

Product name:	ECM1 Rabbit Polyclonal Antibody
Cat number:	ABN10284
Conjugate:	Unconjugated
Size:	100µL
Clone:	Polyclonal
Concentration:	1mg/ml
Host:	Rabbit
Isotype:	IgG
Immunogen:	Synthesized peptide derived from the N-terminal region of human ECM1.
Reactivity:	Human,Rat,Mouse
Applications:	IHC 1:100-1:300,ICC/IF 1:50-1:200,ELISA 1:10000-1:20000
Purification:	Affinity purification
Form:	Liquid
Buffer:	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% New type preservative N.
Storage:	Store at 4°C short term. Aliquot and store at -20°C for 12 months. Avoid freeze/thaw cycles.
Background:	This gene encodes a soluble protein that is involved in endochondral bone formation, angiogenesis, and tumor biology. It also interacts with a variety of extracellular and structural proteins, contributing to the maintenance of skin integrity and homeostasis. Mutations in this gene are associated with lipid proteinosis disorder (also known as hyalinosis cutis et mucosae or Urbach-Wiethe disease) that is characterized by generalized thickening of skin, mucosae and certain viscera. Alternatively spliced transcript variants encoding distinct isoforms have been described for this gene. [provided by RefSeq, Feb 2011],disease:Defects in ECM1 are the cause of lipid proteinosis (LiP) [MIM:247100]; also known as lipid proteinosis of Urbach and Wiethe or hyalinosis cutis et mucosae. LiP is a rare autosomal recessive disorder characterized by generalized thickening of skin, mucosae and certain viscera. Classical features include beaded eyelid papules and laryngeal infiltration leading to hoarseness. Histologically, there is widespread deposition of hyaline material and disruption/reduplication of basement membrane.,