

# BBS1 (H-300): sc-134455

## BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogonadism, 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 mapping to eight genetic loci and encoding eight proteins, BBS1-BBS8. Five BBS genes encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS1 is the protein most commonly involved in Bardet-Biedl syndrome. The BBS1 gene is ubiquitously expressed, with highest abundance in fetal tissues, testes, retina, and adipose tissue. BBS1 is highly conserved in mammals and is inherited in an autosomal recessive manner. Missense mutations in the BBS1 gene account for approximately 80% of all BBS1 mutations.

## CHROMOSOMAL LOCATION

Genetic locus: BBS1 (human) mapping to 11q13.2; Bbs1 (mouse) mapping to 19 A.

## SOURCE

BBS1 (H-300) is a rabbit polyclonal antibody raised against amino acids 291-590 mapping near the C-terminus of BBS1 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. Also available as TransCruz reagent for Gel Supershift and ChIP applications, sc-134455 X, 200 µg/0.1 ml.

## STORAGE

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

## APPLICATIONS

BBS1 (H-300) is recommended for detection of BBS1 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).

BBS1 (H-300) is also recommended for detection of BBS1 in additional species, including equine, canine and porcine.

Suitable for use as control antibody for BBS1 siRNA (h): sc-60249, BBS1 siRNA (m): sc-60250, BBS1 shRNA Plasmid (h): sc-60249-SH, BBS1 shRNA Plasmid (m): sc-60250-SH, BBS1 shRNA (h) Lentiviral Particles: sc-60249-V and BBS1 shRNA (m) Lentiviral Particles: sc-60250-V.

BBS1 (H-300) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

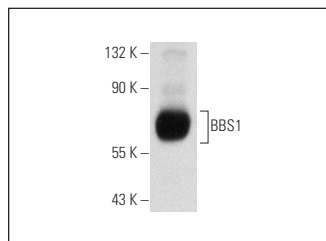
Molecular Weight of BBS1: 65 kDa.

Positive Controls: mouse embryonic brain tissue extract.

## 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.

## DATA



BBS1 (H-300): sc-134455. Western blot analysis of BBS1 expression in mouse embryonic brain tissue extract.

## RESEARCH USE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) or our catalog for detailed protocols and support products.

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Try **BBS1 (F-1): sc-365138**, our highly recommended monoclonal alternative to BBS1 (H-300).