产品名称：Hamartin Rabbit Polyclonal Antibody
产品货号：APRab11891

产品概述（Summary）

产品名称（Production Name）	Hamartin Rabbit Polyclonal Antibody
描述（Description）	Rabbit polyclonal Antibody
宿主（Host）	Rabbit
应用（Application）	WB,ELISA
种属反应性（Reactivity）	Human,Rat,Mouse

产品性能（Performance）

偶联物（Conjugation）	Unconjugated
修饰（Modification）	Unmodified
同种型（Isotype）	IgG
克隆（Clonality）	Polyclonal
形式（Form）	Liquid
存放说明（Storage）	Store at 4°C short term. Aliquot and store at -20°C long term. Avoid freeze/thaw cycles.
储存溶液（Buffer）	Liquid in PBS containing 50% glycerol, 0.5% protective protein and 0.02% New type preservative N.
纯化方式（Purification）	Affinity purification

免疫原信息（Immunogen）

基因名（Gene Name）	TSC1 KIAA0243 TSC
别名（Alternative Names）	tuberous sclerosis 1
基因 ID（Gene ID）	7248.0
蛋白 ID（SwissProt ID）	Q92574.Synthesized peptide derived from Hamartin . at AA range: 360-440

产品应用（Application）

稀释比（Dilution Ratio）	WB 1:500-1:2000,ELISA 1:10000-1:20000
蛋白分子量（Molecular Weight）	130kDa

研究背景（Background）

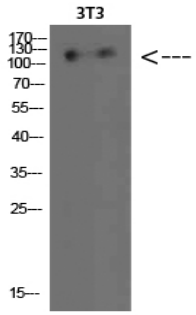
This gene encodes a growth inhibitory protein thought to play a role in the stabilization of tuberin. Mutations in this gene

have been associated with tuberous sclerosis. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jun 2009],disease:Defects in TSC1 are the cause of tuberous sclerosis complex (TSC) [MIM:191100]. The molecular basis of TSC is a functional impairement of the hamartin-tuberin complex. TSC is an autosomal dominant multi-system disorder that affects especially the brain, kidneys, heart, and skin. TSC 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.,disease: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.,domain:The C-terminal putative coiled-coil domain is necessary for interaction with TSC2.,function:Implicated as a tumor suppressor. May have a function in vesicular transport. Interaction between TSC1 and TSC2 may facilitate vesicular docking.,PTM:Phosphorylated upon DNA damage, probably by ATM or ATR.,PTM:Phosphorylation at Ser-505 does not affect interaction with TSC2.,subcellular location:At steady state found in association with membranes.,subunit:Interacts with TSC2, leading to stabilize TSC2. In the absence of TSC2, TSC1 self-aggregates. Interacts with DOCK7.,tissue specificity:Highly expressed in skeletal muscle, followed by heart, brain, placenta, pancreas, lung, liver and kidney. Also expressed in embryonic kidney cells.,

研究领域（Research Area）

mTOR;Insulin_Receptor;

图片（Image Data）



Western Blot analysis of 3T3 cells using Hamartin Polyclonal Antibody diluted at 1:500. Secondary antibody was diluted at 1:20000

注意事项（Note）

For research use only .