



## Rabbit Anti-Phospho-Tuberin (Ser664) antibody

SL5611R

<b>Product Name:</b>	Phospho-Tuberin (Ser664)
<b>Chinese Name:</b>	磷酸化结节性硬化蛋白抗体
<b>Alias:</b>	TSC2(phospho S664);FLJ43106; LAM; TSC2; TSC2_HUMAN; TSC4; Tuberin; Tuberous sclerosis 2 protein.
<b>Organism Species:</b>	Rabbit
<b>Clonality:</b>	Polyclonal
<b>React Species:</b>	Human,Mouse,Rat,Dog,Pig,Horse,
<b>Applications:</b>	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500 (Paraffin sections need antigen repair) not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
<b>Molecular weight:</b>	200kDa
<b>Cellular localization:</b>	cytoplasmicThe cell membrane
<b>Form:</b>	Lyophilized or Liquid
<b>Concentration:</b>	1mg/ml
<b>immunogen:</b>	KLH conjugated Synthesised phosphopeptide derived from human Tuberin around the phosphorylation site of Ser664:PL(p-S)PP
<b>Lsotype:</b>	IgG
<b>Purification:</b>	affinity purified by Protein A
<b>Storage Buffer:</b>	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
<b>Storage:</b>	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. 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.
<b>PubMed:</b>	<a href="#">PubMed</a>
<b>Product Detail:</b>	Tuberin, or TSC2 (Tuberous sclerosis complex), is implicated as a tumor suppressor. It may function in vesicular transport, and may also play a role in the regulation of cell growth arrest and in the regulation of transcription mediated by steroid receptors.

Interaction between hamartin (TSC1) and tuberin may facilitate vesicular docking. It specifically stimulates the intrinsic GTPase activity of the Ras related protein RAP1A and RAB5, suggesting a possible mechanism for its role in regulating cellular growth. Mutations in tuberin lead to constitutive activation of RAP1A in tumors. At least three isoforms of Tuberin exist.

**Function:**

E3 ubiquitin-protein ligase that mediates the polyubiquitination of a number of proteins such as CD3D, CYP3A4, CFTR and APOB for proteasomal degradation. Component of a VCP/p97-AMFR/gp78 complex that participates in the final step of endoplasmic reticulum-associated degradation (ERAD). The VCP/p97-AMFR/gp78 complex is involved in the sterol-accelerated ERAD degradation of HMGCR through binding to the HMGCR-INSIG complex at the ER membrane and initiating ubiquitination of HMGCR. The ubiquitinated HMGCR is then released from the ER by the complex into the cytosol for subsequent destruction. Also acts as a scaffold protein to assemble a complex that couples ubiquitination, retranslocation and deglycosylation. Mediates tumor invasion and metastasis.

**Subunit:**

Interacts with RNF5. Also forms an ERAD complex containing VCP/p97, NGLY1; PSMC1; SAKS1 AND RAD23B required for coupling retrotranslocation, ubiquitination and deglycosylation (By similarity). Interacts with DRL1. Interacts (through a region distinct from the RING finger) with UBE2G2/UBC7. Component of the VCP/p97-AMFR/gp78 complex that enhances VCP/p97 binding to polyubiquitinated proteins for their degradation by the endoplasmic reticulum-associated degradation (ERAD) pathway. Interacts (via the VIM) with VCP/p97. Interacts (via its membrane domain) with INSIG1; the interaction initiates the sterol-mediated ubiquitination and degradation of HMGCR by the ERAD pathway.

**Subcellular Location:**

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

**Tissue Specificity:**

Liver, brain, heart, lymphocytes, fibroblasts, biliary epithelium, pancreas, skeletal muscle, kidney, lung and placenta.

**Post-translational modifications:**

Phosphorylation at Ser-1387, Ser-1418 or Ser-1420 does not affect interaction with TSC1. Phosphorylation at Ser-939 and Thr-1462 by PKB/AKT1 is induced by growth factor stimulation. Phosphorylation by AMPK activates it and leads to negatively regulates the mTORC1 complex. Phosphorylated at Ser-1798 by RPS6KA1; phosphorylation inhibits TSC2 ability to suppress mTORC1 signaling. Phosphorylated by DAPK1.

