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Rabbit Anti-VHL antibody

SL1367R

Product Name:	VHL
Chinese Name:	脑视网膜血管瘤G7蛋白抗体(逢希伯-林道氏病)
Alias:	Von Hippel Lindau; von Hippel-Lindau syndrome protein homolog; Hippel-Lindau disease tumor suppressor VHL; von Hippel-Lindau tumor suppressor isoform 1; VHL; HRCA1; RCA1; VHL1; von Hippel-Lindau disease tumor suppressor isoform 2; pVHL; pVHL; G7 protein; Elongin binding protein; Protein G7; VHL 1; VHL_HUMAN; VHL1; VHLH; Von Hippel Lindau disease tumor suppressor; von Hippel Lindau syndrome; von Hippel Lindau tumor suppressor; Von Hippel-Lindau disease tumor suppressor.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Cow,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800Flow-Cyt=1ug/testIF=1:100-500(Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	24kDa
Cellular localization:	The nucleuscytoplasmicThe cell membrane
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human VHL:101-213/213
Lsotype:	IgG
Purification:	affinity purified by Protein A
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Von Hippel-Lindau syndrome (VHL) is a dominantly inherited familial cancer syndrome

predisposing to a variety of malignant and benign tumors. A germline mutation of this gene is the basis of familial inheritance of VHL syndrome. The protein encoded by this gene is a component of the protein complex that includes elongin B, elongin C, and cullin-2, and possesses ubiquitin ligase E3 activity. This protein is involved in the ubiquitination and degradation of hypoxia-inducible-factor (HIF), which is a transcription factor that plays a central role in the regulation of gene expression by oxygen. RNA polymerase II subunit POLR2G/RPB7 is also reported to be a target of this protein. Alternatively spliced transcript variants encoding distinct isoforms have been observed. [provided by RefSeq, Jul 2008].

Function:
Involved in the ubiquitination and subsequent proteasomal degradation via the von Hippel-Lindau ubiquitination complex. Seems to act as target recruitment subunit in the E3 ubiquitin ligase complex and recruits hydroxylated hypoxia-inducible factor (HIF) under normoxic conditions. Involved in transcriptional repression through interaction with HIF1A, HIF1AN and histone deacetylases. Ubiquitinates, in an oxygen-responsive manner, ADRB2.

Subunit:
Component of the VCB (VHL-Elongin BC-CUL2) complex; this complex acts as a ubiquitin-ligase E3 and directs proteasome-dependent degradation of targeted proteins. Interacts with CUL2; this interaction is dependent on the integrity of the trimeric VBC complex. Interacts (via the beta domain) with HIF1A (via the NTAD domain); this interaction mediates degradation of HIF1A in normoxia and, in hypoxia, prevents ubiqitination and degradation of HIF1A by mediating hypoxia-induced translocation to the nucleus, a process which requires a hypoxia-dependent regulatory signal. Interacts with ADRB2; the interaction, in normoxia, is dependent on hydroxylation of ADRB2 and the subsequent VCB-mediated ubiquitination and degradation of ADRB2. Under hypoxia, hydroxylation, interaction with VHL, ubiquitination and subsequent degradation of ADRB2 are dramatically decreased. Interacts with RNF139, USP33 and PHF17.

Subcellular Location:
Isoform 1: Cytoplasm. Membrane; Peripheral membrane protein. Nucleus. Note=Found predominantly in the cytoplasm and with less amounts nuclear or membrane-associated. Colocalizes with ADRB2 at the cell membrane.
Isoform 3: Cytoplasm. Nucleus. Note=Equally distributed between the nucleus and the cytoplasm but not membrane-associated.

Tissue Specificity:
Expressed in the adult and fetal brain and kidney.

DISEASE:
Defects in VHL are a cause of susceptibility to pheochromocytoma (PCC) [MIM:171300]. A catecholamine-producing tumor of chromaffin tissue of the adrenal medulla or sympathetic paraganglia. The cardinal symptom, reflecting the increased

secretion of epinephrine and norepinephrine, is hypertension, which may be persistent or intermittent.
Defects in VHL are the cause of von Hippel-Lindau disease (VHLD) [MIM:193300]. VHLD is a dominantly inherited familial cancer syndrome characterized by the development of retinal angiomatosis, cerebellar and spinal hemangioblastoma, renal cell carcinoma (RCC), phaeochromocytoma and pancreatic tumors. VHL type 1 is without pheochromocytoma, type 2 is with pheochromocytoma. VHL type 2 is further subdivided into types 2A (pheochromocytoma, retinal angioma, and hemangioblastomas without renal cell carcinoma and pancreatic cyst) and 2B (pheochromocytoma, retinal angioma, and hemangioblastomas with renal cell carcinoma and pancreatic cyst). VHL type 2C refers to patients with isolated pheochromocytoma without hemangioblastoma or renal cell carcinoma. The estimated incidence is 3/100000 births per year and penetrance is 97% by age 60 years.
Defects in VHL are the cause of familial erythrocytosis type 2 (ECYT2) [MIM:263400]; also called VHL-dependent polycythemia or Chuvash type polycythemia. ECYT2 is an autosomal recessive disorder characterized by an increase in serum red blood cell mass, hypersensitivity of erythroid progenitors to erythropoietin, increased erythropoietin serum levels, and normal oxygen affinity. Patients with ECYT2 carry a high risk for peripheral thrombosis and cerebrovascular events.
Defects in VHL are a cause of renal cell carcinoma (RCC) [MIM:144700]. Renal cell carcinoma is a heterogeneous group of sporadic or hereditary carcinoma derived from cells of the proximal renal tubular epithelium. It is subclassified into clear cell renal carcinoma (non-papillary carcinoma), papillary renal cell carcinoma, chromophobe renal cell carcinoma, collecting duct carcinoma with medullary carcinoma of the kidney, and unclassified renal cell carcinoma.

