Anti-human CCM1/Krit1

20140616BB

FOR RESEARCH ONLY! NOT FOR HUMAN USE!

Cat.-no.:	**102-PA25**
Size:	200 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

Synonyms:	Cerebral cavernous malformations1 protein, Krev interaction trapped protein 1

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.

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)

.

Database References Antigen

Protein RefSeq:	NP_004903.2
Uniprot ID:	O00522
mRNA RefSeq:	NM_004912.3

Product Specifications

Species reactivity	human
Clone/Ab feature	rabbit IgG
Cross reactivity	ND
Host	rabbit
Clonality	polyclonal
Purification	Protein A purified
Immunogen	Recombinant human CCM1 (RT #300-054)
Formulation	lyophilized
Buffer	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!

Applications

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

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

Anti-human CCM1

Handling/Applications

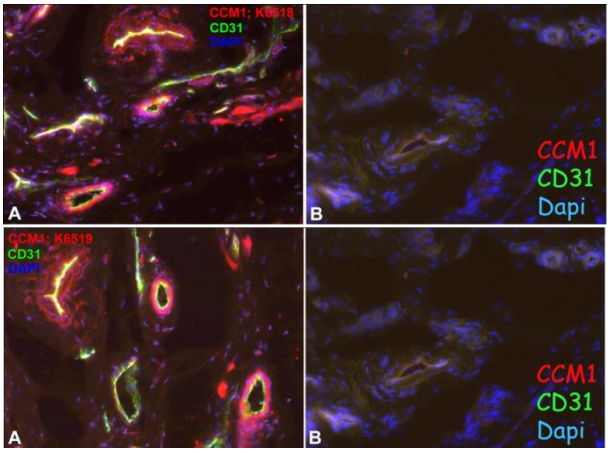

Figure 1: Immunofluorescence staining of human foreskin (cryo-section of unfixed tissue) with anti-CCM1 (red; dilution 1:50) [Cat# 102-PA25]. Costaining of endothelial cells with anti-CD31 (green). Note specific staining in the wall of a subset of vessel. Nuclei counter-stained with Dapi (blue). Specimen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

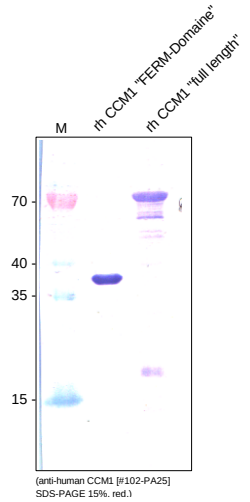

Figure 2: Western analysis of recombinant human CCM-1 (FERM domain) and recombinant human full length CCM-1 using a rabbit polyclonal anti-human CCM-1 antibody [Cat# 102-PA25] generated against the recombinant FERM domain of human CCM-1.