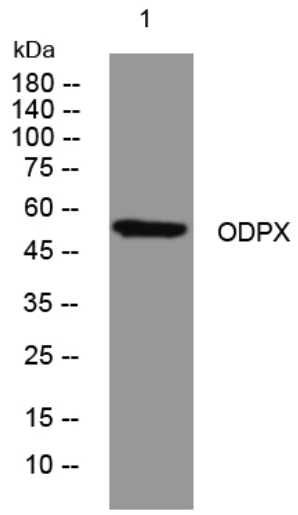
ODPX rabbit pAb

Catalog No	BYab-11272
Isotype	IgG
Reactivity	Human; Mouse
Applications	WB
Gene Name	PDHX PDX1
Protein Name	ODPX
Immunogen	Synthesized peptide derived from human ODPX AA range: 24-74
Specificity	This antibody detects endogenous levels of ODPX at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	WB 1：500-2000
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	
Observed Band	
Cell Pathway	Mitochondrion matrix.
Tissue Specificity	
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.,

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],
matters needing attention	Avoid repeated freezing and thawing!
Usage suggestions	This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



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