

ODPX rabbit pAb

Catalog No :	YT6637
Reactivity :	Human;Mouse
Applications :	WB
Target :	ODPX
Fields :	>>Metabolic pathways
Gene Name :	PDHX PDX1
Protein Name :	ODPX
Human Gene Id :	8050
Human Swiss Prot No :	O00330
Mouse Gene Id :	27402
Mouse Swiss Prot No :	Q8BKZ9
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
Dilution :	WB 1[?]500-2000
Purification :	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
Concentration :	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],

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.,

Subcellular Location : Mitochondrion matrix.

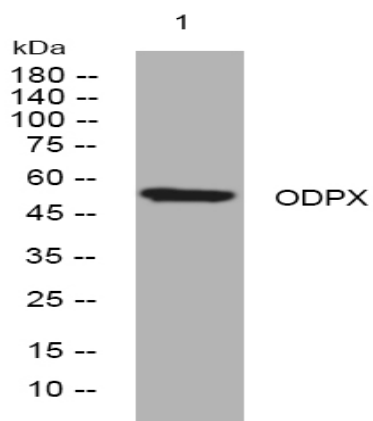
Sort : 11064

No4 : 1

Host : Rabbit

Modifications : Unmodified

Products Images



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