

Rabbit Anti-LMAN1/AF350 Conjugated antibody

SL18304R-AF350

Product Name	Anti-LMAN1/AF350
Chinese Name	AF350 标记的凝集素甘露糖 Binding protein1 抗体
Alias	Endoplasmic reticulum golgi intermediate compartment protein 53; ER-Golgi intermediate compartment 53 kDa protein; ERGIC-53; ERGIC53; ERGIC53 like protein; F5F8D; FMFD1; Gp58; Intracellular mannose specific lectin; Intracellular mannose-specific lectin MR60; Lectin mannose binding 1; Lectin mannose-binding 1; Lman1; LMAN1 like protein; LMAN1_HUMAN; MCFD1; MR60; Protein ERGIC-53.
Research Area	Cell biology immunology Signal transduction
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Human(predicted:Mouse,Rat,Dog,Cow,Horse,Rabbit,Sheep) IF=1:100-500
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight	54kDa
Form	Lyophilized or Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human LMAN1
Lsotype	IgG
Purification	affinity purified by Protein A
Storage Buffer	1M TBS(pH7.4) with 1% BSA, 3% 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 1M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
Product Detail	background: The protein encoded by this gene is a type I integral membrane protein

localized in the intermediate region between the endoplasmic reticulum and the Golgi, presumably recycling between the two compartments. The protein is a mannose-specific lectin and is a member of a novel family of plant lectin homologs in the secretory pathway of animal cells. Mutations in the gene are associated with a coagulation defect. Using positional cloning, the gene was identified as the disease gene leading to combined factor V-factor VIII deficiency, a rare, autosomal recessive disorder in which both coagulation factors V and VIII are diminished. [provided by RefSeq, Jul 2008]

Function:

Mannose-specific lectin. May recognize sugar residues of glycoproteins, glycolipids, or glycosylphosphatidyl inositol anchors and may be involved in the sorting or recycling of proteins, lipids, or both. The LMAN1-MCFD2 complex forms a specific cargo receptor for the ER-to-Golgi transport of selected proteins.

Subcellular Location:

Endoplasmic reticulum-Golgi intermediate compartment membrane. Golgi apparatus membrane. Endoplasmic reticulum membrane.

Tissue Specificity:

Ubiquitous.

Post-translational modifications:

The N-terminal may be partly blocked.

DISEASE:

Defects in LMAN1 are THE cause of factor V and factor VIII combined deficiency type 1 (F5F8D1) [MIM:227300]; also known as multiple coagulation factor deficiency I (MCFD1). F5F8D1 is an autosomal recessive blood coagulation disorder characterized by bleeding symptoms similar to those in hemophilia or parahemophilia, that are caused by single deficiency of FV or FVIII, respectively. The most common symptoms are epistaxis, menorrhagia, and excessive bleeding during or after trauma. Plasma levels of coagulation factors V and VIII are in the range of 5 to 30% of normal.

Similarity:

Contains 1 L-type lectin-like domain.

Database links:

[Entrez Gene: 3998](#) Human

[Entrez Gene: 70361](#) Mouse

[Entrez Gene: 116666](#) Rat

[Omim: 601567](#) Human

[SwissProt: P49257](#) Human

[SwissProt: Q9D0F3](#) Mouse

[SwissProt: Q62902](#) Rat

[Unigene: 465295](#) Human

[Unigene: 290857](#) Mouse

[Unigene: 449042](#) Mouse

[Unigene: 25734](#) Rat

Important Note:

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