Best Tools for immunology Research

LPPRC rabbit pAb

Catalog No :	YT6371
Reactivity :	Human;Mouse;Rat
Applications :	WB;ELISA;IHC
Target :	LPPRC
Gene Name :	LRPPRC LRP130
Protein Name :	LPPRC
Human Gene Id :	10128
Human Swiss Prot No :	P42704
Mouse Gene Id :	72416
Mouse Swiss Prot No :	Q6PB66
Rat Gene Id :	313867
Rat Swiss Prot No :	Q5SGE0
Immunogen :	Synthesized peptide derived from human LPPRC AA range: 1329-1379
Specificity :	This antibody detects endogenous levels of LPPRC at Human/Mouse/Rat
Formulation :	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source :	Polyclonal, Rabbit,IgG
Dilution :	WB 1:500-2000;IHC 1:50-300; ELISA 2000-20000
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.

Concentration : 1 mg/ml

Storage Stability : -15°C to -25°C/1 year(Do not lower than -25°C)

Molecularweight : 153kD

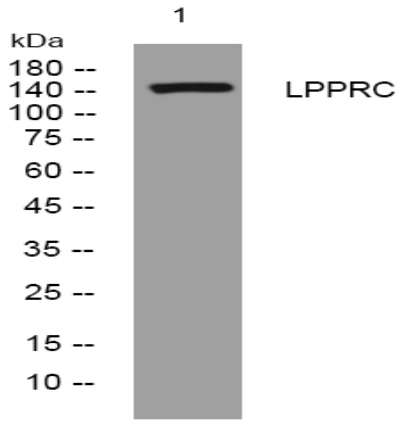
Background : This gene encodes a leucine-rich protein that has multiple pentatricopeptide repeats (PPR). The precise role of this protein is unknown but studies suggest it may play a role in cytoskeletal organization, vesicular transport, or in transcriptional regulation of both nuclear and mitochondrial genes. The protein localizes primarily to mitochondria and is predicted to have an N-terminal mitochondrial targeting sequence. Mutations in this gene are associated with the French-Canadian type of Leigh syndrome. [provided by RefSeq, Mar 2012],

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 (

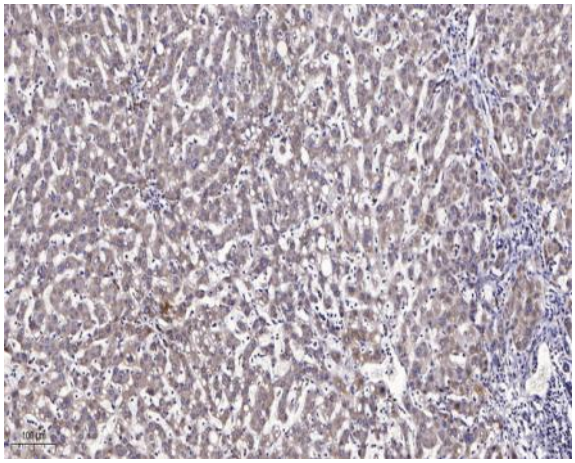
Subcellular Location : Mitochondrion. Nucleus, nucleoplasm. Nucleus inner membrane. Nucleus outer membrane. Seems to be predominantly mitochondrial.

Expression : 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.

Products Images



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).