

Anti-Filamin A (phospho S1458) antibody [EP2309Y] ab68424

RabMAb

2 图像

概述

产品名称	Anti-Filamin A (phospho S1458)抗体[EP2309Y]
描述	兔单克隆抗体[EP2309Y] to Filamin A (phospho S1458)
宿主	Rabbit
经测试应用	适用于: WB, IHC-P 不适用于: Flow Cyt or IP
种属反应性	与反应: Human
免疫原	Synthetic peptide. This information is proprietary to Abcam and/or its suppliers.
阳性对照	WB: HeLa cells. IHC-P: Human stomach and uterus tissue.
常规说明	<p>Our RabMAb[®] technology is a patented hybridoma-based technology for making rabbit monoclonal antibodies. For details on our patents, please refer to RabMAb[®] patents.</p> <p>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.</p> <p>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</p> <p>Mouse, Rat: We have preliminary internal testing data to indicate this antibody may not react with these species. Please contact us for more information.</p>

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
存储溶液	pH: 7.20 Preservative: 0.05% Sodium azide Constituents: 0.1% BSA, 40% Glycerol (glycerin, glycerine), 9.85% Tris glycine, 50% Tissue culture supernatant
纯度	Tissue culture supernatant

克隆	单克隆
克隆编号	EP2309Y
同种型	IgG

应用

The Abpromise guarantee **Abpromise™**承诺保证使用ab68424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
WB		1/1000 - 1/5000. Detects a band of approximately 260 kDa (predicted molecular weight: 280 kDa).
IHC-P		1/50 - 1/100. Perform heat mediated antigen retrieval before commencing with IHC staining protocol.

应用说明 Is unsuitable for Flow Cyt or IP.

靶标

功能 Promotes orthogonal branching of actin filaments and links actin filaments to membrane glycoproteins. Anchors various transmembrane proteins to the actin cytoskeleton and serves as a scaffold for a wide range of cytoplasmic signaling proteins. Interaction with FLNA may allow neuroblast migration from the ventricular zone into the cortical plate. Tethers cell surface-localized furin, modulates its rate of internalization and directs its intracellular trafficking.

组织特异性 Ubiquitous.

疾病相关 Defects in FLNA are the cause of periventricular nodular heterotopia type 1 (PVNH1) [MIM:300049]; also called nodular heterotopia, bilateral periventricular (NHBP or BPNH). PVNH is a developmental disorder characterized by the presence of periventricular nodules of cerebral gray matter, resulting from a failure of neurons to migrate normally from the lateral ventricular proliferative zone, where they are formed, to the cerebral cortex. PVNH1 is an X-linked dominant form. Heterozygous females have normal intelligence but suffer from seizures and various manifestations outside the central nervous system, especially related to the vascular system. Hemizygous affected males die in the prenatal or perinatal period.

Defects in FLNA are the cause of periventricular nodular heterotopia type 4 (PVNH4) [MIM:300537]; also known as periventricular heterotopia Ehlers-Danlos variant. PVNH4 is characterized by nodular brain heterotopia, joint hypermobility and development of aortic dilation in early adulthood.

Defects in FLNA are the cause of otopalatodigital syndrome type 1 (OPD1) [MIM:311300]. OPD1 is an X-linked dominant multiple congenital anomalies disease mainly characterized by a generalized skeletal dysplasia, mild mental retardation, hearing loss, cleft palate, and typical facial anomalies. OPD1 belongs to a group of X-linked skeletal dysplasias known as oto-palatodigital syndrome spectrum disorders that also include OPD2, Melnick-Needles syndrome (MNS), and frontometaphyseal dysplasia (FMD). Remodeling of the cytoskeleton is central to the modulation of cell shape and migration. FLNA is a widely expressed protein that regulates re-organization of the actin cytoskeleton by interacting with integrins, transmembrane receptor complexes and second messengers. Males with OPD1 have cleft palate, malformations of the ossicles causing deafness and milder bone and limb defects than those associated with OPD2.

Obligate female carriers of mutations causing both OPD1 and OPD2 have variable (often milder) expression of a similar phenotypic spectrum.

Defects in FLNA are the cause of otopalatodigital syndrome type 2 (OPD2) [MIM:304120]; also known as cranioorodigital syndrome. OPD2 is a congenital bone disorder that is characterized by abnormally modeled, bowed bones, small or absent first digits and, more variably, cleft palate, posterior fossa brain anomalies, omphalocele and cardiac defects.

