

fibrillin-1 (H-109): sc-20084

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 similar skeletal disorder, congenital contractural arachnodactyly.

REFERENCES

1. Zhang, H., et al. 1994. Structure and expression of fibrillin-2, a novel microfibrillar component preferentially located in elastic matrices. *J. Cell Biol.* 124: 855-863.
2. Dietz, H.C. and Pyeritz, R.E. 1995. Mutations in the human gene for fibrillin-1 (Fbn-1) in the Marfan syndrome and related disorders. *Hum. Mol. Genet.* 4: 1799-1809.

CHROMOSOMAL LOCATION

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

SOURCE

fibrillin-1 (H-109) is a rabbit polyclonal antibody corresponding to amino acids 2726-2834 mapping near 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.

APPLICATIONS

fibrillin-1 (H-109) 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), immunoprecipitation [1-2 µg per 100-500 µg 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).

fibrillin-1 (H-109) is also recommended for detection of fibrillin-1 in additional species, including equine, canine, 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.

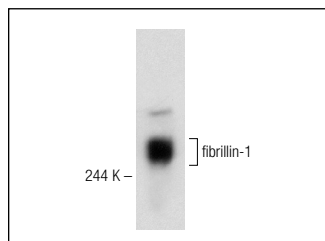
Molecular Weight of fibrillin-1: 330-350 kDa.

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

STORAGE

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

DATA



fibrillin-1 (H-109): sc-20084. Western blot analysis of fibrillin-1 expression in Jurkat whole cell lysate.

SELECT PRODUCT CITATIONS

1. Koli, K., et al. 2004. Disruption of LTBP-4 function reduces TGFβ activation and enhances BMP-4 signaling in the lung. *J. Cell Biol.* 167: 123-133.
2. Tseleni-Balafouta, S., et al. 2006. Fibrillin expression and localization in various types of carcinomas of the thyroid gland. *Mod. Pathol.* 19: 695-700.
3. Martin, L., et al. 2007. Histological skin changes in heterozygote carriers of mutations in ABCC6, the gene causing pseudoxanthoma elasticum. *J. Eur. Acad. Dermatol. Venereol.* 21: 368-373.
4. Moraes-Teixeira Jde, A., et al. 2010. Exercise training enhances elastin, fibrillin and nitric oxide in the aorta wall of spontaneously hypertensive rats. *Exp. Mol. Pathol.* 89: 351-357.
5. Sugawara, Y., et al. 2010. Immunohistochemical localization of elastin, fibrillins and microfibril-associated glycoprotein-1 in the developing periodontal ligament of the rat molar. *J. Periodont. Res.* 45: 52-59.

RESEARCH USE

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

PROTOCOLS

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

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Try **fibrillin-1 (20629): sc-71108**, our highly recommended monoclonal alternative to fibrillin-1 (H-109)