



Pyruvate Dehydrogenase E1 α Rabbit mAb

Catalog No	YP-rAb-18004
Isotype	IgG
Reactivity	Human,Mouse,Rat
Applications	WB,IHC,IF,IP,ELISA
Gene Name	PDHA1 PHE1A
Protein Name	PDHA1 ODP
Purification Process	Protein A
Specificity	Endogenous
Formulation	PBS, 50% glycerol, 0.05% Proclin 300, 0.05%BSA
Source	Monoclonal, Rabbit,IgG
Dilution	IHC 1:200-1:1000; WB 1:1000-1:5000; 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	PDHA1;PDHA1 ODP;PDHA1 ; PHE1A ; Pyruvate dehydrogenase E1 component subunit alpha ; somatic form, mitochondrial ; PDHE1-A type I;PDHA1 PHE1A
Observed Band	43kD
Calculated Molecular Weight	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 one or more areas of the central nervous system, including the brainstem, thalamus, basal ganglia, cerebellum, and spinal cord . The lesions are areas of demyelination, gliosis, necrosis, spongiosis, or capillary proliferation. Clinical symptoms depend on which areas of the central nervous system are involved. The most common underlying cause is a defect in oxidative phosphorylation. LS may be a feature of a deficiency of any of the mitochondrial respiratory chain complexes.,enzyme regulation:E1 activity is regulated by phosphorylation (inactivation) and dephosphorylation (activation) of the alpha subunit.,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).,subunit:Tetramer of 2 alpha and 2 beta subunits.,tissue specificity:Ubiquitous.,

Background

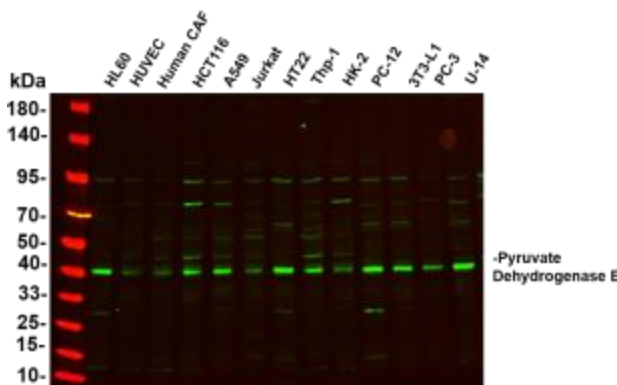
The pyruvate dehydrogenase (PDH) complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2), and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. The PDH complex is composed of multiple copies 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

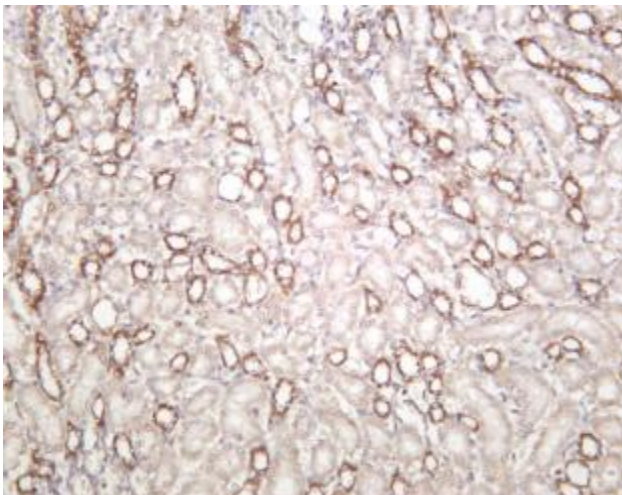
Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

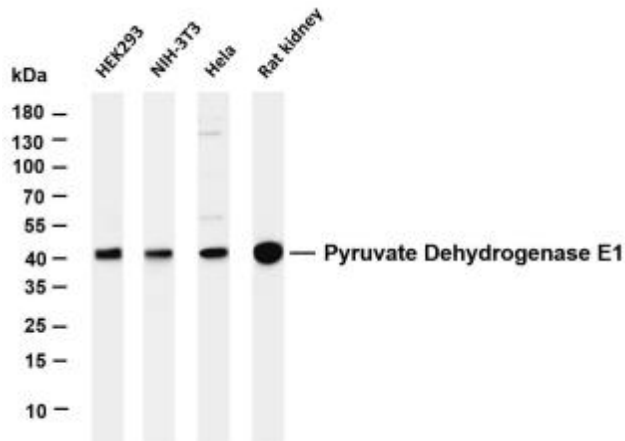


Various whole cell lysates were separated by 4-20% SDS-PAGE, and the primary antibody was used at 4°C, over night with a 1:2500 dilution. The Dylight 800-conjugated Goat anti-Rabbit antibody

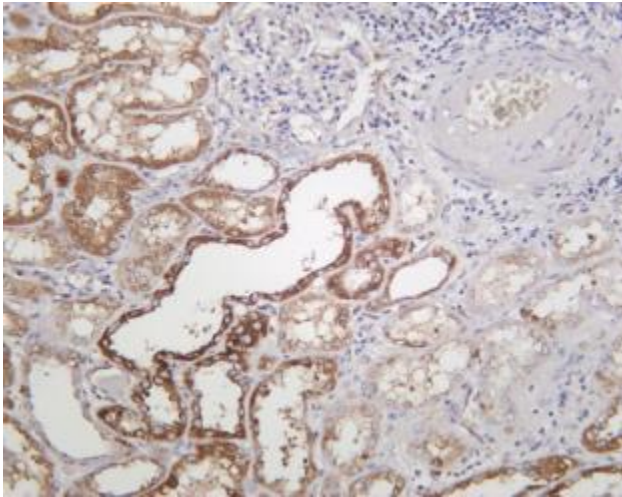


Rat kidney was stained with anti-Pyruvate Dehydrogenase E1 α rabbit antibody

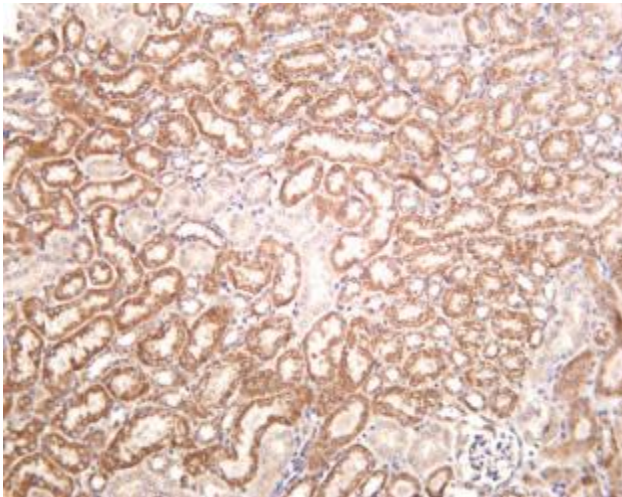




Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-Pyruvate Dehydrogenase E1 α antibody. The HRP-conjugated Goat anti-Rabbit IgG(H + L) antibody was used to detect the antibody. Lane 1: HEK293 Lane 2: NIH-3T3 Lane 3: Hela Lane 4: Rat kidney Predicted band size: 43kDa Observed band size: 43kDa



Human kidney was stained with anti-Pyruvate Dehydrogenase E1 α rabbit antibody



Mouse kidney was stained with anti-Pyruvate Dehydrogenase E1 α rabbit antibody

