

PDHX Rabbit Polyclonal Antibody



CAB6426

Product Information

Size:

20uL, 50uL, 100uL, 200uL

Observed MW:

54kDa

Calculated MW:

29kDa/51kDa/54kDa

Applications:

WB IF IP

Reactivity:

Human, Mouse, Rat

Antibody Information

Recommended dilutions:

WB 1:500 - 1:2000 IF 1:10 -
1:100 IP 1:50 - 1:200

Source:

Rabbit

Isotype:

IgG

Purification:

Affinity purification

Protein 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.

Immunogen information

Gene ID:

8050

Uniprot

O00330

Synonyms:

PDHX; DLDBP; E3BP; OPDX; PDX1; proX

Immunogen:

Recombinant fusion protein containing a sequence corresponding to amino acids 1-300 of human PDHX (NP_003468.2).

Storage:

Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Product Images

