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BBS4 Antibody (Center)

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP7562c

Product Information

Application	WB, E
Primary Accession	Q96RK4
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Clone Names	RB14682
Calculated MW	58282
Antigen Region	337-369

Additional Information

Gene ID	585
Other Names	Bardet-Biedl syndrome 4 protein, BBS4
Target/Specificity	This BBS4 antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 337-369 amino acids from the Central region of human BBS4.
Dilution	WB~~1:1000 E~~Use at an assay dependent concentration.
Format	Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is prepared by Saturated Ammonium Sulfate (SAS) precipitation 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

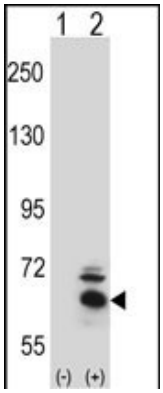
Background

BBS4 contains tetratricopeptide repeats (TPR), similar to O-linked N-acetyglucosamine transferase. Mutations in the gene encoding this protein have been observed in patients with Bardet-Biedl syndrome type 4. BBS4 may play a role in pigmentary retinopathy, obesity, polydactyly, renal malformation and mental retardation.

References

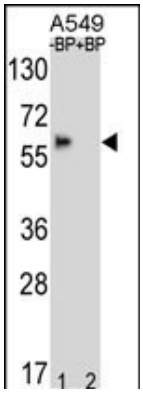
Nachury,M.V.,Cell 129 (6), 1201-1213 (2007)
Ye,X.,DNA Seq. 15 (3), 213-218 (2004)
Kim,J.C.,Nat. Genet. 36 (5), 462-470 (2004)

Images



Western blot analysis of BBS4 (arrow) using rabbit polyclonal BBS4 Antibody (Center) (Cat.#AP7562c). 293 cell lysates (2 ug/lane) either nontransfected (Lane 1) or transiently transfected (Lane 2) with the BBS4 gene.

Western blot analysis of BBS4 Antibody (Center) Pab (Cat.#AP7562c) pre-incubated without(lane 1) and with(lane 2) blocking peptide in A549 cell line lysate. BBS4 Antibody (Center) (arrow) was detected using the purified Pab.



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