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Rabbit Anti-ANTXR2 antibody

SL11071R

Product Name:	ANTXR2
Chinese Name:	炭疽毒素受体2抗体
Alias:	CMG2; ISH; JHF; anthrax toxin receptor 2; ANTR2_HUMAN; Antxr2; Capillary morphogenesis gene 2 protein; CMG 2; CMG-2.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,Dog,Pig,Cow,Horse,Rabbit,
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:	50kDa
Cellular localization:	cytoplasmicThe cell membraneExtracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ANTXR2:101-200/489<Extracellular>
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:	This gene encodes a receptor for anthrax toxin. The protein binds to collagen IV and laminin, suggesting that it may be involved in extracellular matrix adhesion. Mutations in this gene cause juvenile hyaline fibromatosis and infantile systemic hyalinosis. Multiple transcript variants encoding different isoforms have been found for this gene. [provided by RefSeq, Mar 2009].

Function:
Necessary for cellular interactions with laminin and the extracellular matrix.

Subunit:
Binds laminin, and possibly also collagen type IV. Binds to the protective antigen (PA) of Bacillus anthracis in a divalent cation-dependent manner, with the following preference: calcium > manganese > magnesium > zinc. Binding of PA leads to heptamerization of the receptor-PA complex.

Subcellular Location:
Secreted; Cell membrane. Expressed at the cell surface and Endoplasmic reticulum membrane. Expressed predominantly within the endoplasmic reticulum and not at the plasma membrane.

Tissue Specificity:
Expressed in prostate, thymus, ovary, testis, pancreas, colon, heart, kidney, lung, liver, peripheral blood leukocytes, placenta, skeletal muscle, small intestine and spleen.

DISEASE:
Defects in ANTXR2 are the cause of infantile systemic hyalinosis (ISH). This autosomal recessive syndrome is similar to JHF, but has an earlier onset and a more severe course. Symptoms appear at birth or within the first months of life, with painful, swollen joint contractures, osteopenia, osteoporosis and livid red hyperpigmentation over bony prominences. Patients develop multiple subcutaneous skin tumors and gingival hypertrophy. Hyaline deposits in multiple organs, recurrent infections and intractable diarrhea often lead to death within the first 2 years of life. Surviving children may suffer from severely reduced mobility due to joint contractures.

Similarity:
Belongs to the ATR family.
Contains 1 VWFA domain.

SWISS:
P58335

Gene ID:
118429

Database links:

Entrez Gene: 118429Human

Entrez Gene: 71914Mouse

Entrez Gene: 305633Rat

	Omim: 608041Human SwissProt: P58335Human SwissProt: Q6DFX2Mouse Unigene: 162963Human Unigene: 24842Mouse Unigene: 229072Rat **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.