

BBS10 (A-3): sc-518239

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 genes map to multiple loci and encode fourteen proteins, BBS1-BBS14. Many BBS genes encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS10 (Bardet-Biedl syndrome 10), also known as chromosome 12 open reading frame 58, C12orf58 or FLJ23560, is a novel 723 amino acid protein belonging to the TCP-1 chaperonin family. BBS10 localizes to the basal body of primary cilium and assists in protein folding upon ATP hydrolysis. Inhibition of BBS10 has been found to impair ciliogenesis, activate the glycogen synthase kinase 3 pathway and cause peroxisome proliferator-activated receptor nuclear accumulation. The gene encoding BBS10 contains two exons and maps to human chromosome 12q21.2.

REFERENCES

1. Laurier, V., et al. 2006. Pitfalls of homozygosity mapping: an extended consanguineous Bardet-Biedl syndrome family with two mutant genes (BBS2, BBS10), three mutations, but no triallelism. *Eur. J. Hum. Genet.* 14: 1195-1203.
2. Stoetzel, C., et al. 2006. BBS10 encodes a vertebrate-specific chaperonin-like protein and is a major BBS locus. *Nat. Genet.* 38: 521-524.
3. Stoetzel, C., et al. 2007. Identification of a novel BBS gene (BBS12) highlights the major role of a vertebrate-specific branch of chaperonin-related proteins in Bardet-Biedl syndrome. *Am. J. Hum. Genet.* 80: 1-11.
4. Gerth, C., et al. 2008. Retinal morphology in patients with BBS1 and BBS10 related Bardet-Biedl Syndrome evaluated by Fourier-domain optical coherence tomography. *Vision Res.* 48: 392-399.

CHROMOSOMAL LOCATION

Genetic locus: BBS10 (human) mapping to 12q21.2.

SOURCE

BBS10 (A-3) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 444-464 of BBS10 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.

BBS10 (A-3) is available conjugated to agarose (sc-518239 AC), 500 µg/0.25 ml agarose in 1 ml, for IP; to HRP (sc-518239 HRP), 200 µg/ml, for WB, IHC(P) and ELISA; to either phycoerythrin (sc-518239 PE), fluorescein (sc-518239 FITC), Alexa Fluor® 488 (sc-518239 AF488), Alexa Fluor® 546 (sc-518239 AF546), Alexa Fluor® 594 (sc-518239 AF594) or Alexa Fluor® 647 (sc-518239 AF647), 200 µg/ml, for WB (RGB), IF, IHC(P) and FCM; and to either Alexa Fluor® 680 (sc-518239 AF680) or Alexa Fluor® 790 (sc-518239 AF790), 200 µg/ml, for Near-Infrared (NIR) WB, IF and FCM.

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APPLICATIONS

BBS10 (A-3) is recommended for detection of BBS10 of 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).

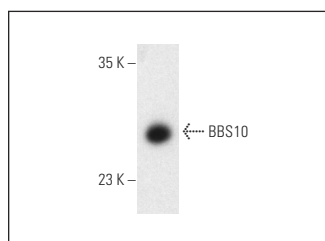
Suitable for use as control antibody for BBS10 siRNA (h): sc-72620, BBS10 shRNA Plasmid (h): sc-72620-SH and BBS10 shRNA (h) Lentiviral Particles: sc-72620-V.

Molecular Weight of BBS10: 81 kDa.

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



BBS10 (A-3): sc-518239. Western blot analysis of truncated human recombinant BBS10. Detection reagent used: m-IgG₁ BP-HRP: sc-525408.

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.