

BBS6 (F-8): sc-390077

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 relocates throughout the PCM and localizes to the intercellular bridge.

REFERENCES

1. Beales, P.L., et al. 2001. Genetic and mutational analyses of a large multi-ethnic Bardet-Biedl cohort reveal a minor involvement of BBS6 and delineate the critical intervals of other loci. *Am. J. Hum. Genet.* 68: 606-616.
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.

CHROMOSOMAL LOCATION

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

SOURCE

BBS6 (F-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 14-37 at the N-terminus of BBS6 of human origin.

PRODUCT

Each vial contains 200 µg IgM kappa light chain 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-390077 X, 200 µg/0.1 ml.

Blocking peptide available for competition studies, sc-390077 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

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

APPLICATIONS

BBS6 (F-8) is recommended for detection of BBS6 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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), immunohistochemistry (including paraffin-embedded sections) (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 siRNA (m): sc-60258, BBS6 shRNA Plasmid (h): sc-60257-SH, BBS6 shRNA Plasmid (m): sc-60258-SH, BBS6 shRNA (h) Lentiviral Particles: sc-60257-V and BBS6 shRNA (m) Lentiviral Particles: sc-60258-V.

BBS6 (F-8) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

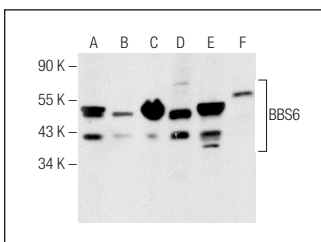
Molecular Weight of BBS6: 62 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204, NTERA-2 cl.D1 whole cell lysate: sc-364181 or SK-LMS-1 cell lysate: sc-3813.

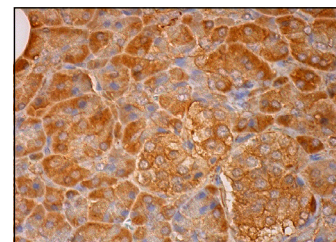
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein L-Agarose: sc-2336 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850. 4) Immunohistochemistry: use m-IgGκ BP-HRP: sc-516102 with DAB, 50X: sc-24982 and Immunohisto-mount: sc-45086, or Organo/Limonene Mount: sc-45087.

DATA



BBS6 (F-8): sc-390077. Western blot analysis of BBS6 expression in MIA PaCa-2 (A), Jurkat (B), SK-LMS-1 (C), IMR-32 (D), WI-38 (E) and NTERA-2 cl.D1 (F) whole cell lysates.



BBS6 (F-8): sc-390077. Immunoperoxidase staining of formalin fixed, paraffin-embedded human pancreas tissue showing cytoplasmic staining of exocrine glandular cells and cytoplasmic and membrane staining of Islets of Langerhans.

RESEARCH USE

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