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Product datasheet

Anti-Fibrillin 1 antibody ab53076

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概述

产品名称	Anti-Fibrillin 1抗体
描述	**兔多克隆抗体**to Fibrillin 1
宿主	Rabbit
经测试应用	**适用于:** ELISA, ICC/IF, IHC-P
种属反应性	**与反应:** Mouse, Human **预测可用于:** Rat
免疫原	Synthetic peptide: STPLYKKKEL NQLEDKYDKD YLSGELGDNL KMKIQVLLH, corresponding to amino acids 2832-2871 of Human Fibrillin 1 Run BLAST with ExPASy Run BLAST with NCBI
常规说明	The Life Science industry has been in the grips of a reproducibility crisis for a number of years. Abcam is leading the way in addressing this with our range of recombinant monoclonal antibodies and knockout edited cell lines for gold-standard validation. 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, along with publications, customer reviews and Q&As

性能

形式	Liquid
存放说明	Shipped at 4°C. Store at -20°C. Stable for 12 months at -20°C.
存储溶液	pH: 7.40 Preservative: 0.02% Sodium azide Constituents: 50% Glycerol, 0.87% Sodium chloride, PBS Without Mg+2 and Ca+2
纯度	Immunogen affinity purified
纯化说明	The antibody was affinity purified from rabbit antiserum by affinity chromatography using epitope-specific immunogen.
克隆	**多克隆**
同种型	IgG

应用

The Abpromise guarantee **Abpromise™承诺保证使用ab53076于以下的经测试应用**

"应用说明"部分 下显示的仅为推荐的起始稀释度;实际最佳的稀释度/浓度应由使用者检定。

应用	Ab评论	说明
ELISA		Use at an assay dependent concentration.
ICC/IF		Use at an assay dependent concentration. PubMed: 23401661
IHC-P	★★★★☆ (1)	Use at an assay dependent concentration.

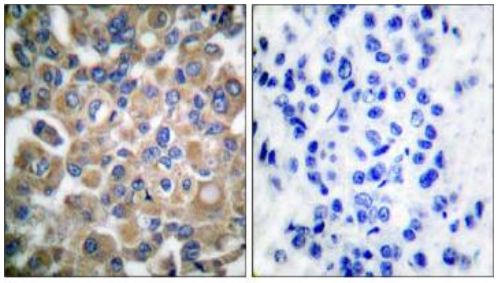
靶标

功能 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.

疾病相关 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.

序列相似性	Belongs to the fibrillin family. Contains 47 EGF-like domains. Contains 9 TB (TGF-beta binding) domains.
翻译后修饰	Forms intermolecular disulfide bonds either with other fibrillin-1 molecules or with other components of the microfibrils.
细胞定位	Secreted > extracellular space > extracellular matrix.

图片



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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