

Rabbit Anti-FREM1 antibody

SL13213R

Product Name:	FREM1
Chinese Name:	Extracellular matrix蛋白FREM1抗体
Alias:	BC037594; BNAR; C9orf143; C9orf145; C9orf154; D430009N09; D630008K06; FLJ25461; FRAS1-related extracellular matrix protein 1; FREM 1; FREM1; FREM1_HUMAN; Heb; MOTA; Protein QBRICK; QBRICK; RGD1306981; RP11-265B7.2; RP23-410K19.1; TILRR.
Organism Species:	Rabbit
Clonality:	Polyclonal
React Species:	Human, Mouse, Rat, Dog, Horse,
Applications:	ELISA=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:	242kDa
Cellular localization:	The cell membraneExtracellular matrixSecretory protein
Form:	Lyophilized or Liquid
Concentration:	lmg/ml
immunogen:	KLH conjugated synthetic peptide derived from human FREM1:1201-1300/2179
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:	<u>PubMed</u>
Product Detail:	FREM1 is a 2,179 amino acid protein that contains one C-type lectin domain, one Calxbeta domain and twelve CSPG repeats. Localized to the basement membrane of embryonic epidermal cells and secreted into extracellular space, FREM1 functions as an extracellular matrix protein that is essential for epidermal adhesion during

embryogenesis and may also participate in epidermal differentiation. FREM1 exists as multiple alternatively spliced isoforms and is encoded by a gene which maps to human chromosome 9. Chromosome 9 contains 145 million base pairs and comprises 4% of the human genome, encoding nearly 900 genes. Hereditary hemorrhagic telangiectasia, which is characterized by harmful vascular defects, and Familial dysautonomia, are both associated with chromosome 9. Notably, chromosome 9 encompasses the largest interferon family gene cluster.

Function:

Extracellular matrix protein that plays a role in epidermal differentiation and is required for epidermal adhesion during embryonic development

Subcellular Location:

Secreted; extracellular space; extracellular matrix; basement membrane. Localizes at the basement membrane zone of embryonic epidermis and hair follicles.

DISEASE:

Defects in FREM1 are the cause of bifid nose with or without anorectal and renal anomalies (BNAR) [MIM:608980]. A bifid nose is a rare congenital deformity due to failure of the paired nasal processes to fuse to a single midline organ during early gestation. BNAR is an autosomal recessive disorder and patients usually present a bifid nose associated with renal and anorectal malformations.

Similarity:

Belongs to the FRAS1 family.

Contains 1 C-type lectin domain.

Contains 1 Calx-beta domain.

Contains 12 CSPG (NG2) repeats.

SWISS:

Q5H8C1

Gene ID:

158326

Database links:

Entrez Gene: 158326Human

Entrez Gene: 329872Mouse

Omim: 608944Human

SwissProt: Q5H8C1Human

SwissProt: Q684R7Mouse

Unigene: 50850Human

Unigene: 242337 Mouse
Important Note: This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

