 4. Ln 2: AP09915PU-N

