abcam

Product datasheet

Anti-Bestrophin/BEST1 antibody [E6-6] ab2182

★★★★★ 7 Abreviews 30 References 2 Images

Overview

Product name	Anti-Bestrophin/BEST1 antibody [E6-6]
Description	Mouse monoclonal [E6-6] to Bestrophin/BEST1
Host species	Mouse
Tested applications	**Suitable for:** WB, IHC-Fr, ICC/IF, IP
Species reactivity	**Reacts with:** Cow, Dog, Human, Pig, Monkey **Predicted to work with:** Non human primates **Does not react with:** Mouse, Rat, Rabbit, Goat
Immunogen	Synthetic peptide corresponding to Human Bestrophin/BEST1 aa 568-585 (C terminal) conjugated to keyhole limpet haemocyanin. Sequence: KDHMDPYWALENRDEAHS Database link: **O76090** **Run BLAST with** **Run BLAST with**
Positive control	IHC-Fr: Pig retinal pigment epithelium tissue. ICC/IF: Bovine retinal pigment epithelium (RPE). WB: Human RPE cell lysate.
General notes	This product has switched from ascites to TCS on 9th September 2020. 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 Constituent: 99% PBS
Purity	Protein A/G purified
Clonality	Monoclonal
Clone number	E6-6
Myeloma	unknown
Isotype	IgG1
Light chain type	kappa

Applications

The Abpromise guarantee Our **Abpromise guarantee** covers the use of ab2182 in the following tested applications.

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

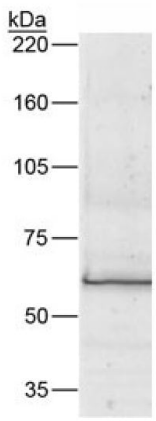
Application	Abreviews	Notes
WB	★★★★★ (3)	1/1000.
IHC-Fr	★★★★★ (2)	Use at an assay dependent concentration.
ICC/IF	★★★★★ (1)	Use at an assay dependent concentration.
IP	★★★★★ (1)	Use at an assay dependent concentration.

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.

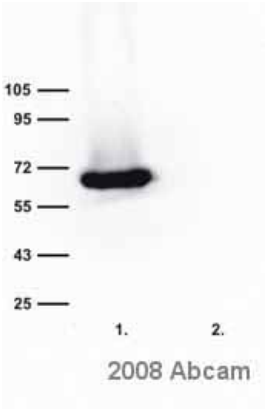
	Defects in BEST1 are the cause of vitreoretinochoroidopathy autosomal dominant (ADVIRC) [MIM:193220]. A disorder characterized by vitreoretinochoroidal dystrophy. The clinical presentation is variable and may be associated with cataract, nanophthalmos, microcornea, shallow anterior chamber, and glaucoma.
Sequence similarities	Belongs to the bestrophin family.
Post-translational modifications	Phosphorylated by PP2A.
Cellular localization	Cell membrane. Basolateral cell membrane.

Images



Western blot - Anti-Bestrophin/BEST1 antibody [E6-6] (ab2182)

Anti-Bestrophin/BEST1 antibody [E6-6] (ab2182) at 1/1000 dilution
+ Human RPE cell lysate



Western blot - Anti-Bestrophin/BEST1 antibody [E6-6] (ab2182)

This image is courtesy of an Abreview submitted by Dr Vladimir Milenkovic

All lanes : Anti-Bestrophin/BEST1 antibody [E6-6] (ab2182) at 1/1000 dilution

Lane 1 : Human RPE, retinal pigment epithelial cell lysate
Lane 2 : Non transfected HEK 293 cell extract

Lysates/proteins at 20 µg per lane.

Secondary
All lanes : HRP-conjugated goat anti-mouse

Developed using the ECL technique.

Performed under reducing conditions.

Observed band size: 67 kDa

Exposure time: 5 minutes

The primary antobody was diluted in PBS/Tween/5%Milk and incubated for 1.5 hours at 25°C.

Our Abpromise to you: Quality guaranteed and expert technical support

- Replacement or refund for products not performing as stated on the datasheet
- Valid for 12 months from date of delivery
- Response to your inquiry within 24 hours
- We provide support in Chinese, English, French, German, Japanese and Spanish
- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

If the product does not perform as described on this datasheet, we will offer a refund or replacement. For full details of the Abpromise, please visit **https://www.abcam.com/abpromise** or contact our technical team.

Terms and conditions

- Guarantee only valid for products bought direct from Abcam or one of our authorized distributors