abcam

Product datasheet

Anti-Hamartin antibody ab25882

2 References 2 图像

概述

产品名称	Anti-Hamartin抗体
描述	**兔多克隆抗体**to Hamartin
宿主	Rabbit
特异性	This antibody will recognise both isoforms of TSC1.
经测试应用	**适用于:** ICC/IF, WB
种属反应性	**与反应:** Mouse
免疫原	Synthetic peptide - a 15 amino acid peptide from the middle region of human TSC1 (Human). Peptide available as **ab95277**.
阳性对照	EL4 cell lysate can be used as positive control
常规说明	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, along with publications, customer reviews and Q&As

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at +4°C.
存储溶液	pH: 7.2 Preservative: 0.02% Sodium azide Constituent: PBS
纯度	Protein A purified
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab25882于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
ICC/IF		Use a concentration of 2 µg/ml.
WB		Use a concentration of 1 µg/ml. Detects a band of approximately 130 kDa (predicted molecular weight: 130 kDa).

靶标

功能 In complex with TSC2, inhibits the nutrient-mediated or growth factor-stimulated phosphorylation of S6K1 and EIF4EBP1 by negatively regulating mTORC1 signaling. Seems not to be required for TSC2 GAP activity towards RHEB. Implicated as a tumor suppressor. Involved in microtubule-mediated protein transport, but this seems to be due to unregulated mTOR signaling.

组织特异性 Highly expressed in skeletal muscle, followed by heart, brain, placenta, pancreas, lung, liver and kidney. Also expressed in embryonic kidney cells.

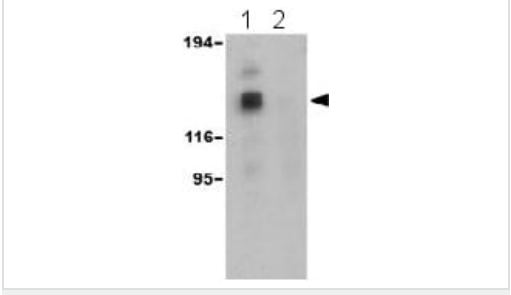
疾病相关 Defects in TSC1 are the cause of tuberous sclerosis type 1 (TSC1) [MIM:191100]. It is an autosomal dominant multi-system disorder that affects especially the brain, kidneys, heart, and skin. TS1C is characterized by hamartomas (benign overgrowths predominantly of a cell or tissue type that occurs 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) [MIM:607341]. 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.

结构域 The C-terminal putative coiled-coil domain is necessary for interaction with TSC2.

翻译后修饰 Phosphorylation at Ser-505 does not affect interaction with TSC2. Phosphorylated upon DNA damage, probably by ATM or ATR.

细胞定位 Cytoplasm. Membrane. At steady state found in association with membranes.

图片

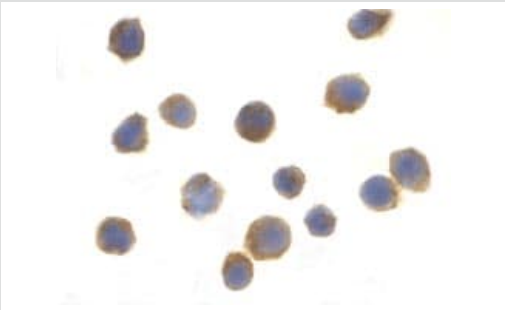


Western blot - Anti-Hamartin antibody (ab25882)

All lanes : Anti-Hamartin antibody (ab25882) at 1 µg/ml

Lane 1 : EL4 cell lysate with absence of blocking peptide
Lane 2 : EL4 cell lysate with presence of blocking peptide

Predicted band size: 130 kDa
Observed band size: 130 kDa



Immunocytochemistry/ Immunofluorescence - Anti-Hamartin antibody (ab25882)

ab25882 at 2µg/ml staining Hamartin in EL4 (mouse) cells by ICC/IF

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