PDHA1 Rabbit pAb

CatalogNo: YT3641

Key Features

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

MW	Isotype
• 43kD (Observed)	• IgG

Storage

Storage*	-15°C to -25°C/1 year (Do not lower than -25°C)
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.

Recommended Dilution Ratios

WB 1:500-1:2000
IHC 1:100-1:300
ELISA 1:40000
IF 1:50-200

Basic Information

Clonality	Polyclonal

Immunogen Information

Immunogen	The antiserum was produced against synthesized peptide derived from human PDHA1. AA range:314-363
Specificity	PDHA1 Polyclonal Antibody detects endogenous levels of PDHA1 protein.

Target Information

Gene name PDHA1 ODPA

Protein Name Pyruvate dehydrogenase E1 component subunit alpha somatic form mitochondrial

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

Contact information

Orders: order@immunoway.com
Support: tech@immunoway.com
Telephone: 877-594-3616 (Toll Free), 408-747-0185
Website: http://www.immunoway.com
Address: 2200 Ringwood Ave San Jose, CA 95131 USA

Please scan the QR code to access additional product information:
PDHA1 Rabbit pAb