

## 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

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<b>Form</b>	PBS, pH7.4, containing 0.01% SKL, 1mM DTT, 5% Trehalose and Proclin300.
<b>Molecular Mass</b>	30.2kDa
<b>Endotoxin</b>	<1.0EU per 1g (determined by the LAL method)
<b>Purity</b>	> 99%
<b>Applications</b>	Positive Control; Immunogen; SDS-PAGE; WB. If bio-activity of the protein is needed, please check active protein.
<b>Stability</b>	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.
<b>Storage</b>	Avoid repeated freeze/thaw cycles. Store at 2-8 centigrade for one month. Aliquot and store at -80 centigrade for 12 months.
<b>Reconstitution</b>	Reconstitute in PBS or others.

## GENE INFORMATION

<b>Gene Name</b>	PDHX pyruvate dehydrogenase complex, component X [ Homo sapiens ]
<b>Official Symbol</b>	PDHX
<b>Synonyms</b>	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-

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

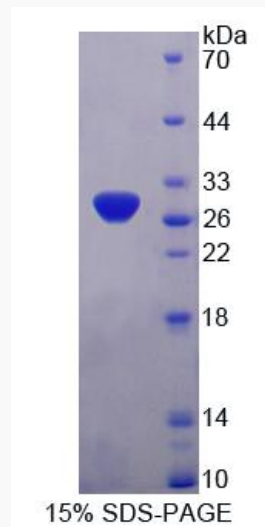
**Gene ID** 8050

**mRNA Refseq** NM\_001135024

**Protein Refseq** NP\_001128496

**MIM** 608769

**UniProt ID** O00330



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