abcam

Product datasheet

Recombinant Human PDHA1 protein ab125602

1 图像

描述

产品名称	重组人PDHA1蛋白
纯度	> 85 % Densitometry.
表达系统	Escherichia coli
Accession	P08559
蛋白长度	Full length protein
无动物成分	No
性质	Recombinant
种属	Human
预测分子量	47 kDa
氨基酸	30 to 390
标签	His tag N-Terminus

技术指标

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.

应用	Functional Studies Western blot SDS-PAGE
形式	Liquid
补充说明	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)

制备和贮存

稳定性和存储

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 (glycerin, glycerine), 1.75% Sodium chloride

常规信息

功能

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).

组织特异性

Ubiquitous.

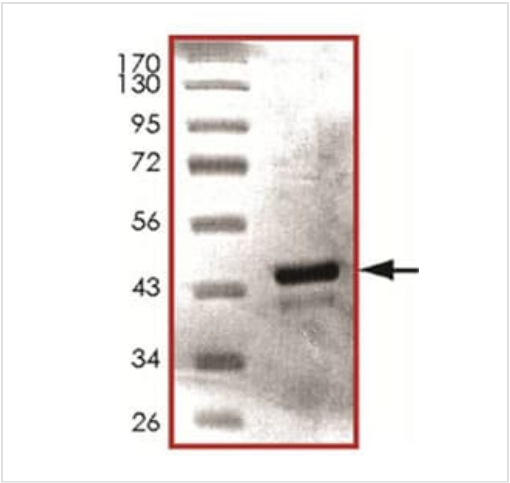
疾病相关

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.

细胞定位

Mitochondrion matrix.

图片



SDS-PAGE - Recombinant Human PDHA1 protein (ab125602)

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- Extensive multi-media technical resources to help you
- We investigate all quality concerns to ensure our products perform to the highest standards

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