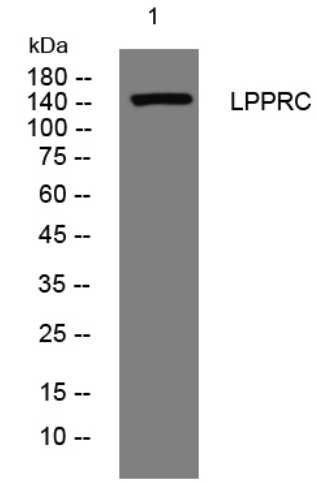
LPPRC rabbit pAb

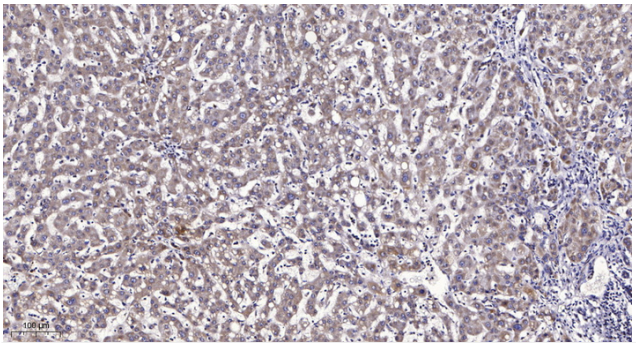
Catalog No	BYab-11006
Isotype	IgG
Reactivity	Human; Mouse;Rat
Applications	WB;ELISA;IHC
Gene Name	LRPPRC LRP130
Protein Name	LPPRC
Immunogen	Synthesized peptide derived from human LPPRC AA range: 1329-1379
Specificity	This antibody detects endogenous levels of LPPRC at Human/Mouse/Rat
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1:500-2000;IHC-p 1:50-300; ELISA 2000-20000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	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 live
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],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

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).