桥粒斑蛋白 2 抗体

产品货号： mlR1749

英文名称： Desmoplakin-II

中文名称： 桥粒斑蛋白 2 抗体

别　　名： 250/210 kDa paraneoplastic pemphigus antigen; Desmoplakin (DPI DPII); Desmoplakin; Desmoplakin I; Desmoplakin II; DESP_HUMAN; DP; DP I; DP II; DPI; DPII; DSP; KPPS2; PPKS 2; PPKS2.

研究领域： 免疫学

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应： Human, Mouse, Rat, Chicken, Cow, Horse,

产品应用： WB=1:500-2000 ELISA=1:500-1000 IHC-P=1:400-800 IHC-F=1:400-800 IF=1:100-500 （石蜡切片需做抗原修复）

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 332kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from human Desmoplakin-II:261-360/2871

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： 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

产品介绍： 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.

Function:

Major high molecular weight protein of desmosomes. Involved in the organization of the desmosomal cadherin-plakoglobin complexes into discrete plasma membrane domains and in the anchoring of intermediate filaments to the desmosomes.

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.

Subcellular Location:

Cell junction, desmosome. Cytoplasm, cytoskeleton. Note=Innermost portion of the desmosomal plaque. Colocalizes with epidermal KRT5-KRT14 and simple KRT8-KRT18 keratins and VIM intermediate filaments network.

Tissue Specificity:

Isoform DPI is apparently an obligate constituent of all desmosomes. Isoform DPII resides predominantly in tissues and cells of stratified origin.

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.

Similarity:

Belongs to the plakin or cytolinker family.

Contains 17 plectin repeats.

Contains 1 SH3 domain.

Contains 6 spectrin repeats.

SWISS:

P15924

Gene ID:

1832

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.