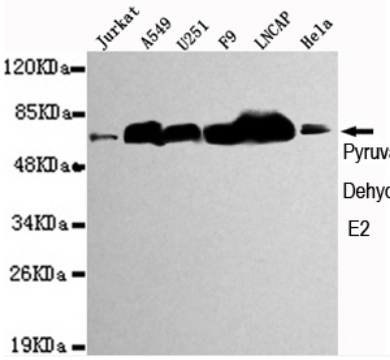
Pyruvate Dehydrogenase E2 mouse mAb

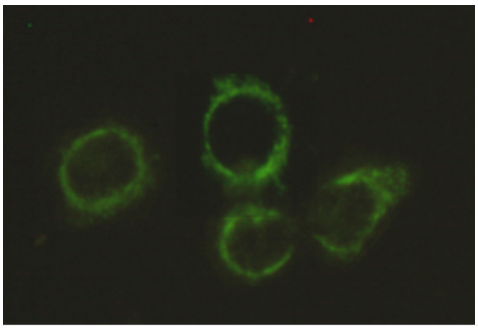
Catalog No	YP-Ab-02364
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB;ICC;IP
Gene Name	dlat
Protein Name	
Immunogen	Purified recombinant human Pyruvate Dehydrogenase E2 protein fragments expressed in E.coli.
Specificity	This antibody detects endogenous levels of Pyruvate Dehydrogenase E2 and does not cross-react with related proteins.
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Monoclonal, Mouse
Purification	The antibody was affinity-purified from mouse ascites by affinity-chromatography using epitope-specific immunogen.
Dilution	wb 1:1000 icc 1:300
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	70 kDa mitochondrial autoantigen of primary biliary cirrhosis;anti DLAT; Dihydrolipoamide acetyltransferase component of pyruvate dehydrogenase complex; Dihydrolipoamide;Dihydrolipoamide S Acetyltransferase;Dihydrolipoamide S-acetyltransferase (E2 component of pyruvate dehydrogenase complex);Dihydrolipoamide S-Acetyltransferase;Dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex;dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex mitochondrial;DLAT;DLAT;DLTA;E2;E2 component of pyruvate dehydrogenase complex;EC 2.3.1.12;M2 antigen complex 70 kDa subunit;M2 Antigen Complex 70kD Subunit;mitochondrial;ODP2_HUMAN;PBC;PDC E2;PDC-E2;PDCE2;Pyruvate dehydrogenase complex component E2;Pyruvate dehydrogenase complex E2 subunit;S acetyltransferase component of pyruvate dehydrogenase complex.
Observed Band	69kD
Cell Pathway	Mitochondrion matrix.

Tissue Specificity	Heart,Keratinocyte carcinoma,Kidney,Liver,Placenta,Testis,
Function	catalytic activity:Acetyl-CoA + enzyme N(6)-(dihydrolipoyl)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
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
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

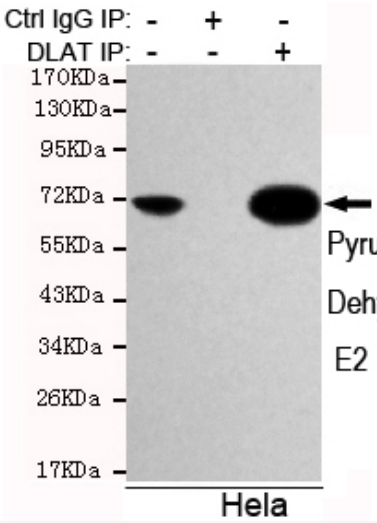
Products Images



Western blot detection of Pyruvate Dehydrogenase E2 in Jurkat,A549,U251,F9,Lncap and Hela cell lysates using Pyruvate Dehydrogenase E2 mouse mAb (1:1000 diluted).Predicted band size:69KDa.Observed band size:69KDa.



Immunocytochemistry stain of Hela using Pyruvate Dehydrogenase E2 mouse mAb (1:300).



Immunoprecipitation analysis of Hela cell lysates using Pyruvate Dehydrogenase E2 mouse mAb.