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Product datasheet for AP26022PU-L

CCM2 Rabbit Polyclonal Antibody

Product data:

Product Type:	Primary Antibodies
Applications:	IF, WB
Recommended Dilution:	**Western blot:** 1-5 µg/ml. **Immunofluorescence:** 1/200. **Immunohistochemistry.**
Reactivity:	Human
Host:	Rabbit
Clonality:	Polyclonal
Immunogen:	Highly pure (> 95%) recombinant Human CCM-2 (Cerebral cavernous malformations 2 protein; aa: Met1-Ala444) from E.coli (*Cat.-No* AR26003PU-N).
Specificity:	This antibody detects recombinant Human CCM-2 in Western Blot and native CCM-2 in Immunohistochemistry.
Formulation:	5mM PBS pH 7.2 without preservatives State: Aff - Purified State: Lyophilized purified Ig fraction
Reconstitution Method:	Restore in sterile water to a concentration of 0.1-1.0 mg/ml.
Purification:	Protein A Chromatography
Conjugation:	Unconjugated
Storage:	Prior to reconstitution store at 2-8°C for one month or dessicated at -20°C for longer. Following reconstitution store undiluted at 2-8°C for one month or (in aliquots) at -20°C for longer. Avoid repeated freezing and thawing.
Stability:	Shelf life: one year from despatch.
Gene Name:	CCM2 scaffolding protein
Database Link:	Entrez Gene 83605 Human Q9BSQ5

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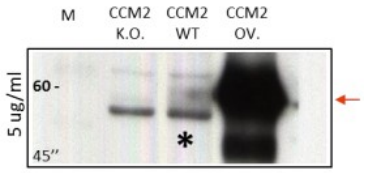
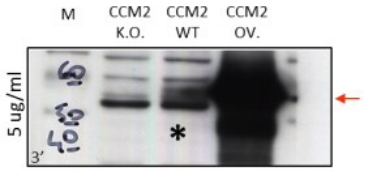
Background: Cerebral cavernous malformations (CCMs) are sporadically acquired or inherited vascular lesions of the central nervous system consisting of clusters of dilated thin-walled blood vessels that predispose individuals to seizures and stroke. Familial CCM is caused by mutations in KRIT1 (CCM1) 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.

Synonyms: CCM2, C7orf22, PP10187

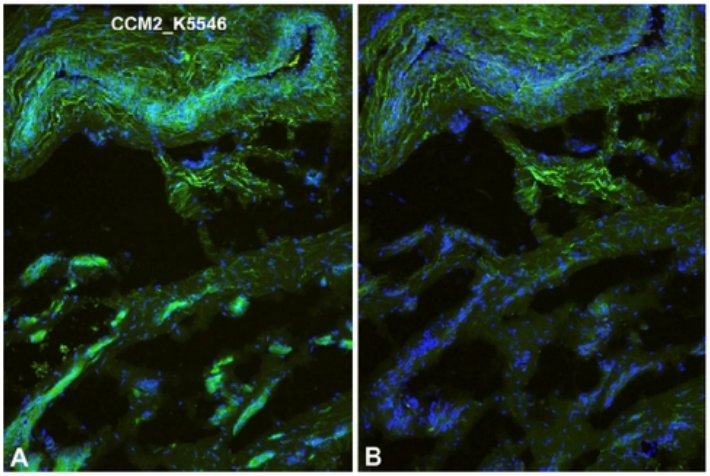
Product images:





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

Western Blot; Analysis of anti-human CCM-2. The experiment was performed by Elisabetta Dejana's group, IFOM-IEO-Campus, Milan Italy



Immunofluorescence staining (green) of Human foreskin (cryo-section of unfixed tissue) with anti-Human CCM2 antibody. (K5546, dilution1/50). 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 anarteriol. 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 GÃ¶ttingen, Germany.