abcam

Product datasheet

Recombinant Human PDHB protein ab152601

1 Image

Description

Product name	Recombinant Human PDHB protein
Expression system	Wheat germ
Protein length	Protein fragment
Animal free	No
Nature	Recombinant
Species	Human
Sequence	LEAAAVLSKEGVECEVINMRTIRPMDMETIEASVMKTNHL VTVEGGWPQF GVGAEICARIMEGPAFNFLDAPAVRVTGADVPMPYAKILE DNSIPQVKDI IFAIKKTLNI
Amino acids	250 to 359
Tags	GST tag N-Terminus

Specifications

Our Abpromise guarantee covers the use of **ab152601** in the following tested applications.

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

Applications	Western blot ELISA
Form	Liquid
Additional notes	This product was previously labelled as Pyruvate Dehydrogenase E1 beta subunit.

Preparation and Storage

Stability and Storage	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

General Info

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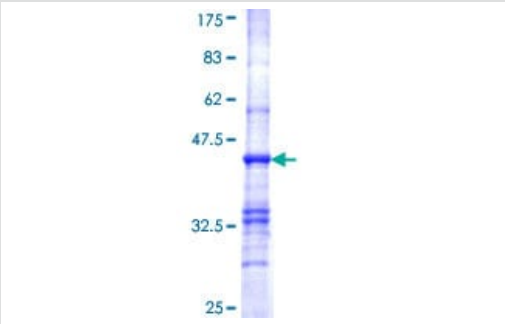
Function	The pyruvate dehydrogenase complex catalyzes the overall conversion of pyruvate to acetyl-CoA and CO(2). It contains multiple copies of three enzymatic components: pyruvate dehydrogenase (E1), dihydrolipoamide acetyltransferase (E2) and lipoamide dehydrogenase (E3).
Involvement in disease	Defects in PDHB are a cause of pyruvate dehydrogenase 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).
Cellular localization	Mitochondrion matrix.

Images



SDS-PAGE - Recombinant Human PDHB protein (ab152601)

ab152601 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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