

## Rabbit Anti-LMBRD1 antibody

SL18309R

Product Name:	LMBRD1
Chinese Name:	核輸出信号相互作用蛋白抗体 人名英格兰 人名英格兰 人名英格兰人姓氏 化乙烯乙烯 人名英格兰人姓氏 化乙烯乙烯 人名英格兰人姓氏 化乙烯乙烯 化乙烯乙烯 化乙烯乙烯 化乙烯乙烯 化乙烯乙烯乙烯乙烯乙烯乙烯乙烯
Alias:	HDAg-L-interacting protein NESI; LMBD1_HUMAN; LMBR1 domain-containing protein 1; lmbrd1; Nuclear export signal-interacting protein; Probable lysosomal cobalamin transporter.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Chicken, Dog, Pig, Cow, 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:	61kDa 🗸 💙
<b>Cellular localization:</b>	cytoplasmic
Form:	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human LMBRD1:21- 120/540 <extracellular></extracellular>
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:	This gene encodes a lysosomal membrane protein that may be involved in the transport and metabolism of cobalamin. This protein also interacts with the large form of the hepatitis delta antigen and may be required for the nucleocytoplasmic shuttling of the hepatitis delta virus. Mutations in this gene are associated with the vitamin B12

metabolism disorder termed, homocystinuria-megaloblastic anemia complementation type F.[provided by RefSeq, Oct 2009]

## **Function:**

Probable lysosomal cobalamin transporter. Required to export cobalamin from lysosomes allowing its conversion to cofactors. Isoform 3 may play a role in the assembly of hepatitis delta virus (HDV).

Subcellular Location: Lysosome membrane.

**Tissue Specificity:** Isoform 3 is expressed in liver.

**Post-translational modifications:** N-glycosylated.

## **DISEASE:**

Defects in LMBRD1 are the cause of methylmalonic aciduria and homocystinuria type cblF (MMAFHC) [MIM:277380]; also known as homocystinuria-megaloblastic anemia complementation type F. MMAFHC is a disorder of cobalamin metabolism characterized by decreased levels of the coenzymes adenosylcobalamin (AdoCbl) and methylcobalamin (MeCbl). It is due to accumulation of free cobalamin in lysosomes, thus hindering its conversion to cofactors. Clinical features include developmental delay, stomatitis, glossitis, seizures and methylmalonic aciduria responsive to vitamin B12.

Similarity:

Belongs to the LIMR family. LMBRD1 subfamily.

SWISS: Q9NUN5

Gene ID: 55788

Database links:

Entrez Gene: 55788 Human

Entrez Gene: 68421 Mouse

Entrez Gene: 246046 Rat

Omim: 612625 Human

SwissProt: Q9NUN5 Human

	SwissProt: Q8K0B2 Mouse
	SwissProt: Q6AZ61 Rat
	Unigene: 271643 Human
	Unigene: 677072 Human
	Unigene: 477783 Mouse
	Unigene: 201860 Rat
	<b>Important Note:</b> This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	
	Paraformaldehyde-fixed, paraffin embedded (Mouse brain); 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 (LMBRD1) Polyclonal Antibody,
	Unconjugated (SL18309R) at 1:400 overnight at 4°C, followed by operating

according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

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