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phospho-TSC1 (Ser505) Rabbit pAb

Catalog # AP99115

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q92574
Other Accession	Q92574
Reactivity	Human, Mouse
Predicted	Rat, Pig, Rabbit, Chicken, Dog, Horse
Host	Rabbit
Clonality	Polyclonal
Calculated MW	129767
Physical State	Liquid
Immunogen	KLH conjugated Synthesised phosphopeptide derived from human TSC1 around the phosphorylation site of Ser505
Epitope Specificity	FD(p-S)PF
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Contains 1 RRM (RNA recognition motif) domain.
SUBUNIT	Interacts with TSC2, leading to stabilize TSC2. In the absence of TSC2, TSC1 self-aggregates. Interacts with DOCK7. Interacts with FBXW5 and TBC1D7.
Post-translational modifications	Phosphorylation at Ser-505 does not affect interaction with TSC2.
DISEASE	Tuberous sclerosis 1 (TSC1) [MIM:191100]: An autosomal dominant multi-system disorder that affects especially the brain, kidneys, heart, and skin. It 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 manifestations include epilepsy, learning difficulties, behavioral problems, and skin lesions. Seizures can be intractable and premature death can occur from a variety of disease-associated causes. Note=The disease is caused by mutations affecting the gene represented in this entry. Focal cortical dysplasia of Taylor balloon cell type (FCDBC) [MIM:607341]: 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. Note=The disease may be caused by mutations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Tuberous sclerosis complex (TSC) is an autosomal dominant genetic disorder characterized by mental retardation and the widespread development of distinctive tumors termed hamartomas. Two different genetic loci have been linked to TSC; one of these loci, the tuberous sclerosis-2 gene (TSC2), encodes a protein called tuberin and the other loci, tuberous sclerosis-1 gene (TSC1), encodes a protein called hamartin. Tuberin and hamartin interact with each other forming a cystoplasmic complex. Hamartin interacts with the ezrin-radixin-moesin (ERM) family of actin-binding proteins and inhibition of

hamartin activity results in loss of cell adhesion. Hamartin is present in most adult tissues with strong expression in brain, heart, and kidney.

Additional Information

Gene ID 7248

Other Names LAM; TSC; TSC1_HUMAN; TSC1; Tuberous sclerosis 1 protein; KIAA0243; TSC1_MOUSE; Tuberous sclerosis 1 protein homolog; TSC1_RAT; TSC complex subunit 1; tuberous sclerosis 1; hamartin; TSC1 (phospho-S505); p-TSC1; phospho-TSC1; TSC1 (phospho-Ser505)

Dilution WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

Storage Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.

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).

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