SANTA CRUZ BIOTECHNOLOGY, INC.

fibrillin-1 (C-19): sc-7540

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The Power to Question

BACKGROUND

The fibrillin family of proteins, including fibrillin-1 (Fbn-1) and fibrillin-2 (Fbn-2), 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 fibrillin-1 gene have been shown to result in Marfan syndrome, a disease characterized by abnormal synthesis, secretion and matrix deposition of fibrillin. Fibrillin-2 is also linked to a rare, yet similiar, skeletal disorder, congenital contractural arachnodactyly.

REFERENCES

1. 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.
2. Zhang, H., Hu, W. and Ramirez, F. 1995. Development expression of fibrillin genes suggests heterogeneity of extracellular microfibrils. J. Cell Biol. 129: 1165-1176.
3. Yin, W., Smiley, E., Germiller, J., Sanguineti, C., Lawton, T., Pereira, L., Ramirez, F. and Bonadio, J. 1995. Primary structure and developmental expression of Fbn-1, the mouse fibrillin gene. J. Biol. Chem. 270: 1798-1806.
4. 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.
5. 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.

CHROMOSOMAL LOCATION

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

SOURCE

fibrillin-1 (C-19) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the C-terminus of fibrillin-1 of human origin.

PRODUCT

Each vial contains 200 µg IgG in 1.0 ml of PBS with < 0.1% sodium azide and 0.1% gelatin.

Blocking peptide available for competition studies, sc-7540 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

RESEARCH USE

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

APPLICATIONS

fibrillin-1 (C-19) is recommended for detection of fibrillin-1 of mouse, rat and 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).

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.

RECOMMENDED SECONDARY REAGENTS

To ensure optimal results, the following support (secondary) reagents are recommended: 1) Western Blotting: use donkey anti-goat IgG-HRP: sc-2020 (dilution range: 1:2000-1:100,000) or Cruz Marker™ compatible donkey anti-goat IgG-HRP: sc-2033 (dilution range: 1:2000-1:5000), Cruz Marker™ Molecular Weight Standards: sc-2035, TBS Blotto A Blocking Reagent: sc-2333 and Western Blotting Luminol Reagent: sc-2048. 2) Immunofluorescence: use donkey anti-goat IgG-FITC: sc-2024 (dilution range: 1:100-1:400) or donkey anti-goat IgG-TR: sc-2783 (dilution range: 1:100-1:400) with UltraCruz™ Mounting Medium: sc-24941.

SELECT PRODUCT CITATIONS

1. Olabisi, O.A., Soto-Nieves, N., Nieves, E., Yang, T.T., Yang, X., Yu, R.Y., Suk, H.Y., Macian, F. and Chow, C.W. 2008. Regulation of transcription factor NFAT by ADP-ribosylation. Mol. Cell. Biol. 28: 2860-2871.
2. Chamberlain, C.M., Ang, L.S., Boivin, W.A., Cooper, D.M., Williams, S.J., Zhao, H., Hendel, A., Folkesson, M., Swedenborg, J., Allard, M.F., McManus, B.M. and Granville, D.J. 2010. Perforin-independent extracellular granzyme B activity contributes to abdominal aortic aneurysm. Am. J. Pathol. 176: 1038-1049.
3. Jimenez-Vergara, A.C., Munoz-Pinto, D.J., Becerra-Bayona, S., Wang, B., Iacob, A. and Hahn, M.S. 2011. Influence of glycosaminoglycan identity on vocal fold fibroblast behavior. Acta Biomater. 7: 3964-3972.

STORAGE

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