abcam

Product datasheet

Biotin Anti-Fibrillin 1 antibody [11C1.3] ab24826

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製品の概要

製品名	Biotin Anti-Fibrillin 1 antibody [11C1.3]
製品の詳細	Biotin Mouse monoclonal [11C1.3] to Fibrillin 1
由来種	Mouse
標識	Biotin
特異性	This antibody recognises fibrillin (Mr 350 kDa).
アプリケーション	**適用あり:** IHC-Fr, ICC
種交差性	**交差種:** Mouse, Human
免疫原	Tissue, cells or virus corresponding to Cow Fibrillin 1. Microfibrils from the zonular apparatus of bovine eye
ポジティブ・コントロール	IHC-Fr: Mouse skin tissue section; ICC: MIA PaCa-2 cells.

製品の特性

製品の状態	Liquid
保存方法	Shipped at 4°C. Store at +4°C short term (1-2 weeks). Upon delivery aliquot. Store at -20°C long term. Avoid freeze / thaw cycle. Store In the Dark.
バッファー	pH: 7.4 Preservative: 0.1% Sodium azide Constituent: PBS
精製度	Affinity purified
ポリ/モノ	モノクローナル
クローン名	11C1.3
アイソタイプ	IgG1
軽鎖の種類	kappa

アプリケーション

The Abpromise guarantee	Abpromise保証は、次のテスト済みアプリケーションにおけるab24826の使用に適用されます

アプリケーションノートには、推奨の開始希釈率がありますが、適切な希釈率につきましてはご検討ください。

アプリケーション	Abreviews	特記事項
IHC-Fr		Use at an assay dependent concentration.
ICC		Use at an assay dependent concentration.

ターゲット情報

機能

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.

関連疾患

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.

配列類似性

Belongs to the fibrillin family.
Contains 47 EGF-like domains.
Contains 9 TB (TGF-beta binding) domains.

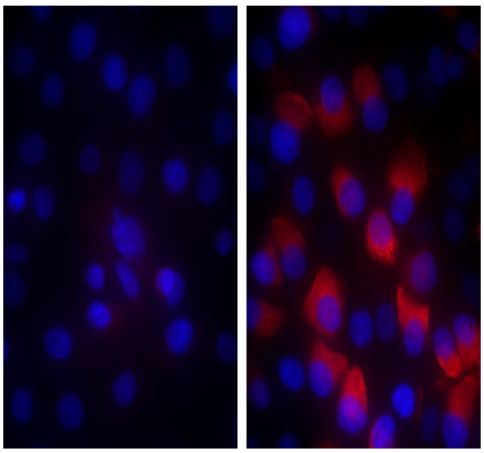
翻訳後修飾

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

細胞内局在

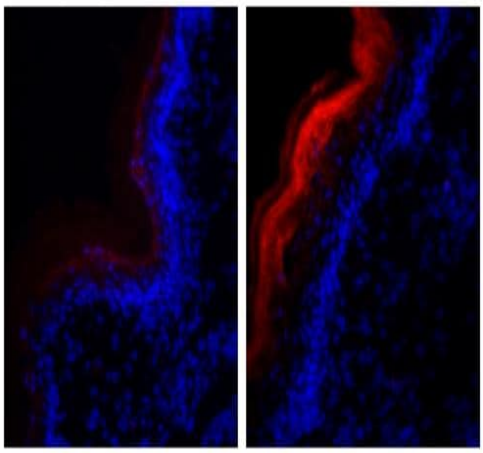
Secreted > extracellular space > extracellular matrix.

画像



Immunocytochemistry - Anti-Fibrillin 1 antibody [11C1.3] (Biotin) (ab24826)

Immunocytochemistry analysis of MIA PaCa-2 (human pancreatic carcinoma cell line) cells labeling Fibrillin 1 with ab24826 (right) followed by Streptavidin-CY3.5 (red). Biotinylated Mouse IgG1 used as the isotype control (left) followed by Streptavidin-CY3.5 (red). Nuclei counterstained with DAPI (blue).



Immunohistochemistry (Frozen sections) - Anti-Fibrillin 1 antibody [11C1.3] (Biotin) (ab24826)

Immunohistochemistry analysis of frozen mouse skin tissue sections labeling Fibrillin 1 with ab24826 (right) followed by Streptavidin-CY3.5 (red). Biotinylated Mouse IgG1 used as the isotype control (left). Nuclei counterstained with DAPI (blue).

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