

## Recombinant Human PHYH Protein

**Catalog Number: PKSH030645**

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

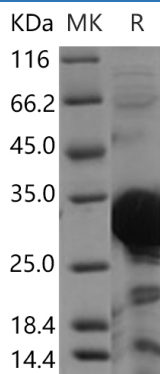
### Description

<b>Species</b>	Human
<b>Source</b>	E.coli-derived Human PHYH protein Ser31-Leu338
<b>Calculated MW</b>	35.6 kDa
<b>Observed MW</b>	26-32 kDa
<b>Accession</b>	O14832
<b>Bio-activity</b>	Not validated for activity

### Properties

<b>Purity</b>	> 80 % as determined by reducing SDS-PAGE.
<b>Endotoxin</b>	Please contact us for more information.
<b>Storage</b>	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.
<b>Shipping</b>	This product is provided as lyophilized powder which is shipped with ice packs.
<b>Formulation</b>	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.
<b>Reconstitution</b>	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.

### For Research Use Only

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