



Qty: 100 µg/200 µl

Mouse anti-Hamartin

Catalog No. 37-0400

Lot No.

Mouse anti-Hamartin

FORM

This monoclonal antibody is supplied as a 200 µl aliquot at a concentration of 0.5 mg/ml in PBS, pH 7.4, containing 0.1% sodium azide. This antibody is highly purified from mouse ascites by protein A chromatography.

CLONE: 5C8A12

ISOTYPE: IgG₁

IMMUNOGEN

Recombinant human Hamartin protein

SPECIFICITY

This antibody is specific for the ~130-150 kDa Hamartin protein, and shows no reactivity with the related Tuberin protein. This specificity was confirmed with Western blot testing on both Hamartin- and Tuberin-transfected cell lysates.

REACTIVITY

Reactivity was confirmed with a Hamartin-transfected 293 cell line.

Sample	ELISA	Immunoprecipitation (Native)	Western Blotting
Human	ND	+++	+++
Immunogen	+++	ND	+++

(Excellent +++, Good++, Poor +, No reactivity 0, Not applicable N/A, Not Determined ND)

USAGE

Working concentrations for specific applications should be determined by the investigator. Appropriate concentrations will be affected by several factors, including secondary antibody affinity, antigen concentration, sensitivity of detection method, temperature and length of incubations, etc. The suitability of this antibody for applications other than those listed below has not been determined. The following concentration ranges are recommended starting points for this product.

ELISA: 0.1-1 µg/ml

Western Blotting: 1-3 µg/ml

Immunoprecipitation: 5-10 µg/ml

STORAGE

Store at 2-8°C for up to one month. Store at -20°C for long-term storage. Avoid repeated freezing and thawing.

(cont'd)

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PI370400

(Rev 10/08) DCC-08-1089

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BACKGROUND

Tuberous sclerosis (TSC) is an autosomal dominant disease caused by mutations in one of two tumor-suppressor genes, TSC1 and TSC2, resulting in seizures, developmental delays, behavioral problems, benign hamartomas and low grade neoplasms predominantly affecting the central nervous system, kidney, skin, lung, and heart. The dysfunction of either gene products of TSC1 and TSC2, hamartin and tuberlin, respectively, may cause the appearance of these tumors.

Hamartin and tuberlin can interact with each other to form a cytoplasmic complex. When bound, hamartin is able to inhibit the ubiquitination of tuberlin, and thus, increasing the cellular level of tuberlin and inhibiting cell growth.¹ Hamartin also interacts with the ezrin-radixin-moesin family of actin-binding proteins. Hamartin affects cell proliferation by deregulating the G1 phase of the cell cycle.² Inhibition of hamartin activity results in the loss of cell adhesion.

Expression studies has shown that hamartin is co-localized with tuberlin in most tissues, including the liver, small and large intestines, prostate, testes, and pancreas.³ However, in the central nervous system, hamartin is mainly localized along neuronal or astrocytic processes, as opposed to tuberlin, which is localized to the perinuclear region of the Purkinje cell.⁴ These studies indicate that hamartin may be important for astrocyte growth control.

REFERENCES

1. Benvenuto G, et al. *Oncogene*. 19(54): 6306-16, 2000.
2. Miloloza A, et al. *Hum Mol Genet*. 9(12): 1721-7, 2000.
3. Johnson MW, et al. *Mod Pathol* 14(3): 202-210, 2001.
4. Gutmann DH, et al. *Acta Neuropathol*. 99(3): 223-30, 2000.

RELATED PRODUCTS

Product	Clone/PAD*	Cat. No.
Mouse anti-Tuberlin	3G9D9	37-0500
Mouse anti-Ezrin	3C12	35-7300
Mouse anti-Actin	ZSA1	03-3100
Protein A	Sepharose® 4B	10-1041
rec-Protein G	Sepharose® 4B	10-1241

*PAD: Polyclonal Antibody Designation

Conjugate	ZyMAX™ Goat x Rabbit IgG (H+L)	ZyMAX™ Goat x Mouse IgG (H+L)
Purified	81-6100	81-6500
FITC	81-6111	81-6511
TRITC	81-6114	81-6514
Cy™3	81-6115	81-6515
Cy™5	81-6116	81-6516
HRP	81-6120	81-6520
AP	81-6122	81-6522
Biotin	81-6140	81-6540

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