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Rabbit Anti-BBS12 antibody

SL11505R

Product Name:	BBS12
Chinese Name:	巴尔得-别德尔综合征相关蛋白12抗体
Alias:	Bardet Biedl syndrome 12 protein; Bardet-Biedl syndrome 12 (human); Bardet-Biedl syndrome 12 protein homolog; BBS12 gene; C4orf24; FLJ35630; FLJ41559; Gm1805; Gm407; Gm721; RP23-137F6.2;BBS12_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Cow,Horse,Rabbit,
Applications:	WB=1:500-2000ELISA=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:	79kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human BBS12:25-100/710
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:	The protein encoded by this gene is part of a complex that is involved in membrane trafficking. The encoded protein is a molecular chaperone that aids in protein folding upon ATP hydrolysis. This protein also plays a role in adipocyte differentiation. Defects in this gene are a cause of Bardet-Biedl syndrome type 12. Two transcript variants encoding the same protein have been found for this gene.

Function:
Probable molecular chaperone. Assists the folding of proteins upon ATP hydrolysis. As part of the BBS/CCT complex may play a role in the assembly of BBSome, a complex involved in ciliogenesis regulating transports vesicles to the cilia. Involved in adipogenic differentiation.

Subunit:
Component of the BBS/CCT complex composed at least of MKKS, BBS10, BBS12, TCP1, CCT2, CCT3, CCT4, CCT5 AND CCT8.

Subcellular Location:
Cell projection, cilium. Note: Located within the basal body of the primary cilium of differentiating preadipocytes.

DISEASE:
Defects in BBS12 are the cause of Bardet-Biedl syndrome type 12 (BBS12) [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. 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. BBS12 seems to be rarely involved in oligogenic inheritance.

Similarity:
Belongs to the TCP-1 chaperonin family. BBS12 subfamily.

SWISS:
Q6ZW61

Gene ID:
166379

Database links:

Entrez Gene: 166379Human

Omim: 610683Human

SwissProt: Q6ZW61Human

Unigene: 400698Human

Important Note:

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