

# DLD Rabbit pAb

DLD Rabbit pAb Catalog # AP57025

#### **Product Information**

**Application** WB, IHC-P, IHC-F, IF

Primary Accession P09622

**Reactivity** Mouse, Dog, Horse, Sheep

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**Tyrosine phosphorylated.

modifications

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

**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.

## **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:

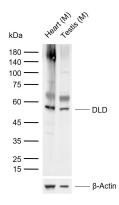
Lane 1: Mouse Heart tissue lysates Lane 2: Mouse Testis tissue lysates

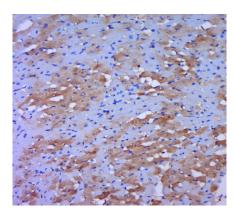
Primary: Anti-DLD (AP57025) at 1/1000 dilution

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.

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