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## Overview

Overview	
Quantity:	0.1 mg
Target:	KRIT1
Binding Specificity:	AA 1-736, N-Term
Reactivity:	Human
Host:	Rabbit
Clonality:	Polyclonal
Application:	Western Blotting (WB), Immunofluorescence (IF)
Product Details	
Immunogen:	Highly pure (>95%) recombinant Human CCM-1 (Met1-Ser736) derived from E. coli fused to a C-
	teminal His-tag (6 x His) (CatNo AR26002PU-N)
Isotype:	IgG
Specificity:	This antibody is anti-His depleated. It detects KRIT1 / CCM1.
Cross-Reactivity (Details):	Species reactivity (tested):Human.
Purification:	Protein A Chromatography
Target Details	
Target:	KRIT1
Alternative Name:	KRIT1 / CCM1 (KRIT1 Products)
Background:	Cerebral Cavernous Malformations (CCM) are frequent vascular abnormalities caused by

mutations in one of the CCM genes. CCM-1 (also known as KRIT1) stabilizes endothelial junctions and is essential for vascular morphogenesis in mouse embryos. However, cellular functions of CCM-1 during the early steps of the CCM pathogenesis remain unknown. It was shown that CCM-1 represents an antiangiogenic protein to keep the human endothelium quiescent. CCM-1 inhibits endothelial proliferation, apoptosis, migration, lumen formation, and sprouting angiogenesis in primary human endothelial cells. CCM-1 strongly induces DLL4-NOTCH signaling, which promotes AKT phosphorylation but reduces phosphorylation of the mitogen-activated protein kinase ERK. Consistently, blocking of NOTCH activity alleviates CCM-1 effects. ERK phosphorylation is increased in human CCM lesions. Transplantation of CCM-1-silenced human endothelial cells into SCID mice recapitulates hallmarks of the CCM pathology and serves as a unique CCM model system. Synonyms: Krev interaction trapped 1

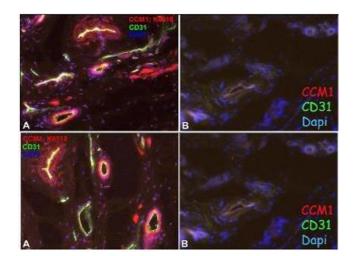
Gene ID:	889
NCBI Accession:	NP_004903
UniProt:	000522
Pathways:	Cell RedoxHomeostasis

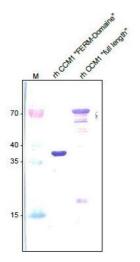
# Application Details

Application Notes:	Optimal working dilution should be determined by the investigator.
Restrictions:	For Research Use only

### Handling

Reconstitution:	Restore in sterile water to a concentration of 0.1-1.0 mg/mL. Centrifuge vial prior to opening.
Buffer:	5 mM PBS, pH 7.2
Handling Advice:	Avoid repeated freezing and thawing.
Storage:	4 °C/-20 °C
Storage Comment:	Prior to reconstitution store at 2-8 °C. Following reconstitution store undiluted at 2-8 °C for one month or (in aliquots) at -20 °C for longer.





#### **Immunofluorescence**

Image 1. Immunofluorescence staining of Human foreskin (Cryo-section of unfixed tissue) with anti-CCM1 Antibody Cat.-No AP26021PU-L (red, dilution: 1/50). Costaining of endothelial cells with anti-CD31 (green). Note specific staining in the wall of a subset of vessel. Nuclei counterstained with Dapi (blue). Specimen provided by Prof. Dr. J. Wilting and Dr. K. Buttler, Goettingen.

#### **Western Blotting**

**Image 2.** Western analysis of recombinant Human CCM-1 (FERM domain) and recombinant Human full length CCM-1 using a Rabbit polyclonal anti-Human CCM-1 antibody Cat.-No AP26021PU generated against the recombinant FERM domain of Human CCM-1.