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Rabbit Anti-BBS10 antibody

SL11512R

Product Name:	BBS10
Chinese Name:	巴尔得-别德尔综合征相关蛋白10抗体
Alias:	Bardet Biedl syndrome 10 protein; Bardet Biedl syndrome 10 protein homolog; C12orf58; FLJ23560; RGD1560748; BBS10_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
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:	81kDa
Cellular localization:	cytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human BBS10:51-130/723
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:	Bardet-Biedl syndrome (BBS) is a pleiotropic genetic disorder characterized by obesity, photoreceptor degeneration, polydactyly, hypogenitalism, renal abnormalities, and developmental delay. Other associated clinical findings in BBS patients include diabetes, hypertension, and congenital heart defects. BBS genes map to multiple loci and encode fourteen proteins, BBS1-BBS14. Many BBS genes encode basal body or cilia proteins, suggesting that BBS is a ciliary dysfunction disorder. BBS10 (Bardet-

Biedl syndrome 10), also known as chromosome 12 open reading fame 58, C12orf58 or FLJ23560, is a novel 723 amino acid protein belonging to the TCP-1 chaperonin family. BBS10 localizes to the basal body of primary cilium and assists in protein folding upon ATP hydrolysis. Inhibition of BBS10 has been found to impair ciliogenesis, activate the glycogen synthase kinase 3 pathway and cause peroxisome proliferator-activated receptor nuclear accumulation. The gene encoding BBS10 contains two exons and maps to human chromosome 12q21.2.

Function:
BBS10 belongs to the TCP-1 chaperonin family. It is a probable molecular chaperone; assist the folding of proteins upon ATP hydrolysis. Defects in BBS10 are the cause of Bardet-Biedl syndrome type 10 (BBS10) [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.

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 BBS10 are the cause of Bardet-Biedl syndrome type 10 (BBS10) [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.

Similarity:
Belongs to the TCP-1 chaperonin family.

SWISS:
Q8TAM1

Gene ID:
79738

Database links:

Entrez Gene: 79738Human

Omim: 610148Human

SwissProt: Q8TAM1Human

	Unigene: 96322Human **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. BBS蛋白是一类研究早期儿童肥胖综合症有关的其中一种。巴尔得-别德尔综合征(Bardet-Biedl syndrome, BBS)的特征为不同程度的肥胖、智力延迟、色素视网膜病变、多指和肾脏异常。