



### Anti-human CCM1/Krit1

20140616BB



**FOR RESEARCH ONLY! NOT FOR HUMAN USE!**

<b>Cat.-no.:</b>	<b>102-PA25S</b>
Size:	100 µg
Lot. No.:	According to product label
Country of origin:	Germany

**Preparation:** Produced from sera of rabbits pre-immunized with highly pure (>95%) recombinant human CCM1 (Leu411-Arg734) from E. coli.

### Target Background

<b>Synonyms:</b>	Cerebral cavernous malformations1 protein, Krev interaction trapped protein 1
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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.

### Database References Antigen

<b>Protein RefSeq:</b>	NP_004903.2
<b>Uniprot ID:</b>	O00522
<b>mRNA RefSeq:</b>	NM_004912.3

### Product Specifications

<b>Species reactivity</b>	human
<b>Clone/Ab feature</b>	rabbit IgG
<b>Cross reactivity</b>	ND
<b>Host</b>	rabbit
<b>Clonality</b>	polyclonal
<b>Purification</b>	Protein A purified
<b>Immunogen</b>	Recombinant human CCM1 (RT #300-054)
<b>Formulation</b>	lyophilized
<b>Buffer</b>	5 mM PBS, pH 7.2

**Stability:** The lyophilized antibody is stable at room temperature for up to 1 month. The reconstituted antibody is stable for at least two weeks at 2-8°C. Frozen aliquots are stable for at least 6 months when stored at -20°C.

**Reconstitution:** Centrifuge vial prior to opening. Reconstitute in sterile water to a concentration of 0.1-1.0 mg/ml.



**AVOID REPEATED FREEZE AND THAW CYCLES!**

### References

1. Verlaan DJ et al, Neurology 26 (2002)
2. Revencu N and Vikkula M, J Med Genet 43 (2006)
3. Yadla et al, S, Neurosurg Focus 29 (2010)
4. Marchuk et al, Hum Mol Genet 12 Spec No 1 (2003)
5. Liu et al, J Vasc Res 48 (2011)

### Applications

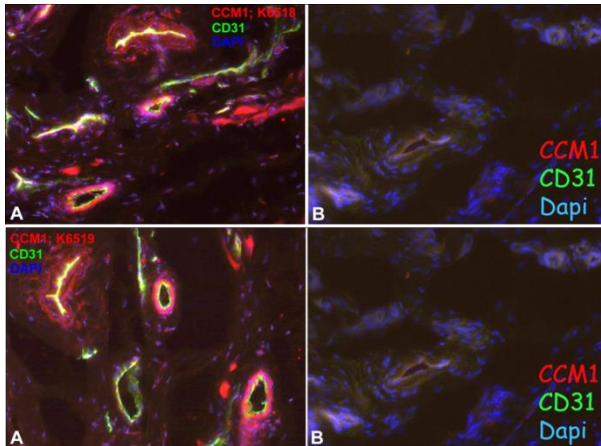
<b>Western Blot:</b>	Use 1-5 µg/ml
<b>IF/IHC</b>	Use 1:200

**NOTE: OPTIMAL DILUTIONS SHOULD BE DETERMINED BY EACH LABORATORY FOR EACH APPLICATION!**



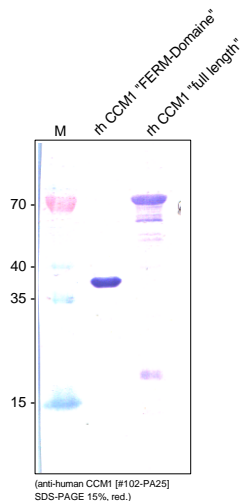
## Anti-human CCM1

### Handling/Applications



**Figure 1: Immunofluorescence staining** of human foreskin (cryo-section of unfixed tissue) with anti-CCM1 (red; dilution 1:50) [Cat# 102-PA25]. Costaining of endothelial cells with anti-CD31 (green). Note specific staining in the wall of a subset of vessel. Nuclei counter-stained with Dapi (blue). Specimen provided by Prof. Dr. J. Wilting and Dr. K. Buttler, Goettingen.

The experiment was performed by the research group of Prof. Dr. J. Wilting, University Göttingen, Germany.



**Figure 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# 102-PA25] generated against the recombinant FERM domain of human CCM-1.