SANTA CRUZ BIOTECHNOLOGY, INC.

Bestrophin (C-14): sc-22027

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.

REFERENCES

1. Best, F. 1905. Ueber eine hereditaere Maculaaffektion. Z. Augenheilk 13: 199-212.
2. Braley, A. E. 1966. Dystrophy of the macula. Am. J. Ophthalmol. 61: 1-24.
3. Petrukhin, K., et al. 1998. Identification of the gene responsible for Best macular dystrophy. Nat. Genet. 19: 241-247.
3. 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.
4. Musarella, M.A. 2001. Molecular genetics of macular degeneration. Doc. Ophthalmol. 102: 165-177.
5. 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.

CHROMOSOMAL LOCATION

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

SOURCE

Bestrophin (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of Bestrophin 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.

Blocking peptide available for competition studies, sc-22027 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

STORAGE

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

APPLICATIONS

Bestrophin (C-14) 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 solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

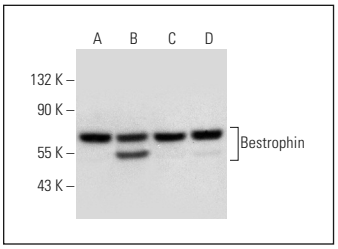
Bestrophin (C-14) is also recommended for detection of Bestrophin in additional species, including equine, canine and porcine.

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.

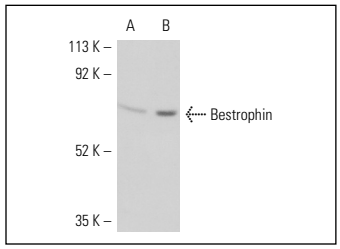
Molecular Weight of Bestrophin: 66 kDa.

Positive Controls: NTERA-2 cl.D1 whole cell lysate: sc-364181, Y79 cell lysate: sc-2240 or SK-N-SH cell lysate: sc-2410.

DATA



Bestrophin (C-14): sc-22027. Western blot analysis of Bestrophin expression in ARPE-19 (**A**), HEK293 (**B**), NTERA-2 cl.D1 (**C**) and PC-3 (**D**) whole cell lysates.



Bestrophin (C-14): sc-22027. Western blot analysis of Bestrophin expression in Y79 (**A**) and SK-N-SH (**B**) whole cell lysates.

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. Gouras, P., et al. 2009. Bestrophin detected in the basal membrane of the retinal epithelium and drusen of monkeys with drusenoid maculopathy. Graefes Arch. Clin. Exp. Ophthalmol. 247: 1051-1056.
3. 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.

RESEARCH USE

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