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Rabbit Anti-BBS9 antibody

SL11511R

Product Name:	BBS9
Chinese Name:	巴尔得-别德尔综合征相关蛋白9抗体
Alias:	B1 antibody; Bardet Biedl syndrome 9; Bardet-Biedl syndrome 9 protein; bbs9; C18 antibody D1 antibody MGC118917; 1 gene protein; Protein PTHB1; PTH-responsive osteosarcoma B1 protein; PTHB1; PTHB1_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	99kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human BBS9:244-320/887
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
Product Detail:	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).

Function:
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.

Subunit:
Part of BBSome complex, that contains BBS1, BBS2, BBS4, BBS5, BBS7, BBS8, BBS9 and BBIP10. The BBSome complex binds to PCM1 and tubulin.

Subcellular Location:
Cytoplasm, cytoskeleton, centrosome. Cell projection, cilium membrane. Cytoplasm. Note=Localizes to nonmembranous centriolar satellites in the cytoplasm.

Tissue Specificity:
Widely expressed. Expressed in adult heart, skeletal muscle, lung, liver, kidney, placenta and brain, and in fetal kidney, lung, liver and brain.

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.

SWISS:
Q3SYG4

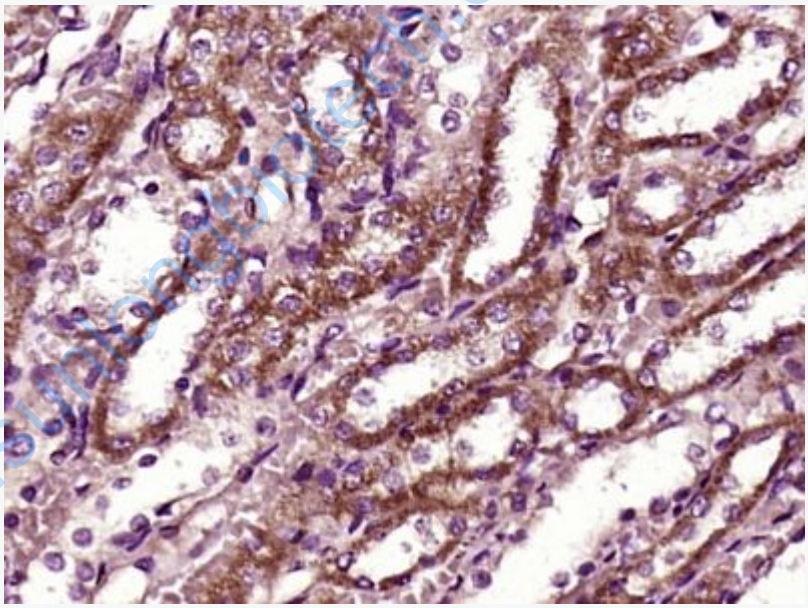
Gene ID:
27241

Database links:

Entrez Gene: 27241Human

Entrez Gene: 319845Mouse

Entrez Gene: 315484Rat

	Omim: 607968Human SwissProt: Q3SYG4Human SwissProt: Q811G0Mouse Unigene: 372360Human Unigene: 176725Mouse Unigene: 92828Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 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 (SL11511R) at 1:400 overnight at 4°C, followed by operating according to SP

	Kit(Rabbit) (sp-0023) instructionsand DAB staining.