



## Mouse Monoclonal Antibody **Pyruvate Dehydrogenase E1 $\alpha$** conjugated to Sepharose Beads

CatalogNo: **ANT8190-S**

Size 200ul

Storage Store at 4 °C for frequent use

### Description

This Antagene antibody is immobilized via covalent binding of primary amino groups to N-hydroxysuccinimide (NHS)-activated sepharose beads. It is useful for immunoprecipitation assays.

## **Pyruvate Dehydrogenase E1 $\alpha$ (ANT0022R) Rabbit mAb**

Formulation: 50% slurry in PBS pH 7.2 with 0.01mg NaN<sub>3</sub> preservative.

### Host Species

- Rabbit
- Human, Mouse, Rat,

### Reactivity

- WB, IHC, IF, IP, ELISA

### Applications

### MW

- 43kD (Calculated)
- 43kD (Observed)

### Isotype

- IgG, Kappa

## **Recommended Dilution Ratios**

### IP

## **Basic Information**

Clonality	Monoclonal
Clone Number	ANT0022R

## **Immunogen Information**

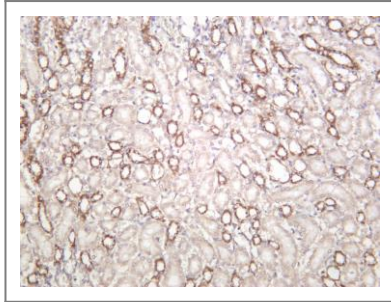
**Sequence** Pyruvate dehydrogenase E1 component subunit alpha somatic form mitochondrial

**Specificity** Endogenous

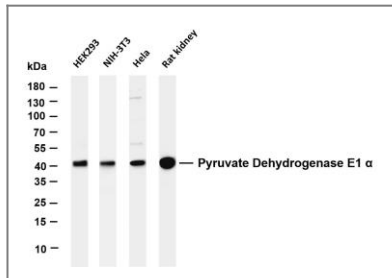
# Target Information

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 ODP		
	Organism	Gene ID	UniProt ID
	Human	<a href="#">5160</a> ;	<a href="#">P08559</a> ;
	Mouse	<a href="#">18597</a> ;	<a href="#">P35486</a> ;
	Rat		<a href="#">P26284</a> ;
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

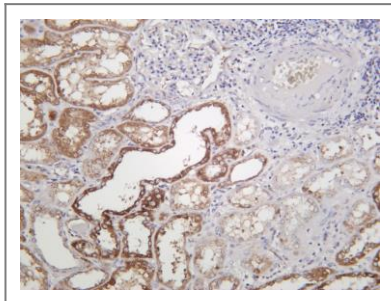


Rat kidney was stained with anti-Pyruvate Dehydrogenase E1  $\alpha$  (ANT0022R) rabbit antibody

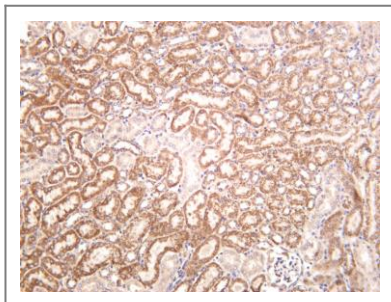


Various whole cell lysates were separated by 4-20% SDS-PAGE, and the membrane was blotted with anti-Pyruvate Dehydrogenase E1  $\alpha$  (ANT0022R) antibody. The HRP-conjugated Goat anti-Rabbit IgG(H + L) antibody was used to detect the antibody. Lane 1: HEK293 Lane 2: NIH-3T3 Lane 3: Hela Lane 4:

Rat kidney Predicted band size: 43kDa Observed band size: 43kDa



Human kidney was stained with anti-Pyruvate Dehydrogenase E1  $\alpha$  (ANT0022R) rabbit antibody



Mouse kidney was stained with anti-Pyruvate Dehydrogenase E1  $\alpha$  (ANT0022R) rabbit antibody