

LRPPRC Recombinant Mouse mAb

Catalog # AP96412

Product Information

Application	WB, IHC-P, IHC-F, IF
Predicted	Human
Host	Mouse
Clonality	Recombinant
Physical State	Liquid
Isotype	IgG2b/Kappa
Purity	affinity purified by Protein G
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 20 PPR (pentatricopeptide) repeats.
SUBUNIT	Interacts with INPP5D/SHIP1 (By similarity). Forms heterodimers composed of LIPR and IL6ST (type I OSM receptor). Also forms heterodimers composed of OSMR and IL6ST (type II OSM receptor). Homodimer. The homodimer binds two molecules of herpes virus IL6. Component of a hexamer of two molecules each of IL6, IL6R and IL6ST. Interacts with HCK.
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 (
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Leucine-rich protein (LRP130) is a cytoplasmic mRNA-binding protein likely to be involved in the processing of mitochondrial DNA transcripts. Defects in the LRPPRC gene that encodes LRP130 result in the French-Canadian type of Leigh syndrome, a severe neurological disorder characterized by lesions in the subcortical region of the brain. LRP130 also interacts with the low-affinity receptor for leukemia inhibitory factor to produce an intracellular signal cascade.

Additional Information

Other Names	CLONE-23970; GP130; LRP130; LSFC; MC4DN5; Lrp157; LPPRC_HUMAN; LRPPRC; 130 kDa leucine-rich protein (LRP 130); LPPRC_RAT; Leucine rich protein 157 (rLRP157); leucine rich pentatricopeptide repeat containing; Leigh syndrome, French-Canadian type (cytochrome oxidase deficiency)
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Dilution	WB=1:500-1000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.
Name	LRPPRC Recombinant Mouse mAb
Function	<p>LRPPRC is thought to play a role in RNA metabolism in both nuclei and mitochondria. In the nucleus, LRPPRC 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. In mitochondria, LRPPRC binds to poly(A) mRNA. LRPPRC may play a role in translation or stability of mitochondrially encoded cytochrome c oxidase (COX) subunits. Defects in LRPPRC are the cause of Leigh syndrome French-Canadian type (LSFC). 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.</p>

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.