

CCM2 Rabbit pAb

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Catalog # AP54633

Product Information

Application	WB
Primary Accession	Q9BSQ5
Reactivity	Human
Predicted	Rat, Mouse, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	48837
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Malcavernin
Epitope Specificity	18-100/444
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBCELLULAR LOCATION	Cytoplasm.
DISEASE	Defects in CCM2 are the cause of cerebral cavernous malformations type 2 (CCM2) [MIM:603284]. Cerebral cavernous malformations (CCMs) are congenital vascular anomalies of the central nervous system that can result in hemorrhagic stroke, seizures, recurrent headaches, and focal neurologic deficits. CCMs have an incidence of 0.1%-0.5% in the general population and are usually present clinically during the 3rd to 5th decades of life. The lesions are characterized by grossly enlarged blood vessels consisting of a single layer of endothelium and without any intervening neural tissue, ranging in diameter from a few millimeters to several centimeters.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Cerebral cavernous malformation (CCM) is an autosomal dominant or sporadic neurovascular disease marked by vascular anomalies located mostly in the central nervous system that can cause stroke, seizures, cerebral hemorrhages, headaches and focal neurologic deficits. CCM is caused by mutations in one of three genes: CCM1, CCM2 or CCM3. CCM1 encodes the protein KRIT1, CCM2 encodes the protein Malcavernin and CCM3 shares its name with the protein it encodes. Malcavernin, also designated cerebral cavernous malformations 2 protein, is a scaffolding protein for MEK kinase-3. Like KRIT1, Malcavernin is expressed in a variety of human organs including the arterial vascular endothelium, pyramidal neurons, astrocytes and their foot processes. In addition, Malcavernin is expressed in various epithelial cells that are required for the formation of the blood-organ barrier. Malcavernin is localized to the cytoplasm but is known to shuttle to and from the nucleus. Due to its lack of a nuclear export signal or nuclear localization signal, it is believed that Malcavernin accomplishes this shuttling via an attachment to KRIT1, which contains a nuclear localization signal. Two isoforms exist for Malcavernin. Isoform 1 represents the full length protein while isoform 2 contains an alternative four amino acid sequence rather than the first 10

residues of isoform 1.

Additional Information

Gene ID	83605
Other Names	Cerebral cavernous malformations 2 protein, Malcavernin, CCM2, C7orf22
Dilution	WB=1:500-2000
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

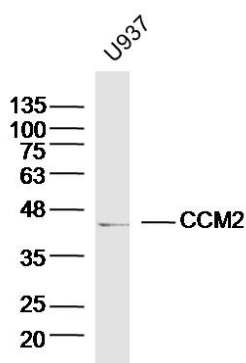
Protein Information

Name	CCM2
Synonyms	C7orf22
Function	Component of the CCM signaling pathway which is a crucial regulator of heart and vessel formation and integrity. May act through the stabilization of endothelial cell junctions (By similarity). May function as a scaffold protein for MAP2K3-MAP3K3 signaling. Seems to play a major role in the modulation of MAP3K3-dependent p38 activation induced by hyperosmotic shock (By similarity).
Cellular Location	Cytoplasm.

Background

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Images



Sample: U937 (human) cell Lysate at 40 ug
Primary: Anti- CCM2 (AP54633) at 1/300 dilution
Secondary: IRDye800CW Goat Anti-Rabbit IgG at 1/20000 dilution
Predicted band size: 49 kD
Observed band size: 45 kD

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