

PDHX Rabbit pAb

货号: B15233

产品信息

反应	Human,Mouse,Rat
宿主	Rabbit
克隆性	Polyclonal
预测反应	
应用	WB IP
推荐浓度	WB: 1:500 - 1:2000 IP: 1:500 - 1:1000
理论分子量	29kDa/51kDa/54kDa
实测分子量	54kDa
形式	Liquid
保存条件	Store at -20°C. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide,50% glycerol,pH7.3.
偶联物	Unconjugated
阳性对照	SW620,HepG2,U-251MG,MCF7,Mouse heart,Mouse testis,Mouse brain,Rat kidney,Rat heart
细胞定位	Mitochondrion matrix
纯化	Affinity purification

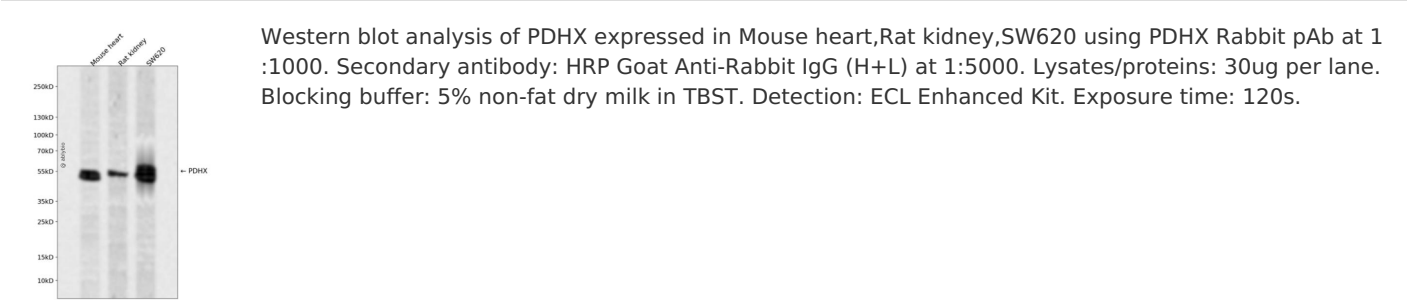
抗原信息

抗原信息	Recombinant fusion protein containing a sequence corresponding to amino acids 1-300 of human PDHX (NP_003468.2).
序列	MAASWRLGCDPRLLRYLVGFPGRRSVGLVKGALGWSVSRGANWRWFHSTQWLRGDPIKILMPSLSPTMEEGNIVKWLK KEGEAVSAGDALCEIETDKAVVTLDASDDGILAKIVVEEGSKNIRLGSIGLIVEEGEDWKHVEIPKDVGPPPPVSKPSEPRP SPEPQISIPVKEHIPGTLRFRLSPAARNILEKHSLDASQGTATGPRGIFTKEDALKLVQLKQTGKITESRPTPAPTATPTAPSP LQATAGPSYPRPVIPPVSTPGQPNAVGTFTFTEIPASNIRRVIAKRLTESKSTVPH

靶点信息

研究背景	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.
基因ID	8050
基因名	PDHX
Swiss	O00330
别名	PDHX;DLDBP;E3BP;OPDX;PDX1;proX

产品验证



实验步骤

访问官网浏览详情: www.ablybio.cn