

LONP1 Rabbit pAb

Catalog # AP99582

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P36776
Other Accession	P36776
Host	Rabbit
Clonality	Polyclonal
Calculated MW	106489
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human LONP1
Epitope Specificity	361-460/959
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the peptidase S16 family. Contains 1 Lon domain.
SUBUNIT	Homohexamer or homoheptamer. Organized in a ring with a central cavity. DNA and RNA binding is stimulated by substrate and inhibited by ATP binding. Interacts with PEO1 and mitochondrial DNA polymerase subunit POLG.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	LONP1 encodes a mitochondrial matrix protein in the Lon family of ATP dependent proteases. It binds a specific sequence in the light and heavy chain promoters of the mitochondrial genome which are involved in regulation of DNA replication and transcription.

Additional Information

Gene ID	9361
Other Names	PRSS15; CODASS; LON; LONP; LonHS; PIM1; hLON; 1200017E13Rik; LONM_BOVIN; LONP1; Lon protease-like protein (LONP); Mitochondrial ATP-dependent protease Lon; Serine protease 15; 3.4.21.53; LONM_HUMAN; LONM_MOUSE; LONM_RAT; lon peptidase 1, mitochondrial; protease, serine, 15
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
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.

Protein Information

Name	LONP1 {ECO:0000255 HAMAP-Rule:MF_03120, ECO:0000312 HGNC:HGNC:9479}
Function	ATP-dependent serine protease that mediates the selective degradation of misfolded, unassembled or oxidatively damaged polypeptides as well as certain short-lived regulatory proteins in the mitochondrial matrix (PubMed:12198491, PubMed:15870080, PubMed:17579211, PubMed:37327776, PubMed:8248235, PubMed:35870450, PubMed:38688959). Endogenous substrates include mitochondrial steroidogenic acute regulatory (StAR) protein, DELE1, helicase Twinkle (TWNK) and the large ribosomal subunit protein MRPL32/bL32m (PubMed:17579211, PubMed:28377575, PubMed:37327776). MRPL32/bL32m is protected from degradation by LONP1 when it is bound to a nucleic acid (RNA), but TWNK is not (PubMed:17579211, PubMed:28377575). In addition to its protease activity, has also an ATP-dependent protein folding chaperone activity and functions in mitochondrial protein folding (PubMed:33431889). Participates in the regulation of mitochondrial gene expression and in the maintenance of the integrity of the mitochondrial genome (PubMed:17420247). Binds to mitochondrial promoters and RNA in a single-stranded, site-specific, and strand-specific manner (PubMed:17420247). May regulate mitochondrial DNA replication and/or gene expression using site-specific, single-stranded DNA binding to target the degradation of regulatory proteins binding to adjacent sites in mitochondrial promoters (PubMed:14739292, PubMed:17420247).
Cellular Location	Mitochondrion matrix {ECO:0000255 HAMAP- Rule:MF_03120, ECO:0000269 PubMed:7961901}
Tissue Location	Duodenum, heart, lung and liver, but not thymus.

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