10320 Camino Santa Fe, Suite G
San Diego, CA 92121
Tel: 858.875.1900 Fax: 858.875.1999

Desmoplakin I+II Rabbit pAb

Catalog # AP99534

Product Information

Application	IHC-P, IHC-F, IF, E
Primary Accession	P15924
Other Accession	P15924
Predicted	Rat, Human, Mouse, Rabbit, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	331774
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human DP-I
Epitope Specificity	271-370/2871
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 plakin or cytolinker family. Contains 17 plectin repeats. Contains 1 SH3 domain. Contains 6 spectrin repeats.
SUBUNIT	Homodimer. Interacts with COL17A1 (via cytoplasmic region). Associates (via C-terminal) with KRT5-KRT14 (via rod region), KRT8-KRT18 and VIM intermediate filaments. Interacts with DSC2.
Post-translational modifications	Ser-2849 is probably phosphorylated by a cAMP-dependent protein kinase. Phosphorylation on Ser-2849 probably affects its association with epidermal, simple cytokeratins and VIM intermediate filaments.Substrate of transglutaminase. Some glutamines and lysines are cross-linked to other desmoplakin molecules, to other proteins such as keratin, envoplakin, periplakin and involucrin, and to lipids like omega-hydroxyceramide.
DISEASE	Defects in DSP are the cause of palmoplantar keratoderma striate type 2 (SPPK2) [MIM:612908]; also known as keratosis palmoplantaris striata II. SPPK2 is characterized by skin thickening in the palms (linear pattern) and the soles (island-like pattern) and flexor aspect of the fingers. Abnormalities of the nails, the teeth and the hair are rarely present. Defects in DSP are the cause of cardiomyopathy dilated with woolly hair and keratoderma (DCWHK) [MIM:605676]; also known as Carvajal syndrome or palmoplantar keratoderma with left ventricular cardiomyopathy and woolly hair. DCWHK is an autosomal recessive cardiocutaneous syndrome characterized by a generalized striate keratoderma particularly affecting the palmoplantar epidermis, woolly hair, and Defects in DSP are the cause of familial arrhythmogenic right ventricular dysplasia type 8 (ARVD8) [MIM:607450]; also known as arrhythmogenic right ventricular cardiomyopathy 8 (ARVC8). ARVD is an autosomal dominant disease characterized by partial degeneration of the myocardium of the right ventricle, electrical instability, and sudden death. It is clinically defined by electrocardiographic and angiographic criteria; pathologic findings, replacement of ventricular myocardium with fatty and fibrous elements, preferentially involve the right ventricular free wall. Defects in DSP are the cause of skin fragility-woolly hair syndrome (SFWHS) [MIM:607655]. SFWHS is an autosomal recessive genodermatosis characterized by focal and diffuse palmoplantar keratoderma, hyperkeratotic

	plaques on the trunk and limbs, and woolly hair with varying degrees of alopecia. Defects in DSP are the cause of epidermolysis bullosa lethal acantholytic (EBLA) [MIM:609638]. EBLA is characterized by severe fragility of skin and mucous membranes. The phenotype is lethal in the neonatal period because of immense transcutaneous fluid loss. Typical features include universal alopecia, neonatal teeth, and nail loss. Histopathology of the skin shows suprabasal clefting and acantholysis throughout the spinous layer, mimicking pemphigus.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Desmosomes are intercellular junctions that form tight links between adjacent cells. Desmoplakin is an obligate component of functional desmosomes that attaches intermediate filaments to desmosomal plaques. It is involved in the organization of desmosomal cadherin-plakoglobin complexes into discrete plasma membrane domains. The N-terminus of desmoplakin is essential for localisation to the desmosome and interaction with plakophilin 1 and plakoglobin. The C-terminus of desmoplakin binds to intermediate filaments. The central region of desmoplakin comprises a coiled-coil rod domain that mediates homodimerisation. There are two isoforms of desmoplakin - desmoplakin I, which is an obligate component of all desmosomes, and desmoplakin II, which is predominantly expressed in tissues and cells of stratified origin. Mutations in the gene encoding desmoplakin result in a number of cardiomyopathies and keratodermas as well as the autoimmune disease paraneoplastic pemphigus.

