

Bestrophin (H-93): sc-292163

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 encodes a 585 amino acid protein.

REFERENCES

- Best, F. 1905. Ueber eine hereditaere Maculaaffektion. Z. Augenheilk 13: 199-212.
- Brale, A. E. 1966. Dystrophy of the macula. Am. J. Ophthalmol. 61: 1-24.
- Marmorstein, A.D., et al. 2000 Bestrophin, the product of the Best vitelliform macular dystrophy gene (VMD2), localizes to the basolateral plasma membrane of the retinal pigment epithelium. Proc. Nat. Acad. Sci. 97: 12758-12763.
- Musarella, M.A. 2001. Molecular genetics of macular degeneration. Doc. Ophthalmol. 102: 165-177.
- Tavsanli, B.C., et al. 2001. Dbest1, a *Drosophila* homolog of human Bestrophin, is not required for viability or photoreceptor integrity. Genesis 31: 130-136.
- Wistow, G., et al. 2002. Expressed sequence tag analysis of human RPE/choroid for the NEIBank Project: over 6000 non-redundant transcripts, novel genes and splice variants. Mol. Vis. 8: 205-220.

CHROMOSOMAL LOCATION

Genetic locus: BEST1 (human) mapping to 11q12.2.

SOURCE

Bestrophin (H-93) is a rabbit polyclonal antibody raised against amino acids 493-585 mapping at the C-terminus of Bestrophin-1 of human origin.

PRODUCT

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

STORAGE

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

APPLICATIONS

Bestrophin (H-93) is recommended for detection of Bestrophin-1 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 solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

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: 68 kDa.

Positive Controls: Y79 cell lysate: sc-2240 or SK-N-SH cell lysate: sc-2410.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use goat anti-rabbit IgG-HRP: sc-2004 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible goat anti-rabbit IgG-HRP: sc-2030 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 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 goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

RESEARCH USE

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

PROTOCOLS

See our web site at www.scbt.com or our catalog for detailed protocols and support products.



Try **Bestrophin (E6-6): sc-32792**, our highly recommended monoclonal alternative to Bestrophin (H-93).