

FBN1 / Fibrillin 1 Antibody (clone 3H6)

Mouse Monoclonal Antibody Catalog # ALS14370

Specification

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Product Information

Application Primary Accession Reactivity Host Clonality Calculated MW

IHC <u>P35555</u> Human Mouse Monoclonal 312kDa KDa

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Additional Information

Gene ID 2200

Other Names Fibrillin-1, FBN1, FBN

Target/Specificity Human Fibrillin 1

Reconstitution & Storage Aliquot and store at -20°C or -80°C. Avoid freeze-thaw cycles.

Precautions FBN1 / Fibrillin 1 Antibody (clone 3H6) is for research use only and not for use in diagnostic or therapeutic procedures.

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Protein Information

Name FBN1 (HGNC:3603)

Synonyms FBN

Function

[Fibrillin-1]: Structural component of the 10-12 nm diameter microfibrils of the extracellular matrix, which conveys both structural and regulatory properties to load-bearing connective tissues (PubMed:1860873, PubMed:1860873, PubMed:15062093). Fibrillin-1-containing microfibrils provide long-term force bearing structural support (PubMed:27026396). In tissues such as the lung, blood vessels and skin, microfibrils form the periphery of the elastic fiber, acting as a scaffold for the deposition of elastin (PubMed:27026396). In addition, microfibrils can occur as elastin-independent networks in tissues such as the ciliary zonule, tendon, cornea and glomerulus where they provide tensile strength and have anchoring roles



(PubMed:27026396). Fibrillin-1 also plays a key role in tissue homeostasis through specific interactions with growth factors, such as the bone morphogenetic proteins (BMPs), growth and differentiation factors (GDFs) and latent transforming growth factor-beta-binding proteins (LTBPs), cell-surface integrins and other extracellular matrix protein and proteoglycan components (PubMed:27026396). Regulates osteoblast maturation by controlling TGF- beta bioavailability and calibrating TGF-beta and BMP levels, respectively (By similarity). Negatively regulates osteoclastogenesis by binding and sequestering an osteoclast differentiation and activation factor TNFSF11 (PubMed:24039232). This leads to disruption of TNFSF11-induced Ca(2+) signaling and impairment of TNFSF11-mediated nuclear translocation and activation of transcription factor NFATC1 which regulates genes important for osteoclast differentiation and function (PubMed:24039232). Mediates cell

href="http://www.uniprot.org/citations/24039232" target="_blank">24039232). Mediates cell adhesion via its binding to cell surface receptors integrins ITGAV:ITGB3 and ITGA5:ITGB1 (PubMed:12807887, PubMed:17158881). Binds heparin and this interaction has an important role in the assembly of microfibrils (PubMed:11461921).

Cellular Location

Secreted. Note=Fibrillin-1 and Asprosin chains are still linked together during the secretion from cells, but are subsequently separated by furin (PubMed:24982166) [Asprosin]: Secreted. Note=Secreted by white adipose tissue and circulates in the plasma.

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Protocols

Provided below are standard protocols that you may find useful for product applications.

- Western Blot
- <u>Blocking Peptides</u>
- Dot Blot
- Immunohistochemistry
- Immunofluorescence
- Immunoprecipitation
- Flow Cytomety
- <u>Cell Culture</u>

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Images





Anti-FBN1 / Fibrillin 1 antibody IHC of human placenta.

FBN1 / Fibrillin 1 Antibody (clone 3H6) - Background

Fibrillins are structural components of 10-12 nm extracellular calcium-binding microfibrils, which occur either in association with elastin or in elastin-free bundles. Fibrillin-1- containing microfibrils provide long-term force bearing structural support. Regulates osteoblast maturation by controlling TGF-beta bioavailability and calibrating TGF-beta and BMP levels, respectively.

FBN1 / Fibrillin 1 Antibody (clone 3H6) - References

Pereira L.V., et al. Hum. Mol. Genet. 2:961-968(1993). Uyeda T., et al.J. Hum. Genet. 49:404-407(2004). Rieder M.J., et al. Submitted (SEP-2009) to the EMBL/GenBank/DDBJ databases. Zody M.C., et al. Nature 440:671-675(2006). Mural R.J., et al. Submitted (JUL-2005) to the EMBL/GenBank/DDBJ databases.