

PDHX polyclonal antibody

Catalog: BS8207

Host: Rabbit

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 anti-mitochondrial 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.

Product:

1mg/ml in PBS with 0.02% sodium azide, 50% glycerol, pH7.2

Molecular Weight:

54kDa

Swiss-Prot:

O00330

Purification&Purity:

The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen and the purity is > 95% (by SDS-PAGE).

Applications:

WB, 1:500 - 1:2000 | IP, 1:50 - 1:200

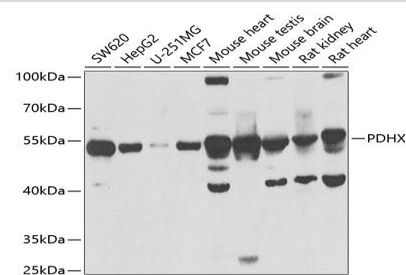
Storage&Stability:

Store at 4 °C short term. Aliquot and store at -20 °C long term. Avoid freeze-thaw cycles.

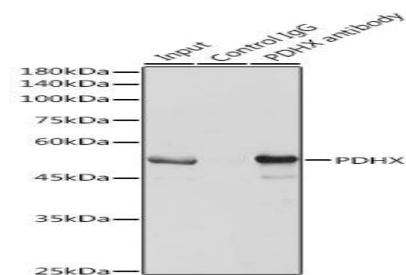
Category:

Polyclonal Antibodies

DATA:



Western blot analysis of extracts of various cell lines, using PDHX antibody at 1:1000 dilution. Secondary antibody: HRP Goat Anti-Rabbit IgG at 1:10000 dilution. Lysates/proteins: 25ug per lane. Blocking buffer: 3% nonfat dry milk in TBST. Detection: ECL Basic Kit. Exposure time: 5s.



Immunoprecipitation analysis of 600ug extracts of Mouse brain cells using 3ug PDHX antibody. Western blot was performed from the immunoprecipitate using PDHX antibody at a dilution of 1:1000.

Note:

For research use only, not for use in diagnostic procedure.

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