

# **BBS9 Polyclonal Antibody**

Purified Rabbit Polyclonal Antibody (Pab) Catalog # AP54530

#### **Product Information**

**Application** WB, IHC-P, IHC-F, IF, ICC, E

Primary Accession
Reactivity
Rat, Pig, Dog
Host
Rabbit
Clonality
Polyclonal
Calculated MW
Physical State

Q3SYG4
Rat, Pig, Dog
Rabbit
Polyclonal
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.

 $\label{lem:cytoplasm.} \textbf{Cytoplasm. Note=Localizes to nonmembranous centriolar satellites in the}$ 

cytoplasm.

**SUBUNIT** Part of BBSome complex, that contains BBS1, BBS2, BBS4, BBS5, BBS7, BBS8,

BBS9 and BBIP10. The BBSome complex binds to PCM1 and tubulin.

**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,

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

Target/Specificity Widely expressed. Expressed in adult heart, skeletal muscle, lung, liver,

kidney, placenta and brain, and in fetal kidney, lung, liver and brain.

**Dilution** WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC=1:100-500,IF=1:100-50

0,ELISA=1:5000-10000

Format 0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce

**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.

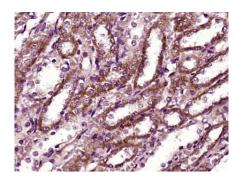
**Cellular Location** Cytoplasm, cytoskeleton, microtubule organizing center, centrosome. Cell

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.

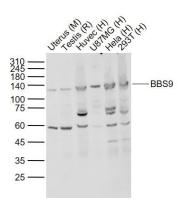
### **Images**



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) instructionsand DAB staining.

#### 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

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