

Rabbit Anti-Fibrillin 2 antibody

SL12166R

Product Name	Fibrillin 2
Chinese Name	原纤维蛋白 2 抗体
Alias	CCA; congenital contractural arachnodactyly (Marfanoid-like); DA9; FBN2; FBN2_HUMAN; fibrillin 2 (congenital contractural arachnodactyly); Fibrillin-2.
Research Area	Cell biology Neurobiology Signal transduction Binding protein Extracellular matrix
Immunogen Species	Rabbit
Clonality	Polyclonal
React Species	Rat(predicted:Human,Mouse,Chicken,Dog,Pig,Cow,Horse,Rabbit,Sheep) IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500 (Paraffin sections need antigen repair)
Applications	not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Theoretical molecular weight	311kDa
Cellular localization	Secretory protein
Form	Liquid
Concentration	1mg/ml
immunogen	KLH conjugated synthetic peptide derived from human Fibrillin 2: 1001-1200/2912
Lsotype	IgG
Purification	affinity purified by Protein A
Buffer Solution	1M TBS(pH7.4) with 1% BSA, 3% Proclin300 and 50% Glycerol.
Storage	Shipped at 4°C. Store at -20 °C for one year. Avoid repeated freeze/thaw cycles.
Attention	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
PubMed	PubMed
Product	Extracellular glycoproteins fibrillin-1 and -2 are major components of connective tissue

Detail

microfibrils. Fibrillin-2 containing microfibrils regulate the early process of elastic fiber assembly in tissue. Mutations in the fibrillin-2 gene resulting in impaired assembly of fibrillin-2 may lead to molecular congenital contractural arachnodactyly. Fibrillin-2 constitutes the backbone of microfibrils which insert directly into the lamina densa of basement membranes. Epithelial cells primarily deposit fibrillin into the extracellular matrix in a nonfibrillar form. Mutations in the 8-cysteine motif of Fibrillin-2 alters its binding to microfibril-associated glycoprotein-1 (MAGP-1), which may increase the severity of congenital contractural arachnodactyly.

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-2-containing microfibrils regulate the early process of elastic fiber assembly. Regulates osteoblast maturation by controlling TGF-beta bioavailability and calibrating TGF-beta and BMP levels, respectively.

Subcellular Location:

Secreted

DISEASE:

Defects in FBN2 are the cause of congenital contractural arachnodactyly (CCA) [MIM:121050]; also known as Beals syndrome or distal arthrogyriposis type 9 (DA9). CCA is a rare, autosomal dominant connective tissue disorder characterized by contractures, arachnodactyly, scoliosis, and crumpled ears. Phenotypically similar to Marfan syndrome, CCA does not affect the aorta and the eyes.

Similarity:

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

SWISS:

P35556

Gene ID:

2201

Database links:

[Entrez Gene: 2201](#) Human

[Entrez Gene: 100047082](#) Mouse

[Entrez Gene: 14119](#) Mouse

[Entrez Gene: 689008](#) Rat

[Omim: 121050](#) Human

[SwissProt: P35556](#) Human

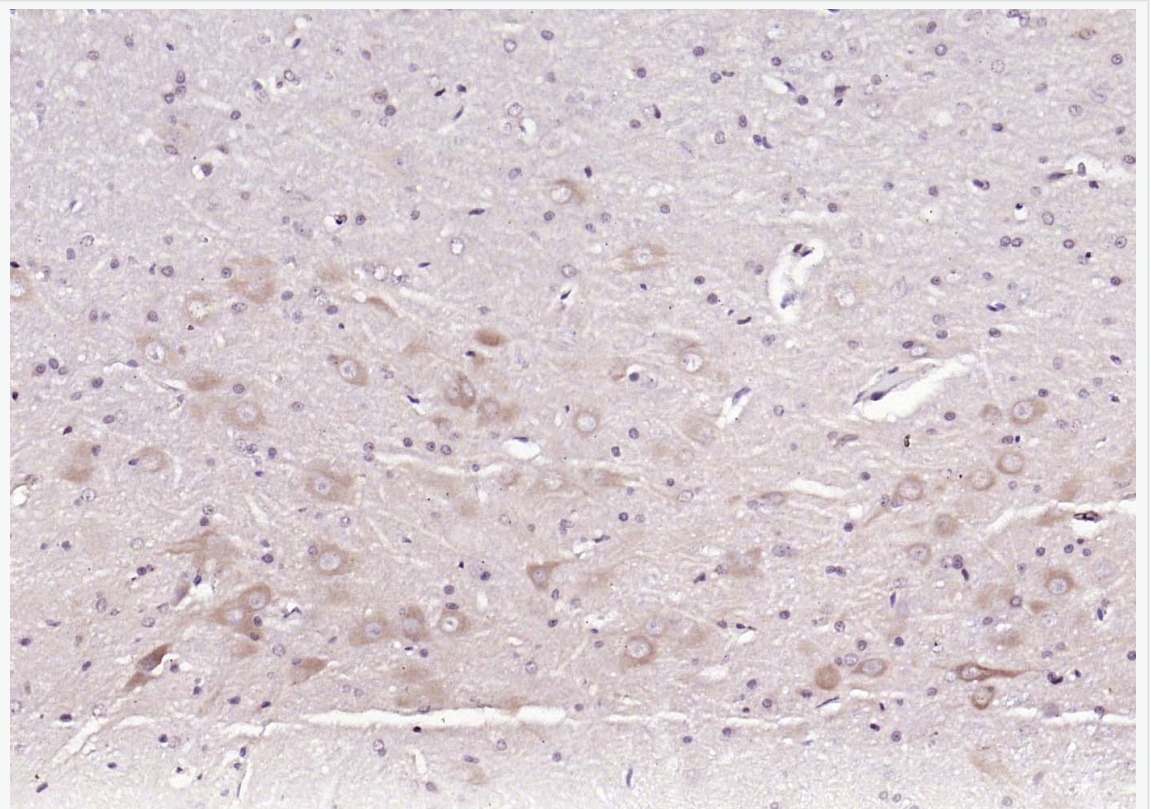
[SwissProt: Q61555](#) Mouse

[Unigene: 519294](#) Human

[Unigene: 20271](#) Mouse

[Unigene: 22906](#) Rat

**Product
Picture**



Paraformaldehyde-fixed, paraffin embedded (rat brain); Antigen retrieval by boiling in sodium citrate buffer (pH6.0) for 15min; Block endogenous peroxidase by 3% hydrogen peroxide for 20 minutes; Blocking buffer (normal goat serum) at 37°C for 30min; Antibody incubation with (Fibrillin 2) Polyclonal Antibody, Unconjugated



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(SL12166R) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.