

BBS4 (1292CT845.130.218): sc-517315

BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogonadism, renal abnormalities, and developmental delay. Other associated clinical findings in BBS patients include 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. 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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CHROMOSOMAL LOCATION

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

SOURCE

BBS4 (1292CT845.130.218) is a mouse monoclonal antibody raised against a recombinant protein corresponding to amino acids 1-240 of BBS4 of human origin.

PRODUCT

Each vial contains 100 µg IgG₁ in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

APPLICATIONS

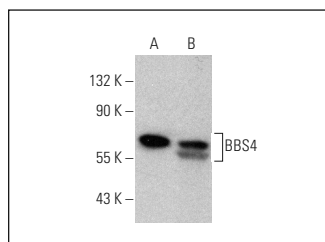
BBS4 (1292CT845.130.218) is recommended for detection of BBS4 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000) and immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)].

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.

Molecular Weight of BBS4: 58 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200 or U-251-MG whole cell lysate: sc-364176.

DATA



BBS4 (1292CT845.130.218): sc-517315. Western blot analysis of BBS4 expression in HeLa (A) and U-251-MG (B) whole cell lysates.

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 for detailed protocols and support products.