

Product datasheet for PH303138

PDHX (NM_003477) Human Mass Spec Standard

Product data:

Product Type:	Mass Spec Standards
Description:	PDHX MS Standard C13 and N15-labeled recombinant protein (NP_003468)
Species:	Human
Expression Host:	HEK293
Expression cDNA Clone or AA Sequence:	RC203138
Predicted MW:	54.1 kDa
Protein Sequence:	>RC203138 protein sequence Red=Cloning site Green=Tags(s)

MAASWRLGCDPRLRLRYLVGFPGRRSVGLVKGALGWSVSRGANWRWFHSTQWLRGDPKIKILMPSLSPTMEE
GNIYKWLKKEGEAVSAGDALCEIETDKAVVTLDA SDDGILAKIVVEEGSKNIRLGSILGLIVEEGEDWKH
VEIPKDVGPPPPVSKPSEPRPSPEPQISIPVKKEHIPGTLRFRLSPAARNILEKHSLDASQGTATGPRGI
FTKEDALKLVQLKQTGKITESRPTAPTATPTAPSPLQATAGPSYPRPVIPPVSTPGQPNVAVGTFTEIPA
SNIRRVIAKRLTESKSTVPHAYATADCDLGA VLKVRQDLVKDDIKVSVNDFI I KAAAVTLKQMPDVNVSW
DGE GPKQLPFIDISVAVATVKGLLTP I I K D A A A K G I Q E I A D S V K A L S K K A R D G K L L P E E Y Q G G S F S I S N L
GMFGIDEFTAVINPPQACILAVGRFRPV L K L T E D E E G N A K L Q Q R Q L I T V T M S S D S R V V D D E L A T R F L K S F
KANLENPIRLA

TRTRPLEQKLI SEEDLAANDILDYKDDDDKV

Tag:	C-Myc/DDK
Purity:	> 80% as determined by SDS-PAGE and Coomassie blue staining
Concentration:	>0.05 µg/µL as determined by microplate BCA method
Labeling Method:	Labeled with [U- 13C6, 15N4]-L-Arginine and [U- 13C6, 15N2]-L-Lysine
Buffer:	25 mM Tris-HCl, 100 mM glycine, pH 7.3
Storage:	Store at -80°C. Avoid repeated freeze-thaw cycles.
Stability:	Stable for 3 months from receipt of products under proper storage and handling conditions.
RefSeq:	<u>NP_003468</u>
RefSeq Size:	2991
RefSeq ORF:	1503

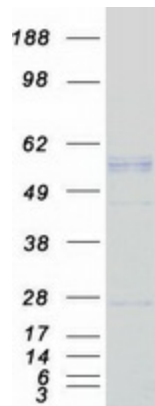


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Synonyms: DLDBP; E3BP; OPDX; PDHXD; PDX1; proX
Locus ID: 8050
UniProt ID: [O00330](#)
Cytogenetics: 11p13

Summary: 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.[provided by RefSeq, Oct 2009]

Product images:



Coomassie blue staining of purified PDHX protein (Cat# [TP303138]). The protein was produced from HEK293T cells transfected with PDHX cDNA clone (Cat# [RC203138]) using MegaTran 2.0 (Cat# [TT210002]).