abcam

Product datasheet

Recombinant Human Pyruvate Dehydrogenase E1-alpha subunit protein ab125602

1 Image

Overview

Product name	Recombinant Human Pyruvate Dehydrogenase E1-alpha subunit protein
Protein length	Full length protein

Description

Nature	Recombinant
Source	Escherichia coli
Amino Acid Sequence	
Accession	P08559
Species	Human
Molecular weight	47 kDa
Amino acids	30 to 390
Tags	His tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab125602** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

Applications	Functional Studies
	Western blot
	SDS-PAGE
Purity	> 85 % Densitometry.
Form	Liquid
Additional notes	ab125602 (Human Pyruvate Dehydrogenase E1-alpha subunit full length protein) can be utilized as a substrate for the following active protein Kinases:
	ab125560 (Active human PDK4 full length protein)
	ab125580 (Active human Mitochondrial Pyruvate dehydrogenase kinase 1 full length protein)

ab125592 (Active human PDK2 full length protein)
ab125606 (Active human PDK3 full length protein)

Preparation and Storage

Stability and Storage

Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles.

pH: 7.00
Preservative: 1.02% Imidazole
Constituents: 0.002% PMSF, 0.81% Sodium phosphate, 0.004% DTT, 25% Glycerol, 1.75% Sodium chloride

General Info

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).

Tissue specificity

Ubiquitous.

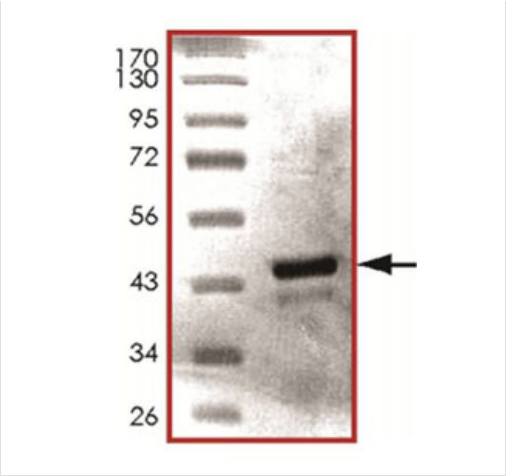
Involvement in disease

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 (X-LS).
Defects in PDHA1 are the cause of X-linked Leigh syndrome (X-LS) [MIM:308930]. X-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.

Cellular localization

Mitochondrion matrix.

Images



SDS-PAGE - Recombinant Human Pyruvate Dehydrogenase E1-alpha subunit protein (ab125602)

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- We investigate all quality concerns to ensure our products perform to the highest standards

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