Bestrophin (E6-6): sc-32792

SANTA CRUZ®
BIOTECHNOLOGY

The Power to Question

BACKGROUND

The retinal pigment epithelium (RPE) and choroid represents a differentiated system of the eye that sustains normal retinal health and function. Best vitelliform macular dystrophy, known as Best disease, is an early-onset autosomal dominant condition in which accumulation of lipofuscin-like material within and beneath the RPE leads to progressive loss of central vision. The lipofuscin-like material in the macular area appears as a yellow mass like the yolk of an egg that later becomes darker and irregular in color, a process known as "scrambling the egg". Best disease is frequently a reflection of mutations in the Bestrophin gene, which encodes a protein containing four putative transmembrane domains and localizes to the basolateral plasma membrane of RPE cells. Human Bestrophin forms oligomeric chloride channels that are sensitive to intracellular calcium. Missense mutations at the Bestrophin locus reduces or abolishes Bestrophin protein mediated membrane current. The human Bestrophin gene maps to chromosome 11q12.3 and encodes a 585 amino acid protein.

CHROMOSOMAL LOCATION

Genetic locus: BEST1 (human) mapping to 11q12.3.

SOURCE

Bestrophin (E6-6) is a mouse monoclonal antibody raised against a synthetic peptide corresponding to the C-terminus of Bestrophin of human origin.

PRODUCT

Each vial contains 200 µg IgG$_1$ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Bestrophin (E6-6) is available conjugated to agarose (sc-32792 AC), 500 µg/ 0.25 ml agarose in 1 ml, for IP; to HRP (sc-32792 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-32792 PE), fluorescein (sc-32792 FITC), Alexa Fluor® 488 (sc-32792 AF488), Alexa Fluor® 546 (sc-32792 AF546), Alexa Fluor® 594 (sc-32792 AF594) or Alexa Fluor® 647 (sc-32792 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-32792 AF680) or Alexa Fluor® 790 (sc-32792 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

Bestrophin (E6-6) is recommended for detection of Bestrophin of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and immunohistochemistry (including paraffin-embedded sections) (starting dilution 1:50, dilution range 1:50-1:500); non cross-reactive with mouse or rat.

Suitable for use as control antibody for Bestrophin siRNA (h): sc-40368, Bestrophin shRNA Plasmid (h): sc-40368-SH and Bestrophin shRNA (h) Lentiviral Particles: sc-40368-V.

Molecular Weight of Bestrophin: 66 kDa.

Positive Controls: Bestrophin (h): 293T Lysate: sc-113613, Y79 cell lysate: sc-2240 or SK-N-SH cell lysate: sc-2410.

RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohistomount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA
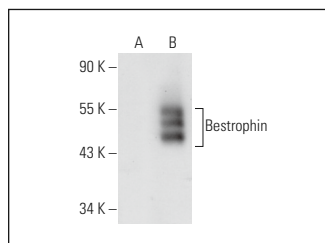
Bestrophin (E6-6): sc-32792. Western blot analysis of Bestrophin expression in non-transfected: sc-117752 (**A**) and human Bestrophin transfected: sc-113613 (**B**) 293T whole cell lysates.
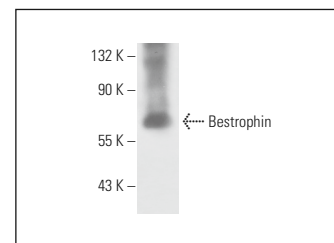
Bestrophin (E6-6): sc-32792. Western blot analysis of Bestrophin expression in porcine retinal epithelium tissue extract.

SELECT PRODUCT CITATIONS

1. Duta, V., et al. 2006. Regulation of basolateral Cl⁻ channels in airway epithelial cells: the role of nitric oxide. J. Membr. Biol. 213: 165-174.

2. Guziewicz, K.E., et al. 2011. Molecular consequences of BEST1 gene mutations in canine multifocal retinopathy predict functional implications for human bestrophinopathies. Invest. Ophthalmol. Vis. Sci. 52: 4497-4505.

3. Hansson, M.L., et al. 2015. Efficient delivery and functional expression of transfected modified mRNA in human embryonic stem cell-derived retinal pigmented epithelial cells. J. Biol. Chem. 290: 5661-5672.

4. Lytvynchuk, L., et al. 2022. Subretinal implantation of human primary RPE cells cultured on nanofibrous membranes in minipigs. Biomedicines 10: 669.

STORAGE

Store at 4° C, **DO NOT FREEZE**. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

Alexa Fluor® is a trademark of Molecular Probes, Inc., Oregon, USA