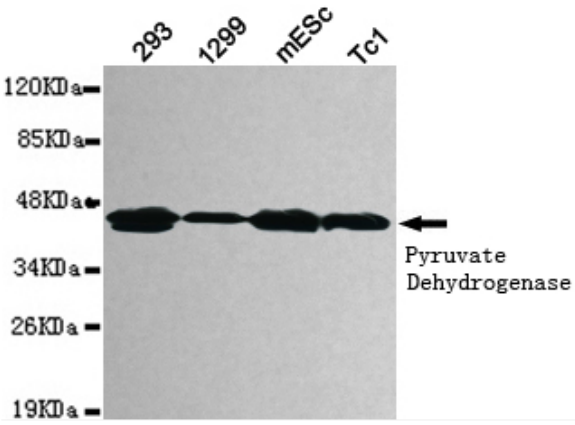
pyruvate dehydrogenase (lipoamide) α 1 mouse mAb

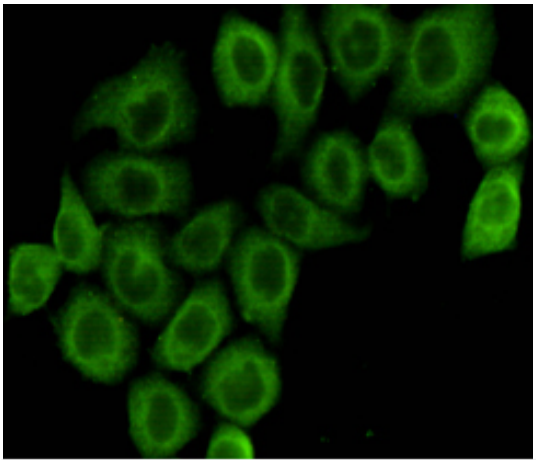
Catalog No	BYab-03449
Isotype	IgG
Reactivity	Human;Mouse
Applications	WB;ICC
Gene Name	pdha1
Protein Name	
Immunogen	Purified recombinant human Pyruvate Dehydrogenase protein fragments expressed in E.coli.
Specificity	This antibody detects endogenous levels of pyruvate dehydrogenase (lipoamide) alpha 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
Purification	The antibody was affinity-purified from mouse ascites by affinity-chromatography using epitope-specific immunogen.
Dilution	wb 1:1000 icc 1:100
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	mitochondrial;ODPA_HUMAN;PDH;PDHA;PDHA1;PDHCE1A;PDHE1 A type I;PDHE1-A type I;PHE1A; Pyruvate Dehydrogenase (lipoamide) alpha 1;Pyruvate dehydrogenase complex, E1 alpha polypeptide 1;Pyruvate Dehydrogenase E1 alpha;Pyruvate dehydrogenase E1 component subunit alpha;Pyruvate dehydrogenase E1 component subunit alpha, somatic form, mitochondrial;somatic form.
Observed Band	43kD
Cell Pathway	Mitochondrion matrix.
Tissue Specificity	Ubiquitous.
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
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],
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 (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).