SunLong Biotech Co.,LTD
Tel: 0086-571- 56623320 Fax:0086-571- 56623318
E-mail:sales@sunlongbiotech.com
www.sunlongbiotech.com

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.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Chicken,Dog,Pig,Cow,Horse,Rabbit,Sheep,
Applications:	ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=1:100-500（Paraffin sections need antigen repair） not yet tested in other applications. optimal dilutions/concentrations should be determined by the end user.
Molecular weight:	311kDa
Cellular localization:	Secretory protein
Form:	Lyophilized or 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
Storage Buffer:	0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.
Storage:	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20°C. When reconstituted in sterile pH 7.4 0.01M PBS or diluent of antibody the antibody is stable for at least two weeks at 2-4 °C.
PubMed:	PubMed
Product Detail:	Extracellular glycoproteins fibrillin-1 and -2 are major components of connective tissue 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 arthrogryposis 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: 2201Human

Entrez Gene: 100047082Mouse

Entrez Gene: 14119Mouse

Entrez Gene: 689008Rat

Omim: 121050Human

SwissProt: P35556Human

SwissProt: Q61555Mouse

Unigene: 519294Human

	Unigene: 20271Mouse Unigene: 22906Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.