Defects in FLNA are the cause of frontometaphyseal dysplasia (FMD) [MIM:305620]. FMD is a congenital bone disease characterized by supraorbital hyperostosis, deafness and digital anomalies.

Defects in FLNA are the cause of Melnick-Needles syndrome (MNS) [MIM:309350]. MNS is a severe congenital bone disorder characterized by typical facies (exophthalmos, full cheeks, micrognathia and malalignment of teeth), flaring of the metaphyses of long bones, s-like curvature of bones of legs, irregular constrictions in the ribs, and sclerosis of base of skull.

Defects in FLNA are the cause of X-linked congenital idiopathic intestinal pseudoobstruction (CIIPX) [MIM:300048]. CIIPX is characterized by a severe abnormality of gastrointestinal motility due to primary qualitative defects of enteric ganglia and nerve fibers. Affected individuals manifest recurrent signs of intestinal obstruction in the absence of any mechanical lesion.

Defects in FLNA are the cause of FG syndrome type 2 (FGS2) [MIM:300321]. FG syndrome (FGS) is an X-linked disorder characterized by mental retardation, relative macrocephaly, hypotonia and constipation.

Defects in FLNA are the cause of terminal osseous dysplasia (TOD) [MIM:300244]. A rare X-linked dominant male-lethal disease characterized by skeletal dysplasia of the limbs, pigmentary defects of the skin and recurrent digital fibroma during infancy. A significant phenotypic variability is observed in affected females.

Defects in FLNA are the cause of cardiac valvular dysplasia X-linked (CVDX) [MIM:314400]. A rare X-linked heart disease characterized by mitral and/or aortic valve regurgitation. The histologic features include fragmentation of collagenous bundles within the valve fibrosa and accumulation of proteoglycans, which produces excessive valve tissue leading to billowing of the valve leaflets.

序列相似性

Belongs to the filamin family.
Contains 1 actin-binding domain.
Contains 2 CH (calponin-homology) domains.
Contains 24 filamin repeats.

结构域

Comprised of a NH2-terminal actin-binding domain, 24 internally homologous repeats and two hinge regions. Repeat 24 and the second hinge domain are important for dimer formation.

翻译后修饰

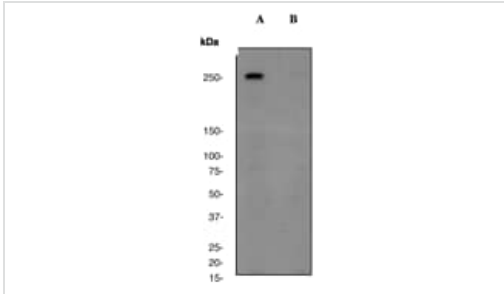
Phosphorylated upon DNA damage, probably by ATM or ATR (By similarity). Phosphorylation extent changes in response to cell activation.

The N-terminus is blocked.

细胞定位

Cytoplasm > cell cortex. Cytoplasm > cytoskeleton.

图片



Western blot - Anti-Filamin A (phospho S1458) antibody [EP2309Y] (ab68424)

All lanes : Anti-Filamin A (phospho S1458) antibody [EP2309Y] (ab68424) at 1/5000 dilution

Lane 1 : HeLa cell lysate - untreated

Lane 2 : HeLa cell lysate - treated with AP

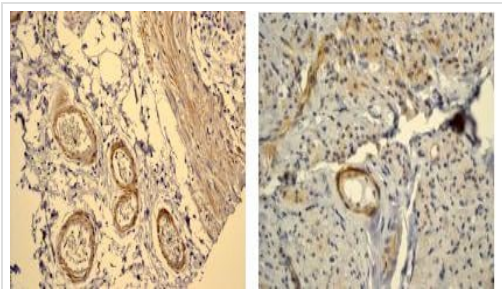
Lysates/proteins at 10 µg per lane.

Secondary

All lanes : HRP-labelled Goat anti-Rabbit antibody at 1/2000 dilution

Predicted band size: 280 kDa

Observed band size: 260 kDa



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Filamin A (phospho S1458) antibody [EP2309Y] (ab68424)

Immunohistochemical analysis of paraffin-embedded human stomach (left) and human uterus tissue (right) using ab68424 at 1/10 dilution. Heat-mediated antigen retrieval was performed prior to commencing with IHC protocol and an HRP-conjugated polymerized secondary antibody was used.

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