

Fibrillin 2 Rabbit pAb

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Catalog # AP54758

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P35556
Reactivity	Rat
Predicted	Pig, Dog, Human, Mouse, Rabbit, Chicken, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	314775
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Fibrillin 2
Epitope Specificity	1001-1200/2912
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	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.

Additional Information

Gene ID	2201
Other Names	Fibrillin-2, Placensin, FBN2 {ECO:0000303 PubMed:32329225, ECO:0000312 HGNC:HGNC:3604}
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500

Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.
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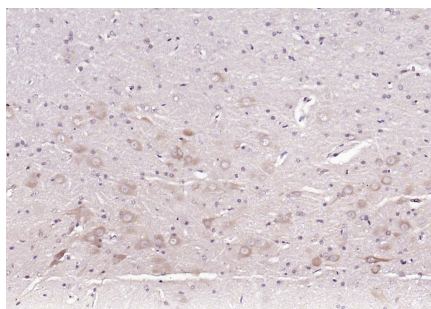
Protein Information

Name	FBN2 {ECO:0000303 PubMed:32329225, ECO:0000312 HGNC:HGNC:3604}
Function	[Fibrillin-2]: 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.
Cellular Location	Secreted. Note=Fibrillin-2 and Placensin chains are still linked together during the secretion from cells, but are subsequently separated by furin [Placensin]: Secreted. Note=Secreted by placental cells
Tissue Location	Almost exclusively expressed in placenta (PubMed:32329225). Expressed at much lower level in other tissues (PubMed:32329225). Expressed in fetal eye (18 weeks)in the retinal pigment epithelium (RPE), the choroid, Bruch's membrane and in the sclera (PubMed:24899048). Not expressed in the neural retina (PubMed:24899048).

Background

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Images



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 (AP54758) at 1:200 overnight at 4°C, followed by operating according to SP Kit(Rabbit) (sp-0023) instructions and DAB staining.

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