SWISS:
P40337

Gene ID:
7428

Database links:

Entrez Gene: 7428Human

Entrez Gene: 22346Mouse

Omim: 608537Human

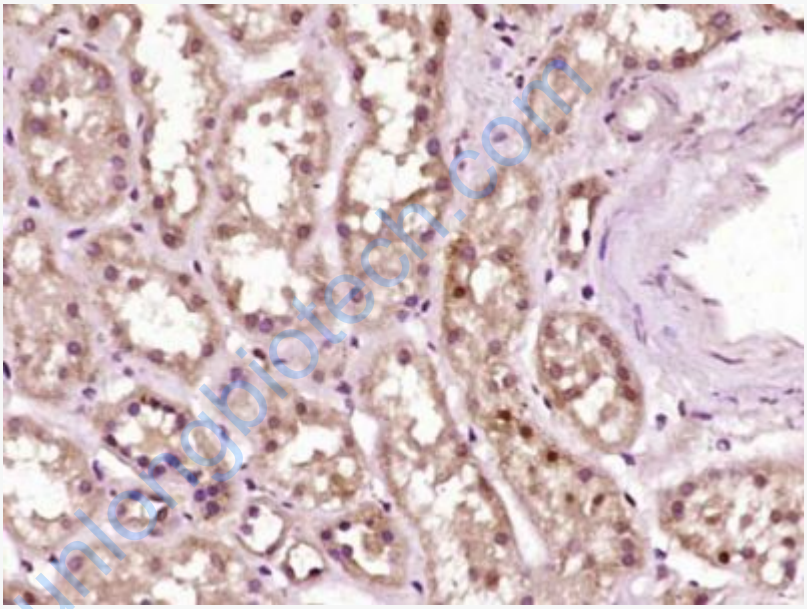
SwissProt: P40337Human

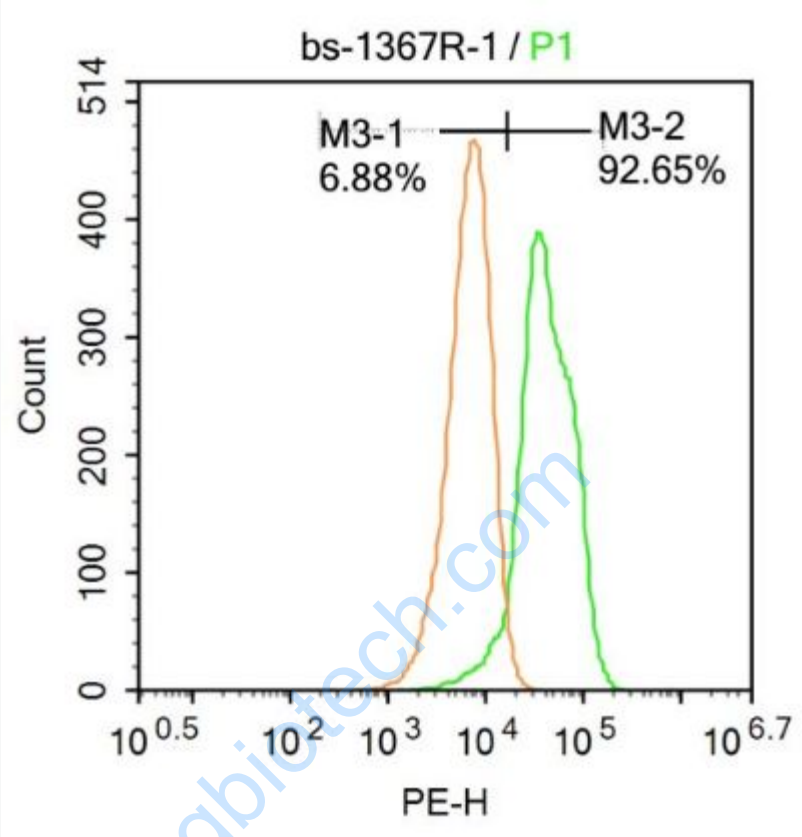
SwissProt: P40338Mouse

Unigene: 517792Human

Unigene: 607789Human

Unigene: 29407Mouse

	Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. von Hippel-Lindau是一种Tumour抑制因子，在细胞对氧的感受过程中发挥关键作用，VHL蛋白除了调节血管生成外还在调节细胞的生长和生存、对调节细胞周期、Apoptosis和Extracellular matrix方面起重要作用。
Picture:	 Paraformaldehyde-fixed, paraffin embedded (Human kidney); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (VHL) Polyclonal Antibody, Unconjugated (SL1367R) at 1:400 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructionsand DAB staining.



Blank control:Molt-4.

Primary Antibody (green line): Rabbit Anti-VHL antibody (SL1367R)

Dilution: 1μg /10^6 cells;

Isotype Control Antibody (orange line): Rabbit IgG .

Secondary Antibody : Goat anti-rabbit IgG-AF647

Dilution: 1μg /test.

Protocol

The cells were fixed with 4% PFA (10min at room temperature)and then permeabilized with 90% ice-cold methanol for 20 min at-20℃. The cells were then incubated in 5%BSA to block non-specific protein-protein interactions for 30 min at at room temperature .Cells stained with Primary Antibody for 30 min at room

	temperature. The secondary antibody used for 40 min at room temperature. Acquisition of 20,000 events was performed.