

Anti-Von Hippel Lindau/VHL antibody ab28434

4 References

製品の概要

製品名	Anti-Von Hippel Lindau/VHL antibody
製品の詳細	Rabbit polyclonal to Von Hippel Lindau/VHL
由来種	Rabbit
アプリケーション	適用あり: WB
種交差性	交差種: Human
免疫原	Recombinant full length protein corresponding to Human Von Hippel Lindau/VHL.
ポジティブ・コントロール	HeLa nuclear extract
特記事項	<p>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.</p> <p>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</p>

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles.
バッファー	pH: 7.9
精製度	Protein A purified
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

アプリケーション

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アプリケーション	Abreviews	特記事項
WB		Use at an assay dependent concentration.

ターゲット情報

機能	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.
組織特異性	Expressed in the adult and fetal brain and kidney.
パスウェイ	Protein modification; protein ubiquitination.
関連疾患	<p>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.</p> <p>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 angiomas, cerebellar and spinal hemangioblastoma, renal cell carcinoma (RCC), pheochromocytoma 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.</p> <p>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.</p> <p>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.</p>
ドメイン	The Elongin BC complex binding domain is also known as BC-box with the consensus [APST]-L-x(3)-C-x(3)-[AILV].
細胞内局在	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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