Recombinant Human PHYH Protein

Catalog Number: PKSH030645

Note: Centrifuge before opening to ensure complete recovery of vial contents.

Description	
Species	Human
Source	E.coli-derived Human PHYH protein Ser31-Leu338
Calculated MW	35.6 kDa
Observed MW	26-32 kDa
Accession	O14832
Bio-activity	Not validated for activity
Properties	
Purity	> 80 % as determined by reducing SDS-PAGE.
Endotoxin	Please contact us for more information.
Storage	Generally, lyophilized proteins are stable for up to 12 months when stored at -20 to -80
	°C. Reconstituted protein solution can be stored at 4-8°C for 2-7 days. Aliquots of
	reconstituted samples are stable at $< -20^{\circ}$ C for 3 months.
Shipping	This product is provided as lyophilized powder which is shipped with ice packs.
Formulation	Lyophilized from sterile 20mM mops, 10% glycerol, 2mM DDT, 1mM EDTA, 0.2mM
	PMSF, 0.2M NaCl, pH 7.2
	Normally 5% - 8% trehalose, mannitol and 0.01% Tween 80 are added as protectants
	before lyophilization.
	Please refer to the specific buffer information in the printed manual.
Reconstitution	Please refer to the printed manual for detailed information.
Data	
KDa MK R	



> 80 % as determined by reducing SDS-PAGE.

Background

PHYH belongs to the family of iron(II)-dependent oxygenases, which typically incorporate one atom of dioxygen into the substrate and one atom into the succinate carboxylate group. PHYH is expressed in liver, kidney, and T-cells, but not in spleen, brain, heart, lung and skeletal muscle. It converts phytanoyl-CoA to 2-hydroxyphytanoyl-CoA. Defects in PHYH can cause Refsum disease (RD). RD is an autosomal recessive disorder characterized clinically by a tetrad of abnormalities: retinitis pigmentosa, peripheral neuropathy, cerebellar ataxia, and elevated protein levels in the cerebrospinal fluid (CSF). Patients exhibit accumulation of the branched-chain fatty acid, phytanic acid, in blood and tissues.

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