

DLAT Rabbit pAb

Catalog # AP98751

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	P10515
Other Accession	P10515
Reactivity	Rat, Human, Mouse
Predicted	Pig, Cat, Rabbit, Zebrafish, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	68997
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human PDCE2
Epitope Specificity	231-330/647
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the 2-oxoacid dehydrogenase family. Contains 2 lipoyl-binding domains.
DISEASE	Note=Primary biliary cirrhosis is a chronic, progressive cholestatic liver disease characterized by the presence of antimitochondrial autoantibodies in patients' serum. It manifests with inflammatory obliteration of intra-hepatic bile duct, leading to liver cell damage and cirrhosis. Patients with primary biliary cirrhosis show autoantibodies against the E2 component of pyruvate dehydrogenase complex. Defects in DLAT are the cause of pyruvate dehydrogenase E2 deficiency (PDHE2 deficiency) [MIM:245348]; also known as lactic acidemia due to defect of E2 lipoyl transacetylase of the pyruvate dehydrogenase complex. Pyruvate dehydrogenase (PDH) deficiency is a major cause of primary lactic acidosis and neurological dysfunction in infancy and early childhood. In this form of PDH deficiency episodic dystonia is the major neurological manifestation, with other more common features of pyruvate dehydrogenase deficiency, such as hypotonia and ataxia, being less prominent.
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 component E2 of the multi-enzyme pyruvate dehydrogenase complex (PDC). PDC resides in the inner mitochondrial membrane and catalyzes the conversion of pyruvate to acetyl coenzyme A. The protein product of this gene, dihydrolipoamide acetyltransferase, accepts acetyl groups formed by the oxidative decarboxylation of pyruvate and transfers them to coenzyme A. Dihydrolipoamide acetyltransferase is the antigen for antimitochondrial antibodies. These autoantibodies are present in nearly 95% of patients with the autoimmune liver disease primary biliary cirrhosis (PBC). In PBC, activated T lymphocytes attack and destroy epithelial cells in the bile duct where this protein is abnormally distributed and overexpressed. PBC eventually leads to cirrhosis and liver failure. Mutations in this gene are also a cause of pyruvate dehydrogenase E2 deficiency which causes primary lactic acidosis in infancy and early childhood.[provided by

Additional Information

Gene ID	1737
Other Names	PDC-E2; PDCE2; DLTA; E2; PBC; 6332404G05Rik; noa; nrb; wu:fc14f10; wu:fc21f08; ODP2_BOVIN; DLAT; Dihydrolipoamide acetyltransferase component of pyruvate dehydrogenase complex; Pyruvate dehydrogenase complex component E2 (PDC-E2 PDCE2); 2.3.1.12; ODP2_HUMAN; 70 kDa mitochondrial autoantigen of primary biliary cirrhosis (PBC); M2 antigen complex 70 kDa subunit; ODP2_MOUSE; ODP2_RAT; dihydrolipoamide S-acetyltransferase; dihydrolipoyllysine-residue acetyltransferase; E2 component of pyruvate dehydrogenase complex; dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex, mitochondrial
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	DLAT (HGNC:2896)
Synonyms	DLTA
Function	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). Within this complex, the catalytic function of this enzyme is to accept, and to transfer to coenzyme A, acetyl groups from acetyl- lipoyl moiety generated by the pyruvate dehydrogenase, leading to acetyl-CoA formation (Probable).
Cellular Location	Mitochondrion matrix {ECO:0000250 UniProtKB:P08461}

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