

Hamartin (TSC1) Antibody (Center)
Affinity Purified Rabbit Polyclonal Antibody (Pab)
Catalog # AP6359C

Specification

Hamartin (TSC1) Antibody (Center) - Product Information

Application	IF, WB, IHC-P,E
Primary Accession	O92574
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Isotype	Rabbit IgG
Calculated MW	129767
Antigen Region	401-430

Hamartin (TSC1) Antibody (Center) - Additional Information

Gene ID 7248

Other Names

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

Target/Specificity

This Hamartin (TSC1) antibody is generated from rabbits immunized with a KLH conjugated synthetic peptide between 401-430 amino acids from the Central region of human Hamartin (TSC1).

Dilution

IF~~1:10~50
WB~~1:1000
IHC-P~~1:10~50

Format

Purified polyclonal antibody supplied in PBS with 0.09% (W/V) sodium azide. This antibody is purified through a protein A column, followed by peptide affinity purification.

Storage

Maintain refrigerated at 2-8°C for up to 2 weeks. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

Precautions

Hamartin (TSC1) Antibody (Center) is for research use only and not for use in diagnostic or therapeutic procedures.

Hamartin (TSC1) Antibody (Center) - 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 relocates to the cytosol (PubMed:[24529379](#)).

Tissue Location

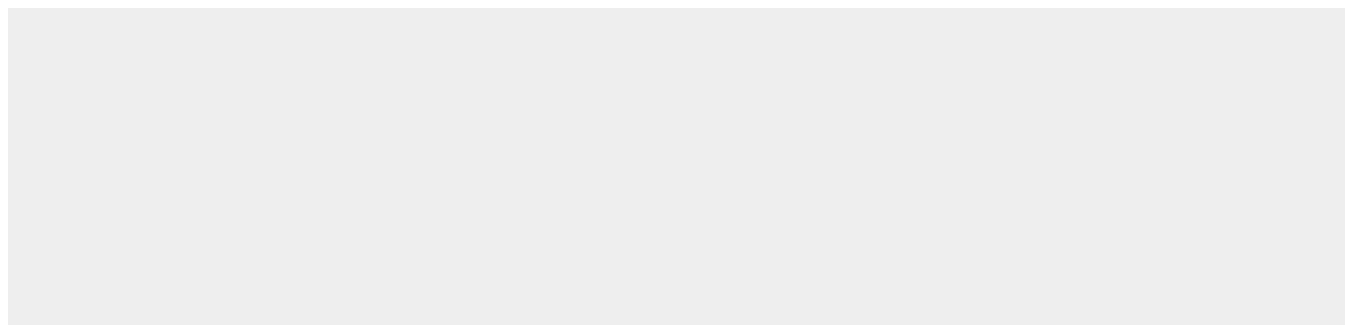
Highly expressed in skeletal muscle, followed by heart, brain, placenta, pancreas, lung, liver and kidney (PubMed:[9242607](#)). Also expressed in embryonic kidney cells (PubMed:[9242607](#)).

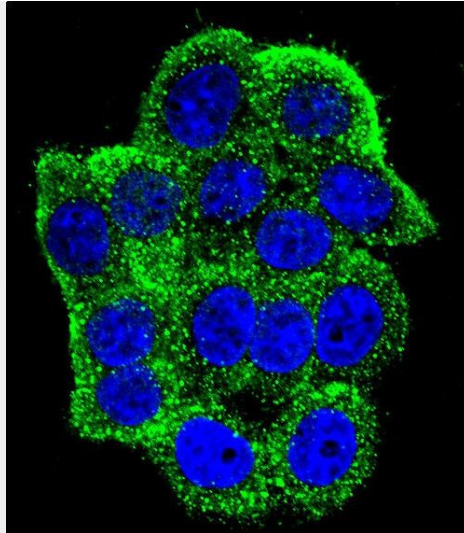
Hamartin (TSC1) Antibody (Center) - Protocols

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

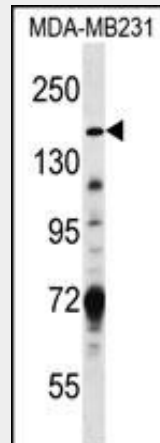
- [Western Blot](#)
- [Blocking Peptides](#)
- [Dot Blot](#)
- [Immunohistochemistry](#)
- [Immunofluorescence](#)
- [Immunoprecipitation](#)
- [Flow Cytometry](#)
- [Cell Culture](#)

Hamartin (TSC1) Antibody (Center) - Images

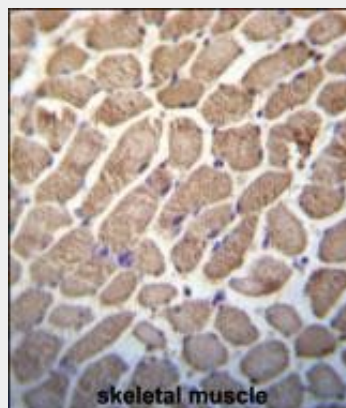




Confocal immunofluorescent analysis of Hamartin (TSC1) Antibody (Center)(Cat#AP6359c) with Hela cell followed by Alexa Fluor 488-conjugated goat anti-rabbit IgG (green). DAPI was used to stain the cell nuclear (blue).



TSC1 Antibody (Center) (Cat. #AP6359c) western blot analysis in MDA-MB231 cell line lysates (35ug/lane). This demonstrates the TSC1 antibody detected the TSC1 protein (arrow).



Hamartin (TSC1) Antibody (Center) (Cat. #AP6359c) immunohistochemistry analysis in formalin fixed and paraffin embedded human skeletal muscle followed by peroxidase conjugation of the secondary antibody and DAB staining. This data demonstrates the use of Hamartin (TSC1) Antibody (Center) for immunohistochemistry. Clinical relevance has not been evaluated.

Hamartin (TSC1) Antibody (Center) - 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 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 (Center) - 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).