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PRODUCT INFORMATION

PDHE1-A (human, recombinant)

Item No. 42229

Overview and Properties

Synonyms:	Pyruvate Dehydrogenase E1, Pyruvate Dehydrogenase E1 Component Subunit α
Source:	Recombinant human N-terminal His-tagged PDHE1-A expressed in *E. coli*
Amino Acids:	30-390
Uniprot No.:	P08559
Molecular Weight:	47 kDa
Storage:	-80°C (as supplied)
Stability:	≥1 year
Purity:	≥70% estimated by SDS-PAGE
Supplied in:	50 mM sodium phosphate, pH 7.0, with 300 mM sodium chloride, 150 mM imidazole, 0.1 mM PMSF, 0.25 mM DTT, and 25% glycerol

Information represents the product specifications. Batch specific analytical results are provided on each certificate of analysis.

Description

Pyruvate dehydrogenase E1 component subunit α (PDHE1-A), also known as pyruvate dehydrogenase E1, is a dehydrogenase and member of the 2-oxo acid dehydrogenase family.[1,2] It is a tetramer composed of a cleavable mitochondrial signaling sequence and two α- and β domains with active sites between each of the α/β domain interfaces.[2] It is a member of the mitochondrial pyruvate dehydrogenase complex and catalyzes the oxidative decarboxylation of pyruvate, which is the first reaction in the enzymatic conversion of pyruvate to acetyl-CoA. Phosphorylation of serine 203, -264, or -271 inhibits PDHE1-A enzymatic activity by blocking active site associations with its cofactor thiamine pyrophosphate (Item No. 20254) and serves as a reversible regulatory mechanism for PDHE1-A and pyruvate dehydrogenase complex activity.[3] Dichloroacetate-induced phosphorylation of PDHE1-A reduces the viability of HSC-2 and HSC-3 oral squamous cell carcinoma cells.[4] Mutations in *PDHA1*, the gene encoding PDHE1-A, are associated with lactic acidosis, developmental delay, cerebral atrophy, seizures, and elevated urine and cerebrospinal fluid (CSF) levels of pyruvate in patients with the inborn error of metabolism pyruvate dehydrogenase complex deficiency (PDCD).[5,6] Cayman's PDHE1-A (human, recombinant) protein can be used or enzyme activity assays. This protein has a calculated molecular weight of 47 kDA.

References

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