

# fibrillin-1 (2Q629): sc-71108

## BACKGROUND

The fibrillin family of proteins, including fibrillin-1 (FBN1) and fibrillin-2 (FBN2), are integral components of a distinct subset of extracellular microfibrils. Microfibrils are found in elastic tissues where they facilitate elastic fiber formation and in nonelastic tissue where they support the association of the epithelial cells with the interstitial matrix. Characteristic of the fibrillin proteins are the epidermal growth factor (EGF)-like motifs, which contain a consensus sequence for calcium binding. This calcium association may be critical for protein-protein interactions and stabilization of the microfibrils. Mutations of the FBN1 gene have been shown to result in Marfan syndrome, a disease characterized by abnormal synthesis, secretion and matrix deposition of fibrillin. FBN2 is also linked to a rare, yet similar, skeletal disorder, congenital contractural arachnodactyly.

## REFERENCES

- Zhang, H., Apfelroth, S.D., Hu, W., Davis, E.C., Sanguineti, C., Bonadio, J., Mecham, R.P. and Ramirez, F. 1994. Structure and expression of fibrillin-2, a novel microfibrillar component preferentially located in elastic matrices. *J. Cell Biol.* 124: 855-863.
- Zhang, H., Hu, W. and Ramirez, F. 1995. Development expression of fibrillin genes suggests heterogeneity of extracellular microfibrils. *J. Cell Biol.* 129: 1165-1176.
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- Dietz, H.C. and Pyeritz, R.E. 1995. Mutations in the human gene for fibrillin-1 (FBN1) in the Marfan syndrome and related disorders. *Hum. Mol. Genet.* 4: 1799-1809.
- Putnam, E.A., Zhang, H., Ramirez, F. and Milewicz, D.M. 1995. Fibrillin-2 (FBN2) mutations result in the Marfan-like disorder, congenital contractural arachnodactyly. *Nat. Genet.* 11: 456-458.
- Reinhardt, D.P., Mechling, D.E., Boswell, B.A., Keene, D.R., Sakai, L.Y. and Bachinger, H.P. 1997. Calcium determines the shape of fibrillin. *J. Biol. Chem.* 272: 7368-7373.

## CHROMOSOMAL LOCATION

Genetic locus: FBN1 (human) mapping to 15q21.1; Fbn1 (mouse) mapping to 2 F1.

## SOURCE

fibrillin-1 (2Q629) is a mouse monoclonal antibody raised against microfibrils from zonular apparatus of the eye of bovine origin.

## PRODUCT

Each vial contains 100 µg IgG<sub>1</sub> in 1.0 ml PBS with < 0.1% sodium azide and 0.1% gelatin.

## RESEARCH USE

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

## APPLICATIONS

fibrillin-1 (2Q629) is recommended for detection of fibrillin-1 of human origin by Western Blotting (starting dilution 1:200, dilution range 1:100-1:1000), 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).

fibrillin-1 (2Q629) is also recommended for detection of fibrillin-1 in additional species, including bovine and porcine.

Suitable for use as control antibody for fibrillin-1 siRNA (h): sc-43117, fibrillin-1 siRNA (m): sc-43118, fibrillin-1 shRNA Plasmid (h): sc-43117-SH, fibrillin-1 shRNA Plasmid (m): sc-43118-SH fibrillin-1 shRNA (h) Lentiviral Particles: sc-43117-V and fibrillin-1 shRNA (m) Lentiviral Particles: sc-43118-V.

Molecular Weight of fibrillin-1: 330-350 kDa.

Positive Controls: Jurkat whole cell lysate: sc-2204.

## SELECT PRODUCT CITATIONS

- Shieh, D.B., Li, R.Y., Liao, J.M., Chen, G.D. and Liou, Y.M. 2010. Effects of genistein on  $\beta$ -catenin signaling and subcellular distribution of actin-binding proteins in human umbilical CD105-positive stromal cells. *J. Cell. Physiol.* 223: 423-434.

## STORAGE

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

## PROTOCOLS

See our web site at [www.scbt.com](http://www.scbt.com) for detailed protocols and support products.