

# fibrillin-2 (N-20): sc-28136

## BACKGROUND

Extracellular glycoproteins fibrillin-1 and -2 are major components of connective tissue microfibrils. Fibrillin-2 containing microfibrils regulate the early process of elastic fiber assembly in tissue. Mutations in the fibrillin-2 gene resulting in impaired assembly of fibrillin-2 may lead to molecular congenital contractural arachnodactyly. Fibrillin-2 constitutes the backbone of microfibrils which insert directly into the lamina densa of basement membranes. Epithelial cells primarily deposit fibrillin into the extracellular matrix in a nonfibrillar form. Mutations in the 8-cysteine motif of fibrillin-2 alters its binding to microfibril-associated glycoprotein-1 (MAGP-1), which may increase the severity of congenital contractural arachnodactyly.

## REFERENCES

1. Mariencheck, M.C., et al. 1995. Fibrillin-1 and fibrillin-2 show temporal and tissue-specific regulation of expression in developing elastic tissues. *Connect. Tissue Res.* 31: 87-97.
2. Dzamba, B., et al. 2001. Assembly of epithelial cell fibrillins. *J. Invest. Dermatol.* 117: 1612-1620.
3. Lin, G., et al. 2002. Homo- and heterotypic fibrillin-1 and -2 interactions constitute the basis for the assembly of microfibrils. *J. Biol. Chem.* 277: 50795-50804.
4. Quondamatteo, F., et al. 2002. Fibrillin-1 and fibrillin-2 in human embryonic and early fetal development. *Matrix Biol.* 21: 637-646.
5. Ritty, T.M., et al. 2003. Fibrillin-1 and -2 contain heparin-binding sites important for matrix deposition and that support cell attachment. *Biochem. J.* 375: 425-432.
6. Werneck, C.C., et al. 2004. Identification of a major microfibril-associated glycoprotein-1-binding domain in fibrillin-2. *J. Biol. Chem.* 279: 23045-23051.
7. Tsuruga, E., et al. 2005. Microfibril-associated glycoprotein-1 and fibrillin-2 are associated with tropoelastin deposition *in vitro*. *Int. J. Biochem. Cell Biol.* 37: 120-129.
8. SWISS-PROT/TrEMBL (Q61555). World Wide Web URL: <http://www.expasy.ch/sprot/sprot-top.html>

## CHROMOSOMAL LOCATION

Genetic locus: FBN2 (human) mapping to 5q23.3.

## SOURCE

fibrillin-2 (N-20) is an affinity purified goat polyclonal antibody raised against a peptide mapping at the N-terminus of fibrillin-2 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-28136 P, (100 µg peptide in 0.5 ml PBS containing < 0.1% sodium azide and 0.2% BSA).

## APPLICATIONS

fibrillin-2 (N-20) is recommended for detection of fibrillin-2 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).

Suitable for use as control antibody for fibrillin-2 siRNA (h): sc-45971, fibrillin-2 shRNA Plasmid (h): sc-45971-SH and fibrillin-2 shRNA (h) Lentiviral Particles: sc-45971-V.

Molecular Weight of fibrillin-2: 350 kDa.

## 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.

## STORAGE

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

## RESEARCH USE

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

## PROTOCOLS

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

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Try **fibrillin-2 (H-10): sc-393968**, our highly recommended monoclonal alternative to fibrillin-2 (N-20).