Recombinant Human PHYH Protein

Cat.NO.: TP06370

3th Edition

Synonyms:LN1;LNAP1;PAHX;PHYH1;RD

Description: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.

Form:PBS

Molecular Weight:35.6 kDa

Sequences: Ser31-Leu338

Purity:> 95% by HPLC

Concentration:

Endotoxin Level:<1.0 EU per 1 ug of protein (determined by LAL method)

Storage: Can be stored at +4°C short term (1-2 weeks). For long term storage, aliquot and store at -20°C or -70°C. Avoid repeated freezing and thawing cycles.