

BBS4 Antibody (Center)

Purified Mouse Monoclonal Antibody (Mab) Catalog # AM2250b

Specification

BBS4 Antibody (Center) - Product Information

Application Primary Accession Reactivity Host Clonality Isotype IHC, WB,E <u>Q96RK4</u> Human, Mouse, Rat Mouse Monoclonal IgG1

BBS4 Antibody (Center) - Additional Information

Gene ID 585

Other Names Bardet-Biedl syndrome 4 protein, BBS4

Target/Specificity

This BBS4 antibody is generated from a mice immunized with a recombinant protein between 1-240 amino acids from human BBS4.

Dilution IHC~~1:250 WB~~1:1000

Format

Purified monoclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein G column, followed by dialysis against PBS.

Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions BBS4 Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

BBS4 Antibody (Center) - Protein Information

Name BBS4

Function The BBSome complex is thought to function as a coat complex required for sorting of specific membrane proteins to the primary cilia. The BBSome complex is required for ciliogenesis but is dispensable for centriolar satellite function. This ciliogenic function is mediated in part by the Rab8 GDP/GTP exchange factor, which localizes to the basal body and contacts the BBSome.



Rab8(GTP) enters the primary cilium and promotes extension of the ciliary membrane. Firstly the BBSome associates with the ciliary membrane and binds to RAB3IP/Rabin8, the guanosyl exchange factor (GEF) for Rab8 and then the Rab8-GTP localizes to the cilium and promotes docking and fusion of carrier vesicles to the base of the ciliary membrane. The BBSome complex, together with the LTZL1, controls SMO ciliary trafficking and contributes to the sonic hedgehog (SHH) pathway regulation. Required for proper BBSome complex assembly and its ciliary localization. Required for microtubule anchoring at the centrosome but not for microtubule nucleation. May be required for the dynein-mediated transport of pericentriolar proteins to the centrosome.

Cellular Location

Cytoplasm, cytoskeleton, microtubule organizing center, centrosome. Cell projection, cilium membrane. Cytoplasm. Cytoplasm, cytoskeleton, microtubule organizing center, centrosome, centriolar satellite. Cell projection, cilium, flagellum {ECO:0000250|UniProtKB:Q8C1Z7}. Cell projection, cilium {ECO:0000250|UniProtKB:Q8C1Z7}. Note=Localizes to the pericentriolar material. Centrosomal localization requires dynein (By similarity) Localizes to the connecting cilium of photoreceptor cells (By similarity). {ECO:0000250|UniProtKB:Q8C1Z7}

Tissue Location

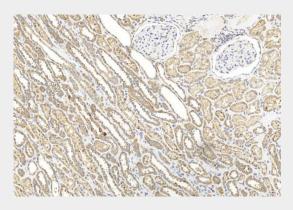
Ubiquitously expressed. The highest level of expression is found in the kidney

BBS4 Antibody (Center) - Protocols

Provided below are standard protocols that you may find useful for product applications.

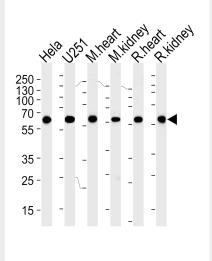
- <u>Western Blot</u>
- Blocking Peptides
- <u>Dot Blot</u>
- <u>Immunohistochemistry</u>
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

BBS4 Antibody (Center) - Images



Immunohistochemical analysis of paraffin-embedded Human kidney section using BBS4 (Cat#am2250b). am2250b was diluted at 1:250 dilution. A undiluted biotinylated goat polyvalent antibody was used as the secondary, followed by DAB staining.





Western blot analysis of lysates from Hela, U251 cell line, mouse heart and kidney, rat heart and kidney tissue (from left to right), using BBS4 Antibody (Cat. #AM2250b). AM2250b was diluted at 1:1000 at each lane. A goat anti-mouse IgG H&L(HRP) at 1:3000 dilution was used as the secondary antibody. Lysates at 35µg per lane.

BBS4 Antibody (Center) - Background

The BBSome complex is thought to function as a coat complex required for sorting of specific membrane proteins to the primary cilia. The BBSome complex is required for ciliogenesis but is dispensable for centriolar satellite function. This ciliogenic function is mediated in part by the Rab8 GDP/GTP exchange factor, which localizes to the basal body and contacts the BBSome. Rab8(GTP) enters the primary cilium and promotes extension of the ciliary membrane. Firstly the BBSome associates with the ciliary membrane and binds to RAB3IP/Rabin8, the guanosyl exchange factor (GEF) for Rab8 and then the Rab8-GTP localizes to the cilium and promotes docking and fusion of carrier vesicles to the base of the ciliary membrane. The BBSome complex, together with the LTZL1, controls SMO ciliary trafficking and contributes to the sonic hedgehog (SHH) pathway regulation. Required for proper BBSome complex assembly and its ciliary localization. Required for microtubule anchoring at the centrosome but not for microtubule nucleation. May be required for the dynein-mediated transport of pericentriolar proteins to the centrosome.

BBS4 Antibody (Center) - References

Mykytyn K., et al.Nat. Genet. 28:188-191(2001). Ye X., et al.DNA Seq. 15:213-218(2004). Ota T., et al.Nat. Genet. 36:40-45(2004). Zody M.C., et al.Nature 440:671-675(2006). Badano J.L., et al.Nature 439:326-330(2006).