Product Name: PDHA1/2 (Phospho-Ser293/291) Rabbit Polyclonal Antibody
Catalog #: APRab05838

Summary

Production Name	PDHA1/2 (Phospho-Ser293/291) Rabbit Polyclonal Antibody
Description	Rabbit Polyclonal Antibody
Host	Rabbit
Application	WB
Reactivity	Human,Mouse,Rat

Performance

Conjugation	Unconjugated
Modification	Phospho Antibody
Isotype	IgG
Clonality	Polyclonal
Form	Liquid
Storage	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
Buffer	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Purification	Affinity purification

Immunogen

Gene Name	PDHA1 PHE1A
Alternative Names	Pyruvate dehydrogenase E1 component subunit alpha, somatic form, mitochondrial (EC 1.2.4.1) (PDHE1-A type I)
Gene ID	5160.0
SwissProt ID	P08559.

Application

Dilution Ratio	WB 1:500-2000
Molecular Weight	43kD

Background

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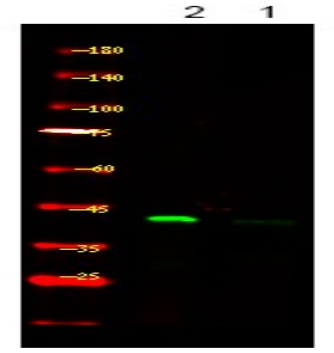
catalytic activity:Pyruvate + [dihydrolipoyllysine-residue acetyltransferase] lipoyllysine = [dihydrolipoyllysine-residue acetyltransferase] S-acetyldihydrolipoyllysine + CO(2).,cofactor:Thiamine pyrophosphate.,disease:Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 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.,

Research Area

Image Data



Western Blot analysis of 1 HepG2 cell 2, LPS 100ng/mL 30min treated ,using primary antibody at 1:1000 dilution. Secondary antibody was diluted at 1:10000

Note

For research use only.

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