

PDHX Rabbit pAb

Catalog No.: A6426

Basic Information

Observed MW

54kDa

Calculated MW

54kDa

Category

Primary antibody

Applications

WB,IP,ELISA

Cross-Reactivity

Human, Mouse, Rat

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 Dilutions

WB 1:500 - 1:2000**IP** 0.5µg-4µg antibody for
400µg-600µg extracts of
whole cells**ELISA** Recommended starting
concentration is 1 µg/mL.
Please optimize the
concentration based on
your specific assay
requirements.

Immunogen Information

Gene ID

8050

Swiss Prot

O00330

Immunogen

Recombinant protein (or fragment). This information is considered to be commercially sensitive.

Synonyms

E3BP; OPDX; PDX1; proX; DLDBP; PDHDX; PDHX

Contact

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Product Information

Source

Rabbit

Isotype

IgG

Purification

Affinity purification

Storage

Store at -20°C. Avoid freeze / thaw cycles.

Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Validation Data

