SANTA CRUZ BIOTECHNOLOGY, INC.

BBS4 (E-20): sc-49386

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The Power to Question

BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, renal abnormalities and developmental delay. Other associated clinical findings in BBS patients include diabetes, hypertension and congenital heart defects. BBS is a heterogeneous disorder mapping to eight genetic loci and encoding eight proteins, BBS1-BBS8. Five BBS proteins encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS4 is expressed in the olfactory epithelium and localizes to the centriolar satellites of centrosomes and basal bodies of primary cilia. BBS4 regulates the p150 subunit of the dynein transport machinery (DCTN1) to attract pericentriolar material-1 protein (PCM1) and its associated components to the satellites. Loss of BBS4 is correlated with obesity caused by abnormal lipid profiles, liver dysfunction, elevated Insulin and abnormal leptin levels.

REFERENCES

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3. Dollfus, H., et al. 2005. Update on Bardet-Biedl syndrome. J. Fr. Ophtalmol. 28: 106-112.
4. Karmous-Benailly, H., et al. 2005. Antenatal presentation of Bardet-Biedl syndrome may mimic Meckel syndrome. Am. J. Hum. Genet. 76: 493-504.
5. Ahmad, J., et al. 2005. DFNB48, a new nonsyndromic recessive deafness locus, maps to chromosome 15q23-q25.1. Hum. Genet. 116: 407-412.
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7. Iannaccone, A., et al. 2005. Clinical evidence of decreased olfaction in Bardet-Biedl syndrome caused by a deletion in the BBS4 gene. Am. J. Med. Genet. A 132: 343-346.
8. Nakane, T., et al. 2005. No evidence for triallelic inheritance of MKKS/BBS loci in Amish Mckusick-Kaufman syndrome. Am. J. Med. Genet. A 138: 32-34.
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CHROMOSOMAL LOCATION

Genetic locus: BBS4 (human) mapping to 15q24.1; Bbs4 (mouse) mapping to 9 B.

SOURCE

BBS4 (E-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping near the N-terminus of BBS4 of human origin.

STORAGE

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

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-49386 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

Available as TransCruz reagent for Gel Supershift and ChIP applications, sc-49386 X, 200 µg/0.1 ml.

APPLICATIONS

BBS4 (E-20) is recommended for detection of BBS4 (Bardet-Biedl syndrome 4 protein) of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

BBS4 (E-20) is also recommended for detection of BBS4 (Bardet-Biedl syndrome 4 protein) in additional species, including equine, canine, bovine, porcine and avian.

Suitable for use as control antibody for BBS4 siRNA (h): sc-60255, BBS4 siRNA (m): sc-60256, BBS4 shRNA Plasmid (h): sc-60255-SH, BBS4 shRNA Plasmid (m): sc-60256-SH, BBS4 shRNA (h) Lentiviral Particles: sc-60255-V and BBS4 shRNA (m) Lentiviral Particles: sc-60256-V.

BBS4 (E-20) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of BBS4: 58 kDa.

Positive Controls: Mouse heart extract: sc-2254, rat heart extract: sc-2393 or rat kidney extract: sc-2394.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (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.