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DLDD/Lipoamide Dehydrogenase Polyclonal Antibody

Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP57025

Product Information

Application	WB, IHC-P, IHC-F, IF, ICC, E
Primary Accession	P09622
Reactivity	Rat, Dog, Bovine
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

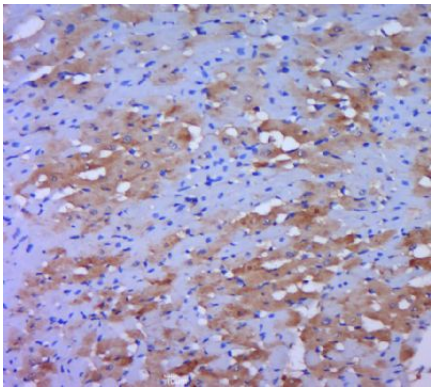
Gene ID	1738
Other Names	Dihydrolipoyl dehydrogenase, mitochondrial, 1.8.1.4, Dihydrolipoamide

	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,ICC=1:100-500,IF=1:100-500,ELISA=1:5000-10000
Format	0.01M TBS(pH7.4) with 1% BSA, 0.09% (W/V) sodium azide and 50% Glyce
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
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).
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.

Images



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.