

[Go to Product page](#)

## Datasheet for ABIN7317253

**PHYH Protein**

## Overview

Quantity:	100 µg
Target:	PHYH
Origin:	Human
Source:	Escherichia coli (E. coli)
Protein Type:	Recombinant

## Product Details

Purpose:	Recombinant Human PHYH Protein
Sequence:	Ser31-Leu338
Characteristics:	A DNA sequence encoding the human PHYH (O14832) (Ser31-Leu338) was expressed, with a N-terminal Met.
Purity:	> 80 % as determined by reducing SDS-PAGE.

## Target Details

Target:	PHYH
Alternative Name:	PHYH ( <a href="#">PHYH Products</a> )
Background:	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

## Target Details

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.  
Synonym: LN1, LNAP1, PAHX, PHYH1, RD

Molecular Weight: 35.6 kDa

UniProt: [O14832](#)

Pathways: [Monocarboxylic Acid Catabolic Process](#)

## Application Details

Restrictions: For Research Use only

## Handling

Format: Lyophilized

Reconstitution: Please refer to the printed manual for detailed information.

Buffer: Lyophilized from sterile 20 mM mops, 10 % glycerol, 2 mM DDT, 1 mM EDTA, 0.2 mM PMSF, 0.2M NaCl, pH 7.2

Storage: 4 °C, -20 °C, -80 °C

Storage Comment: 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.