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EFEMP2 Rabbit pAb

Catalog # AP99624

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	O95967
Other Accession	O95967
Predicted	Rat, Pig, Human, Mouse, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	49405
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human EFEMP2/Fibulin 4
Epitope Specificity	331-443/443
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.
DISEASE	Defects in EFEMP2 are a cause of cutis laxa autosomal recessive type 1 (ARCL1) [MIM:219100]. Hereditary cutis laxa refers to a heterogeneous group of connective tissue disorders characterized by cutaneous abnormalities and variable systemic manifestations. The most constant clinical feature is loose skin, sagging over the face and trunk. Hereditary cutis laxa is inherited in both autosomal dominant and autosomal recessive modes. ARCL1 shows the most severe phenotype and has the poorest prognosis. In addition to the skin, internal organs enriched in elastic fibers, such as the lung and arteries, are affected
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Fibulin proteins contribute to normal development of elastic fiber systems in various types of organs that require elasticity, such as vasculature, lung and skin. Fibulin-4, also known as EFEMP2 (EGF-containing fibulin-like extracellular matrix protein 2), MBP1 or UPH1 is a 443 amino acid secreted protein that contains six EGF-like calcium-binding domains and belongs to the fibulin family. Expressed ubiquitously with highest expression in heart, Fibulin-4 is essential for connective tissue development and elastic fiber formation, and may also play an important role in vascular patterning and collagen biosynthesis. Defects in the gene encoding Fibulin-4 are associated with autosomal recessive cutis laxa type I (CL type I), a connective tissue disorder that is inherited in both an autosomal dominant and an autosomal recessive manner and is characterized by inelastic tissue in all affected areas of the body.

Additional Information

Gene ID	30008

Other Names	ARCL1B; FBLN4; MBP1; UPH1; FBLN4_HUMAN; EFEMP2; Fibulin-4 (FIBL-4); Protein UPH1; EGF-like fibulin extracellular matrix protein 2; EGF containing fibulin extracellular matrix protein 2; EGF-containing fibulin-like extracellular matrix protein 2; EGF containing fibulin like extracellular matrix protein 2; fibulin 4
Dilution	WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	EFEMP2 (HGNC:3219)
Synonyms	FBLN4
Function	Plays a crucial role in elastic fiber formation in tissue, and in the formation of ultrastructural connections between elastic laminae and smooth muscle cells in the aorta, therefore participates in terminal differentiation and maturation of smooth muscle cell (SMC) and in the mechanical properties and wall integrity maintenance of the aorta (PubMed:27339457). In addition, is involved in the control of collagen fibril assembly in tissue throught proteolytic activation of LOX leading to cross- linking of collagen and elastin (By similarity). Also promotes ELN coacervation and participates in the deposition of ELN coacervates on to microfibrils but also regulates ELN cross-linking through LOX interaction (PubMed:18973305, PubMed:19570982). Moreover adheres to the cells through heparin binding in a calcium-dependent manner and regulates vascularlar smooth muscle cells proliferation through angiotensin signaling (PubMed:23782690).
Cellular Location	Secreted, extracellular space, extracellular matrix Secreted, extracellular space, extracellular matrix, basement membrane {ECO:0000250|UniProtKB:Q9WVJ9}. Note=Localizes on the microfibrils surrounding ELN cores. {ECO:0000250|UniProtKB:Q9WVJ9}

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