

Recombinant Human PDHA1 protein ab125602

画像数 1

製品の詳細

製品名	Recombinant Human PDHA1 protein
精製度	> 85 % Densitometry.
発現系	Escherichia coli
アクセッション番号	<u>P08559</u>
タンパク質長	Full length protein
Animal free	No
由来	Recombinant
生物種	Human
予測される分子量	47 kDa
領域	30 to 390
タグ	His tag N-Terminus

特性

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.

アプリケーション	Functional Studies Western blot SDS-PAGE
製品の状態	Liquid
備考	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)

前処理および保存

保存方法および安定性

Shipped on dry ice. Upon delivery aliquot and store at -80°C. 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

関連情報

機能

The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO₂. It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).

組織特異性

Ubiquitous.

関連疾患

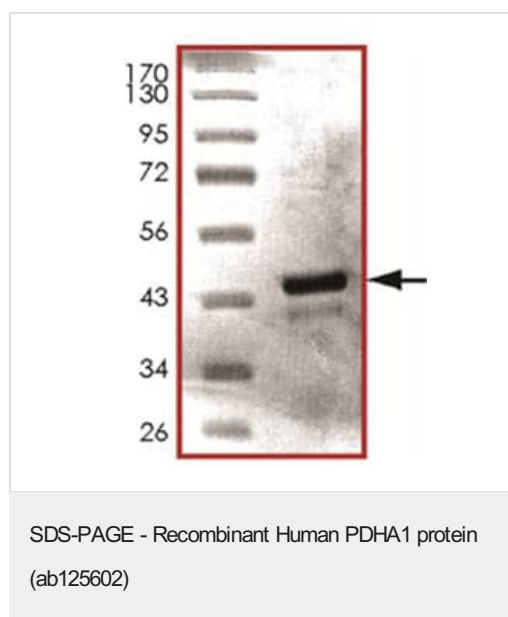
Defects in PDHA1 are a cause of pyruvate decarboxylase E1 component deficiency (PDHE1 deficiency) [MIM:312170]. PDHE1 deficiency is the most common enzyme defect in patients with primary lactic acidosis. It is associated with variable clinical phenotypes ranging from neonatal death to prolonged survival 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.

細胞内局在

Mitochondrion matrix.

画像



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