

BBS2 (A-12): sc-365355

BACKGROUND

Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, 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.

CHROMOSOMAL LOCATION

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

SOURCE

BBS2 (A-12) is a mouse monoclonal antibody raised against amino acids 55-300 mapping near the N-terminus of BBS2 of human origin.

PRODUCT

Each vial contains 200 µg IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

BBS2 (A-12) is available conjugated to agarose (sc-365355 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-365355 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-365355 PE), fluorescein (sc-365355 FITC), Alexa Fluor® 488 (sc-365355 AF488), Alexa Fluor® 546 (sc-365355 AF546), Alexa Fluor® 594 (sc-365355 AF594) or Alexa Fluor® 647 (sc-365355 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-365355 AF680) or Alexa Fluor® 790 (sc-365355 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

BBS2 (A-12) is recommended for detection of BBS2 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).

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.

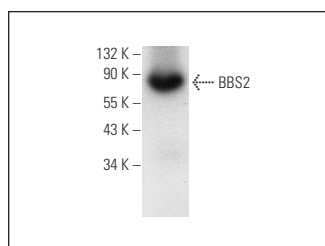
Molecular Weight of BBS2: 80 kDa.

Positive Controls: NTERA-2 cl.D1 whole cell lysate: sc-364181 or SW-13 cell lysate: sc-24778.

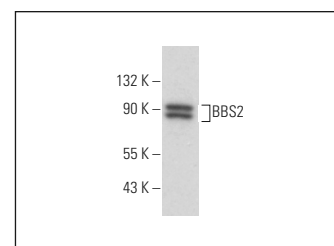
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 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 m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

DATA



BBS2 (A-12): sc-365355. Western blot analysis of BBS2 expression in NTERA-2 cl.D1 whole cell lysate.



BBS2 (A-12): sc-365355. Western blot analysis of BBS2 expression in SW-13 whole cell lysate.

SELECT PRODUCT CITATIONS

- Klinger, M., et al. 2014. The novel centriolar satellite protein SSX2IP targets Cep290 to the ciliary transition zone. *Mol. Biol. Cell* 25: 495-507.
- Barbelanne, M., et al. 2015. Nephrocystin proteins NPHP5 and Cep290 regulate BBSome integrity, ciliary trafficking and cargo delivery. *Hum. Mol. Genet.* 24: 2185-2200.
- Datta, P., et al. 2015. Accumulation of non-outer segment proteins in the outer segment underlies photoreceptor degeneration in Bardet-Biedl syndrome. *Proc. Natl. Acad. Sci. USA* 112: E4400-E4409.
- Desai, P.B., et al. 2020. Ubiquitin links smoothed to intraflagellar transport to regulate Hedgehog signaling. *J. Cell Biol.* 219: e201912104.
- Prasai, A., et al. 2020. The BBSome assembly is spatially controlled by BBS1 and BBS4 in human cells. *J. Biol. Chem.* 295: 14279-14290.
- Hsu, Y., et al. 2021. Photoreceptor cilia, in contrast to primary cilia, grant entry to a partially assembled BBSome. *Hum. Mol. Genet.* 30: 87-102.

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.