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Rabbit Anti-Bestrophin antibody

SL11040R

Product Name:	Bestrophin
Chinese Name:	卵黄状黄斑病蛋白抗体
Alias:	BEST 1; BEST1; BEST-1; BEST; Best macular dystrophy; BEST1; BEST1_HUMAN; Bestrophin 1; Bestrophin-1; Bestrophin1; BMD; mBest1; TU15B; Vitelliform macular dystrophy 2; Vitelliform macular dystrophy; Vitelliform macular dystrophy protein 2; VMD 2; VMD2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	64kDa
Cellular localization:	The cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human Bestrophin:251-350/585
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Best vitelliform macular dystrophy, known as Best disease, is an early-onset autosomal dominant condition in which accumulation of lipofuscin-like material within and beneath the RPE leads to progressive loss of central vision. Best disease is frequently a reflection of mutations in the Bestrophin gene, which encodes a protein containing four

	putative transmembrane domains and localizes to the basolateral plasma membrane of RPE cells. Human Bestrophin forms oligomeric chloride channels that are sensitive to intracellular calcium. Missense mutations at the Bestrophin locus reduces or abolishes Bestrophin protein mediated membrane current. Bestrophin Bestrophin 2,Bestrophin 3, and Bestrophin 4 are transmembrane proteins that contain a high percentage of aromatic residues, a conserved RFP (Arg-Phe-Pro) motif and they function as anion channels. **Function:** Forms calcium-sensitive chloride channels. Highly permeable to bicarbonate. **Subunit:** Tetramer or pentamers. May interact with PPP2CB and PPP2R1B. **Subcellular Location:** Cell membrane. Basolateral cell membrane. **Tissue Specificity:** Predominantly expressed in the basolateral membrane of the retinal pigment epithelium. **Post-translational modifications:** Phosphorylated by PP2A. **DISEASE:** Defects in BEST1 are the cause of vitelliform macular dystrophy type 2 (VMD2) ; also known as Best macular dystrophy (BMD). VMD2 is an autosomal dominant form of macular degeneration that usually begins in childhood or adolescence. VMD2 is characterized by typical 'egg-yolk' macular lesions due to abnormal accumulation of lipofuscin within and beneath the retinal pigment epithelium cells. Progression of the disease leads to destruction of the retinal pigment epithelium and vision loss. Defects in BEST1 are the cause of retinitis pigmentosa type 50 (RP50) . A retinal dystrophy belonging to the group of pigmentary retinopathies. RP is characterized by retinal pigment deposits visible on fundus examination and primary loss of rod photoreceptor cells followed by secondary loss of cone photoreceptors. Patients typically have night vision blindness and loss of midperipheral visual field. As their condition progresses, they lose their far peripheral visual field and eventually central vision as well. **Similarity:** Belongs to the bestrophin family. **SWISS:** O76090 **Gene ID:** 7439

	Database links: Entrez Gene: 7439Human Entrez Gene: 24115Mouse Entrez Gene: 293735Rat Omim: 607854Human SwissProt: O76090Human SwissProt: O88870Mouse Unigene: 524910Human Unigene: 712676Human Unigene: 31577Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.