

Filamin 3 (D-6): sc-166484

BACKGROUND

Filamins are Actin-binding proteins which contain an N-terminal Actin-binding domain, a membrane glycoprotein domain and a C-terminal self-association domain. Filamins help reshape the cytoskeleton by forming flexible cross-links between two Actin filaments, which maintain membrane integrity during force application. Filamins also participate in signal transduction pathways associated with cell motility, adhesion, differentiation and survival, and force transduction. The Filamin family is comprised of Filamin 1, Filamin 2 and Filamin 3. Filamin 3, also designated Filamin B and β -Filamin, is a form of Filamin that plays a role in endochondral ossification, vertebral segmentation and joint formation. The interaction of Filamin 3 with Filamin 1 may allow neuroblast migration into the cortical plate from the ventricular zone. Mutations in the gene that encodes for Filamin 3, FLNB, are associated with five human skeletal disorders, specifically, autosomal dominant Larsen syndrome, spondylarcarpotarsal syndrome, type I atelosteogenesis, type III atelosteogenesis and Boomerang dysplasia as well as the neurologic disorder periventricular heterotopia.

REFERENCES

1. Takafuta, T., et al. 1998. Human β -Filamin is a new protein that interacts with the cytoplasmic tail of glycoprotein Ib α . *J. Biol. Chem.* 273: 17531-17538.
2. Online Mendelian Inheritance in Man, OMIM[™]. 2002. Johns Hopkins University, Baltimore, MD. MIM Number: 603381. World Wide Web URL: <http://www.ncbi.nlm.nih.gov/omim/>
3. Krakow, D., et al. 2004. Mutations in the gene encoding Filamin B disrupt vertebral segmentation, joint formation and skeletogenesis. *Nat. Genet.* 36: 405-410.
4. Bicknell, L.S., et al. 2005. Mutations in FLNB cause boomerang dysplasia. *J. Med. Genet.* 42: e43.

CHROMOSOMAL LOCATION

Genetic locus: FLNB (human) mapping to 3p14.3; Flnb (mouse) mapping to 14 A1.

SOURCE

Filamin 3 (D-6) is a mouse monoclonal antibody specific for an epitope mapping between amino acids 985-1013 within an internal region of Filamin 3 of human origin.

PRODUCT

Each vial contains 200 μ g IgG₁ kappa light chain in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-166484 P, (100 μ g peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% stabilizer protein).

STORAGE

Store at 4° C, ****DO NOT FREEZE****. Stable for one year from the date of shipment. Non-hazardous. No MSDS required.

APPLICATIONS

Filamin 3 (D-6) is recommended for detection of Filamin 3 of human origin and Filamin β of mouse and rat origin by Western Blotting (starting dilution 1:100, dilution range 1:100-1:1000), immunoprecipitation [1-2 μ g per 100-500 μ g of total protein (1 ml of cell lysate)], immunofluorescence (starting dilution 1:50, dilution range 1:50-1:500) and solid phase ELISA (starting dilution 1:30, dilution range 1:30-1:3000).

Suitable for use as control antibody for Filamin 3 siRNA (h): sc-60641, Filamin 3 siRNA (m): sc-60642, Filamin 3 shRNA Plasmid (h): sc-60641-SH, Filamin 3 shRNA Plasmid (m): sc-60642-SH, Filamin 3 shRNA (h) Lentiviral Particles: sc-60641-V and Filamin 3 shRNA (m) Lentiviral Particles: sc-60642-V.

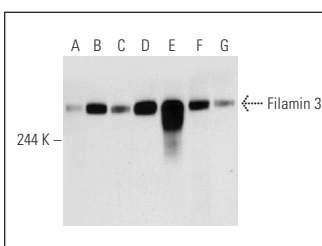
Molecular Weight of Filamin 3: 280 kDa.

Positive Controls: HeLa whole cell lysate: sc-2200, NIH/3T3 whole cell lysate: sc-2210 or U-87 MG cell lysate: sc-2411.

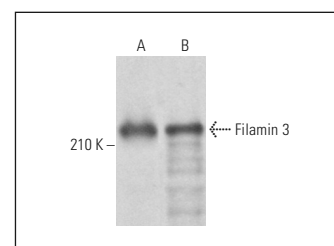
RECOMMENDED SUPPORT REAGENTS

To ensure optimal results, the following support reagents are recommended: 1) Western Blotting: use m-IgG κ BP-HRP: sc-516102 or m-IgG κ BP-HRP (Cruz Marker): sc-516102-CM (dilution range: 1:1000-1:10000), Cruz Marker[™] Molecular Weight Standards: sc-2035, UltraCruz[®] Blocking Reagent: sc-516214 and Western Blotting Luminol Reagent: sc-2048. 2) Immunoprecipitation: use Protein A/G PLUS-Agarose: sc-2003 (0.5 ml agarose/2.0 ml). 3) Immunofluorescence: use m-IgG κ BP-FITC: sc-516140 or m-IgG κ BP-PE: sc-516141 (dilution range: 1:50-1:200) with UltraCruz[®] Mounting Medium: sc-24941 or UltraCruz[®] Hard-set Mounting Medium: sc-359850.

DATA



Filamin 3 (D-6): sc-166484. Western blot analysis of Filamin 3 expression in HeLa (A), Jurkat (B), K-562 (C), U-87 MG (D), NIH/3T3 (E), C6 (F) and KNRK (G) whole cell lysates.



Filamin 3 (D-6): sc-166484. Western blot analysis of Filamin 3 expression in HUV-EC-C (A) and A549 (B) whole cell lysates.

RESEARCH USE

For research use only, not for use in diagnostic procedures.

PROTOCOLS

See our web site at www.scbt.com for detailed protocols and support products.