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Rabbit Anti-EFEMP2 antibody

SL13059R

Product Name:	EFEMP2
Chinese Name:	纤连蛋白4抗体
Alias:	Fibulin 4; EFEMP2; EGF containing fibulin like extracellular matrix protein 2; EGF-containing fibulin-like extracellular matrix protein 2; FBLN 4; FBLN4; FBLN4_HUMAN; FIBL 4; FIBL-4; FIBL4; Fibulin4; Fibulin-4; MBP 1; MBP1; Mutant p53 binding protein 1; Protein UPH1; UPH 1; UPH1; UPH1 protein.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Sheep,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800ICC=1:100-500IF=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:	47kDa
Cellular localization:	Secretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human EFEMP2/Fibulin 4:331-443/443
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:	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.

Subcellular Location:
Secreted.

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

Similarity:
Belongs to the fibulin family.
Contains 6 EGF-like domains.

SWISS:
O95967

Gene ID:
30008

Database links:

Entrez Gene: 30008Human

Omim: 604633Human

SwissProt: O95967Human

Unigene: 170622Human

Important Note:
This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.