



Anti-human CCM2

20140401BB



FOR RESEARCH ONLY! NOT FOR HUMAN USE!

Cat.-no.:	102-PA26S
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 CCM2 (Met1-Ala464) from E. coli.

Target Background

Synonyms:	Cerebral cavernous malformations1 2 protein
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Cerebral cavernous malformations (CCMs) are sporadically acquired or inherited vascular lesions of the central nervous system consisting of clusters of dilated thin-walled blood vessels that predispose individuals to seizures and stroke. Familial CCM is caused by mutations in KRIT1 (CCM1) or in malcavernin (CCM2). The roles of the CCM proteins in the pathogenesis of the disorder remain largely unknown. It was shown that the CCM1 gene product, KRIT1, interacts with the CCM2 gene product, malcavernin. Analogous to the established interactions of CCM1 and beta1 integrin with ICAP1, the CCM1/CCM2 association is dependent upon the phosphotyrosine binding (PTB) domain of CCM2. A familial CCM2 missense mutation abrogates the CCM1/CCM2 interaction, suggesting that loss of this interaction may be critical in CCM pathogenesis. CCM2 and ICAP1 bound to CCM1 via their respective PTB domains differentially influence the subcellular localization of CCM1. The data indicate that the genetic heterogeneity observed in familial CCM may reflect mutation of different molecular members of a coordinated signaling complex.

References

1. Plummer et al, Curr Neurol Neurosci Rep 5 (2005)
2. Dashti et al, Neurosurg Focus 21 (2006)
3. Revencu N and Vikkula M, J Med Genet 43 (2006)
4. Yadla et al, S, Neurosurg Focus 29 (2010)
5. Verlaan DJ et al, Neurology 26 (2002)

Database References Antigen

Protein RefSeq:	NP_001025006.1
Uniprot ID:	Q9BSQ5
mRNA RefSeq:	NM_001029835.2

Product Specifications

Species reactivity	human
Clone/Ab feature	rabbit IgG
Cross reactivity	ND
Host	rabbit
Clonality	polyclonal
Purification	Protein A purified
Immunogen	Recombinant human CCM2 (RT #300-055)
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 (IF)

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



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Handling/Applications

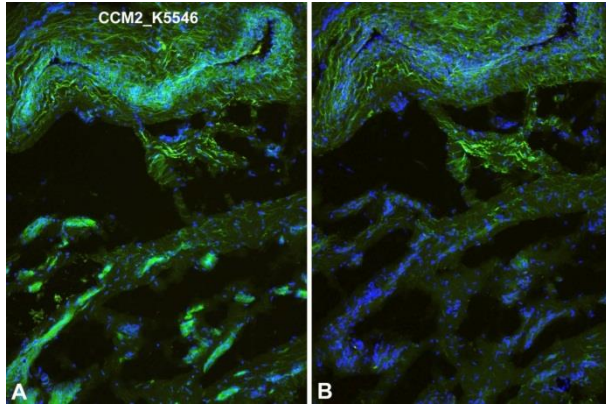
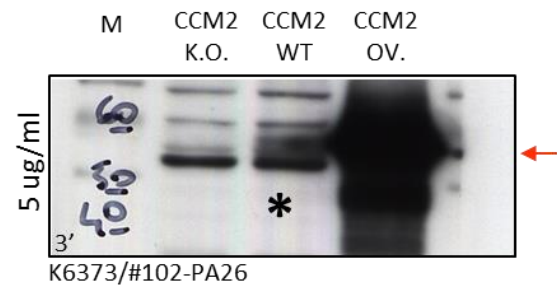
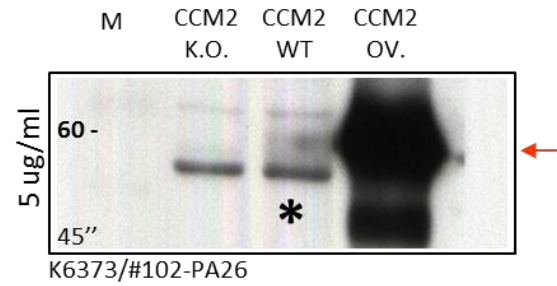


Figure 1: Immunofluorescence staining (green) of human foreskin (cryo-section of unfixed tissue) with anti-human CCM2 (K5546; dilution 1:50) [Cat# 102-PA26]. A) Note specific staining in the wall of microvessels. B) Negative control of a consecutive section. Note non-specific fluorescence in elastic fibres in the adventitia of an arterioli. Nuclei counter-stained with Dapi (blue). Specimen provided by Prof. Dr. J. Wilting, Goettingen.

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



K.O. = knock out (completely deleted)
K.D. = knock down (not completely deleted)
OV. = over expressed in COS1 cells

Figure 2. Western Analysis of anti-human CCM-2.

The experiment was performed by Elisabetta Dejana's group, IFOM-IEO-Campus, Milan Italy