G-Biosciences Image not found or type unknown

Immunotag™ PDC-E2 Monoclonal Antibody

Antibody Specification

Catalog No.	ITM1071
Product Description	Immunotag™ PDC-E2 Monoclonal Antibody
Size	50 µg, 100 µg
Conjugation	HRP, Biotin, FITC, Alexa Fluor® 350, Alexa Fluor® 405, Alexa Fluor® 488, Alexa Fluor® 555, Alexa Fluor® 594, Alexa Fluor® 647
IMPORTANT NOTE	This product is custom manufactured with a lead time of 3-4 weeks. Once in production, this item cannot be cancelled from an order and is not eligible for return.
Target Protein	PDC-E2
Clonality	Monoclonal
Storage/Stability	-20°C/1 year
Application	WB
Recommended Dilution	Western Blot: 1/1000 - 1/2000. Not yet tested in other applications.
Concentration	1 mg/ml
Reactive Species	Human,Rb
Host Species	Mouse
Immunogen	Purified recombinant human PDC-E2 protein fragments expressed in Ecoli
Specificity	PDC-E2 Monoclonal Antibody detects endogenous levels of PDC-E2 protein.
Purification	Affinity purification
Form	Purified mouse monoclonal in buffer containing 0.1M Tris-Glycine (pH 7.4, 150 mM NaCl) with 0.2% sodium azide, 50% glycerol.
Gene Name	DLAT
Accession No.	P10515 Q8BMF4 P08461
Alternate Names	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

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Description	dihydrolipoamide S-acetyltransferase(DLAT) Homo sapiens 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
Cell Pathway/ Category	Glycolysis / Gluconeogenesis,Citrate cycle (TCA cycle),Pyruvate metabolism,
Protein Expression	Heart,Keratinocyte carcinoma,Kidney,Liver,Placenta,Testis,
Subcellular Localization	mitochondrion,mitochondrial matrix,mitochondrial pyruvate dehydrogenase complex,myelin sheath,pyruvate dehydrogenase complex,
Protein 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 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.,
Usage	For Research Use Only! Not for diagnostic or therapeutic procedures.