Additional Information

Gene ID	1832
Other Names	DESP_HUMAN; DSP; DP; 250/210 kDa paraneoplastic pemphigus antigen; DESP_MOUSE; desmoplakin; desmoplakin (DPI, DPII)
Dilution	IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000
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	DSP (HGNC:3052)
Function	A component of desmosome cell-cell junctions which are required for positive regulation of cellular adhesion (PubMed:25733715). Critical for cell-cell adhesion in early stage blastocysts and progression through proamniotic cavity formation (By similarity). Not required for preimplantation morphogenic process in blastocysts (By similarity). Required for keratin filament anchoring at the desmosome junction and subsequent organization of the keratin intermediate filament network within the cytoplasm (By similarity). Required for anchoring of desmosomes to the microtubule architecture, via its interaction with NIN (By similarity). Promotes microtubule- mediated GJA1/CX43 trafficking to cell membranes via its interaction with MAPRE1/EB1, thereby facilitating gap junction intracellular communication (PubMed:25225338). Plays a key role in adhesion and organization of the dermal epithelial barrier (PubMed:26604139). Critical for

	the maintenance of the neural tube structure following formation and organization of the neuroepithelium (By similarity). Facilitates outgrowth and repair of motor neuron fibers in regenerating axons following injury, probably by promoting recruitment of a complex containing DSP, CDH2, VIM and JUP to the outgrowth tips (By similarity). Critical for the normal formation of heart and myocardial tissue during early embryogenesis (By similarity). Also required for development of vascular capillary structures and intact endothelial cell barriers (By similarity). Regulates profibrotic gene expression in cardiomyocytes via activation of the MAPK14/p38 MAPK signaling cascade and increase in TGFB1 protein abundance (By similarity). Maintains cardiac rhythmicity by ensuring correct cell-cell adhesion within the sinoatrial node, via stabilization of protein components of both desmosome and Gap junctions (By similarity). Involved in maintaining the protein stability and recruitment of GJA1 to functional gap junctions, via inhibition of KRAS-mediated MAPK1/MAPK3 phosphorylation of GJA1 (By similarity). Negative regulator of cell cycle progression and differentiation in keratinocytes, potentially via inhibition of MAPK and phosphoinositide-3-kinase (PI3K) signaling pathways (PubMed:17475244). Mediates the interaction between the desmosome and COP9 signalosome complex (CSN) protein complex (PubMed:28891468). As a result of this interaction, promotes keratinocyte differentiation via deneddylation of EGFR resulting in a reduction in EGFR protein stabilization and translocation away from the cell membrane (PubMed:28891468). Required for the maintenance of protein abundance of desmosome junction components DSG1, DSG2, DSC2, DSC3, PKP1, PKP2 and PKP3 (PubMed:17475244, PubMed:26073755). Required for the survival and maintenance of germ cells in the gonads during embryonic development (By similarity). Binds to telomere DNA (via C-terminus) and acts to prevent telomere damage and maintain telomere length via its interaction with TRF2 (PubMed:31595153).
Cellular Location	Cell projection, axon {ECO:0000250|UniProtKB:E9Q557}. Cell junction, desmosome. Cell membrane. Cytoplasm. Nucleus. Note=Localizes to desmosome precursor particles in the cytoplasm (PubMed:25208567). Localizes to the cytoplasm in undifferentiated keratinocytes however becomes localizes to both lateral and tricellular cell-cell contacts as differentiation progresses and as epithelial sheet formation completes (By similarity) Localizes to the interface of the cortical keratin network and endodermal cell periphery in developing embryos (By similarity) {ECO:0000250|UniProtKB:E9Q557, ECO:0000269|PubMed:25208567}
Tissue Location	Expressed in oral mucosa (at protein level) (PubMed:30479852). Expressed in arrector pili muscle (at protein level) (PubMed:29034528). Expressed in the heart (at protein level) (PubMed:18662195, PubMed:32410525). Expressed in keratinocytes of the skin (at protein level) (PubMed:26604139) [Isoform DPII]: Resides predominantly in tissues and cells of stratified origin

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