

ODPX Rabbit Polyclonal Antibody

Description

Product type	Primary Antibody
Code	BT-AP11651
Host	Rabbit
Isotype	IgG
Size	100ul, 50ul, 20ul
Immunogen	Synthesized peptide derived from human ODPX
Mol wt	55110
Species reactivity	Human, Mouse
Clonality	Polyclonal
Recommended application	WB
Concentration	1 mg/ml
Full name	ODPX
Synonyms	ODPX

This product is for research use only, not for use in human, therapeutic or diagnostic procedure.

Background

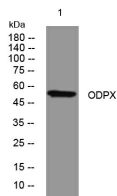
The pyruvate dehydrogenase (PDH) complex is located in the mitochondrial matrix and catalyzes the conversion of pyruvate to acetyl coenzyme A. The PDH complex thereby links glycolysis to Krebs cycle. The PDH complex contains three catalytic subunits| E1| E2| and E3| two regulatory subunits| E1 kinase 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.

Recommended Dilution

WB: 1: 500 - 1: 2000

Not yet tested in other applications.

Images



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

Storage

-20°C for 1 year