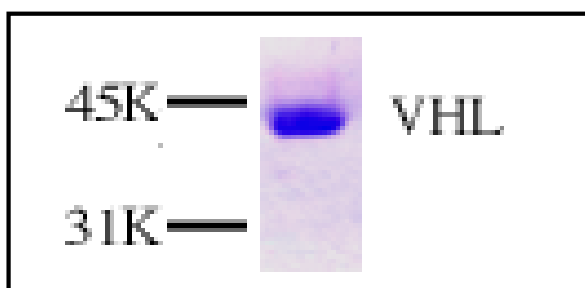


VHL

von Hippel-Lindau Tumor Suppressor Protein
 human, recombinant, Sf9 insect cells

Cat. No.	Amount
PR-768	5 µg



For *in vitro* use only
 Quality guaranteed for 12 months
 Store at -80°C

Avoid freeze / thaw cycles

Form

Liquid. Supplied in 20 mM Tris-HCl pH 8.0, 100 mM KCl, 0.2 mM EDTA, 1 mM DTT and 20% glycerol.

Activity

30-100 ng are required for *in vitro* transcription assays and protein-protein interaction assays.

Application

Purified VHL protein has been used for *in vitro* transcriptional activation and protein-protein interaction assays.

Molecular Weight

42 kDa

Purity

> 95% by SDS-PAGE

Description

Von Hippel-Lindau (VHL) disease is a hereditary cancer with a predilection for the central nervous system and retina. The von Hippel-Lindau tumor suppressor gene is mutated in families with von Hippel-Lindau disease and encodes a protein (VHL) of 213 amino acids with an acidic pentapeptide motif in the N-terminus. Mutations in the VHL gene result in constitutive expression of many hypoxia-induced genes, at least in part because of increases in the cellular level of hypoxia-inducible transcription factor HIF-1α. VHL protein binds to elongin B, elongin C, and Cul2 to form a stable complex that targets Hypoxia Inducible Factors (HIFs) for degradation and transcriptional regulation. In addition, VHL protein has also been shown to interact with specific Protein Kinase C isoforms, histone deacetylases and HIF-1 inhibitor (FIH-1).

The wild type human VHL protein (213 amino acids) was expressed in baculovirus system and purified using an affinity column and FPLC chromatography under non-denaturing condition.

Purified VHL protein is greater than 90% homogeneous and contains no detectable protease, DNase, and RNase activity.

Selected References:

- Singh *et al.* (2001) von Hippel-Lindau disease. *Survey of Ophthalmology* **46**:117.
 Latif *et al.* (1993) Identification of the von Hippel-Lindau disease tumor suppressor gene. *Science* **260**:1317.
 Linehan *et al.* (1995) Identification of the von Hippel-Lindau (VHL) gene. Its role in renal cancer. *J. Am. Med. Assoc.* **273**:564.
 Duan *et al.* (1995) Characterization of the VHL tumor suppressor gene product: localization, complex formation, and the effect of natural inactivating mutations. *Proc. Natl. Acad. Sci. USA* **92**:6459.
 Kamura *et al.* (2000) Activation of HIF1α ubiquitination by a reconstituted von Hippel-Lindau (VHL) tumor suppressor complex. *Proc. Natl. Acad. Sci. USA* **97**:10430.
 Duan *et al.* (1995) Inhibition of transcription elongation by the VHL tumor suppressor protein. *Science* **269**:1402.
 Iwai *et al.* (1999) Identification of the von Hippel-Lindau tumor suppressor protein as part of an active E3 ubiquitin ligase complex. *Proc. Natl. Acad. Sci. USA* **96**:12436.
 Pal *et al.* (1997) The von Hippel-Lindau gene product inhibits vascular permeability factor/vascular endothelial growth factor expression in renal cell carcinoma by blocking protein kinase C pathways. *J. Biol. Chem.* **272**:27509.
 Mahon *et al.* (2001) FIH-1: a novel protein that interacts with HIF-1α and VHL to mediate repression of HIF-1 transcriptional activity. *Genes & Dev.* **15**:2675.