

PDHX Conjugated Antibody

Catalog No: #C38904

Package Size: #C38904-AF350 100ul #C38904-AF405 100ul #C38904-AF488 100ul
 #C38904-AF555 100ul #C38904-AF594 100ul #C38904-AF647 100ul
 #C38904-AF680 100ul #C38904-AF750 100ul #C38904-Biotin 100ul

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Description

Product Name	PDHX Conjugated Antibody
Host Species	Rabbit
Clonality	Polyclonal
Species Reactivity	Hu
Specificity	The antibody detects endogenous level of total PDHX antibody.
Immunogen Description	Recombinant protein of human PDHX.
Conjugates	Biotin AF350 AF405 AF488 AF555 AF594 AF647 AF680 AF750
Other Names	E3BP; OPDX; PDX1; proX; DLDBP;
Accession No.	Swiss-Prot#:O00330NCBI Gene ID:8050
Uniprot	O00330
GeneID	8050;
Excitation Emission	AF350: 346nm/442nm AF405: 401nm/421nm AF488: 493nm/519nm AF555: 555nm/565nm AF594: 591nm/614nm AF647: 651nm/667nm AF680: 679nm/702nm AF750: 749nm/775nm
Calculated MW	54
Formulation	0.01M Sodium Phosphate, 0.25M NaCl, pH 7.6, 5mg/ml Bovine Serum Albumin, 0.02% Sodium Azide
Storage	Store at 4°C in dark for 6 months

Application Details

Suggested Dilution:

AF350 conjugated: most applications: 1: 50 - 1: 250

AF405 conjugated: most applications: 1: 50 - 1: 250

AF488 conjugated: most applications: 1: 50 - 1: 250

AF555 conjugated: most applications: 1: 50 - 1: 250

AF594 conjugated: most applications: 1: 50 - 1: 250

AF647 conjugated: most applications: 1: 50 - 1: 250

AF680 conjugated: most applications: 1: 50 - 1: 250

AF750 conjugated: most applications: 1: 50 - 1: 250

Biotin conjugated: working with enzyme-conjugated streptavidin, most applications: 1: 50 - 1: 1,000

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.

Note: This product is for in vitro research use only