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Hamartin (TSC1) Antibody (S505) Blocking peptide

Synthetic peptide
Catalog # BP6359d

Specification

Hamartin (TSC1) Antibody (S505) Blocking peptide - Product Information

Primary Accession Q92574

Hamartin (TSC1) Antibody (S505) Blocking peptide - Additional Information

Gene ID 7248

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

Target/Specificity
The synthetic peptide sequence used to generate the antibody AP6359d was selected from the S505 region of human TSC1. A 10 to 100 fold molar excess to antibody is recommended. Precise conditions should be optimized for a particular assay.

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) Antibody (S505) Blocking peptide - Protein Information

Name TSC1

Synonyms KIAA0243, TSC

Hamartin (TSC1) Antibody (S505) Blocking peptide - Background

TSC1 is implicated as a tumor suppressor and 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) Antibody (S505) Blocking peptide - References

Wu, J., et al., J. Cutan. Pathol. 31(5):383-387 (2004).Lewis, J.C., et al., J. Med. Genet. 41(3):203-207 (2004).J, et al., J. Child Neurol. 19(2):102-106 (2004).Murthy, V., et al., J. Biol. Chem. 279(2):1351-1358 (2004).Astrinidis, A., et al., J. Biol. Chem. 278(51):51372-51379 (2003).

Function

In complex with TSC2, inhibits the nutrient-mediated or growth factor-stimulated phosphorylation of S6K1 and EIF4EBP1 by negatively regulating mTORC1 signaling (PubMed:12271141, PubMed:28215400). Seems not to be required for TSC2 GAP activity towards RHEB (PubMed:15340059). Implicated as a tumor suppressor. Involved in microtubule-mediated protein transport, but this seems to be due to unregulated mTOR signaling (By similarity). 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

Cytoplasm. Membrane; Peripheral membrane protein. Note=At steady state found in association with membranes.

Tissue Location

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

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Hamartin (TSC1) Antibody (S505) Blocking peptide - Protocols

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

- Blocking Peptides