Product datasheet

Anti-Fibrillin 1 antibody ab53076

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: ICC/IF, IHC-P, ELISA
Species reactivity: Reacts with: Mouse, Human
Predicted to work with: Rat
Immunogen: Synthetic peptide: STPLYKKEL NQLEDKYDKD YLSGELGDNL KMKIQVLLH, corresponding to amino acids 2832-2871 of Human Fibrillin 1
Positive control: Human breast carcinoma tissue

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

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.
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
Forms intermolecular disulfide bonds either with other fibrillin-1 molecules or with other
modifications components of the microfibrils.

Cellular localization Secreted > extracellular space > extracellular matrix.

Images

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

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