

BBS2 (H-246): sc-99057

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 that maps to eight genetic loci and encodes eight proteins, BBS1-BBS8. Five BBS genes encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS2 is a 721-amino acid protein that is evolutionarily conserved and is expressed in a broad range of tissues including: brain, kidney, adrenal gland, and thyroid gland. Loss of BBS2 may be involved in defects in social interactions as well as infertility. BBS2 retinopathy involves normal retina development followed by apoptotic death of photoreceptors, the primary ciliated cells of the retina.

REFERENCES

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7. 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.
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CHROMOSOMAL LOCATION

Genetic locus: BBS2 (human) mapping to 16q12.2; Bbs2 (mouse) mapping to 8 C5.

SOURCE

BBS2 (H-246) is a rabbit polyclonal antibody raised against amino acids 55-300 mapping near the N-terminus of BBS2 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.

APPLICATIONS

BBS2 (H-246) is recommended for detection of BBS2 of mouse, rat and 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).

BBS2 (H-246) is also recommended for detection of BBS2 in additional species, including equine, canine, bovine and porcine.

Suitable for use as control antibody for BBS2 siRNA (h): sc-60251, BBS2 siRNA (m): sc-60252, BBS2 shRNA Plasmid (h): sc-60251-SH, BBS2 shRNA Plasmid (m): sc-60252-SH, BBS2 shRNA (h) Lentiviral Particles: sc-60251-V and BBS2 shRNA (m) Lentiviral Particles: sc-60252-V.

Molecular Weight of BBS2: 80 kDa.

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). 3) Immunofluorescence: use goat anti-rabbit IgG-FITC: sc-2012 (dilution range: 1:100-1:400) or goat anti-rabbit IgG-TR: sc-2780 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.



Try **BBS2 (A-12): sc-365355**, our highly recommended monoclonal alternative to BBS2 (H-246).