

Recombinant Human BBS1 protein ab157954

1 Image

Description

Product name	Recombinant Human BBS1 protein		
Expression system	Wheat germ		
Accession	<u>Q4G0L2</u>		
Protein length	Full length protein		
Animal free	No		
Nature	Recombinant		
Species	Human		
Sequence	MSPGPQLWHLLQALVSMCIRISDPTSSSAYPNCLQILWNK TFGTRPKRET AEEPLSIQSLRFLQLELSEMEAFVNQHKSNSIKRQTVITTM TTLKKNLAD EDAVSCLVLGTENKELLVLDPEAFTILAKMSLPSVPVFLE VSGQFDVEFR LAAACRNGNMYLRRDSKHPKYCIELSAQPVGILRVHKVLVV GSTQDSLH GFTHKGKKLWTVQMPAAILTMNLLEQHSRGLQAVMAGLA NGEVRIYRDKA LLNVIHTPDAVTSLCFGRYGREDNTLIMTTRGGGLIILKIRT AMFVEGG SEVGPPPAQAMKLNVPKTRLYVDQTLREREAGTAMHRA FQTDLYLLRLR AARAYLQALESSLSPSTTAREPLKLHAVVQGLGPTFKLT LHLQNTSTTR PVLGLLVCFLYNEALYSLPRAFFKVPLLVPGLNYPLETFVE SLSNKGISD IIKVGPALVPRGR		
Amino acids	1 to 463		
Tags	GST tag N-Terminus		

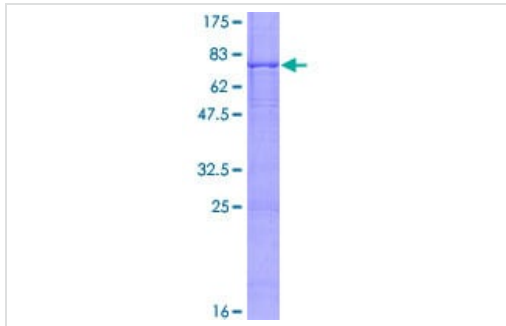
Specifications

Our **Abpromise guarantee** covers the use of **ab157954** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Western blot
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	ELISA
Form	Liquid
Additional notes	
Preparation and Storage	
Stability and Storage	<p>Shipped on dry ice. Upon delivery aliquot and store at -80°C. Avoid freeze / thaw cycles.</p> <p>pH: 8.00</p> <p>Constituents: 0.31% Glutathione, 0.79% Tris HCl</p>
General Info	
Function	<p>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 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.</p>
Tissue specificity	<p>Highly expressed in the kidney. Also found in fetal tissue, testis, retina, adipose tissue, heart, skeletal muscle and pancreas.</p>
Involvement in disease	<p>Defects in BBS1 are a cause of Bardet-Biedl syndrome type 1 (BBS1) [MIM:209900]. 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 consanguinity 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).</p>
Cellular localization	<p>Cell projection > cilium membrane. Cytoplasm. Localizes to nonmembranous centriolar satellites in the cytoplasm.</p>
Images	



SDS-PAGE - Recombinant Human BBS1 protein
(ab157954)

ab157954 on a 12.5% SDS-PAGE stained with Coomassie Blue.

Please note: All products are "FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC PROCEDURES"

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