abcam

Product datasheet

Recombinant Human Bestrophin/BEST1 protein ab152797

1 图像

描述

产品名称	重组人Bestrophin/BEST1蛋白
表达系统	Wheat germ
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
序列	MFEKLTLYCDSYIQLIPISFVLGFYVTLVVTRWWNQYENLPW PDRLMSLV SGFVEGKDEQGRLLRRTLIRYANLGNVLILRSVSTAVYKRFP SAQHLVQA GFMTPAEHKQLEKLSLPHNMFWVPWVWFANLSMKAWLGGRIR DPILLQSL LNEMNTLRTQCGHLYAYDWISIPLVYTQVVTVAVYSFFLTCL VGRQFLNP AKAYPGHELDLVVPVFTFLQFFFYVGWLKVAEQLINPFGEDD DDFETNWI VDRNLQVSLLAVDEMHQDLPRMEPDMYWNKPEPQPPYTAASA QFRRVSFM GSTFNISLNKEEMEFQPNQEDEEDAHAGIIGRFLGLQSHDHH PPRANSRT KLLWPKRESLLHEGLPKNHKAAKQNVRGQEDNKAWKLKAVDA FKSAPLYQ RPGYYSAPQTPLSPTPMFFPLEPSAPSKLHSVTGIDTKDKSL KTVSSGAK KSFELLSESDGALMEHPEVSQVRRKTVEFNLTDMPEIPENHL KEPLEQSP TNIHTTLKDHMDPYWALENRSVLHLNQGHCIALCPTPASLAL SLPFLHNF LGFHHCQSTLDLRPALAWGIYLATFTGILGKCSGPFLTSPWY HPEDFLGP GEGR
预测分子量	96 kDa including tags
氨基酸	1 to 604

技术指标

技术指标

Our **Abpromise guarantee** covers the use of **ab152797** in the following tested applications.

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

应用	ELISA SDS-PAGE Western blot
形式	Liquid
补充说明	This product was previously labelled as Bestrophin.

制备和贮存

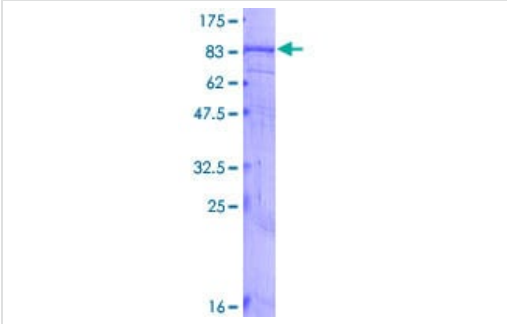
稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

常规信息

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



SDS-PAGE - Recombinant Human Bestrophin/BEST1 protein (ab152797)

12.5% SDS-PAGE showing ab152797 stained with Coomassie Blue.

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