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Fibulin 5 Rabbit pAb

Catalog # AP98639

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9UBX5
Other Accession	Q9UBX5
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	50180
Physical State	Liquid
Immunogen	Recombinant human Fibulin 5 protein
Epitope Specificity	199-448/448
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the fibulin family.Contains 6 EGF-like domains.
SUBUNIT	Homodimer.
DISEASE	Cutis laxa, autosomal dominant, 2 (ADCL2) [MIM:614434]: A connective tissue disorder characterized by loose, hyperextensible skin with decreased resilience and elasticity leading to a premature aged appearance. Face, hands, feet, joints, and torso may be differentially affected. Additional variable clinical features are gastrointestinal diverticula, hernia, and genital prolapse. Rare manifestations are pulmonary artery stenosis, aortic aneurysm, bronchiectasis, and emphysema. Note=The disease is caused by mutations affecting the gene represented in this entry. Cutis laxa, autosomal recessive, 1A (ARCL1A) [MIM:219100]: A connective tissue disorder characterized by loose, hyperextensible skin with decreased resilience and elasticity leading to a premature aged appearance. Face, hands, feet, joints, and torso may be differentially affected. The clinical spectrum of autosomal recessive cutis laxa is highly heterogeneous with respect to organ involvement and severity. Type I autosomal recessive cutis laxa is a specific, life-threatening disorder with organ involvement, lung atelectasis and emphysema, diverticula of the gastrointestinal and genitourinary systems, and vascular anomalies. Associated cranial anomalies, late closure of the fontanel, joint laxity, hip dislocation, and inguinal hernia have been observed but are uncommon. Note=The disease is caused by mutations affecting the gene represented in this entry.Age-related macular degeneration 3 (ARMD3) [MIM:608895]: A form of age-related macular degeneration, a multifactorial eye disease and the most common cause of irreversible vision loss in the developed world. In most patients, the disease is manifest as ophthalmoscopically visible yellowish accumulations of protein and lipid that lie beneath the retinal pigment epithelium and within an elastin-containing structure known as Bruch membrane. Note=Disease susceptibility is associated with variations affecting the gene represented in this entry.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Fibulin 5: A protein that belongs to a family of extracellular proteins

expressed in the basement membranes of blood vessels. Fibulin 5 may be essential for the polymerization of elastin. Missense mutations in FBLN5, the gene that encodes fibulin 5, appear responsible for 1-2% of cases of age-related macular degeneration (AMD). FBLN5 is located on chromosome 14 in band 14q32.1. See also: Fibulin 3. May play a role in vascular growth and maturation during development and in lesions of injured vessels.

Additional Information

Gene ID	10516
Other Names	ADCL2; ARCL1A; ARMD3; CMT1H; DANCE; EVEC; FIBL-5; HNARMD; UP50; A55; FBLN5_HUMAN; FBLN5; Developmental arteries and neural crest EGF-like protein (Dance); Urine p50 protein (UP50); FBLN5_MOUSE; fibulin 5; embryonic vascular EGF-like repeat-containing protein; developmental arteries and neural crest EGF-like
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,ICC/IF=1:50-200
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.

Protein Information

Name	FBLN5
Synonyms	DANCE
Function	Essential for elastic fiber formation, is involved in the assembly of continuous elastin (ELN) polymer and promotes the interaction of microfibrils and ELN (PubMed:18185537). Stabilizes and organizes elastic fibers in the skin, lung and vasculature (By similarity). Promotes adhesion of endothelial cells through interaction of integrins and the RGD motif. Vascular ligand for integrin receptors which may play a role in vascular development and remodeling (PubMed:10428823). May act as an adapter that mediates the interaction between FBN1 and ELN (PubMed:17255108).
Cellular Location	Secreted. Secreted, extracellular space, extracellular matrix. Note=co-localizes with ELN in elastic fibers.
Tissue Location	Expressed in skin fibroblasts (at protein level) (PubMed:17035250). Expressed predominantly in heart, ovary, and colon but also in kidney, pancreas, testis, lung and placenta. Not detectable in brain, liver, thymus, prostate, or peripheral blood leukocytes (PubMed:10428823).

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