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DSPP Rabbit pAb

Catalog # AP97763

Product Information

Application	WB, IHC-P, IHC-F, IF
Primary Accession	Q9NZW4
Other Accession	Q9NZW4
Reactivity	Human
Host	Rabbit
Clonality	Polyclonal
Calculated MW	131151
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Dentin sialoprotein
Epitope Specificity	31-130/1301
Isotype	IgG
Purity	affinity purified by Protein A
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SUBUNIT	Interacts with FBLN7.
Post-translational modifications	DSP is glycosylated.
DISEASE	Defects in DSPP are the cause of deafness autosomal dominant type 39 with dentinogenesis imperfecta 1 (DFNA39/DGI1) [MIM:605594]. Affected individuals present DGI1 associated with early onset progressive sensorineural high-frequency hearing loss. Defects in DSPP are the cause of dentinogenesis imperfecta type 1 (DGI1) [MIM:125490]; also known as dentinogenesis imperfecta Shields type 2 (DGI2). DGI1 is an autosomal dominant disorder in which both the primary and the permanent teeth are affected. It occurs with an incidence of 1:8000 live births. The teeth are amber and opalescent, the pulp chamber being obliterated by abnormal dentin. The enamel, although unaffected, tends to fracture, which makes dentin undergo rapid attrition, leading to shortening of the teeth. Defects in DSPP are a cause of dentinogenesis imperfecta Shields type 3 (DGI3) [MIM:125500]. Patients with DGI3 do not have stigmata of osteogenesis imperfecta. The finding that a single defects in the DSPP gene causes both phenotypic patterns of DGI2 and DGI3 strongly supports the conclusion that these two disorders are not separate diseases but rather the phenotypic variation of a single genetic defect. Defects in DSPP are the cause of dentin dysplasia type 2 (DTDP2) [MIM:125420]; also known as dentin dysplasia Shields type 2. DTDP2 is an autosomal dominant disorder in which mineralization of the dentine of the primary teeth is abnormal. On the basis of the phenotypic overlap between, and shared chromosomal location with DGI2 it has been proposed that DTDP2 and DGI2 are allelic. From the results of recent studies, it is clear that different types of mutations in DSPP lead to the two different phenotypes.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene encodes two principal proteins of the dentin extracellular matrix of the tooth. The preproprotein is secreted by odontoblasts and cleaved into dentin sialoprotein and dentin phosphoprotein. Dentin phosphoprotein is thought to be involved in the biomineralization process of dentin. Mutations

in this gene have been associated with dentinogenesis imperfecta-1; in some individuals, dentinogenesis imperfecta occurs in combination with an autosomal dominant form of deafness. Allelic differences due to repeat polymorphisms have been found for this gene. [provided by RefSeq, Jul 2008]

Additional Information

Gene ID	1834
Other Names	DFNA39; DGI1; DMP3; DPP; DSP; DSPP_HUMAN; DSPP; dentin sialophosphoprotein
Dilution	WB=1:500-2000,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	DSPP
Function	DSP may be an important factor in dentinogenesis. DPP may bind high amount of calcium and facilitate initial mineralization of dentin matrix collagen as well as regulate the size and shape of the crystals.
Cellular Location	Secreted, extracellular space, extracellular matrix
Tissue Location	Expressed in teeth. DPP is synthesized by odontoblast and transiently expressed by pre-ameloblasts

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