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Rabbit Anti-ADAMTSL2 antibody

SL5862R

Product Name:	ADAMTSL2
Chinese Name:	整合素样金属蛋白酶与凝血酶样2蛋白抗体
Alias:	ADAMTS like 2; ADAMTS like protein 2; ADAMTS-like protein 2; ADAMTSL 2; ADAMTSL-2; ADAMTSL2; ATL2_HUMAN.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human,Mouse,Rat,
Applications:	WB=1:500-2000ELISA=1:500-1000IHC-P=1:400-800IHC-F=1:400-800IF=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:	102kDa
Cellular localization:	Secretory protein
Form:	Lyophilized or Liquid
Concentration:	1mg/ml
immunogen:	KLH conjugated synthetic peptide derived from human ADAMTSL2:522-580/951
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:	ADAMTS (A Disintegrin And Metalloproteinase Domain with Thrombospondin type 1 Modules) is a family of zinc-dependent proteases that are implicated in a variety of normal and pathological conditions, including arthritis and cancer. ADAMTS protein family members contain an amino-terminal propeptide domain, a metalloproteinase domain, a disintegrin-like domain and a carboxy-terminus that contains a varying number of Thrombospondin type 1 (TSP-1) motifs. ADAMTS-L2 (ADAMTS-like

protein 2) is a 951 amino acid secreted protein that is highly expressed in lung, kidney and liver. Mutations in the gene encoding ADAMTS are the cause of geleophysic dysplasia, an autosomal recessive disorder characterized by cardiac vavular anomalies, short stature, thick skin and brachydactyly. In individuals affected with geleophysic dysplasia, there is a significant increase in total active TGF-beta 1 and nuclear locations of p-SAMD2 in fibroblasts. Interestingly, ADAMTS-L2 interacts with LTBP-1, a glycoprotein that is part of the platelet-derived TGF-beta 1 complex.

Function:
Defects in ADAMTSL2 are the cause of geleophysic dysplasia [MIM:231050]. Geleophysic dysplasia is an autosomal recessive disorder characterized by short stature, brachydactyly, thick skin and cardiac valvular anomalies often responsible for an early death.

Subcellular Location:
Secreted.

Post-translational modifications:
Glycosylated (By similarity). Can be O-fucosylated by POFUT2 on a serine or a threonine residue found within the consensus sequence C1-X(2)-(S/T)-C2-G of the TSP type-1 repeat domains where C1 and C2 are the first and second cysteine residue of the repeat, respectively. Fucosylated repeats can then be further glycosylated by the addition of a beta-1,3-glucose residue by the glucosyltransferase, B3GALTL. Fucosylation mediates the efficient secretion of ADAMTS family members. Also can be C-glycosylated with one or two mannose molecules on tryptophan residues within the consensus sequence W-X-X-W of the TPRs, and N-glycosylated. These other glycosylations can also facilitate secretion (By similarity).

DISEASE:
Defects in ADAMTSL2 are the cause of geleophysic dysplasia (GLPD) [MIM:231050]. Geleophysic dysplasia is an autosomal recessive disorder characterized by short stature, brachydactyly, thick skin and cardiac valvular anomalies often responsible for an early death.

Similarity:
Contains 1 PLAC domain.
Contains 7 TSP type-1 domains.

SWISS:
Q86TH1

Gene ID:
9719

Database links:

	Entrez Gene: 9719Human Entrez Gene: 77794Mouse Omim: 612277Human SwissProt: Q86TH1Human SwissProt: Q7TSK7Mouse Unigene: 522543Human Unigene: 330088Mouse **Important Note:** This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. Extracellular matrix蛋白