

PDC-E2 Monoclonal Antibody

Catalog No: YM1071

Reactivity: Human;Rabbit

Applications: WB

Target: PDC-E2

Fields: >>Glycolysis / Gluconeogenesis;>>Citrate cycle (TCA cycle);>>Pyruvate

metabolism;>>Metabolic pathways;>>Carbon metabolism

Gene Name: DLAT

Protein Name: Dihydrolipoyllysine-residue acetyltransferase component of pyruvate

dehydrogenase complex mitochondrial

Human Gene Id: 1737

Human Swiss Prot P10515

No:

Mouse Swiss Prot

No:

Rat Swiss Prot No: P08461

Immunogen: Purified recombinant human PDC-E2 protein fragments expressed in E.coli.

Specificity: PDC-E2 Monoclonal Antibody detects endogenous levels of PDC-E2 protein.

Formulation : Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Source: Monoclonal, Mouse

Q8BMF4

Dilution: WB 1:1000 - 1:2000. Not yet tested in other applications.

Purification : Affinity purification

Concentration: 1 mg/ml

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Storage Stability: -15°C to -25°C/1 year(Do not lower than -25°C)

Molecularweight: 69kD

Cell Pathway : __Glycolysis / Gluconeogenesis;Citrate cycle (TCA cycle);Pyruvate metabolism;

Background: 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 enventually leads to cirrhosis

and liver failure. Mutations in this gene are also a cause of pyruvate dehydrogenase E2 deficiency which causes primary lact

Function: catalytic activity:Acetyl-CoA + enzyme N(6)-(dihydrolipovl)lysine = CoA +

enzyme N(6)-(S-acetyldihydrolipoyl)lysine.,cofactor:Binds 2 lipoyl cofactors covalently.,disease:Defects in DLAT are the cause of pyruvate dehydrogenase E2 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.,disease:Primary biliary cirrhosis is a chronic, progressive cholestatic liver disease characterized by the presence of antimitochondrial

autoantibodies in patients' serum. It ma

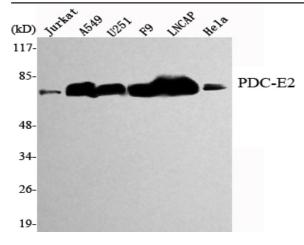
Subcellular Location:

Mitochondrion matrix.

Expression: Heart, Keratinocyte carcinoma, Kidney, Liver, Placenta, Testis,

Products Images

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Western Blot analysis using PDC-E2 Monoclonal Antibody against Jurkat, A549, U251, F9, LNCAP, HeLa cell lysate.