

BBS8 (C-14): sc-49806

BACKGROUND

Bardet-Biedl syndrome (BBS) is a heterogeneous pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogonadism, renal abnormalities, developmental delay, diabetes, hypertension and congenital heart defects. 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. Mutations in BBS8, also designated tetratricopeptide repeat protein (TTC8), probably account for only a minority (2%) of BBS families, underlining the difficulty of genotyping heterogeneous conditions. The identification of BBS8 provides the key to the pathogenesis of the condition as a primary ciliary disorder.

REFERENCES

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4. Dollfus, H., Verloes, A., Bonneau, D., Cossee, M., Perrin-Schmitt, F., Brandt, C., Flament, J. and Mandel, J.L. 2005. Update on Bardet-Biedl syndrome. *J. Fr. Ophthalmol.* 28: 106-112.

CHROMOSOMAL LOCATION

Genetic locus: TTC8 (human) mapping to 14q31.3; Ttc8 (mouse) mapping to 12 E.

SOURCE

BBS8 (C-14) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of BBS8 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-49806 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.

RESEARCH USE

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

APPLICATIONS

BBS8 (C-14) is recommended for detection of BBS8 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); cross-reacts with isoforms 1-5.

BBS8 (C-14) is also recommended for detection of BBS8 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for BBS8 siRNA (h): sc-60261, BBS8 siRNA (m): sc-60262, BBS8 shRNA Plasmid (h): sc-60261-SH, BBS8 shRNA Plasmid (m): sc-60262-SH, BBS8 shRNA (h) Lentiviral Particles: sc-60261-V and BBS8 shRNA (m) Lentiviral Particles: sc-60262-V.

BBS8 (C-14) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

Molecular Weight of BBS8: 62 kDa.

Positive Controls: LNCaP cell lysate: sc-2231, SK-BR-3 cell lysate: sc-2218 or mouse liver extract: sc-2256.

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.

PROTOCOLS

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