



Pyruvate Dehydrogenase E2 Rabbit mAb

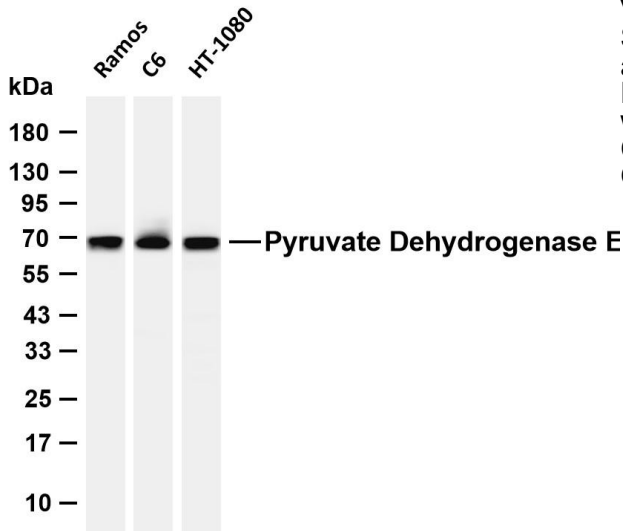
Catalog No	YP-rAb-17083
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,IP,ELISA
Gene Name	DLAT
Protein Name	
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:500-1:2000; WB 1:2000-1:10000; IF 1:200-1:1000; ELISA 1:5000-1:20000; IP 1:50-1:200; Note: For IHC, we suggest antigen retrieval with TE buffer pH 9.0
Concentration	0.5 mg/ml
Purity	≥90%
Storage Stability	-15° C to -25° C/1 year(Do not lower than -25° C)
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 ; 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
Calculated Molecular Weight	69kD



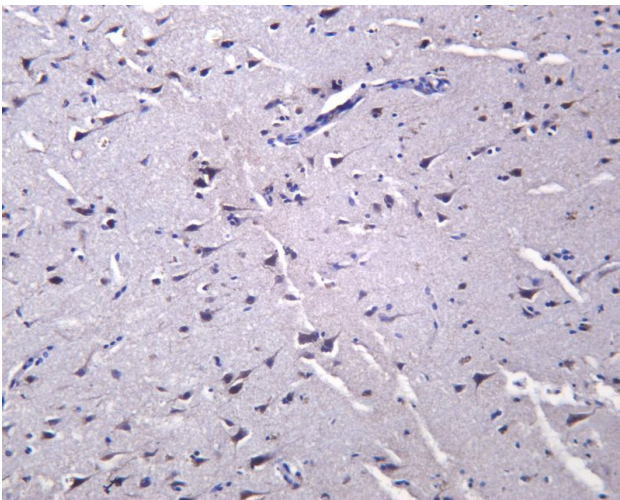


Cell Pathway	Mitochondrion matrix.
Tissue Specificity	Heart,Keratinocyte carcinoma,Kidney,Liver,Placenta,Testis,
Function	<p>Catalytic activity:Acetyl-CoA + enzyme N(6)-(dihydrolipoyl)lysine = CoA + enzyme N(6)-(S-acetyldihydrolipoyl)lysine.,cofactor:Binds 2 lipoyl cofactors covalently.,Disease:Defects 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 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.,Function:The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2). It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).,sequence Caution:Contaminating sequence. Sequence of unknown origin in the N-terminal part.,similarity:Belongs to the 2-oxoacid dehydrogenase family.,similarity:Contains 1 lipoyl-binding domain.,similarity:Contains 2 lipoyl-binding domains.,subunit:20 to 30 alpha(2)-beta(2) tetramers of E1 + 6 homodimers of E3 + 60 copies of E2.,</p>
Background	<p>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 lact</p>
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.

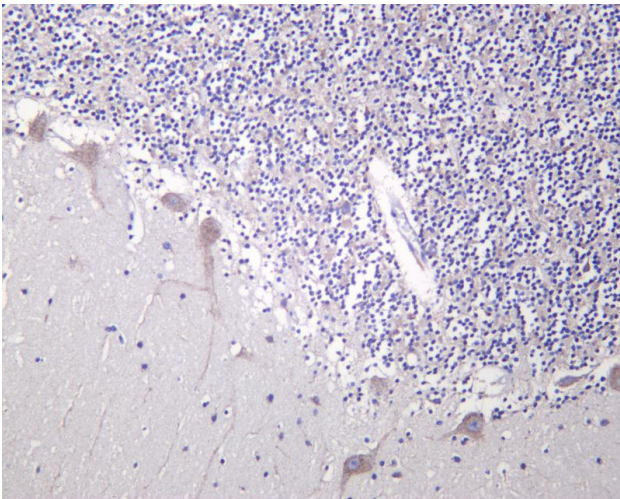




Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-Pyruvate Dehydrogenase E2 antibody. The HRP-conjugated Goat anti-Rabbit IgG (H + L) antibody was used to detect the antibody. Lane 1: Ramos Lane 2: C6 Lane 3: HT-1080 Predicted band size: 69kDa Observed band size: 69kDa



Human brain was stained with anti-Pyruvate Dehydrogenase E2 Rabbit antibody



Human cerebellum was stained with anti-Pyruvate Dehydrogenase E2 Rabbit antibody

