abcam

Product datasheet

Anti-Fibrillin 1 antibody ab53076

1 Abreviews 4 References 1 Image

Overview

Product name	Anti-Fibrillin 1 antibody
Description	Rabbit polyclonal to Fibrillin 1
Host species	Rabbit
Specificity	Fibrillin 1 antibody detects endogenous levels of total Fibrillin 1 protein.
Tested applications	**Suitable for:** IHC-P
Species reactivity	**Reacts with:** Human **Predicted to work with:** Rat
Immunogen	Synthetic peptide: STPLYKKKEL NQLEDKYDKD YLSGELGDNL KMKIQVLLH, corresponding to amino acids 2832-2871 of Human Fibrillin 1 Run BLAST with ExPASy Run BLAST with NCBI
General notes	Reproducibility is key to advancing scientific discovery and accelerating scientists' next breakthrough. Abcam is leading the way with our range of recombinant antibodies, knockout-validated antibodies and knockout cell lines, all of which support improved reproducibility. We are also planning to innovate the way in which we present recommended applications and species on our product datasheets, so that only applications & species that have been tested in our own labs, our suppliers or by selected trusted collaborators are covered by our Abpromise™ guarantee. In preparation for this, we have started to update the applications & species that this product is Abpromise guaranteed for. We are also updating the applications & species that this product has been "predicted to work with," however this information is not covered by our Abpromise guarantee. Applications & species from publications and Abreviews that have not been tested in our own labs or in those of our suppliers are not covered by the Abpromise guarantee. Please check that this product meets your needs before purchasing. If you have any questions, special requirements or concerns, please send us an inquiry and/or contact our Support team ahead of purchase. Recommended alternatives for this product can be found below, as well as customer reviews and Q&As.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
Storage buffer	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: 50% Glycerol, 0.87% Sodium chloride, PBS Without Mg+2 and Ca+2
Purity	Immunogen affinity purified
Purification notes	The antibody was affinity purified from rabbit antiserum by affinity chromatography using epitope-specific immunogen.
Clonality	Polyclonal
Isotype	IgG

Applications

Our Abpromise guarantee covers the use of **ab53076** in the following tested applications.

The application notes include recommended starting dilutions; optimal dilutions/concentrations should be determined by the end user.

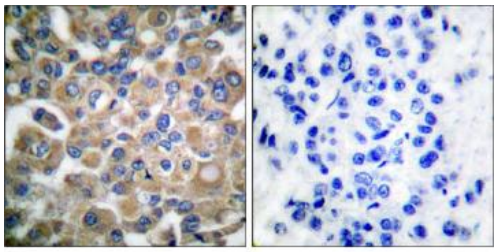
Application	Abreviews	Notes
IHC-P	★★★★☆	Use at an assay dependent concentration.

Target

Function	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.
Involvement in disease	Defects in FBN1 are a cause of Marfan syndrome (MFS) [MIM:154700]. MFS is an autosomal dominant disorder that affects the skeletal, ocular, and cardiovascular systems. A wide variety of skeletal abnormalities occurs with MFS, including scoliosis, chest wall deformity, tall stature, abnormal joint mobility. Ectopia lentis occurs in up to about 80% of MFS patients and is almost always bilateral. The leading cause of premature death in MFS patients is progressive dilation of the aortic root and ascending aorta, causing aortic incompetence and dissection. Note=The majority of the more than 600 mutations in FBN1 currently known are point mutations, the rest are frameshifts and splice site mutations. Marfan syndrome has been suggested in at least 2 historical figures, Abraham Lincoln and Paganini. Defects in FBN1 are a cause of isolated ectopia lentis (EL) [MIM:129600]. The symptoms of this autosomal dominant fibrillinopathy overlap with those of Marfan syndrome, with the exclusion of the skeletal and cardiovascular manifestations. Defects in FBN1 are the cause of Weill-Marchesani syndrome autosomal dominant (ADWMS) [MIM:608328]. A rare connective tissue disorder characterized by short stature, brachydactyly, joint stiffness, and eye abnormalities including microspherophakia, ectopia lentis, severe myopia and glaucoma. Defects in FBN1 are a cause of Shprintzen-Goldberg craniosynostosis syndrome (SGS) [MIM:182212]. SGS is a very rare syndrome characterized by a marfanoid habitus, craniosynostosis, characteristic dysmorphic facial features, skeletal and cardiovascular

	abnormalities, mental retardation, developmental delay and learning disabilities. Defects in FBN1 are a cause of overlap connective tissue disease (OCTD) [MIM:604308]. A heritable disorder of connective tissue characterized by involvement of the mitral valve, aorta, skeleton, and skin. MASS syndrome is closely resembling both the Marfan syndrome and the Barlow syndrome. However, no dislocation of the lenses or aneurysmal changes occur in the aorta, and the mitral valve prolapse is by no means invariable. Defects in FBN1 are a cause of stiff skin syndrome (SSKS) [MIM:184900]. It is a syndrome characterized by hard, thick skin, usually over the entire body, which limits joint mobility and causes flexion contractures. Other occasional findings include lipodystrophy and muscle weakness.
Sequence similarities	Belongs to the fibrillin family. Contains 47 EGF-like domains. Contains 9 TB (TGF-beta binding) domains.
Post-translational modifications	Forms intermolecular disulfide bonds either with other fibrillin-1 molecules or with other components of the microfibrils.
Cellular localization	Secreted > extracellular space > extracellular matrix.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Fibrillin 1 antibody (ab53076)

Immunohistochemical analysis of paraffin-embedded human breast carcinoma tissue
using ab53076 at 1/50 dilution, with and without immunizing peptide.

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