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Hamartin Antibody

Rabbit mAb
Catalog # AP92337

Product Information

Application	WB, IHC, IF, FC, ICC, IHF
Primary Accession	Q92574
Reactivity	Human
Clonality	Monoclonal
Other Names	Hamartin; kiaa0243; LAM; TSC; Tsc1; Tuberous sclerosis 1;
Isotype	Rabbit IgG
Host	Rabbit
Calculated MW	129767

Additional Information

Dilution	WB 1:1000~1:5000 IHC 1:50~1:200 ICC/IF 1:50~1:200 FC 1:200
Purification	Affinity-chromatography
Immunogen	A synthesized peptide derived from human Hamartin
Description	In complex with TSC2, inhibits the nutrient-mediated or growth factor-stimulated phosphorylation of S6K1 and EIF4EBP1 by negatively regulating mTORC1 signaling. Seems not to be required for TSC2 GAP activity towards RHEB. Implicated as a tumor suppressor. Involved in microtubule-mediated protein transport, but this seems to be due to unregulated mTOR signaling.
Storage Condition and Buffer	Rabbit IgG in phosphate buffered saline , pH 7.4, 150mM NaCl, 0.02% sodium azide and 50% glycerol. Store at +4°C short term. Store at -20°C long term. Avoid freeze / thaw cycle.

Protein Information

Name	TSC1 {ECO:0000303|PubMed:9242607, ECO:0000312|HGNC:HGNC:12362}
Function	Non-catalytic component of the TSC-TBC complex, a multiprotein complex that acts as a negative regulator of the canonical mTORC1 complex, an evolutionarily conserved central nutrient sensor that stimulates anabolic reactions and macromolecule biosynthesis to promote cellular biomass generation and growth (PubMed:12172553, PubMed:12271141, PubMed:12906785, PubMed:15340059, PubMed:24529379, PubMed:28215400). The TSC-TBC complex acts as a GTPase-activating protein (GAP) for the small GTPase RHEB, a direct activator of the protein kinase activity of mTORC1 (PubMed:12906785, PubMed:15340059, PubMed:24529379). In absence of nutrients, the TSC-TBC complex inhibits mTORC1, thereby preventing phosphorylation of ribosomal protein S6 kinase (RPS6KB1 and RPS6KB2) and EIF4EBP1 (4E-BP1) by the mTORC1 signaling (PubMed:12271141, PubMed:24529379, PubMed:28215400,

PubMed:33215753). The TSC-TBC complex is inactivated in response to nutrients, relieving inhibition of mTORC1 (PubMed:12172553, PubMed:24529379). Within the TSC-TBC complex, TSC1 stabilizes TSC2 and prevents TSC2 self-aggregation (PubMed:10585443, PubMed:28215400). Acts as a tumor suppressor (PubMed:9242607). Involved in microtubule- mediated protein transport via its ability to regulate mTORC1 signaling (By similarity). Also acts as a co-chaperone for HSP90AA1 facilitating HSP90AA1 chaperoning of protein clients such as kinases, TSC2 and glucocorticoid receptor NR3C1 (PubMed:29127155). Increases ATP binding to HSP90AA1 and inhibits HSP90AA1 ATPase activity (PubMed:29127155). Competes with the activating co-chaperone AHSA1 for binding to HSP90AA1, thereby providing a reciprocal regulatory mechanism for chaperoning of client proteins (PubMed:29127155). Recruits TSC2 to HSP90AA1 and stabilizes TSC2 by preventing the interaction between TSC2 and ubiquitin ligase HERC1 (PubMed:16464865, PubMed:29127155).

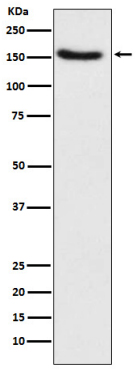
Cellular Location

Lysosome membrane; Peripheral membrane protein. Cytoplasm, cytosol Note=Recruited to lysosomal membranes in a RHEB-dependent process in absence of nutrients (PubMed:24529379). In response to nutrients, the complex dissociates from lysosomal membranes and relocalizes to the cytosol (PubMed:24529379).

Tissue Location

Highly expressed in skeletal muscle, followed by heart, brain, placenta, pancreas, lung, liver and kidney (PubMed:9242607). Also expressed in embryonic kidney cells (PubMed:9242607).

Images



Western blot analysis of Hamartin expression in HeLa cell lysate.

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