

Recombinant Human PDHX Protein, His-tagged

Cat. No. PDHX-756H **Lot. No.** (See product label)

SPECIFICATION

Product Overview	Recombinant Human PDHX(Val220~Thr468) fused with His tag at N-terminal was expressed in E. coli.
Species	Human
Source	E.coli
ProteinLength	Val220~Thr468
Description	The pyruvate dehydrogenase (PDH) complex is located in the mitochondrial matrix and catalyzes the conversion of pyruvate to acetyl coenzyme A. The PDH complex thereby links glycolysis to Krebs cycle. The PDH complex contains three catalytic subunits, E1, E2, and E3, two regulatory subunits, E1 kinase and E1 phosphatase, and a non-catalytic subunit, E3 binding protein (E3BP). This gene encodes the E3 binding protein subunit; also known as component X of the pyruvate dehydrogenase complex. This protein tethers E3 dimers to the E2 core of the PDH complex. Defects in this gene are a cause of pyruvate dehydrogenase deficiency which results in neurological dysfunction and lactic acidosis in infancy and early childhood. This protein is also a minor antigen for antimitochondrial antibodies. These autoantibodies are present in nearly 95% of patients with the autoimmune liver disease primary biliary cirrhosis (PBC). In PBC, activated T lymphocytes attack and destroy epithelial cells in the bile duct where this protein is abnormally distributed and overexpressed. PBC eventually leads to cirrhosis and liver failure. Alternative splicing results in multiple transcript variants encoding distinct isoforms

 Tel: 1-631-559-9269 1-516-512-3133

 Email: info@creative-biomart.com  Fax: 1-631-938-8127

 45-1 Ramsey Road, Shirley, NY 11967, USA

Form	PBS, pH7.4, containing 0.01% SKL, 1mM DTT, 5% Trehalose and Proclin300.
Molecular Mass	30.2kDa
Endotoxin	<1.0EU per 1g (determined by the LAL method)
Purity	> 99%
Applications	Positive Control; Immunogen; SDS-PAGE; WB. If bio-activity of the protein is needed, please check active protein.
Stability	The thermal stability is described by the loss rate. The loss rate was determined by accelerated thermal degradation test, that is, incubate the protein at 37 centigrade for 48h, and no obvious degradation and precipitation were observed. The loss rate is less than 5% within the expiration date under appropriate storage condition.
Storage	Avoid repeated freeze/thaw cycles. Store at 2-8 centigrade for one month. Aliquot and store at -80 centigrade for 12 months.
Reconstitution	Reconstitute in PBS or others.

GENE INFORMATION

Gene Name	PDHX pyruvate dehydrogenase complex, component X [Homo sapiens]
Official Symbol	PDHX
Synonyms	PDHX; pyruvate dehydrogenase complex, component X; pyruvate dehydrogenase protein X component, mitochondrial; DLDBP; E3BP; OPDX; PDX1; proX; pyruvate dehydrogenase complex, E3-binding protein subunit; lipoyl-containing pyruvate dehydrogenase complex component X; pyruvate dehydrogenase complex, lipoyl-

 Tel: 1-631-559-9269 1-516-512-3133

 Email: info@creative-biomart.com  Fax: 1-631-938-8127

 45-1 Ramsey Road, Shirley, NY 11967, USA

containing component X; dihydrolipoamide dehydrogenase-binding protein of pyruvate dehydrogenase complex;

Gene ID	8050
mRNA Refseq	NM_001135024
Protein Refseq	NP_001128496
MIM	608769
UniProt ID	O00330

