

BBS9 (N-15): sc-82536



The Power to Question

BACKGROUND

BBS9 (Bardet-Biedl syndrome 9), also known as B1, D1, C18 or PTHB1, is an 887 amino acid protein that localizes to both the cytoplasm and the centrosome and exists as six alternatively spliced isoforms. Expressed in a wide variety of tissues, including liver, lung, heart, brain and skeletal muscle, BBS9 functions as a component of the multi-protein BBSome complex which is required for ciliogenesis and is regulated by GDP/GTP exchange factors. Defects in the gene encoding BBS9 are associated with the pathogenesis of Bardet-Biedl syndrome type 9 (BBS9), an autosomal recessive disorder that is characterized by severe pigmentary retinopathy, early onset obesity, polydactyly, hypogenitalism, renal malformation and mental retardation. Additionally, chromosomal aberrations involving the BBS9 gene may play a role in the formation of Wilms' tumor 5 (WT5).

REFERENCES

- Adams, A.E., Rosenblatt, M. and Suva, L.J. 1999. Identification of a novel parathyroid hormone-responsive gene in human osteoblastic cells. *Bone* 24: 305-313.
- Vernon, E.G., Malik, K., Reynolds, P., Powlesland, R., Dallosso, A.R., Jackson, S., Henthorn, K., Green, E.D. and Brown, K.W. 2003. The parathyroid hormone-responsive B1 gene is interrupted by a t(1;7)(q42;p15) breakpoint associated with Wilms' tumour. *Oncogene* 22: 1371-1380.
- Nishimura, D.Y., Swiderski, R.E., Searby, C.C., Berg, E.M., Ferguson, A.L., Hennekam, R., Merin, S., Weleber, R.G., Biesecker, L.G., Stone, E.M. and Sheffield, V.C. 2005. Comparative genomics and gene expression analysis identifies BBS9, a new Bardet-Biedl syndrome gene. *Am. J. Hum. Genet.* 77: 1021-1033.
- Online Mendelian Inheritance in Man, OMIM™. 2005. Johns Hopkins University, Baltimore, MD. MIM Number: 607968. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
- Nachury, M.V., Loktev, A.V., Zhang, Q., Westlake, C.J., Peränen, J., Merdes, A., Slusarski, D.C., Scheller, R.H., Bazan, J.F., Sheffield, V.C. and Jackson, P.K. 2007. A core complex of BBS proteins cooperates with the GTPase Rab8 to promote ciliary membrane biogenesis. *Cell* 129: 1201-1213.
- Forti, E., Aksanov, O. and Birk, R.Z. 2007. Temporal expression pattern of Bardet-Biedl syndrome genes in adipogenesis. *Int. J. Biochem. Cell Biol.* 39: 1055-1062.
- Kang, H., Lee, S.K., Kim, M.H., Song, J., Bae, S.J., Kim, N.K., Lee, S.H. and Kwack, K. 2008. Parathyroid hormone-responsive B1 gene is associated with premature ovarian failure. *Hum. Reprod.* 23: 1457-1465.

CHROMOSOMAL LOCATION

Genetic locus: BBS9 (human) mapping to 7p14.3; Bbs9 (mouse) mapping to 9 A3.

STORAGE

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

SOURCE

BBS9 (N-15) is an affinity purified goat polyclonal antibody raised against a peptide mapping within an internal region of BBS9 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.

Blocking peptide available for competition studies, sc-82536 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

APPLICATIONS

BBS9 (N-15) is recommended for detection of BBS9 of mouse, rat and human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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); non cross-reactive with other BBS family members.

Suitable for use as control antibody for BBS9 siRNA (h): sc-72622, BBS9 siRNA (m): sc-72623, BBS9 shRNA Plasmid (h): sc-72622-SH, BBS9 shRNA Plasmid (m): sc-72623-SH, BBS9 shRNA (h) Lentiviral Particles: sc-72622-V and BBS9 shRNA (m) Lentiviral Particles: sc-72623-V.

Molecular Weight of BBS9: 99 kDa.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (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) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

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.