abcam

Product datasheet

Anti-Bestrophin/BEST1 antibody ab14927

6 Abreviews 7 References

Overview

Product name	Anti-Bestrophin/BEST1 antibody
Description	Rabbit polyclonal to Bestrophin/BEST1
Host species	Rabbit
Tested applications	**Suitable for:** ICC, IP, ICC/IF, WB
Species reactivity	**Reacts with:** Mouse, Rat, Cow, Human **Predicted to work with:** Cynomolgus monkey
Immunogen	Synthetic peptide corresponding to Human Bestrophin/BEST1 aa 1-100. Database link: **O76090** Run BLAST with Run BLAST with
General notes	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: Tris glycine, 0.5% BSA, 30% Glycerol
Purity	Immunogen affinity purified
Clonality	Polyclonal
Isotype	IgG

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab14927 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Application	Abreviews	Notes
ICC	★★★★★ (1)	Use at an assay dependent concentration.
IP	★★★★☆ (1)	Use at an assay dependent concentration.
ICC/IF	★★★★☆ (1)	Use at an assay dependent concentration.
WB	★★★★☆ (2)	Use at an assay dependent concentration.

Target

Function Forms calcium-sensitive chloride channels. Highly permeable to bicarbonate.

Tissue specificity Predominantly expressed in the basolateral membrane of the retinal pigment epithelium.

Involvement in disease Defects in BEST1 are the cause of vitelliform macular dystrophy type 2 (VMD2) [MIM:153700]; 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) [MIM:613194]. 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.
Defects in BEST1 are a cause of adult-onset vitelliform macular dystrophy (AVMD) [MIM:608161]. AVMD is a rare autosomal dominant disorder with incomplete penetrance and highly variable expression. Patients usually become symptomatic in the fourth or fifth decade of life with a protracted disease of decreased visual acuity.
Defects in BEST1 are the cause of bestrophinopathy autosomal recessive (ARB) [MIM:611809]. A retinopathy characterized by central visual loss, an absent electro-oculogram light rise, and a reduced electroretinogram.
Defects in BEST1 are the cause of vitreoretinochoroidopathy autosomal dominant (ADVIRC) [MIM:193220]. A disorder characterized by vitreoretinochoroidal dystrophy. The clinical presentation is variable and may be associated with cataract, nanophthalmos, microcornea, shallow anterior chamber, and glaucoma.

Sequence similarities Belongs to the bestrophin family.

Post-translational modifications Phosphorylated by PP2A.

Cellular localization Cell membrane. Basolateral cell membrane.

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