

BBS4 (F-24): sc-130699

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 mapping to eight genetic loci and encoding eight proteins, BBS1-BBS8. Five BBS proteins 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.

SOURCE

BBS4 (F-24) is a purified rabbit polyclonal antibody raised against a peptide mapping within an internal region of BBS4 of human origin.

PRODUCT

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

APPLICATIONS

BBS4 (F-24) is recommended for detection of BBS4 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)] and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for BBS4 siRNA (h): sc-60255, BBS4 shRNA Plasmid (h): sc-60255-SH and BBS4 shRNA (h) Lentiviral Particles: sc-60255-V.

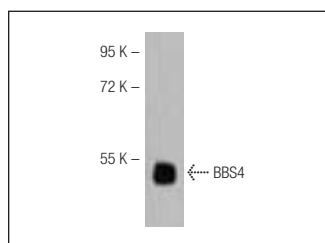
Molecular Weight of BBS4: 58 kDa.

Positive Controls: 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).

DATA



BBS4 (F-24): sc-130699. Western blot analysis of BBS4 expression in HeLa whole cell lysate.

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.