

FBN1 Antibody (Internal region)

Peptide-affinity purified goat antibody Catalog # AF3039a

## Specification

# FBN1 Antibody (Internal region) - Product Information

Application Primary Accession Other Accession

Predicted Host Clonality Concentration Isotype Calculated MW E <u>P35555</u> <u>NP\_000129.2</u>, <u>2200</u>, <u>14118 (mouse)</u>, <u>83727</u> (<u>rat)</u> Human, Mouse, Rat, Dog, Cow Goat Polyclonal 0.5 mg/ml IgG 312298

# FBN1 Antibody (Internal region) - Additional Information

Gene ID 2200

Other Names Fibrillin-1, FBN1, FBN

**Format** 0.5 mg/ml in Tris saline, 0.02% sodium azide, pH7.3 with 0.5% bovine serum albumin

Storage

Maintain refrigerated at 2-8°C for up to 6 months. For long term storage store at -20°C in small aliquots to prevent freeze-thaw cycles.

**Precautions** FBN1 Antibody (Internal region) is for research use only and not for use in diagnostic or therapeutic procedures.

### FBN1 Antibody (Internal region) - 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:<a href="http://www.uniprot.org/citations/1860873" target="\_blank">1860873</a>, PubMed:<a href="http://www.uniprot.org/citations/1860873" target="\_blank">1860873</a>, PubMed:<a href="http://www.uniprot.org/citations/15062093" target="\_blank">15062093</a>). Fibrillin-1-containing microfibrils provide long-term force bearing structural support (PubMed:<a



href="http://www.uniprot.org/citations/27026396" target="\_blank">27026396</a>). 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:<a

href="http://www.uniprot.org/citations/27026396" target="\_blank">27026396</a>). 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:<a href="http://www.uniprot.org/citations/27026396" target=" blank">27026396</a>). 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: <a href="http://www.uniprot.org/citations/27026396" target="\_blank">27026396</a>). 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:<a href="http://www.uniprot.org/citations/24039232" target=" blank">24039232</a>). 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:<a

href="http://www.uniprot.org/citations/24039232" target="\_blank">24039232</a>). Mediates cell adhesion via its binding to cell surface receptors integrins ITGAV:ITGB3 and ITGA5:ITGB1 (PubMed:<a href="http://www.uniprot.org/citations/12807887" target="\_blank">12807887</a>, PubMed:<a href="http://www.uniprot.org/citations/12807881" target="\_blank">12807887</a>, PubMed:<a href="http://www.uniprot.org/citations/17158881" target="\_blank">17158881</a>). Binds heparin and this interaction has an important role in the assembly of microfibrils (PubMed:<a href="http://www.uniprot.org/citations/11461921" target="\_blank">11461921</a>).

#### **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 Antibody (Internal region) - Protocols

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

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

# FBN1 Antibody (Internal region) - Images

### FBN1 Antibody (Internal region) - References

Mutation of ACTA2 gene as an important cause of familial and nonfamilial nonsyndromatic thoracic aortic aneurysm and/or dissection (TAAD). Morisaki H, Akutsu K, Ogino H, Kondo N, Yamanaka I, Tsutsumi Y, Yoshimuta T, Okajima T, Matsuda H, Minatoya K, Sasaki H, Tanaka H, Ishibashi-Ueda H, Morisaki T, Human mutation 2009 Jul : . PMID: 19639654