

Recombinant Human PHYH Protein

Catalog Number:PKSH030645



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

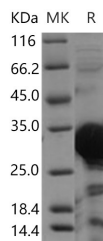
Description

Synonyms	LN1;LNAP1;PAHX;PHYH1;RD
Species	Human
Expression Host	E.coli
Sequence	Ser31-Leu338
Accession	O14832
Calculated Molecular Weight	35.6 kDa
Observed molecular weight	26-32 kDa
Tag	None

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°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 % Tween80 are added as protectants before lyophilization. Please refer to the specific buffer informa
Reconstitution	Please refer to the printed manual for detailed information.

Data



> 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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