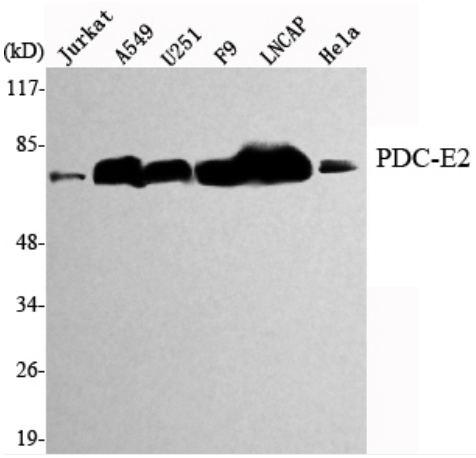
PDC-E2 Monoclonal Antibody

Catalog No	BYab-02352
Isotype	IgG
Reactivity	Human;Rabbit
Applications	WB
Gene Name	DLAT
Protein Name	Dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex mitochondrial
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	Purified mouse monoclonal in buffer containing 0.1M Tris-Glycine (pH 7.4, 150 mM NaCl) with 0.2% sodium azide, 50% glycerol.
Source	Monoclonal, Mouse
Purification	Affinity purification
Dilution	Western Blot: 1/1000 - 1/2000. Not yet tested in other applications.
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	DLAT; DLTA; Dihydrolipoyllysine-residue acetyltransferase component of pyruvate dehydrogenase complex; mitochondrial; 70 kDa mitochondrial autoantigen of primary biliary cirrhosis; PBC; Dihydrolipoamide acetyltransferase component of pyruva
Observed Band	
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 analysis using PDC-E2 Monoclonal Antibody against Jurkat, A549, U251, F9, LNCAP, HeLa cell lysate.