

BBS8 (E-2): sc-271009

BACKGROUND

Bardet-Biedl syndrome (BBS) is a heterogeneous pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, renal abnormalities, developmental delay, diabetes, hypertension and congenital heart defects. 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. Mutations in BBS8, also designated tetratricopeptide repeat protein (TTC8), probably account for only a minority (2%) of BBS families, underlining the difficulty of genotyping heterogeneous conditions. The identification of BBS8 provides the key to the pathogenesis of the condition as a primary ciliary disorder.

CHROMOSOMAL LOCATION

Genetic locus: TTC8 (human) mapping to 14q31.3; Ttc8 (mouse) mapping to 12 E.

SOURCE

BBS8 (E-2) is a mouse monoclonal antibody raised against amino acids 232-531 mapping at the C-terminus of BBS8 of human origin.

PRODUCT

Each vial contains 200 µg IgG_{2b} kappa 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-271009 X, 200 µg/0.1 ml.

BBS8 (E-2) is available conjugated to agarose (sc-271009 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-271009 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-271009 PE), fluorescein (sc-271009 FITC), Alexa Fluor® 488 (sc-271009 AF488), Alexa Fluor® 546 (sc-271009 AF546), Alexa Fluor® 594 (sc-271009 AF594) or Alexa Fluor® 647 (sc-271009 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-271009 AF680) or Alexa Fluor® 790 (sc-271009 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

APPLICATIONS

BBS8 (E-2) is recommended for detection of BBS8 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 BBS8 siRNA (h): sc-60261, BBS8 siRNA (m): sc-60262, BBS8 shRNA Plasmid (h): sc-60261-SH, BBS8 shRNA Plasmid (m): sc-60262-SH, BBS8 shRNA (h) Lentiviral Particles: sc-60261-V and BBS8 shRNA (m) Lentiviral Particles: sc-60262-V.

BBS8 (E-2) X TransCruz antibody is recommended for Gel Supershift and ChIP applications.

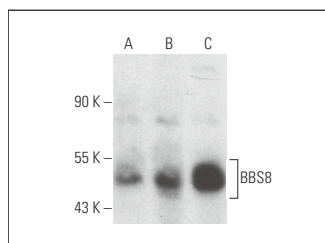
Molecular Weight of BBS8: 62 kDa.

Positive Controls: LNCaP whole cell lysate: sc-2231, SHP-77 whole cell lysate: sc-364258 or SK-BR-3 cell lysate: sc-2218.

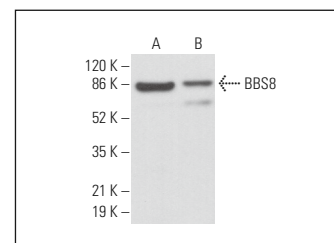
RECOMMENDED SUPPORT REAGENTS

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



BBS8 (E-2): sc-271009. Western blot analysis of BBS8 expression in SKBR-3 (A), LNCaP (B) and SHP-77 (C) whole cell lysates.



BBS8 (E-2): sc-271009. Western blot analysis of BBS8 expression in NIH/3T3 (A) and SH-SY5Y (B) whole cell lysates. Detection reagent used: m-IgGκ BP-HRP: sc-516102.

SELECT PRODUCT CITATIONS

- Murphy, D., et al. 2015. Alternative splicing shapes the phenotype of a mutation in BBS8 to cause nonsyndromic retinitis pigmentosa. *Mol. Cell Biol.* 35: 1860-1870.
- Kunova Bosakova, M., et al. 2019. Fibroblast growth factor receptor influences primary cilium length through an interaction with intestinal cell kinase. *Proc. Natl. Acad. Sci. USA* 116: 4316-4325.
- 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.

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