Best Tools for immunology Research

pyruvate dehydrogenase (lipoamide) α 1 mouse mAb

Catalog No :	YM1234
Reactivity :	Human;Mouse
Applications :	WB;ICC
Target :	PDHA1
Fields :	>>Glycolysis / Gluconeogenesis;>>Citrate cycle (TCA cycle);>>Pyruvate metabolism;>>Metabolic pathways;>>Carbon metabolism;>>HIF-1 signaling pathway;>>Glucagon signaling pathway;>>Central carbon metabolism in cancer;>>Diabetic cardiomyopathy
Gene Name :	pdha1
Human Gene Id :	5160
Human Swiss Prot No :	P08559
Mouse Swiss Prot No :	P35486
Immunogen :	Purified recombinant human Pyruvate Dehydrogenase protein fragments expressed in E.coli.
Specificity :	This antibody detects endogenous levels of pyruvate dehydrogenase (lipoamide) alpha 1 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
Dilution :	wb 1:1000 icc 1:100
Purification :	The antibody was affinity-purified from mouse ascites by affinity-chromatography using epitope-specific immunogen.
Concentration :	1 mg/ml

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

Observed Band : 43kD

Cell Pathway : Glycolysis / Gluconeogenesis;Citrate cycle (TCA cycle);Valine; leucine and isoleucine biosynthesis;Pyruvate metabolism;Butanoate metabolism;

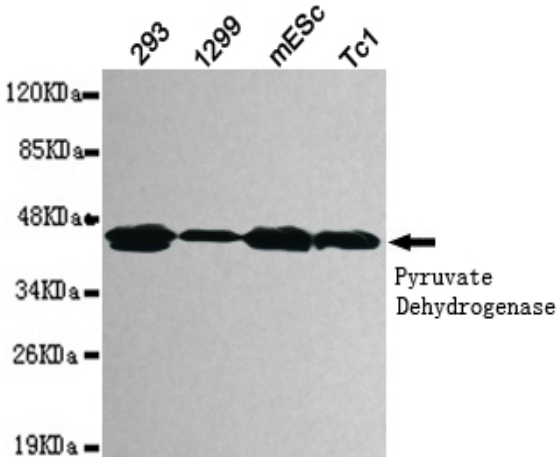
Background : The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2), and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. The PDH complex is composed of multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3). The E1 enzyme is a heterotetramer of two alpha and two beta subunits. This gene encodes the E1 alpha 1 subunit containing the E1 active site, and plays a key role in the function of the PDH complex. Mutations in this gene are associated with pyruvate dehydrogenase E1-alpha deficiency and X-linked Leigh syndrome. Alternatively spliced transcript variants encoding different isoforms have been found for this gene.[provided by RefSeq, Mar 2010],

Function : catalytic activity:Pyruvate + [dihydrolipoyllysine-residue acetyltransferase] lipoyllysine = [dihydrolipoyllysine-residue acetyltransferase] S-acetyldihydrolipoyllysine + CO(2).,cofactor:Thiamine pyrophosphate.,disease:Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival complicated by developmental delay, seizures, ataxia, apnea, and in some cases to an X-linked form of Leigh syndrome (LS) (Leigh encephalomyelopathy).,disease:Defects in PDHA1 are the cause of X-linked Leigh syndrome (LS) [MIM:308930]. LS is an early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in o

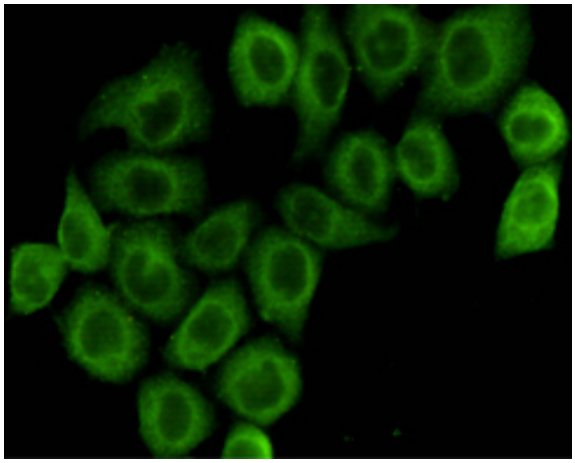
Subcellular Location : Mitochondrion matrix.

Expression : Ubiquitous.

Products Images



Western blot detection of pyruvate dehydrogenase (lipoamide) alpha 1 in 293,1299,mEsc and Tc1 cell lysates using pyruvate dehydrogenase (lipoamide) alpha 1 mouse mAb (1:1000 diluted).Predicted band size: 43KDa.Observed band size: 43KDa.



Immunocytochemistry staining of HeLa cells fixed with 4% Paraformaldehyde and using anti-pyruvate dehydrogenase (lipoamide) alpha 1 mouse mAb(dilution 1:100).