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FAM20C Rabbit pAb

Catalog # AP98852

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	Q8IXL6
Other Accession	Q8IXL6
Reactivity	Rat
Predicted	Human, Mouse, Monkey, Chicken, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	66234
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human FAM20C
Epitope Specificity	351-450/584
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the FAM20 family.
DISEASE	Defects in FAM20C are the cause of Raine syndrome (RNS) [MIM:259775]. RNS is an autosomal recessive osteosclerotic bone dysplasia with neonatal lethal outcome. Clinical features include generalized osteosclerosis, craniofacial dysplasia and microcephaly.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The FAM20 proteins are a family of secreted proteins that regulate differentiation and function of hematopoietic and other tissues. FAM20C, also known as DMP4 (Dentin matrix protein 4), is a 570 amino acid secreted protein that binds calcium and may play a role in dentin mineralization. Defects in the gene encoding FAM20C are the cause of Raine syndrome (Lethal osteosclerotic bone dysplasia), an autosomal recessive osteosclerotic bone dysplasia, that is characterized by generalized osteosclerosis, microencephaly and craniofacial dysplasia. Usually, affected individuals survive only days or weeks. The mutations of the FAM20C gene include four nonsynomous base changes and four splice-site changes that have a detrimental affect on splicing.

Additional Information

Gene ID	56975
Other Names	DMP-4; DMP4; G-CK; GEF-CK; RNS; mKIAA4081; RGD1311980; FA20C_HUMAN; FAM20C; Dentin matrix protein 4 (DMP-4); Golgi casein kinase; Golgi-enriched fraction casein kinase (GEF-CK); 2.7.11.1; FA20C_MOUSE; Kiaa4081; FAM20C golgi associated secretory pathway kinase; family with sequence similarity 20, member C; FAM20C, golgi associated secretory pathway kinase; dentin matrix protein 4

Dilution	IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500
Storage	Store at -20 °C for one year. Avoid repeated freeze/thaw cycles. 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.

Protein Information

Name	FAM20C (HGNC:22140)
Function	Golgi serine/threonine protein kinase that phosphorylates secretory pathway proteins within Ser-x-Glu/pSer motifs and plays a key role in biomineralization of bones and teeth (PubMed:22582013, PubMed:23754375, PubMed:25789606). Constitutes the main protein kinase for extracellular proteins, generating the majority of the extracellular phosphoproteome (PubMed:26091039). Mainly phosphorylates proteins within the Ser-x-Glu/pSer motif, but also displays a broader substrate specificity (PubMed:26091039). Phosphorylates ERO1A, enhancing its activity which is required to maintain endoplasmic reticulum redox homeostasis and for oxidative protein folding (PubMed:29858230, PubMed:34349020). During endoplasmic reticulum stress, phosphorylates P4HB/PDIA1 which induces a functional switch, causing P4HB to change from an oxidoreductase to a molecular chaperone (PubMed:32149426). This is critical to maintain ER proteostasis and reduce cell death under ER stress (PubMed:32149426). Phosphorylation of P4HB also promotes its interaction with ERN1, leading to reduced activity of ERN1, a key sensor for the endoplasmic reticulum unfolded protein response (PubMed:32149426). Required for osteoblast differentiation and mineralization (PubMed:34349020). Phosphorylates casein as well as a number of proteins involved in biomineralization such as AMELX, AMTN, ENAM and SPP1/OPN (PubMed:22582013, PubMed:25789606, PubMed:34349020). In addition to its role in biomineralization, also plays a role in lipid homeostasis, wound healing and cell migration and adhesion (PubMed:26091039).
Cellular Location	Golgi apparatus membrane; Single-pass type II membrane protein. Secreted. Endoplasmic reticulum. Note=Resides in the Golgi apparatus membrane and is secreted following propeptide cleavage (PubMed:34349020). Retained in the endoplasmic reticulum (ER) in response to ER stress where it phosphorylates P4HB (PubMed:32149426)
Tissue Location	Widely expressed..

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