abcam

Product datasheet

Recombinant Human Filamin B protein ab158452

1 图像

描述

产品名称	重组人Filamin B蛋白
表达系统	Wheat germ
Accession	O75369
蛋白长度	Protein fragment
无动物成分	No
性质	Recombinant
种属	Human
序列	SAIPKASSDASKVTSKGAGLSKAFVGQKSSFLVDCSKAGSNM LLIGVHGP TTPCEEVSMKHVGNQQYNVTYVVKERGDYVLAVKWGEEHIPG SPFHVTVP
氨基酸	2503 to 2602
标签	GST tag N-Terminus

技术指标

Our **Abpromise guarantee** covers the use of **ab158452** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

应用	Western blot ELISA
形式	Liquid
补充说明	

制备和贮存

稳定性和存储	Shipped on dry ice. Upon delivery aliquot and store at -80ºC. Avoid freeze / thaw cycles. pH: 8.00 Constituents: 0.31% Glutathione, 0.79% Tris HCl

常规信息

功能

Connects cell membrane constituents to the actin cytoskeleton. May promote orthogonal branching of actin filaments and links actin filaments to membrane glycoproteins. Anchors various transmembrane proteins to the actin cytoskeleton. Interaction with FLNA may allow neuroblast migration from the ventricular zone into the cortical plate. Various interactions and localizations of isoforms affect myotube morphology and myogenesis. Isoform 6 accelerates muscle differentiation in vitro.

组织特异性

Ubiquitous. Isoform 1 and isoform 2 are expressed in placenta, bone marrow, brain, umbilical vein endothelial cells (HUVEC), retina and skeletal muscle. Isoform 1 is predominantly expressed in prostate, uterus, liver, thyroid, stomach, lymph node, small intestine, spleen, skeletal muscle, kidney, placenta, pancreas, heart, lung, platelets, endothelial cells, megakaryocytic and erythroleukemic cell lines. Isoform 2 is predominantly expressed in spinal cord, platelet and Daudi cells. Also expressed in thyroid adenoma, neurofibrillary tangles (NFT), senile plaques in the hippocampus and cerebral cortex in Alzheimer disease (AD). Isoform 3 and isoform 6 are expressed predominantly in lung, heart, skeletal muscle, testis, spleen, thymus and leukocytes. Isoform 4 and isoform 5 are expressed in heart.

疾病相关

Note=Interaction with FLNA may compensate for dysfunctional FLNA homodimer in the periventricular nodular heterotopia (PVNH) disorder.
Defects in FLNB are the cause of atelosteogenesis type 1 (AO1) [MIM:108720]; also known as giant cell chondrodysplasia or spondylohumerofemoral hypoplasia. Atelosteogenesis are lethal short-limb skeletal dysplasias with vertebral abnormalities, disharmonious skeletal maturation, poorly modeled long bones and joint dislocations.
Defects in FLNB are the cause of atelosteogenesis type 3 (AO3) [MIM:108721]. Atelosteogenesis are short-limb lethal skeletal dysplasias with vertebral abnormalities, disharmonious skeletal maturation, poorly modeled long bones and joint dislocations. In AO3 recurrent respiratory insufficiency and/or infections usually result in early death.
Defects in FLNB are the cause of boomerang dysplasia (BOOMD) [MIM:112310]. This is a perinatal lethal osteochondrodysplasia characterized by absence or underossification of the limb bones and vertebre. Boomerang dysplasia is distinguished from atelosteogenesis on the basis of a more severe defect in mineralisation, with complete absence of ossification in some limb elements and vertebral segments.
Defects in FLNB are the cause of Larsen syndrome (LRS) [MIM:150250]. An osteochondrodysplasia characterized by large-joint dislocations and characteristic craniofacial abnormalities. The cardinal features of the condition are dislocations of the hip, knee and elbow joints, with equinovarus or equinovalgus foot deformities. Spatula-shaped fingers, most marked in the thumb, are also present. Craniofacial anomalies include hypertelorism, prominence of the forehead, a depressed nasal bridge, and a flattened midface. Cleft palate and short stature are often associated features. Spinal anomalies include scoliosis and cervical kyphosis. Hearing loss is a well-recognized complication.
Defects in FLNB are the cause of spondylocarpotarsal synostosis syndrome (SCT) [MIM:272460]; also known as spondylocarpotarsal syndrome (SCT) or congenital synspondylism or vertebral fusion with carpal coalition or congenital scoliosis with unilateral unsegmented bar. The disorder is characterized by short stature and vertebral, carpal and tarsal fusions.

序列相似性

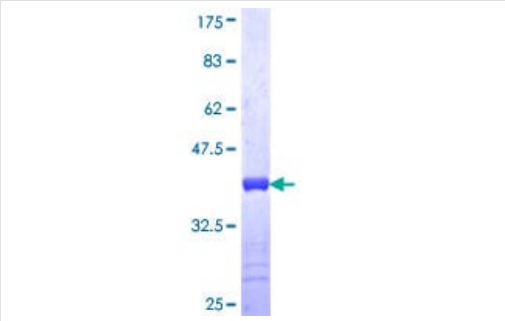
Belongs to the filamin family.
Contains 1 actin-binding domain.
Contains 2 CH (calponin-homology) domains.
Contains 24 filamin repeats.

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. The first hinge region prevents binding to ITGA and ITGB subunits.

翻译后修饰	ISGylation prevents ability to interact with the upstream activators of the JNK cascade and inhibits IFNA-induced JNK signaling.
细胞定位	Cytoplasm > cytoskeleton. Polarized at the periphery of myotubes; Cytoplasm > cytoskeleton. Predominantly localized at actin stress fibers and Cytoplasm > cell cortex. Cytoplasm > cytoskeleton. Cytoplasm > myofibril > sarcomere > Z line. In differentiating myotubes, isoform 1, isoform 2 and isoform 3 are localized diffusely throughout the cytoplasm with regions of enrichment at the longitudinal actin stress fiber. In differentiated tubes, isoform 1 is also detected within the Z-lines.

图片



SDS-PAGE - Recombinant Human Filamin B protein (ab158452)

ab158452 on a 12.5% SDS-PAGE stained with Coomassie Blue.

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