# Pyruvate Dehydrogenase E1 α (ANT0022R) Rabbit mAb

CatalogNo: ANT8190 Recombinant

Formulation: PBS,50%glycerol,0.05%Proclin 300,0.05%BSA
Quantity : 100 ug/vial

| Host Species | Reactivity | Applications |
|---|---|---|
| • Rabbit | • Human,Mouse,Rat, | • WB,IHC,IF,IP,ELISA |

| MW | Isotype |
|---|---|
| • 43kD (Calculated)<br>43kD (Observed) | • IgG,Kappa |

## Recommended Dilution Ratios

**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,**

## Storage

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

## Basic Information

| | |
|---|---|
| **Clonality** | Monoclonal |
| **Clone Number** | ANT0022R |

## Immunogen Information

| | |
|---|---|
| **Squence** | Pyruvate dehydrogenase E1 component subunit alpha somatic form mitochondrial |
| **Specificity** | Endogenous |

# Target Information

**Gene name** >>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

**Protein Name** PDHA1 ODPA

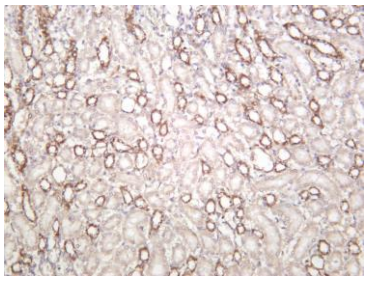
| Organism | Gene ID | UniProt ID |
|---|---|---|
| Human | 5160; | P08559; |
| Mouse | 18597; | P35486; |
| Rat | | P26284; |

**Cellular Localization** Mitochondrion matrix

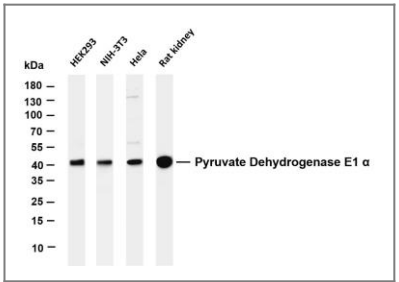
**Tissue specificity** Ubiquitous.

**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.,

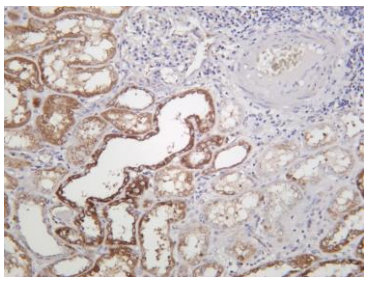
## Validation Data



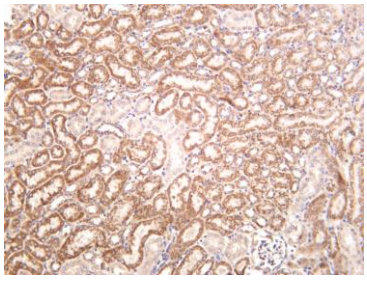
Rat kidney was stained with anti-Pyruvate Dehydrogenase E1 α (ANT0022R) rabbit antibody




Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-Pyruvate Dehydrogenase E1 α (ANT0022R) 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 (ANT0022R) rabbit antibody



Mouse kidney was stained with anti-Pyruvate Dehydrogenase E1 α (ANT0022R) rabbit antibody

For Research use only, not for diagnostics and clinical use
Contact Antagene Inc Tel 1-866-964-2589 Email: info@antageneinc.com