

# hamartin (A-2): sc-514817

## BACKGROUND

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 cytoplasmic 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.

## REFERENCES

1. The European Chromosome 16 Tuberous Sclerosis Consortium. 1993. Identification and characterization of the tuberous sclerosis gene on chromosome 16. *Cell* 75: 1305-1315.
2. van Slegtenhorst, M., et al. 1997. Identification of the tuberous sclerosis gene TSC1 on chromosome 9q34. *Science* 277: 805-808.
3. Young, J. and Povey, S. 1998. The genetic basis of tuberous sclerosis. *Mol. Med. Today* 4: 313-319.
4. Plank, T.L., et al. 1998. Hamartin, the product of the tuberous sclerosis 1 (TSC1) gene, interacts with tuberin and appears to be localized to cytoplasmic vesicles. *Cancer Res.* 58: 4766-4770.
5. Nellist, M., et al. 1999. Characterization of the cytosolic tuberin-hamartin complex. Tuberin is a cytosolic chaperone for hamartin. *J. Biol. Chem.* 274: 35647-35652.
6. Plank, T.L., et al. 1999. The expression of hamartin, the product of the TSC1 gene, in normal human tissues and in TSC1- and TSC2-linked angiomyolipomas. *Mod. Pathol.* 12: 539-545.
7. Lamb, R.F., et al. 2000. The TSC1 tumour suppressor hamartin regulates cell adhesion through ERM proteins and the GTPase Rho. *Nat. Cell Biol.* 2: 281-287.

## CHROMOSOMAL LOCATION

Genetic locus: TSC1 (human) mapping to 9q34.13; Tsc1 (mouse) mapping to 2 A3.

## SOURCE

hamartin (A-2) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 1136-1164 at the C-terminus of hamartin of human origin.

## PRODUCT

Each vial contains 200 µg IgG<sub>2a</sub> kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-514817 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

## APPLICATIONS

hamartin (A-2) is recommended for detection of hamartin of mouse, rat and human origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 µg per 100-500 µg of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for hamartin siRNA (h): sc-37437, hamartin siRNA (m): sc-37438, hamartin shRNA Plasmid (h): sc-37437-SH, hamartin shRNA Plasmid (m): sc-37438-SH, hamartin shRNA (h) Lentiviral Particles: sc-37437-V and hamartin shRNA (m) Lentiviral Particles: sc-37438-V.

Molecular Weight of hamartin: 130 kDa.

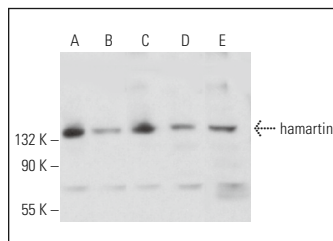
Positive Controls: HeLa whole cell lysate: sc-2200, IMR-32 cell lysate: sc-2409 or WI-38 whole cell lysate: sc-364260.

## RECOMMENDED SUPPORT REAGENTS

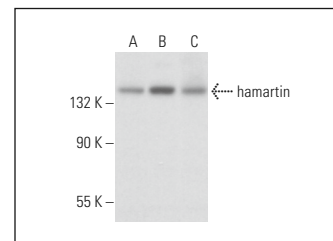
To ensure optimal results, the following support reagents are recommended:

- 1) Western Blotting: use m-IgGκ BP-HRP: sc-516102 or m-IgGκ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker™ Molecular Weight Standards: sc-2035, UltraCruz® Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048.
- 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml).
- 3) Immunofluorescence: use m-IgGκ BP-FITC: sc-516140 or m-IgGκ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz® Mounting Medium: sc-24941 or UltraCruz® Hard-set Mounting Medium: sc-359850.

## DATA



hamartin (A-2): sc-514817. Western blot analysis of hamartin expression in HeLa (A), NIH/3T3 (B), SK-MEL-24 (C) and C6 (D) whole cell lysates and mouse brain tissue extract (E).



hamartin (A-2): sc-514817. Western blot analysis of hamartin expression in SJRH30 (A), IMR-32 (B) and WI-38 (C) whole cell lysates.

## SELECT PRODUCT CITATIONS

1. Liu, W., et al. 2021. The expression of TRIM6 activates the mTORC1 pathway by regulating the ubiquitination of TSC1-TSC2 to promote renal fibrosis. *Front. Cell Dev. Biol.* 8: 616747.

## STORAGE

Store at 4° C, \*\*DO NOT FREEZE\*\*. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

## RESEARCH USE

For research use only, not for use in diagnostic procedures.