SANTA CRUZ BIOTECHNOLOGY, INC.

BBS6 (N-14): sc-49799

SANTA CRUZ BIOTECHNOLOGY

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. 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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2. Badano, J.L., et al. 2003. Heterozygous mutations in BBS1, BBS2 and BBS6 have a potential epistatic effect on Bardet-Biedl patients with two mutations at a second BBS locus. Hum. Mol. Genet. 12: 1651-1659.
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.
4. Dollfus, H., et al. 2005. Update on Bardet-Biedl syndrome. J. Fr. Ophtalmol. 28: 106-112.
5. Heon, E., et al. 2005. Ocular phenotypes of three genetic variants of Bardet-Biedl syndrome. Am. J. Med. Genet. A 132A: 283-287.
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.

SOURCE

BBS6 (N-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of BBS6 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. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-49799 X, 200 µg/0.1 ml.

Blocking peptide available for competition studies, sc-49799 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

BBS6 (N-14) is recommended for detection of BBS6 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).

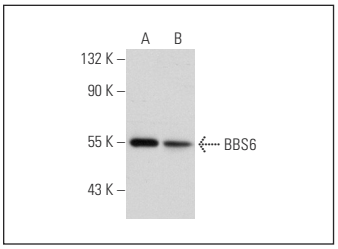
Suitable for use as control antibody for BBS6 siRNA (h): sc-60257, BBS6 shRNA Plasmid (h): sc-60257-SH and BBS6 shRNA (h) Lentiviral Particles: sc-60257-V.

BBS6 (N-14) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

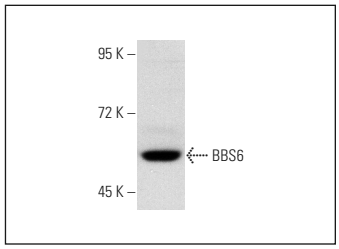
Molecular Weight of BBS6: 62 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, SK-N-SH cell lysate: sc-2410 or BBS6 (h): 293T Lysate: sc-113970.

DATA



BBS6 (N-14): sc-49799. Western blot analysis of BBS6 expression in non-transfected: sc-117752 (**A**) and human BBS6 transfected: sc-113970 (**B**) 293T whole cell lysates.



BBS6 (N-14): sc-49799. Western blot analysis of BBS6 expression in SK-N-SH whole cell lysate.

RESEARCH USE

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