

### Anti-Bestrophin/BEST1 antibody ab14927

★★★★★ [6 Abreviews](#) [7 References](#)

#### 概述

产品名称	Anti-Bestrophin/BEST1抗体
描述	兔多克隆抗体to Bestrophin/BEST1
宿主	Rabbit
经测试应用	适用于: ICC, IP, ICC/IF, WB
种属反应性	与反应: Mouse, Rat, Cow, Human 预测可用于: Cynomolgus monkey 
免疫原	Synthetic peptide corresponding to Human Bestrophin/BEST1 aa 1-100. Database link: <a href="#">O76090</a>

 [Run BLAST with](#)

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#### 常规说明

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

#### 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 Preservative: 0.02% Sodium azide Constituents: Tris glycine, 0.5% BSA, 30% Glycerol
纯度	Immunogen affinity purified
克隆	多克隆
同种型	IgG

#### 应用

## The Abpromise guarantee

**Abpromise™** 承诺保证使用ab14927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
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.

## 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 vitreoretinchoroidopathy autosomal dominant (ADVIRC) [MIM:193220]. A disorder characterized by vitreoretinchoroidal 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.

**Please note:** All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

### **Our Abpromise to you: Quality guaranteed and expert technical support**

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- 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.

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