

## Product datasheet for **TA361472**

### PDHX Rabbit Polyclonal Antibody

#### Product data:

Product Type:	Primary Antibodies
Applications:	WB
Reactivity:	Human
Host:	Rabbit
Clonality:	Polyclonal
Immunogen:	The immunogen is a synthetic peptide directed towards the middle region of human PDHX
Specificity:	<b>Expected reactivity:</b> Human
Formulation:	Liquid. Purified antibody supplied in 1x PBS buffer with 0.09% (w/v) sodium azide and 2% sucrose. <i>Note that this product is shipped as lyophilized powder to China customers.</i>
Concentration:	lot specific
Purification:	Affinity purified
Conjugation:	Unconjugated
Storage:	For short term use, store at 2-8°C up to 1 week. For long term storage, store at -20°C in small aliquots to prevent freeze-thaw cycles.
Stability:	Shelf life: one year from despatch.
Predicted Protein Size:	55 kDa
Gene Name:	pyruvate dehydrogenase complex component X
Database Link:	<a href="#">NP_001128496.1</a> <a href="#">Entrez Gene 8050 Human</a> <a href="#">O00330</a>



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**Background:**

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.

**Synonyms:**

DLDBP; E3BP; OPDX; PDX1; proX

**Product images:**