**DISEASE:**

Defects in TSC2 are the cause of tuberous sclerosis type 2 (TSC2) [MIM:613254]. TSC2 is 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 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 TSC2 are a cause of lymphangiomyomatosis (LAM) [MIM:606690]. LAM is a progressive and often fatal lung disease characterized by a diffuse proliferation of abnormal smooth muscle cells in the lungs. It affects almost exclusively young women and can occur as an isolated disorder or in association with tuberous sclerosis complex.

**Similarity:**

Contains 1 Rap-GAP domain.

**SWISS:**

P49815

**Gene ID:**

7249

**Database links:**

[Entrez Gene: 7249](#)Human

[Entrez Gene: 22084](#)Mouse

[Oimim: 191092](#)Human

[SwissProt: P49815](#)Human

[SwissProt: Q61037](#)Mouse

[Unigene: 90303](#)Human

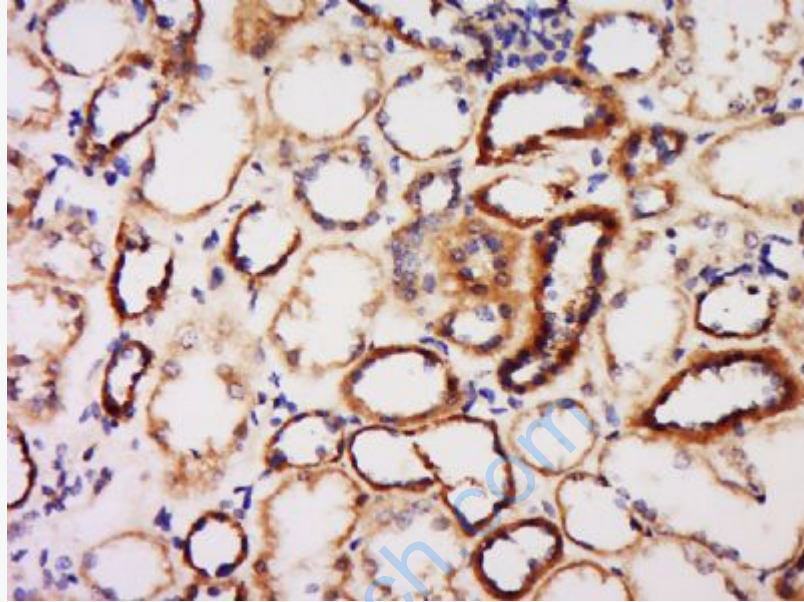
[Unigene: 30435](#)Mouse

**Important Note:**

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

Tuberin 为180kD-220的蛋白产生于结节性硬化(TSC-2)位于染色体16的基因。结节性硬化为常染色体疾病已知为斑痣性错构瘤病其特征为广泛性发展呈良性生长,在许多组织与器官中描述为错构瘤。Tuberin被认为在GTPase活动性蛋白中起作用调节细胞内吞与作为Tumour抑制子。Tuberin低水平广泛性表达在大多数组织中,而在皮层神经元、小脑浦肯野氏细胞、脊索运动神经元、

胰岛B细胞、心肌、肾脏与皮肤的小血管中表达增加。



**Picture:**

Tissue/cell: human kidney tissue; 4% Paraformaldehyde-fixed and paraffin-embedded;

Antigen retrieval: citrate buffer ( 0.01M, pH 6.0 ), Boiling bathing for 15min; Block endogenous peroxidase by 3% Hydrogen peroxide for 30min; Blocking buffer (normal goat serum,C-0005) at 37°C for 20 min;

Incubation: Anti-TSC2 Polyclonal Antibody, Unconjugated(SL5611R) 1:500, overnight at 4°C, followed by conjugation to the secondary antibody(SP-0023) and DAB(C-0010) staining