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Rabbit Anti-NUBPL antibody

SL19505R

Product Name:	NUBPL
Chinese Name:	核苷酸Binding protein样NUBPL抗体
Alias:	C14orf127; FLJ12660; huInd1; IND1 homolog; Iron-sulfur protein NUBPL; Nubpl; NUBPL_HUMAN; nucleotide binding protein like; Nucleotide-binding protein-like.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Cow,Horse,Sheep,
Applications:	WB=1:500-2000ELISA=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:	34kDa
Cellular localization:	cytoplasmic
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human NUBPL:1-100/319
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 member of the Mrp/NBP35 ATP-binding proteins family. The encoded protein is required for the assembly of the respiratory chain NADH dehydrogenase (complex I), an oligomeric enzymatic complex located in the inner mitochondrial membrane. Mutations in this gene cause mitochondrial complex I deficiency. Alternative splicing results in multiple transcript variants. [provided by RefSeq, May 2014]

Function:
Required for the assembly of the mitochondrial membrane respiratory chain NADH dehydrogenase (Complex I). May deliver of one or more Fe-S clusters to complex I subunits.

Subcellular Location:
Mitochondrion.

Tissue Specificity:
Highest expression in liver and kidney. expressed at significant levels in small intestine and brain (at protein level).

DISEASE:
Defects in NUBPL are a cause of mitochondrial complex I deficiency (MT-C1D) [MIM:252010]. A disorder of the mitochondrial respiratory chain that causes a wide range of clinical manifestations from lethal neonatal disease to adult-onset neurodegenerative disorders. Phenotypes include macrocephaly with progressive leukodystrophy, non-specific encephalopathy, cardiomyopathy, myopathy, liver disease, Leigh syndrome, Leber hereditary optic neuropathy, and some forms of Parkinson disease.

Similarity:
Belongs to the Mrp/NBP35 ATP-binding proteins family.

SWISS:
Q8TB37

Gene ID:
80224

Database links:

Entrez Gene: 80224 Human

Entrez Gene: 76826 Mouse

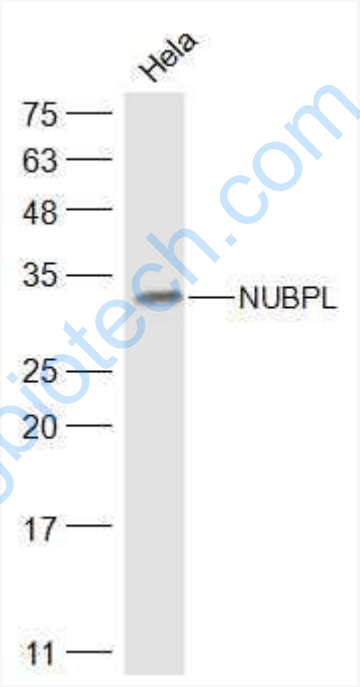
Entrez Gene: 299008 Rat

Omim: 613621 Human

SwissProt: Q8TB37 Human

SwissProt: Q9CWD8 Mouse

Unigene: 288981 Human

	Unigene: 244781 Mouse Unigene: 13455 Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Picture:	 Sample: Hela(Human) Cell Lysate at 30 ug Primary: Anti-NUBPL (SL19505R) at 1/500 dilution Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution Predicted band size: 34 kD Observed band size: 34 kD