

BBS9 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP54530

Specification

SUBUNIT

BBS9 Polyclonal Antibody - Product Information

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

Primary Accession
Reactivity
Rost
Clonality
Calculated MW
Rat, Pig, Dog
Rabbit
Polyclonal
Polyclonal

Calculated MW 99 KDa Physical State Liquid

Immunogen KLH conjugated synthetic peptide derived

laG

from human BBS9

Epitope Specificity 244-320/887

Isotype
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.
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. This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Background Descriptions

Important Note

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).

BBS9 Polyclonal Antibody - 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

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<span class ="dilution_WB">WB~~1:1000</span><br \><span class
="dilution_IHC-P">IHC-P~~N/A</span><br \><span class
="dilution_IHC-F">IHC-F~~N/A</span><br \><span class
="dilution_IF">IF~~1:50~200</span><br \><span class ="dilution_ICC">ICC~~N/A</span><br \><span class ="dilution_ICC">ICC~~N/A</span><br \><span class ="dilution_ICC">ICC~~N/A</span><br \><span class ="dilution_ICC">ICC~~N/A</span>
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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.

BBS9 Polyclonal Antibody - 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.

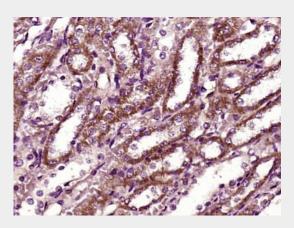


BBS9 Polyclonal Antibody - Protocols

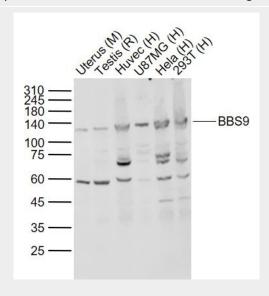
Provided below are standard protocols that you may find useful for product applications.

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

BBS9 Polyclonal Antibody - 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 (bs-11511R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and 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





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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 (bs-11511R) 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