

## ODPX rabbit pAb

<b>Catalog No :</b>	YT6637
<b>Reactivity :</b>	Human;Mouse
<b>Applications :</b>	WB
<b>Target :</b>	ODPX
<b>Fields :</b>	>>Metabolic pathways
<b>Gene Name :</b>	PDHX PDX1
<b>Protein Name :</b>	ODPX
<b>Human Gene Id :</b>	8050
<b>Human Swiss Prot No :</b>	O00330
<b>Mouse Gene Id :</b>	27402
<b>Mouse Swiss Prot No :</b>	Q8BKZ9
<b>Immunogen :</b>	Synthesized peptide derived from human ODPX AA range: 24-74
<b>Specificity :</b>	This antibody detects endogenous levels of ODPX at Human/Mouse
<b>Formulation :</b>	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
<b>Source :</b>	Polyclonal, Rabbit,IgG
<b>Dilution :</b>	WB 1[?]500-2000
<b>Purification :</b>	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
<b>Concentration :</b>	1 mg/ml

**Storage Stability :** -15°C to -25°C/1 year(Do not lower than -25°C)

**Molecularweight :** 55kD

**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],

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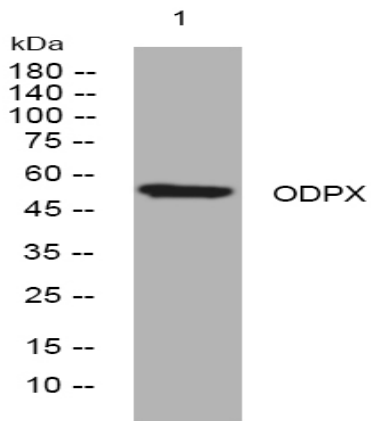
**Function :** disease:Defects in PDHX are a cause of lacticacidemia [MIM:245349].,function:Required for anchoring dihydrolipoamide dehydrogenase (E3) to the dihydrolipoamide transacetylase (E2) core of the pyruvate dehydrogenase complexes of eukaryotes. This specific binding is essential for a functional PDH complex.,similarity:Belongs to the 2-oxoacid dehydrogenase family.,similarity:Contains 1 lipoyl-binding domain.,subunit:Eukaryotic pyruvate dehydrogenase complexes are organized about a core consisting of the oligomeric dihydrolipoamide acetyl-transferase, around which are arranged multiple copies of pyruvate dehydrogenase, dihydrolipoamide dehydrogenase and protein X bound by non-covalent bonds.,

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**Subcellular Location :** Mitochondrion matrix.

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## Products Images



Western blot analysis of lysates from 3T3 cells, primary antibody was diluted at 1:1000, 4° over night