

BBS9 Rabbit pAb

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Catalog # AP54530

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q3SYG4
Reactivity	Rat, Mouse
Predicted	Pig, Dog, Human, Rabbit, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	99280
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human BBS9
Epitope Specificity	244-320/887
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm, cytoskeleton, centrosome. Cell projection, cilium membrane. Cytoplasm. Note=Localizes to nonmembranous centriolar satellites in the cytoplasm.
DISEASE	Defects in BBS9 are a cause of Bardet-Biedl syndrome type 9 (BBS9) [MIM:209900]. Bardet-Biedl syndrome (BBS) is a genetically heterogeneous, autosomal recessive disorder characterized by usually severe pigmentary retinopathy, early onset obesity, polydactyly, hypogenitalism, renal malformation and mental retardation.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	BBS9 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).

Additional Information

Gene ID	27241
Other Names	Protein PTHB1, Bardet-Biedl syndrome 9 protein, Parathyroid hormone-responsive B1 gene protein, BBS9, PTHB1

Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

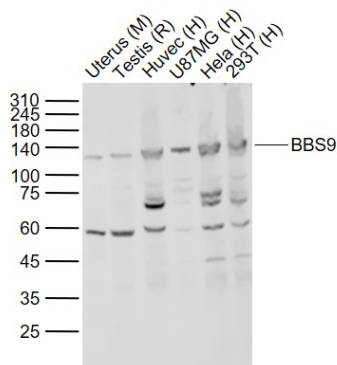
Protein Information

Name	BBS9
Synonyms	PTHB1
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. Required for proper BBSome complex assembly and its ciliary localization.
Cellular Location	Cytoplasm, cytoskeleton, microtubule organizing center, centrosome. Cell projection, cilium membrane. Cytoplasm Cytoplasm, cytoskeleton, microtubule organizing center, centrosome, centriolar satellite
Tissue Location	Widely expressed. Expressed in adult heart, skeletal muscle, lung, liver, kidney, placenta and brain, and in fetal kidney, lung, liver and brain.

Background

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Images



Sample:

Lane 1: Uterus (Mouse) Lysate at 40 ug

Lane 2: Testis (Rat) Lysate at 40 ug

Lane 3: Huvec (Human) Lysate at 30 ug

Lane 4: U87MG (Human) Lysate at 30 ug

Lane 5: Hela (Human) Lysate at 30 ug

Lane 6: 293T (Human) Lysate at 30 ug

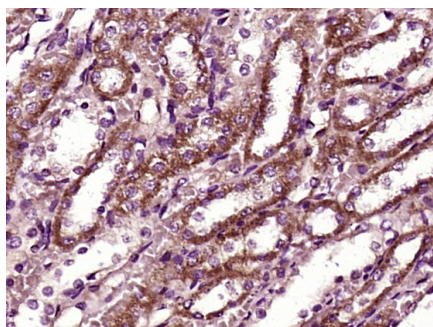
Primary: Anti-BBS9 (AP54530) at 1/1000 dilution

Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution

Predicted band size: 100-110 kD

Observed band size: 120 kD

Paraformaldehyde-fixed, paraffin embedded (Rat kidney);
Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer



(normal goat serum) at 37°C for 30min; Antibody incubation with (BBS9) Polyclonal Antibody, Unconjugated (AP54530) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

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