

Human PDHA1 / C54G Protein1 (aa 30-390)

Catalog Number: 14567-HNCB



Sino Biological Inc.
Biological Solution Specialist

General Information

Gene Name Synonym:

PDHA; PDHAD; PDHCE1A; PHE1A; Pdha-1

Protein Construction:

A DNA sequence encoding the human PDHA1 (P08559-1) (Phe30-Ser390) was fused with two additional amino acids (Gly & Pro) at the N-terminus.

Source: Human

Expression Host: Baculovirus-Insect Cells

QC Testing

Purity: > 85 % as determined by SDS-PAGE

Endotoxin:

< 1.0 EU per µg of the protein as determined by the LAL method

Stability:

Samples are stable for up to twelve months from date of receipt at -70 °C

Predicted N terminal: Gly

Molecular Mass:

The recombinant human PDHA1 consists of 363 amino acids and has a calculated molecular mass of 40.4 kDa. The recombinant protein migrates as an approximately 45 kDa band in SDS-PAGE under reducing conditions.

Formulation:

Lyophilized from sterile 20mM Tris, 500mM NaCl, 10% glycerol, pH 8.0.

Normally 5 % - 8 % trehalose, mannitol and 0.01% Tween80 are added as protectants before lyophilization. Specific concentrations are included in the hardcopy of COA. Please contact us for any concerns or special requirements.

Usage Guide

Storage:

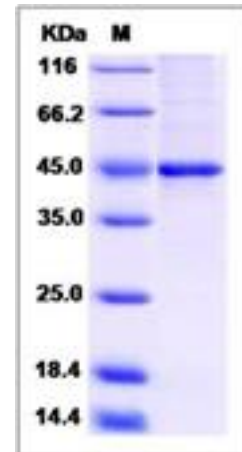
Store it under sterile conditions at -20°C to -80°C upon receiving. Recommend to aliquot the protein into smaller quantities for optimal storage.

Avoid repeated freeze-thaw cycles.

Reconstitution:

Detailed reconstitution instructions are sent along with the products.

SDS-PAGE:



Protein Description

PDHA1, also known as C54G1, is an alpha subunit of pyruvate dehydrogenase. Pyruvate dehydrogenase, together with dihydrolipoamide acetyltransferase and lipoamide dehydrogenase, composes the pyruvate dehydrogenase (PDH) complex. The PDH complex is a nuclear-encoded mitochondrial multienzyme complex that catalyzes the overall conversion of pyruvate to acetyl-CoA and CO₂, and provides the primary link between glycolysis and the tricarboxylic acid (TCA) cycle. PDHA1 plays a key role in the function of the PDH complex. Defects in PDHA1 can cause pyruvate dehydrogenase E1-alpha deficiency. Defects in PDHA1 also are the cause of X-linked Leigh syndrome (X-LS). X-LS is an early-onset progressive neurodegenerative disorder with a characteristic neuropathology consisting of focal, bilateral lesions in one or more areas of the central nervous system, including the brainstem, thalamus, basal ganglia, cerebellum, and spinal cord.

References

1. Quintana E. et al., 2010, Clin Genet. 77 (5): 474-82.
2. Ah Mew N. et al., 2011, Pediatr Neurol. 45 (1): 57-9.
3. Pinheiro A. et al., 2012, Gene. 506 (1): 173-8.

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