SANTA CRUZ BIOTECHNOLOGY, INC.

BBS6 (H-300): sc-134456

BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, renal abnormalities, and developmental delay. BBS patients also have an increased risk of developing diabetes, hypertension, and congenital heart defects. BBS is a heterogeneous disorder; BBS genes map to eight genetic loci and encode eight proteins, BBS1-BBS8. Five BBS genes encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS6 is a Group II chaperonin-like protein that has evolved recently in animals from CCT/TRiC, a subunit of the eukaryotic chaperonin. Most of BBS6 localizes to the pericentriolar material (PCM), a proteinaceous tube surrounding centrioles. During interphase, BBS6 is restricted to the lateral surfaces of the PCM, but during mitosis, it relocalizes throughout the PCM and localizes to the intercellular bridge.

REFERENCES

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3. Andersen, K.L., et al. 2005. Variation of the McKusick-Kaufman gene and studies of relationships with common forms of obesity. J. Clin. Endocrinol. Metab. 90: 225-230.
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6. Hichri, H., et al. 2005. Testing for triallelism: analysis of six BBS genes in a Bardet-Biedl syndrome family cohort. Eur. J. Hum. Genet. 13: 607-616.
7. Karmous-Benailly, H., et al. 2005. Antenatal presentation of Bardet-Biedl syndrome may mimic Meckel syndrome. Am. J. Hum. Genet. 76: 493-504.
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CHROMOSOMAL LOCATION

Genetic locus: MKKS (human) mapping to 20p12.2; Mkks (mouse) mapping to 2 F3.

SOURCE

BBS6 (H-300) is a rabbit polyclonal antibody raised against amino acids 271-570 mapping at the C-terminus of BBS6 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.

APPLICATIONS

BBS6 (H-300) is recommended for detection of BBS6 of mouse, rat and 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).

BBS6 (H-300) is also recommended for detection of BBS6 in additional species, including canine.

Molecular Weight BBS6: 62 kDa.

Positive Controls: SK-N-SH cell lysate: sc-2410 or HeLa whole cell lysate: sc-2200.

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.

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.

PROTOCOLS

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