



fibrillin-1 (2726-2834): sc-4542 WB

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 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

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6. 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.

SOURCE

fibrillin-1 (2726-2834) is expressed in *E. coli* as a 39 kDa tagged fusion protein corresponding to amino acids 2726-2834 of fibrillin-1 of human origin.

PRODUCT

fibrillin-1 (2726-2834) is purified from bacterial lysates (>98%) by glutathione agarose affinity chromatography; supplied as 10 µg in 0.1 ml SDS-PAGE loading buffer.

APPLICATIONS

fibrillin-1 (2726-2834) is suitable as a Western blotting control for sc-7540 and sc-20084.

STORAGE

Store at -20° C; stable for one year from the date of shipment.

RESEARCH USE

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