abcam

Product datasheet

Anti-Von Hippel Lindau/VHL antibody ab28434

4 References

Overview

Product name Anti-Von Hippel Lindau/VHL antibody

Description Rabbit polyclonal to Von Hippel Lindau/VHL

Host species Rabbit

Tested applications Suitable for: WB

Species reactivity Reacts with: Human

Immunogen Recombinant full length protein corresponding to Human Von Hippel Lindau/VHL.

Positive control HeLa nuclear extract

General notes

The Life Science industry has been in the grips of a reproducibility crisis for a number of years.

Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets

your needs before purchasing.

If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be

found below, along with publications, customer reviews and Q&As

Properties

Form Liquid

Storage instructions Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.

Storage buffer pH: 7.9

Purity Protein A purified

Clonality Polyclonal

Isotype IgG

Applications

The Abpromise guarantee Our Abpromise guarantee covers the use of ab28434 in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

1

Application	Abreviews	Notes
WB		Use at an assay dependent concentration.

Target

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.

Tissue specificity

Expressed in the adult and fetal brain and kidney.

Pathway

Protein modification; protein ubiquitination.

Involvement in 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 erythrocytosis familial 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.

Domain

The Elongin BC complex binding domain is also known as BC-box with the consensus [APST]-L-x(3)-C-x(3)-[AlLV].

Cellular localization

Cytoplasm. Membrane. Nucleus. Found predominantly in the cytoplasm and with less amounts nuclear or membrane-associated and Cytoplasm. Nucleus. Equally distributed between the nucleus and the cytoplasm but not membrane-associated.

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