



# pyruvate dehydrogenase (lipoamide) $\alpha$ 1 mouse mAb

|                                    |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                           |
|------------------------------------|---------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Catalog No</b>                  | YP-Ab-03449                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>Isotype</b>                     | IgG                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
| <b>Reactivity</b>                  | Human;Mouse                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>Applications</b>                | WB;ICC                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                    |
| <b>Gene Name</b>                   | pdha1                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Immunogen</b>                   | Purified recombinant human Pyruvate Dehydrogenase protein fragments expressed in E.coli.                                                                                                                                                                                                                                                                                                                                                                                                                                  |
| <b>Specificity</b>                 | This antibody detects endogenous levels of pyruvate dehydrogenase (lipoamide) alpha 1 and does not cross-react with related proteins.                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Formulation</b>                 | Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.                                                                                                                                                                                                                                                                                                                                                                                                                                                   |
| <b>Source</b>                      | Monoclonal, Mouse                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                         |
| <b>Purification</b>                | The antibody was affinity-purified from mouse ascites by affinity-chromatography using epitope-specific immunogen.                                                                                                                                                                                                                                                                                                                                                                                                        |
| <b>Dilution</b>                    | wb 1:1000 icc 1:100                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
| <b>Concentration</b>               | 1 mg/ml                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                   |
| <b>Purity</b>                      | $\geq 90\%$                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>Storage Stability</b>           | -20°C/1 year                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                              |
| <b>Synonyms</b>                    | PDHA1;mitochondrial;ODPA_HUMAN;PDH;PDHA;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;somatic form; somatic form.                                                                                                                                                                                                                       |
| <b>Observed Band</b>               | 43kD                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                      |
| <b>Calculated Molecular Weight</b> | 43kD                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                      |
| <b>Cell Pathway</b>                | Mitochondrion matrix.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Tissue Specificity</b>          | Ubiquitous.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>Function</b>                    | 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<sub>2</sub>, 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],

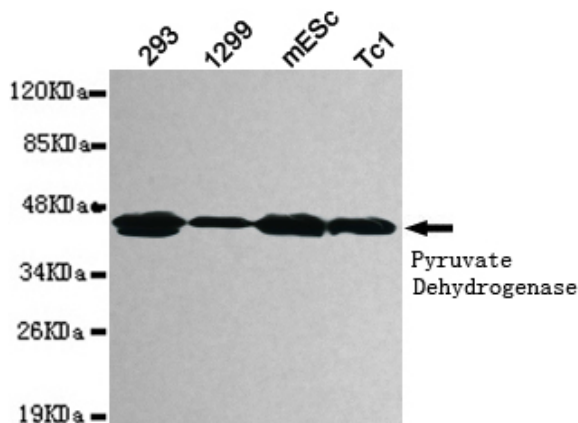
## matters needing attention

Avoid repeated freezing and thawing!

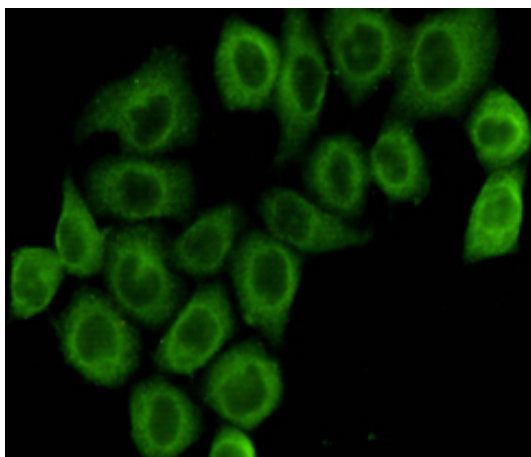
## Usage suggestions

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: 43 kDa. Observed band size: 43 kDa.



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