

# **BBS4** Antibody (Center)

Purified Mouse Monoclonal Antibody (Mab) Catalog # AM2250b

## **Product Information**

Application WB, IHC-P, E Primary Accession Q96RK4

**Reactivity** Human, Mouse, Rat

**Host** Mouse **Clonality** Monoclonal

Isotype IgG1

**Clone Names** 1292CT845.130.218

Calculated MW 58282

### **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** WB~~1:1000 IHC-P~~1:250 E~~Use at an assay dependent concentration.

**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.

#### **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

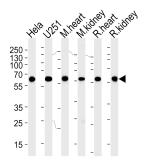
## **Background**

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.

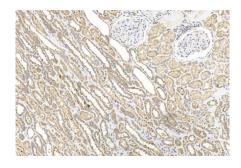
#### References

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).

## **Images**



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.



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.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.