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VHL

Reactivity: Human Mouse Rat

Tested applications:WB IHC IF

Recommended Dilution:WB 1:500 - 1:2000 IHC 1:50 - 1:200 IF 1:20 - 1:100

Calculated MW:24kDa

Observed MW: Refer to Figures

Immunogen:

Recombinant protein of human VHL

Storage Buffer:

Store at -20. Avoid freeze / thaw cycles. Buffer: PBS with 0.02% sodium azide, 50% glycerol, pH7.3.

Concentration:

b

Synonym:

Von Hippel-Lindau disease tumor suppressor; pVHL; Protein G7; VHL

Catalog #:A0377

Antibody Type:

Polyclonal Antibody

Species: Rabbit

Gene ID:7428

Isotype: IgG

Swiss Prot:P40337

Purity: Affinity purification

For research use only.

Background:

The Von Hippel-Lindau (VHL) protein is a substrate recognition component of an E3 ubiquitin ligase complex containing elongin BC (TCEB1 and TCEB2), cullin 1 (CUL1), and RING-box protein 1 (RBX1) (1,2,3). VHL protein has been shown to exist as three distinct isoforms resulting from alternatively spliced transcript variants (4). Loss of VHL protein function results in a dominantly inherited familial cancer syndrome that manifests as angiomas of the retina, hemangioblastomas of the central nervous system, renal clear-cell carcinomas and pheochromocytomas (4). Under normoxic conditions, VHL directs the ubiquitylation and subsequent proteosomal degradation of the hypoxia inducible factor HIF alpha, maintaining very low levels of HIF alpha in the cell. Cellular exposure to hypoxic conditions, or loss of VHL protein function, results in increased HIF alpha protein levels and increased expression of HIF-induced gene products, many of which are angiogenesis factors such as vascular endothelial growth factor (VEGF). Thus, loss of VHL protein function is believed to contribute to the formation of highly vascular neoplasias (4). In addition to HIF alpha, VHL is known to regulate the ubiquitylation of several other proteins, including tat-binding protein 1 (TBP-1), the atypical protein kinase C lambda (aPKC), and two subunits of the multiprotein RNA Polymerase II complex (RPB1 and RPB7) (5,6,7,8). Interactions with elongin BC, RPB1, RPB7 and the pVHL-associated KRAB-A domain containing protein (VHLaK) suggest that VHL may also play a more direct role in transcriptional repression.

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