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Product datasheet

Anti-Fibrillin 1 antibody [3H6] ab124334

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Overview

Product name	Anti-Fibrillin 1 antibody [3H6]
Description	Mouse monoclonal [3H6] to Fibrillin 1
Host species	Mouse
Tested applications	**Suitable for:** WB, IHC-P, Sandwich ELISA
Species reactivity	**Reacts with:** Human
Immunogen	Recombinant fragment, corresponding to amino acids 2772-2872 of Human Fibrillin 1 with a proprietary tag (NP_000129).
Positive control	Human placenta tissue; Recombinant tagged Fibrillin 1
General notes	Abcam is committed to meeting high standards of ethical manufacturing and has decided to discontinue this product by June 2019 as it has been generated by the ascites method. We are sorry for any inconvenience this may cause.

Properties

Form	Liquid
Storage instructions	Shipped at 4°C. Upon delivery aliquot and store at -20°C or -80°C. Avoid repeated freeze / thaw cycles.
Storage buffer	pH: 7.4 Constituent: 99% PBS
Purity	Protein A purified
Clonality	Monoclonal
Clone number	3H6
Isotype	IgG2a
Light chain type	kappa

Applications

Our Abpromise guarantee covers the use of **ab124334** 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
WB		1/500 - 1/1000. Predicted molecular weight: 312 kDa.
IHC-P		Use a concentration of 5 µg/ml.
Sandwich ELISA		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.

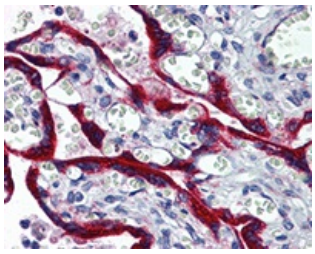
Post-translational modifications

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

Cellular localization

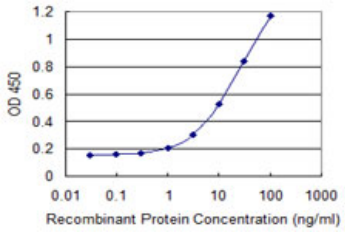
Secreted > extracellular space > extracellular matrix.

Images



Immunohistochemistry (Formalin/PFA-fixed paraffin-embedded sections) - Anti-Fibrillin 1 antibody [3H6] (ab124334)

ab124334, at 5 µg/ml staining Fibrillin 1 in formalin-fixed, paraffin-embedded Human Placenta tissue by Immunohistochemistry.



Sandwich ELISA - Anti-Fibrillin 1 antibody [3H6] (ab124334)

Detection limit for recombinant tagged Fibrillin 1 is 0.3 ng/ml as a capture antibody.

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