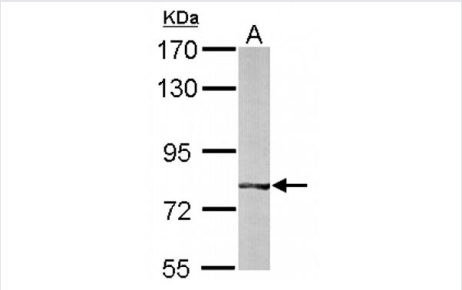
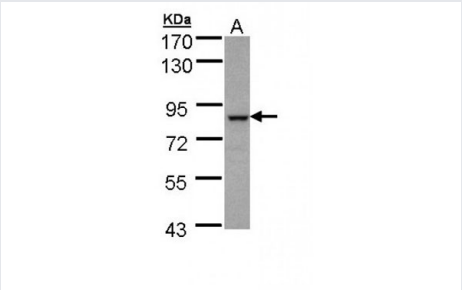
BBS7 Antibody – Rabbit Polyclonal

PRODUCT OVERVIEW

Product Name	BBS7, Polyclonal Antibody
Full Product Name	BBS7 antibody
Product Gene Name	anti-BBS7 antibody
Product Synonym Names	Polyclonal BBS7; Anti-BBS7; BBS-7; Bardet-Biedl syndrome 7; BBS7; BBS 7; BBS2-like 1; BBS2L1; BBS2-like protein 1; FLJ10715; Bardet-Biedl syndrome 7 protein
Clonality	Polyclonal
Isotype	IgG
Host	Rabbit
Form/Format	Supplied as a concentrated soloution containing 0.1M Tris, 0.1M Glycine, 10% Glycerol (pH 7.0). 0.01% Thimerosal was added as a preservative.
Purity/Purification	BBS7 antibody was purified by antigen-affinity chromatography
Concentration	1 mg/ml (lot specific)
Preparation and Storage	Store at 4 degree C for short term storage. Aliquot and store at -20 degree C for long term storage. Avoid repeated freeze/thaw cycles.
Molecular Weight	80 kDa
OMIM	607590
3D Structure	ModBase 3D Structure for Q8IWZ6
Cross-Reactivity	Human, Mouse
Product Categories/Family for anti-BBS7 antibody	Differentiation & Development; Purified Polyclonal Antibodies
Applications Tested/Suitable for anti-BBS7 antibody	Western Blot (WB)
Application Notes for anti-BBS7 antibody	WB: 1:500-1:3000
Related Product Information for anti-BBS7 antibody	MBS837514 is an antibody targeting BBS7. Also known as BBS7 antibody. Host: Rabbit. Affinity purified Rabbit polyclonal BBS7 antibody
Other Notes	Small volumes of anti-BBS7 antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.



Western Blot (WB) of anti-BBS7 antibody
Western blot analysis of 50 ug of whole cell lysate (A: mouse liver) using a 7.5 % SDS PAGE gel and BBS7 antibody at a dilution of 1:1000



Western Blot (WB) of anti-BBS7 antibody
Western blot analysis of 30 ug of whole cell lysate (A: A431) using a 7.5 % SDS PAGE gel and BBS7 antibody at a dilution of 1:1000

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for BBS7. It may not necessarily be applicable to this product.

NCBI GI #	29029557
NCBI GeneID	55212
NCBI Accession #	NP_789794.1
UniProt Primary Accession #	Q8IWZ6
UniProt Secondary Accession #	Q4W5P8; Q8N581; Q9NVI4
UniProt Related Accession #	Q8IWZ6
GenBank Nucleotide #	NM_176824.2
NCBI Official Full Name	Bardet-Biedl syndrome 7 protein isoform a
NCBI Official Synonym Full Names	Bardet-Biedl syndrome 7
NCBI Official Symbol	BBS7
NCBI Official Synonym Symbols	BBS2L1
UniProt Protein Name	Bardet-Biedl syndrome 7 protein
UniProt Synonym Protein Names	BBS2-like protein 1
Protein Family	Bardet-Biedl syndrome 7 protein
UniProt Gene Name	BBS7
UniProt Synonym Gene Names	BBS2L1
UniProt Entry Name	BBS7_HUMAN
NCBI Protein Information	Bardet-Biedl syndrome 7 protein
NCBI Summary for BBS7	This gene encodes one of eight proteins that form the BBSome complex containing BBS1, BBS2, BBS4, BBS5, BBS7, BBS8, BBS9 and BBIP10. The BBSome complex is believed to recruit Rab8(GTP) to the primary cilium and promote ciliogenesis. The BBSome complex assembly is mediated by a complex composed of three chaperonin-like BBS proteins (BBS6, BBS10, and BBS12) and CCT/TRiC family chaperonins. Mutations in this gene are implicated in Bardet-Biedl syndrome, a genetic disorder whose symptoms include obesity, retinal degeneration, polydactyly and nephropathy; however, mutations in this gene and the BBS8 gene are thought to play a minor role and mutations in chaperonin-like BBS genes are found to be a major contributor to disease development in a multiethnic Bardet-Biedl syndrome patient population. Two transcript variants encoding distinct isoforms have been identified for this gene.[provided by RefSeq, Oct 2014]

UniProt Comments for BBS7	BBS7: 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. Ciliary dysfunction leads to a broad spectrum of disorders, collectively termed ciliopathies. Overlapping clinical features include retinal degeneration, renal cystic disease, skeletal abnormalities, fibrosis of various organ, and a complex range of anatomical and functional defects of the central and peripheral nervous system. The ciliopathy range of diseases includes Meckel-Gruber syndrome, Bardet-Biedl syndrome, Joubert syndrome, nephronophtisis, Senior-Loken syndrome, and Jeune asphyxiating thoracic dystrophy among others. Single-locus allelism is insufficient to explain the variable penetrance and expressivity of such disorders, leading to the suggestion that variations across multiple sites of the ciliary proteome, including BBS7, influence the clinical outcome. Defects in BBS7 are a cause of Bardet-Biedl syndrome type 7 (BBS7). Bardet-Biedl syndrome (BBS) is a genetically heterogeneous disorder characterized by usually severe pigmentary retinopathy, early onset obesity, polydactyly, hypogenitalism, renal malformation and mental retardation. Secondary features include diabetes mellitus, hypertension and congenital heart disease. A relatively high incidence of BBS is found in the mixed Arab populations of Kuwait and in Bedouin tribes throughout the Middle East, most likely due to the high rate of consaguinity in these populations and a founder effect. Inheritance is autosomal recessive, but three mutated alleles (two at one locus, and a third at a second locus) may be required for disease manifestation in some cases (triallelic inheritance). 2 isoforms of the human protein are produced by alternative splicing. Chromosomal Location of Human Ortholog: 4q27 Cellular Component: centrosome; axoneme; nucleus; cytosol Molecular Function: protein binding Biological Process: limb development; fat cell differentiation; smoothened signaling pathway; organelle organization and biogenesis; pigment granule aggregation in cell center; digestive tract morphogenesis; palate development; melanosome transport; regulation of transcription from RNA polymerase II promoter; protein transport; eye development; visual perception; sensory cilium biogenesis; positive regulation of proteasomal ubiquitin-dependent protein catabolic process; brain development; heart looping; determination of left/right symmetry Disease: Bardet-biedl Syndrome 7
Research Articles on BBS7	**1.** Observational study of genetic testing. (HuGE Navigator)
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