

# **Anti-Human CCM-1**

Synonyms: CCM-1, Cerebral cavernous malformations protein 1, KRIT1, KRIT1, ankyrin repeat containing, CAM

#### PLEASE NOTE: ALWAYS CENTRIFUGE VIAL BEFORE OPENING

Size Order# Lot# Expiry Date

100 μg 4384.852.100 200 μg 4384.852.200

Please enquire for bulk quantities and other vial sizes.

## **Description**

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.

Source RabbitClone AB-Sbccju!JH

### **Biological Activity**

Western Blot: Use 1-5 μg/ml

#### Reconstitution

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

Usage: For research use only. Not for use in diagnostic or therapeutic procedures. Not for human use.