

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**Synonyms** KIAA0243, TSC**Function**In complex with TSC2, inhibits the
nutrient-mediated or growth
factor-stimulated phosphorylation of S6K1
and EIF4EBP1 by negatively regulating**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 impairment 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
dysplasias 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**

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).

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

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

Provided below are standard protocols that you

may find useful for product applications.

- [Blocking Peptides](#)