

ODPX rabbit pAb

Catalog_no :	AT6637
Applications :	WB
Reactivity :	Human, Mouse
Category :	抗原抗体
Size :	100µg/50µg/20µg
Gene_name :	PDHX PDX1
Protein_name :	ODPX
Humangene_id :	8050
Humanswissprot_no :	O00330
Mousegene_id :	27402
Mouseswissprot_no :	Q8BKZ9
Immunogen :	Synthesized peptide derived from human ODPX
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 :	Rabbit
Dilution :	WB 1 : 500-2000
Purification :	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Concentration :	1 mg/ml
Storage_stability :	-20°C/1 year
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],
