

Rabbit Anti-DLDD/Lipoamide Dehydrogenase antibody

SL18295R

ipoamide ehydrogenase oranched chain aplha- onent of pyruvate in keto acid otein; Glycine cleavage reductase; Lipoyl HUMP00000206746; HE3; Pyruvate
1:400-800ICC=1:100-
/Lipoamide
Glycerol.

Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
PubMed:	PubMed
	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]
	Function: Lipoamide dehydrogenase is a component of the glycine cleavage system as well as of the alpha-ketoacid dehydrogenase complexes. Involved in the hyperactivation of spermatazoa during capacitation and in the spermatazoal acrosome reaction.
	Subcellular Location: Mitochondrion matrix.
	Post-translational modifications: Tyrosine phosphorylated.
Product Detail:	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.
	Belongs to the class-I pyridine nucleotide-disulfide oxidoreductase family. SWISS: P09622
	Gene ID: 1738
	Database links:
	Entrez Gene: 1738 Human
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