LPPRC Rabbit pAb

CatalogNo: YT6371

Key Features

Host Species
- Rabbit

Reactivity
- Human,Mouse,Rat

Applications
- WB,ELISA,IHC

MW
- 153kD (Calculated)

Isotype
- IgG

Recommended Dilution Ratios

WB 1:500-2000
IHC 1:50-300
ELISA 1:2000-20000

Storage

Storage*	-15°C to -25°C/1 year(Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	Synthesized peptide derived from human LPPRC AA range: 1329-1379
Specificity	This antibody detects endogenous levels of LPPRC at Human/Mouse/Rat

Target Information

Gene name	LRPPRC LRP130

Protein Name LPPRC

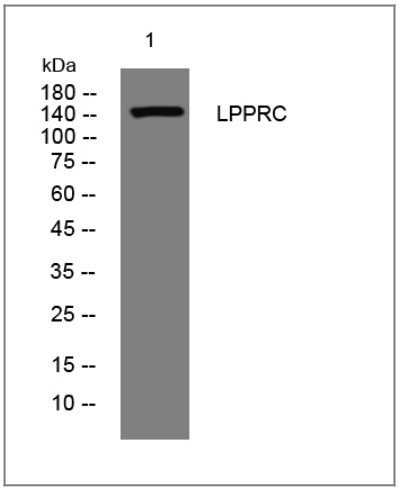
Organism	Gene ID	UniProt ID
Human	10128;	P42704;
Mouse	72416;	Q6PB66;
Rat	313867;	Q5SGE0;

Cellular Localization Mitochondrion. Nucleus, nucleoplasm. Nucleus inner membrane. Nucleus outer membrane. Seems to be predominantly mitochondrial.

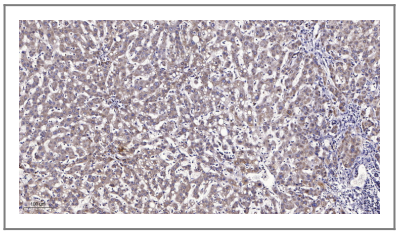
Tissue specificity Expressed ubiquitously. Expression is highest in heart, skeletal muscle, kidney and liver, intermediate in brain, non-mucosal colon, spleen and placenta, and lowest in small intestine, thymus, lung and peripheral blood leukocytes.

Function Disease:Defects in LRPPRC are the cause of Leigh syndrome French-Canadian type (LSFC) [MIM:220111]. Leigh syndrome is a severe neurological disorder characterized by bilaterally symmetrical necrotic lesions in subcortical brain regions that is commonly associated with systemic cytochrome c oxidase (COX) deficiency. In the Saguenay-Lac Saint Jean region of Quebec province in Canada, a biochemically distinct form of Leigh syndrome with COX deficiency has been described. Patients have been observed to have a developmental delay, hypotonia, mild facial dysmorphism, chronic well-compensated metabolic acidosis, and high mortality due to episodes of severe acidosis and coma. Enzyme activity was close to normal in kidney and heart, 50% of normal in fibroblasts and skeletal muscle, and nearly absent in brain and liver. LSFC patients show reduced (<30%) levels of LRPPRC in both fibroblast and liver mitochondria and a specifically reduced translation of COX subunits MT-CO1/COXI and MT-CO3 (COXIII).,Function:May play a role in RNA metabolism in both nuclei and mitochondria. In the nucleus binds to HNRPA1-associated poly(A) mRNAs and is part of nmRNP complexes at late stages of mRNA maturation which are possibly associated with nuclear mRNA export. May bind mature mRNA in the nucleus outer membrane. In mitochondria binds to poly(A) mRNA. Plays a role in translation or stability of mitochondrially encoded cytochrome c oxidase (COX) subunits. May be involved in transcription regulation. Cooperates with PPARGC1A to regulate certain mitochondrially encoded genes and gluconeogenic genes and may regulate docking of PPARGC1A to transcription factors. Seems to be involved in the transcription regulation of the multidrug-related genes MDR1 and MVP. Part of a nuclear factor that binds to the invMED1 element of MDR1 and MVP gene promoters. Binds single-stranded DNA.,sequence Caution:Translation N-terminally extended.,similarity:Contains 20 PPR (pentatricopeptide) repeats.,subcellular location:Seems to be predominantly mitochondrial.,subunit:Interacts with CECR2, HEBP2, MAP1S, RMP/C19orf2 and UXT. Interacts with PPARGC1A. Interacts with FOXO1 (By similarity) Component of mRNP complexes associated with HNRPA1.,tissue specificity:Expressed ubiquitously. Expression is highest in heart, skeletal muscle, kidney and liver, intermediate in brain, non-mucosal colon, spleen and placenta, and lowest in small intestine, thymus, lung and peripheral blood leukocytes.,

Validation Data



Western blot analysis of lysates from Hela cells, primary antibody was diluted at 1:1000, 4°over night



Immunohistochemical analysis of paraffin-embedded human liver cancer. 1, Antibody was diluted at 1:200(4° overnight). 2, Tris-EDTA,pH9.0 was used for antigen retrieval. 3,Secondary antibody was diluted at 1:200(room temperature, 45min).

Contact information

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