abcam

Product datasheet

Recombinant Human Von Hippel Lindau/VHL protein ab48702

画像数 1

製品の詳細

製品名	Recombinant Human Von Hippel Lindau/VHL protein
精製度	> 85 % SDS-PAGE. Von Hippel Lindau/VHL ß-domain (1-154) was overexpressed in E.coli and purified by using conventional chromatography techniques.
発現系	Escherichia coli
タンパク質長	Protein fragment
Animal free	No
由来	Recombinant
生物種	Human
配列	MGSSHHHHHH SSGLVPRGSH MPRRAENWDE AEVGAEEAGV EEYGPEEDGG EESGAEESGP EESGPEELGA EEEMEAGRPR PVLRSVNSRE PSQVIFCNRS PRVVLPVWLN FDGEPQPYPT LPPGTGRRIH SYRGHLWLFR DAGTHDGLLV NQTELFVPSL NVDGQPIFAN ITLP
予測される分子量	19 kDa including tags
領域	1 to 154
タグ	His tag N-Terminus

特性

Our **Abpromise guarantee** covers the use of **ab48702** in the following tested applications.

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

アプリケーション	SDS-PAGE
製品の状態	Liquid

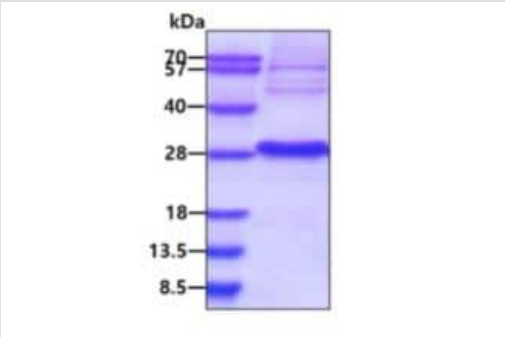
前処理および保存

保存方法および安定性	Shipped at 4°C. Upon delivery aliquot and store at -20°C. Avoid freeze / thaw cycles. pH: 7.40 Constituents: PBS, 0.0154% DTT, 0.0584% EDTA

関連情報

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	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.
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.

画像



SDS-PAGE - Recombinant Human Von Hippel Lindau/VHL protein (ab48702)

3ug by SDS-PAGE under reducing condition and visualized by coomassie blue stain

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