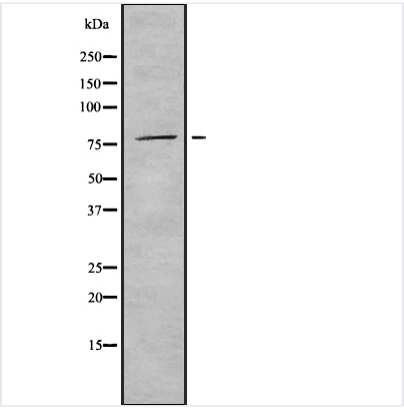
PNPase Antibody

PRODUCT OVERVIEW

Product Name	PNPase, Antibody
Full Product Name	PNPase Antibody
Product Gene Name	anti-PNPase antibody
Molecular Weight	85,951 Da
OMIM	610316
3D Structure	ModBase 3D Structure for Q8TCS8
ISO Certification	Manufactured in an ISO 9001:2015 Certified Laboratory.
Related Product Information for anti-PNPase antibody	MBS8513203 is an antibody targeting PNPase. Also known as PNPase Antibody.
Other Notes	Small volumes of anti-PNPase antibody vial(s) may occasionally become entrapped in the seal of the product vial during shipment and storage. If necessary, briefly centrifuge the vial on a tabletop centrifuge to dislodge any liquid in the container`s cap. Certain products may require to ship with dry ice and additional dry ice fee may apply.



Western Blot (WB) of anti-PNPase antibody
Western blot analysis PNPase using HeLa whole cell lysates

ADDITIONAL DETAILS

NCBI/Uniprot data below describe general gene information for PNPase. It may not necessarily be applicable to this product.

NCBI GI #	188528628
NCBI GeneID	87178
NCBI Accession #	NP_149100.2
UniProt Primary Accession #	Q8TCS8
UniProt Secondary Accession #	Q53SU0; Q68CN1; Q7Z7D1; Q8IWX1; Q96T05; Q9BRU3; Q9BVX0
UniProt Related Accession #	Q8TCS8
GenBank Nucleotide #	NM_033109.4
NCBI Official Full Name	polyribonucleotide nucleotidyltransferase 1, mitochondrial

NCBI Official Synonym Full Names	polyribonucleotide nucleotidyltransferase 1
NCBI Official Symbol	PNPT1
NCBI Official Synonym Symbols	OLD35; DFNB70; PNPASE; old-35; COXPD13
UniProt Protein Name	Polyribonucleotide nucleotidyltransferase 1, mitochondrial
UniProt Synonym Protein Names	3'-5' RNA exonuclease OLD35; PNPase old-35; Polynucleotide phosphorylase 1; PNPase 1; Polynucleotide phosphorylase-like protein
Protein Family	Polyribonucleotide nucleotidyltransferase
UniProt Gene Name	PNPT1
UniProt Synonym Gene Names	PNPASE; PNPase 1
NCBI Protein Information	polyribonucleotide nucleotidyltransferase 1, mitochondrial
NCBI Summary for PNPase	The protein encoded by this gene belongs to the evolutionary conserved polynucleotide phosphorylase family comprised of phosphate dependent 3'-to-5' exoribonucleases implicated in RNA processing and degradation. This enzyme is predominantly localized in the mitochondrial intermembrane space and is involved in import of RNA to mitochondria. Mutations in this gene have been associated with combined oxidative phosphorylation deficiency-13 and autosomal recessive nonsyndromic deafness-70. Related pseudogenes are found on chromosomes 3 and 7. [provided by RefSeq, Dec 2012]
UniProt Comments for PNPase	RNA-binding protein implicated in numerous RNA metabolic processes. Catalyzes the phosphorolysis of single-stranded polyribonucleotides processively in the 3'-to-5' direction. Mitochondrial intermembrane factor with RNA-processing exoribonulease activity. Component of the mitochondrial degradosome (mtEXO) complex, that degrades 3' overhang double-stranded RNA with a 3'-to-5' directionality in an ATP-dependent manner. Required for correct processing and polyadenylation of mitochondrial mRNAs. Plays a role as a cytoplasmic RNA import factor that mediates the translocation of small RNA components, like the 5S RNA, the RNA subunit of ribonuclease P and the mitochondrial RNA-processing (MRP) RNA, into the mitochondrial matrix. Plays a role in mitochondrial morphogenesis and respiration; regulates the expression of the electron transport chain (ETC) components at the mRNA and protein levels. In the cytoplasm, shows a 3'-to-5' exoribonuclease mediating mRNA degradation activity; degrades c-myc mRNA upon treatment with IFNB1/IFN-beta, resulting in a growth arrest in melanoma cells. Regulates the stability of specific mature miRNAs in melanoma cells; specifically and selectively degrades miR-221, preferentially. Plays also a role in RNA cell surveillance by cleaning up oxidized RNAs. Binds to the RNA subunit of ribonuclease P, MRP RNA and miR-221 microRNA.
Research Articles on PNPase	**1.** The PNPT1 p.Arg136His and p.Pro140Leu variants in our subject showed 1) the mutated amino acids are highly conserved and our structural analysis supported them to be functionally deleterious; 2) The carrier frequencies of the variants are very low in populations (1:60.000), with no homozygous carriers found; 3) The RC complex amounts in the subject's myoblasts were functionally rescued by expression of wild-type PNPT1.
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