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DLD Rabbit pAb

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Catalog # AP57025

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P09622
Reactivity	Mouse, Rat
Predicted	Human, Dog, Horse, Sheep, Xenopus
Host	Rabbit
Clonality	Polyclonal
Calculated MW	54177
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Lipoamide Dehydrogenase
Epitope Specificity	241-340/509
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Mitochondrion matrix.
SIMILARITY	Belongs to the class-I pyridine nucleotide-disulfide oxidoreductase family.
Post-translational modifications	Tyrosine phosphorylated.
DISEASE	Note=Defects in DLD are involved in the development of congenital infantile lactic acidosis. Defects in DLD are a cause of maple syrup urine disease (MSUD) [MIM:248600]. MSUD is characterized by mental and physical retardation, feeding problems and a maple syrup odor to the urine. The keto acids of the branched-chain amino acids are present in the urine, resulting from a block in oxidative decarboxylation.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes a member of the class-I pyridine nucleotide-disulfide oxidoreductase family. The encoded protein has been identified as a moonlighting protein based on its ability to perform mechanistically distinct functions. In homodimeric form, the encoded protein functions as a dehydrogenase and is found in several multi-enzyme complexes that regulate energy metabolism. However, as a monomer, this protein can function as a protease. Mutations in this gene have been identified in patients with E3-deficient maple syrup urine disease and lipoamide dehydrogenase deficiency. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jan 2014]

Additional Information

Gene ID	1738

Other Names	Dihydrolipoyl dehydrogenase, mitochondrial, 1.8.1.4, Dihydrolipoamide dehydrogenase, Glycine cleavage system L protein, DLD, GCSL, LAD, PHE3
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=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.

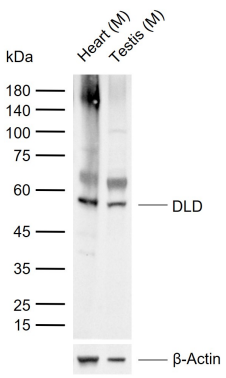
Protein Information

Name	DLD (HGNC:2898)
Synonyms	GCSL, LAD, PHE3
Function	Lipoamide dehydrogenase is a component of the glycine cleavage system as well as an E3 component of three alpha-ketoacid dehydrogenase complexes (pyruvate-, alpha-ketoglutarate-, and branched- chain amino acid-dehydrogenase complex) (PubMed:15712224, PubMed:16442803, PubMed:16770810, PubMed:17404228, PubMed:20160912, PubMed:20385101). The 2-oxoglutarate dehydrogenase complex is mainly active in the mitochondrion (PubMed:29211711). A fraction of the 2-oxoglutarate dehydrogenase complex also localizes in the nucleus and is required for lysine succinylation of histones: associates with KAT2A on chromatin and provides succinyl-CoA to histone succinyltransferase KAT2A (PubMed:29211711). In monomeric form may have additional moonlighting function as serine protease (PubMed:17404228). Involved in the hyperactivation of spermatazoa during capacitation and in the spermatazoal acrosome reaction (By similarity). The pyruvate dehydrogenase (PDH) complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2), and thereby links cytoplasmic glycolysis and the mitochondrial tricarboxylic acid (TCA) cycle (Probable). It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and dihydrolipoamide dehydrogenase (E3) (Probable). The E3 subunit catalyzes reoxidation of the dihydrolipoyl moiety on lipoyl-bearing domains (LBDs) of E2 with NAD+ as the ultimate electron acceptor (PubMed:16442803, PubMed:16770810, PubMed:20160912, PubMed:20385101).
Cellular Location	Mitochondrion matrix. Nucleus. Cell projection, cilium, flagellum {ECO:0000250|UniProtKB:Q811C4}. Cytoplasmic vesicle, secretory vesicle, acrosome. Note=Mainly localizes in the mitochondrion. A small fraction localizes to the nucleus, where the 2- oxoglutarate dehydrogenase complex is required for histone succinylation.

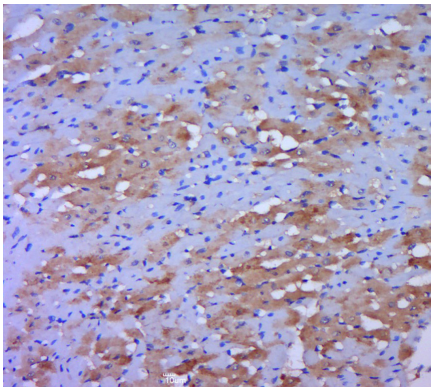
Background

This gene encodes a member of the class-I pyridine nucleotide-disulfide oxidoreductase family. The encoded protein has been identified as a moonlighting protein based on its ability to perform mechanistically distinct functions. In homodimeric form, the encoded protein functions as a dehydrogenase and is found in several multi-enzyme complexes that regulate energy metabolism. However, as a monomer, this protein can function as a protease. Mutations in this gene have been identified in patients with E3-deficient maple syrup urine disease and lipoamide dehydrogenase deficiency. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jan 2014]

Images



Sample:
Lane 1: Mouse Heart tissue lysates
Lane 2: Mouse Testis tissue lysates
Primary: Anti-DLD (AP57025) at 1/1000 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 50 kDa
Observed band size: 52 kDa



Paraformaldehyde-fixed, paraffin embedded (rat heart tissue); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (DLDD) Polyclonal Antibody, Unconjugated (AP57025) at 1:400 overnight at 4°C, followed by a conjugated secondary (sp-0023) for 20 minutes and DAB staining.