SANTA CRUZ BIOTECHNOLOGY, INC.

BBS7 (E-8): sc-390403

SANTA CRUZ BIOTECHNOLOGY

The Power to Question

BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, 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; 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. The BBS2 gene contains two overlapping genes: BBS2L1 and BBS2L2. BBSL1 was re-named BBS7, whereas BBS2L2 independently funcitons as BBS1. BBS7 contains 672 amino acids and is expressed at low to moderate levels in most human tissues.

REFERENCES

1. Mykytyn, K., et al. 2002. Identification of the gene (BBS1) most commonly involved in Bardet-Biedl syndrome, a complex human obesity syndrome. Nat. Genet. 31: 435-438.
2. Online Mendelian Inheritance in Man, OMIM™. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 607590. World Wide Web URL: http://www.ncbi.nlm.nih.gov/omim/
3. Badano, J.L., et al. 2003. Identification of a novel Bardet-Biedl syndrome protein, BBS7, that shares structural features with BBS1 and BBS2. Am. J. Hum. Genet. 72: 650-658.
4. 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.

CHROMOSOMAL LOCATION

Genetic locus: BBS7 (human) mapping to 4q27; Bbs7 (mouse) mapping to 3 B.

SOURCE

BBS7 (E-8) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 483-521 within an internal region of BBS7 of human origin.

PRODUCT

Each vial contains 200 µg IgG_1 lambda light chain 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-390403 X, 200 µg/0.1 ml.

BBS7 (E-8) is available conjugated to agarose (sc-390403 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-390403 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-390403 PE), fluorescein (sc-390403 FITC), Alexa Fluor® 488 (sc-390403 AF488), Alexa Fluor® 546 (sc-390403 AF546), Alexa Fluor® 594 (sc-390403 AF594) or Alexa Fluor® 647 (sc-390403 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-390403 AF680) or Alexa Fluor® 790 (sc-390403 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

Blocking peptide available for competition studies, sc-390403 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

APPLICATIONS

BBS7 (E-8) is recommended for detection of BBS7 of mouse, rat and human origin by Western Blotting (starting dilution 1:100, 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).

BBS7 (E-8) is also recommended for detection of BBS7 in additional species, including canine, bovine and porcine.

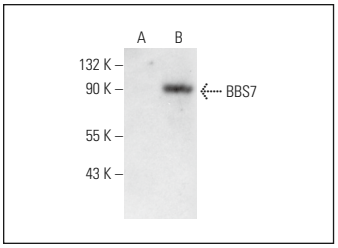
Suitable for use as control antibody for BBS7 siRNA (h): sc-60259, BBS7 siRNA (m): sc-60260, BBS7 shRNA Plasmid (h): sc-60259-SH, BBS7 shRNA Plasmid (m): sc-60260-SH, BBS7 shRNA (h) Lentiviral Particles: sc-60259-V and BBS7 shRNA (m) Lentiviral Particles: sc-60260-V.

BBS7 (E-8) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

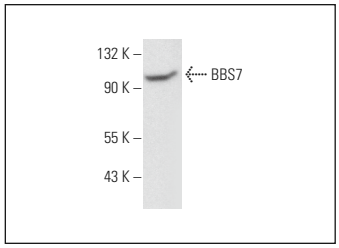
Molecular Weight of BBS7: 80 kDa.

Positive Controls: BBS7 (m): 293T Lysate: sc-118689 or RPE-J cell lysate: sc-24771.

DATA



BBS7 (E-8): sc-390403. Western blot analysis of BBS7 expression in non-transfected: sc-117752 (**A**) and mouse BBS7 transfected: sc-118689 (**B**) 293T whole cell lysates.



BBS7 (E-8): sc-390403. Western blot analysis of BBS7 expression in RPE-J whole cell lysate. Detection reagent used: m-IgGλ BP-HRP (Cruz Marker): sc-516132-CM.

SELECT PRODUCT CITATIONS

1. Barbelanne, M., et al. 2015. Nephrocystin proteins NPHP5 and Cep290 regulate BBSome integrity, ciliary trafficking and cargo delivery. Hum. Mol. Genet. 24: 2185-2200.

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.