

### LMBRD1 Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP57032

# **Specification**

# **LMBRD1** Polyclonal Antibody - Product Information

Application Primary Accession

Reactivity
Host
Clonality
Calculated MW
Physical State
Immunogen

immunogen

**Epitope Specificity** 

Isotype **Purity** 

affinity purified by Protein A

Buffer

SUBCELLULAR LOCATION

**SIMILARITY** 

Post-translational modifications

**DISEASE** 

IHC-P, IHC-F, IF, ICC

Q9NUN5

Rat, Pig, Dog, Bovine

Rabbit Polyclonal 61 KDa Liquid

KLH conjugated synthetic peptide derived

from human LMBRD1

21-120/540

laG

0.01M TBS (pH7.4) with 1% BSA, 0.02%

Proclin300 and 50% Glycerol.

Lysosome membrane.

Belongs to the LIMR family. LMBRD1

subfamily. N-glycosylated.

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.

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

# Important Note

### **Background Descriptions**

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]



## **LMBRD1** Polyclonal Antibody - Additional Information

#### **Gene ID 55788**

#### **Other Names**

Lysosomal cobalamin transport escort protein LMBD1, LMBD1, HDAg-L-interacting protein NESI, LMBR1 domain-containing protein 1, Nuclear export signal-interacting protein, LMBRD1 (<a href="http://www.genenames.org/cgi-bin/gene\_symbol\_report?hgnc\_id=23038" target="blank">HGNC:23038</a>), C6orf209, NESI

## Target/Specificity

Isoform 3 is expressed in liver.

#### **Dilution**

```
<span class ="dilution_IHC-P">IHC-P~~N/A</span><br \> <span class
="dilution_IHC-F">IHC-F~~N/A</span><br \> <span class
="dilution_IF">IF~~1:50~200</span><br \> <span class ="dilution_ICC">ICC~~N/A</span>
```

### **Storage**

Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

## LMBRD1 Polyclonal Antibody - Protein Information

Name LMBRD1 (HGNC:23038)

Synonyms C6orf209, NESI

#### **Function**

Lysosomal membrane chaperone required to export cobalamin (vitamin B12) from the lysosome to the cytosol, allowing its conversion to cofactors (PubMed:<a href="http://www.uniprot.org/citations/19136951" target="\_blank">19136951</a>). Targets ABCD4 transporter from the endoplasmic reticulum to the lysosome (PubMed:<a href="http://www.uniprot.org/citations/27456980" target="\_blank">27456980</a>). Then forms a complex with lysosomal ABCD4 and cytoplasmic MMACHC to transport cobalamin across the lysosomal membrane (PubMed:<a href="http://www.uniprot.org/citations/25535791" target="\_blank">25535791</a>). Acts as an adapter protein which plays an important role in mediating and regulating the internalization of the insulin receptor (INSR) (By similarity). Involved in clathrin-mediated endocytosis of INSR via its interaction with adapter protein complex 2 (By similarity). Essential for the initiation of gastrulation and early formation of mesoderm structures during embryogenesis (By similarity).

### **Cellular Location**

Endoplasmic reticulum membrane. Lysosome membrane; Multi-pass membrane protein. Cell membrane {ECO:0000250|UniProtKB:Q8K0B2}; Multi-pass membrane protein. Cytoplasmic vesicle, clathrin-coated vesicle {ECO:0000250|UniProtKB:Q8K0B2}

### **Tissue Location**

Isoform 3 is expressed in liver.

## **LMBRD1** Polyclonal Antibody - Protocols

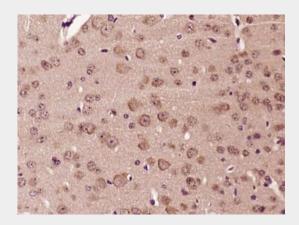




Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- Blocking Peptides
- Dot Blot
- <u>Immunohistochemistry</u>
- Immunofluorescence
- <u>Immunoprecipitation</u>
- Flow Cytomety
- Cell Culture

# LMBRD1 Polyclonal Antibody - Images



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 (bs-18309R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.