Product datasheet

Anti-Fibrillin 1 antibody ab231094

Anti-Fibrillin 1 antibody

Rabbit polyclonal to Fibrillin 1

Rabbit

適用あり: WB, IHC-P

交差種: Rat, Human

交差が予測される動物種: Mouse, Cow, Pig

Recombinant fragment (His-tag) corresponding to Rat Fibrillin 1 aa 168-364. Expressed in E.coli. N-terminal tag.

Sequence:

```
CTYGFTGPQCRQDRYRTGPGCFTVSNQCMQGQLSGIVCTKT
LCCATVGRAW
```

```
GHPCEMCPAQPHPCRGFIPNIRTGACQDVDECAIPLGCL
QGGNCINTVT
```

```
SFECKCPAGHKFNEVSQKDIDEKCDSTIGVCDGECTNT
VSSYFCKCPP
```

```
GFTSPDGDVRCVRPGCYTALTNGRCSNQLPQSITKMQ
CCCDVGHR
```

Database link: Q9WUH8

IHC-P: Rat lung tissue. WB: Rat lung and bone marrow lysates; Human lung lysate; Recombinant rat Fibrillin 1 protein.

製品の状態

Liquid

保存方法


バッファー

pH: 7.40

Preservative: 0.02% Sodium azide

Constituents: PBS, 50% Glycerol

精製度

Immunogen affinity purified
ab231094 was purified by antigen-specific affinity chromatography followed by Protein A affinity chromatography.

**IgG**

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**Application**

Our Abpromise guarantee covers the use of ab231094 in the following tested applications.

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

<table>
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<th>Application</th>
<th>Abreviews</th>
<th>Note</th>
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<tbody>
<tr>
<td>WB</td>
<td>🌟🌟🌟🌟🌟</td>
<td>Use a concentration of 0.2 - 2 µg/ml. Predicted molecular weight: 312 kDa.</td>
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<tr>
<td>IHC-P</td>
<td></td>
<td>Use a concentration of 5 - 20 µg/ml.</td>
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**Target Information**

**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.

**Related Diseases**

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.

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

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

Secreted > extracellular space > extracellular matrix.

Paraffin-embedded rat lung tissue stained for Fibrillin 1 using ab231094 at 20 µg/ml in immunohistochemical analysis. DAB staining.

Anti-Fibrillin 1 antibody (ab231094) at 1 µg/ml + Rat lung lysate

**Predicted band size**: 312 kDa
Anti-Fibrillin 1 antibody (ab231094) at 2 µg/ml + Rat bone marrow lysate

**Predicted band size:** 312 kDa

Anti-Fibrillin 1 antibody (ab231094) at 2 µg/ml + Human lung lysate

**Predicted band size:** 312 kDa
Western blot - Anti-Fibrillin 1 antibody (ab231094) at 2 µg/ml + Recombinant rat Fibrillin 1 protein

**Predicted band size:** 312 kDa

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