# abcam

## Product datasheet

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

★★★★★ **7 Abreviews**   **30 References**   2 图像

## 概述

| | |
|---|---|
| 产品名称 | Anti-Bestrophin/BEST1抗体[E6-6] |
| 描述 | 小鼠单克隆抗体[E6-6] to Bestrophin/BEST1 |
| 宿主 | Mouse |
| 经测试应用 | **适用于：** WB, IHC-Fr, ICC/IF, IP |
| 种属反应性 | **与反应：** Cow, Dog, Human, Pig, Monkey |
| | **预测可用于：** Non human primates ⚠ **不与反应：** Mouse, Rat, Rabbit, Goat |
| 免疫原 | Synthetic peptide corresponding to Human Bestrophin/BEST1 aa 568-585 (C terminal) conjugated to keyhole limpet haemocyanin.<br>Sequence:<br><br>KDHMDPYWALENRDEAHS<br><br>Database link: **O76090**<br><br>🖼 **Run BLAST with**     🖼 **Run BLAST with** |
| 阳性对照 | IHC-Fr: Pig retinal pigment epithelium tissue. ICC/IF: Bovine retinal pigment epithelium (RPE). WB: Human RPE cell lysate. |
| 常规说明 | This product has switched from ascites to TCS on 9th September 2020.<br><br>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.<br><br>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 |

## 性能

| | |
|---|---|
| 形式 | Liquid |
| 存放说明 | Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles. |
| 存储溶液 | pH: 7.40<br>Preservative: 0.02% Sodium azide |

|  | Constituent: 99% PBS |
|---|---|
| 纯度 | Protein A/G purified |
| 克隆 | 单克隆 |
| 克隆编号 | E6-6 |
| 骨髓瘤 | unknown |
| 同种型 | IgG1 |
| 轻链类型 | kappa |

## 应用

**The Abpromise guarantee**      **Abpromise™承诺保证使用ab2182于以下的经测试应用**

"应用说明"部分 下显示的仅为**推荐的起始稀释度**；实际**最佳的稀释度/浓度应由使用者**检定。

| 应用 | Ab评论 | 说明 |
|---|---|---|
| **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. |

## 靶标

| 功能 | Forms calcium-sensitive chloride channels. Highly permeable to bicarbonate. |
|---|---|
| 组织特异性 | Predominantly expressed in the basolateral membrane of the retinal pigment epithelium. |
| 疾病相关 | 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.

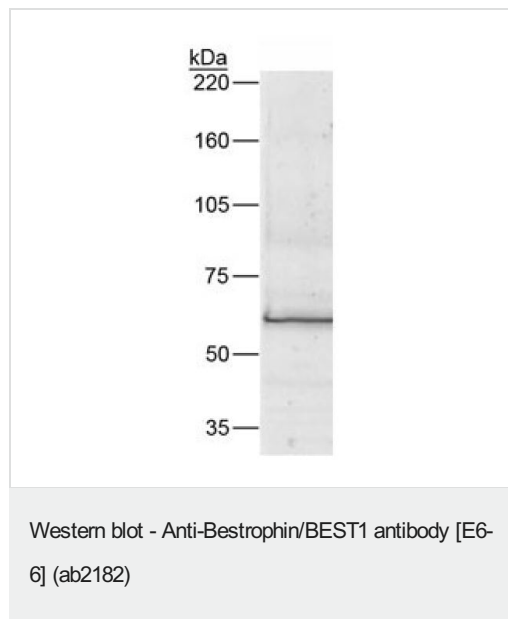| | |
|---|---|
| **序列相似性** | Belongs to the bestrophin family. |
| **翻译后修饰** | Phosphorylated by PP2A. |
| **细胞定位** | Cell membrane. Basolateral cell membrane. |

## 图片



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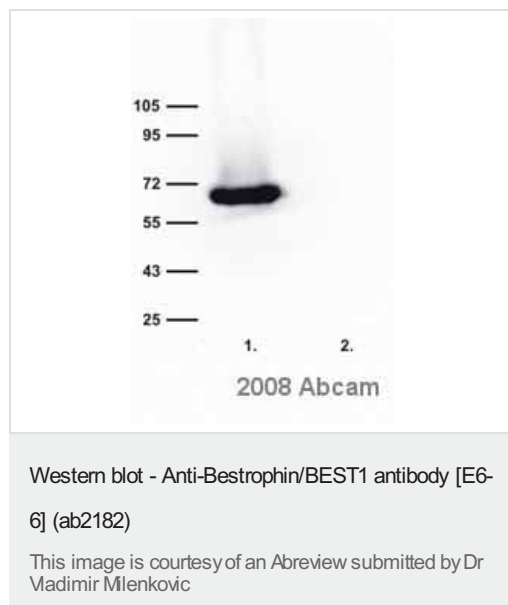
**All lanes :** Anti-Bestrophin/BEST1 antibody [E6-6] (ab2182) at 1/1000 dilution

**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.cn/abpromise** or contact our technical team.

## Terms and conditions

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