

Product datasheet for **RC203138L2V**

PDHX (NM_003477) Human Tagged ORF Clone Lentiviral Particle

Product data:

Product Type:	Lentiviral Particles
Product Name:	PDHX (NM_003477) Human Tagged ORF Clone Lentiviral Particle
Symbol:	PDHX
Synonyms:	DLDBP; E3BP; OPDX; PDHXD; PDX1; proX
Mammalian Cell Selection:	None
Vector:	pLenti-C-mGFP (PS100071)
Tag:	mGFP
ACCN:	NM_003477
ORF Size:	1503 bp
ORF Nucleotide Sequence:	The ORF insert of this clone is exactly the same as(RC203138).
OTI Disclaimer:	The molecular sequence of this clone aligns with the gene accession number as a point of reference only. However, individual transcript sequences of the same gene can differ through naturally occurring variations (e.g. polymorphisms), each with its own valid existence. This clone is substantially in agreement with the reference, but a complete review of all prevailing variants is recommended prior to use. More info
OTI Annotation:	This clone was engineered to express the complete ORF with an expression tag. Expression varies depending on the nature of the gene.
RefSeq:	NM_003477.1
RefSeq Size:	2991 bp
RefSeq ORF:	1506 bp
Locus ID:	8050
UniProt ID:	O00330
Cytogenetics:	11p13
Domains:	biotin_lipoyl, 2-oxoacid_dh, e3_binding
MW:	54.1 kDa



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Gene 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]