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Hamartin (TSC1) Blocking Peptide (C-term)

Synthetic peptide
Catalog # BP6359b

Specification

Hamartin (TSC1) Blocking Peptide (C-term) - Product Information

Primary Accession Q92574

Hamartin (TSC1) Blocking Peptide (C-term) - Additional Information

Gene ID 7248

Other Names
Hamartin, Tuberous sclerosis 1 protein, TSC1, KIAA0243, TSC

Target/Specificity
The synthetic peptide sequence is selected from aa 881~895 of HUMAN TSC1

Format
Peptides are lyophilized in a solid powder format. Peptides can be reconstituted in solution using the appropriate buffer as needed.

Storage
Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C.

Precautions
This product is for research use only. Not for use in diagnostic or therapeutic procedures.

Hamartin (TSC1) Blocking Peptide (C-term) - 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:12906785, PubMed:12271141, PubMed:28215400, PubMed:15340059, PubMed:24529379). 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). 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).

Hamartin (TSC1) Blocking Peptide (C-term) - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Blocking Peptides

Hamartin (TSC1) Blocking Peptide (C-term) - Images

Hamartin (TSC1) Blocking Peptide (C-term) - Background

Implicated as a tumor suppressor. May have a function in vesicular transport. Interaction between TSC1 and TSC2 may facilitate vesicular docking.
Defects in TSC1 are the cause of tuberous sclerosis complex (TSC). The molecular basis of TSC is a functional impairement of the hamartin-tuberin complex. TSC is an autosomal dominant multi-system disorder that affects especially the brain, kidneys, heart, and skin. TSC is characterized by hamartomas (benign overgrowths predominantly of a cell or tissue type that occurs normally in the organ) and hamartias (developmental abnormalities of tissue combination). Clinical symptoms can range from benign hypopigmented macules of the skin to profound mental retardation with intractable seizures to premature death from a variety of disease-associated causes.

Defects in TSC1 may be a cause of focal cortical dysplasia of Taylor balloon cell type (FCDBC). FCDBC is a subtype of cortical displasias linked to chronic intractable epilepsy. Cortical dysplasias display a broad spectrum of structural changes, which appear to result from changes in proliferation, migration, differentiation, and apoptosis of neuronal precursors and neurons during cortical development.

Hamartin (TSC1) Blocking Peptide (C-term) - References

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