

# Anti-Filamin A antibody ab51217

★★★★★ [8 Abreviews](#) [13 References](#) [画像数 1](#)

### 製品の概要

製品名	Anti-Filamin A antibody
製品の詳細	Rabbit polyclonal to Filamin A
由来種	Rabbit
アプリケーション	<b>適用あり:</b> WB
種交差性	<b>交差種:</b> Human
免疫原	A synthesized non-phosphopeptide derived from human Filamin A around the phosphorylation site of serine 2152 (A-P-S <sup>P</sup> -V-A).
特記事項	<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&amp;As</p>

### 製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
バッファー	<p>pH: 7.40</p> <p>Preservative: 0.02% Sodium azide</p> <p>Constituents: 50% Glycerol (glycerin, glycerine), 0.87% Sodium chloride, PBS</p>
精製度	Immunogen affinity purified
特記事項(精製)	The antibody was affinity-purified from rabbit antiserum by affinity-chromatography using epitope-specific immunogen.
ポリ/モノ	ポリクローナル
アイソタイプ	IgG

### アプリケーション

## The Abpromise guarantee

**Abpromise保証は、次のテスト済みアプリケーションにおけるab51217の使用に適用されます**

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
WB	★★★★★ (4)	1/300 - 1/1000. Detects a band of approximately 281 kDa (predicted molecular weight: 281 kDa).

## ターゲット情報

### 機能

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.

### 組織特異性

Ubiquitous.

### 関連疾患

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-palato-digital 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.

#### 配列類似性

Belongs to the filamin family.

Contains 1 actin-binding domain.

Contains 2 CH (calponin-homology) domains.

Contains 24 filamin repeats.

#### ドメイン

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.

#### 翻訳後修飾

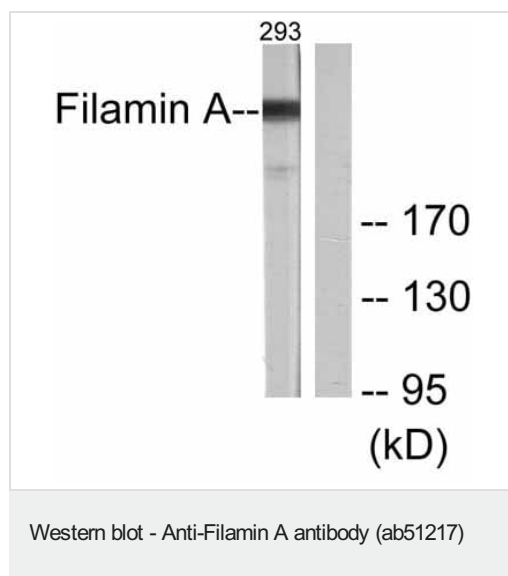
Phosphorylated upon DNA damage, probably by ATM or ATR (By similarity). Phosphorylation extent changes in response to cell activation.

The N-terminus is blocked.

#### 細胞内局在

Cytoplasm > cell cortex. Cytoplasm > cytoskeleton.

#### 画像



**All lanes :** Anti-Filamin A antibody (ab51217) at 1/300 dilution

**Lane 1 :** Extracts of 293 cells treated with EGF (200ng/ml, 5mins).

**Lane 2 :** Extracts of 293 cells treated with EGF (200ng/ml, 5mins) and peptide.

**Predicted band size:** 281 kDa

**Observed band size:** 281 kDa

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