

## Product datasheet

# Recombinant Human Bestrophin/BEST1 protein ab152797

[1 Image](#)

### Description

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<b>Product name</b>	Recombinant Human Bestrophin/BEST1 protein
<b>Expression system</b>	Wheat germ
<b>Protein length</b>	Full length protein
<b>Animal free</b>	No
<b>Nature</b>	Recombinant
<b>Species</b>	Human
<b>Sequence</b>	MF EKLTLYCDSYIQLIPISFVLGFYVTLVVTRWWNQYENLP WPDRLMSLV SGFVEGKDEQGRLRRLIRYANLGNVLILRSVSTAVYKRF PSAQHLVQA GFMPAEHKQLEKLSLPHNMFVWPVWVWFANLSMKAWL GGRIRDPIILLQSL LNEMNLTQTQCGHLYAYDWISIPLVYTQVTVAVYSFFLTC LVGRQFLNP AKAYPGHELDLVVPVFTFLQFFFYVGWLKVAEQLINPFGE DDDDFETNWI VDRNLQVSLLAVIDEMHQDLPRMEPDMYWNKPEPQPPYT AASAQFRRVSFM GSTFNISLNKEEMEFQPNQEDEEDAHAGIIGRFLGLQSHD HHPPRANSRT KLLWPKRESLLHEGLPKNHKAAKQNVRGQEDNKAWKLN AVDAFKSAPLYQ RPGYYSAPQTPLSPTPMFFPLEPSAPSKLHVSVDGIDTKDK SLKTVSSGAK KSFELLSESDGALMEHPEVSQVRRKTVFNLDMPEIPE NHLKEPLEQSP TNIHTTLKDHMDPYWALENRSVLHLNQGHICALCPTPASLA LSLPFLHNF LGFHHCQSTLDRPALAWGYLATFTGILGKCSGPFLTSPW YHPEDFLGP GEGR
<b>Predicted molecular weight</b>	96 kDa including tags
<b>Amino acids</b>	1 to 604

## Specifications

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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.

<b>Applications</b>	ELISA SDS-PAGE Western blot
<b>Form</b>	Liquid
<b>Additional notes</b>	This product was previously labelled as Bestrophin.

## Preparation and Storage

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<b>Stability and Storage</b>	Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl
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## General Info

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<b>Function</b>	Forms calcium-sensitive chloride channels. Highly permeable to bicarbonate.
<b>Tissue specificity</b>	Predominantly expressed in the basolateral membrane of the retinal pigment epithelium.
<b>Involvement in disease</b>	<p>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.</p> <p>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.</p> <p>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.</p> <p>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.</p> <p>Defects in BEST1 are the cause of vitreoretinopathology autosomal dominant (ADVIRC) [MIM:193220]. A disorder characterized by vitreoretinopathology. The clinical presentation is variable and may be associated with cataract, nanophthalmos, microcornea, shallow anterior chamber, and glaucoma.</p>
<b>Sequence similarities</b>	Belongs to the bestrophin family.
<b>Post-translational</b>	Phosphorylated by PP2A.

## modifications

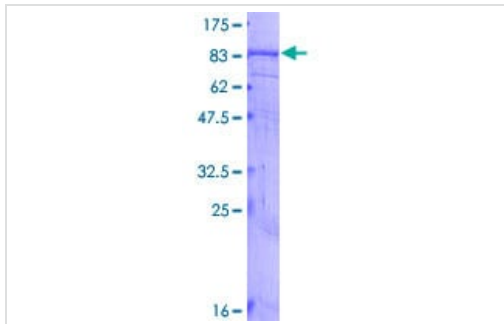
## Cellular localization

Cell membrane. Basolateral cell membrane.

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## Images

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

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

**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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