

### Product datasheet

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# ARG57546 anti-PDHX / Pyruvate dehydrogenase antibody [1E11]

Package: 50 μl Store at: -20°C

#### Summary

Product Description Mouse Monoclonal antibody [1E11] recognizes PDHX / Pyruvate dehydrogenase

Tested Reactivity Hu
Tested Application WB

Host Mouse

**Clonality** Monoclonal

Clone 1E11

Isotype IgG1, kappa

Target Name PDHX / Pyruvate dehydrogenase

Species Human

Immunogen Recombinant Human PDHX (aa. 54-501) purified from E. coli.

Conjugation Un-conjugated

Alternate Names Lipoyl-containing pyruvate dehydrogenase complex component X; OPDX; proX; E3-binding protein;

Dihydrolipoamide dehydrogenase-binding protein of pyruvate dehydrogenase complex; E3BP; PDX1;

Pyruvate dehydrogenase protein X component, mitochondrial; DLDBP

#### **Application Instructions**

Application table	Application	Dilution
	WB	1:1000
Application Note	* The dilutions indicate recommended starting dilutions and the optimal dilutions or concentrations should be determined by the scientist.	

#### **Properties**

Form Liquid

Purification Purification with Protein A.

Buffer PBS (pH 7.4), 0.02% Sodium azide and 10% Glycerol.

Preservative 0.02% Sodium azide

Stabilizer 10% Glycerol

Concentration 1 mg/ml

Storage instruction For continuous use, store undiluted antibody at 2-8°C for up to a week. For long-term storage, aliquot

and store at -20°C. Storage in frost free freezers is not recommended. Avoid repeated freeze/thaw cycles. Suggest spin the vial prior to opening. The antibody solution should be gently mixed before use.

Note For laboratory research only, not for drug, diagnostic or other use.

#### Bioinformation

Gene Symbol

PDHX

Gene Full Name

pyruvate dehydrogenase complex, component X

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.[provided by RefSeq, Oct 2009]

Function

Required for anchoring dihydrolipoamide dehydrogenase (E3) to the dihydrolipoamide transacetylase (E2) core of the pyruvate dehydrogenase complexes of eukaryotes. This specific binding is essential for a functional PDH complex. [UniProt]

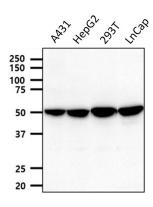
Calculated Mw

54 kDa

PTM

Delipoylated at Lys-97 by SIRT4, delipoylation decreases the PHD complex activity. [UniProt]

#### **Images**



## ARG57546 anti-PDHX / Pyruvate dehydrogenase antibody [1E11] WB image

Western blot: 40  $\mu$ g of A431, HepG2, 293T and LnCap cell lysates stained with ARG57546 anti-PDHX / Pyruvate dehydrogenase antibody [1E11] at 1:1000 dilution.