



DESP rabbit pAb

Catalog No	YP-Ab-08942
Isotype	IgG
Reactivity	Human; Mouse
Applications	IHC;IF
Gene Name	DSP
Immunogen	Synthesized peptide derived from human DESP AA range: 1546-1596
Specificity	This antibody detects endogenous levels of DESP at Human/Mouse
Formulation	Liquid in PBS containing 50% glycerol, 0.5% BSA and 0.02% sodium azide.
Source	Polyclonal, Rabbit,IgG
Purification	The antibody was affinity-purified from rabbit serum by affinity-chromatography using specific immunogen.
Dilution	IHC-p 1:50-200. IF 1:50-200
Concentration	1 mg/ml
Purity	≥90%
Storage Stability	-20°C/1 year
Synonyms	DSP;DESP;DP;Desmoplakin
Observed Band	
Calculated Molecular Weight	
Cell Pathway	Cell junction, desmosome . Cytoplasm, cytoskeleton . Cell membrane . Innermost portion of the desmosomal plaque. Colocalizes with epidermal KRT5-KRT14 and simple KRT8-KRT18 keratins and VIM intermediate filaments network (PubMed:12802069). Localizes at the intercalated disk in cardiomyocytes (By similarity). .
Tissue Specificity	Expressed in oral mucosa (at protein level) (PubMed:30479852). Expressed in arrector pili muscle (at protein level) (PubMed:29034528). ; [Isoform DPI]: Apparently an obligate constituent of all desmosomes.; [Isoform DPII]: Resides predominantly in tissues and cells of stratified origin.
Function	disease:Defects in DSP are the cause of dilated cardiomyopathy 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 dilated left ventricular cardiomyopathy.,



disease: 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

Background

This gene encodes a protein that anchors intermediate filaments to desmosomal plaques and forms an obligate component of functional desmosomes. Mutations in this gene are the cause of several cardiomyopathies and keratodermas, including skin fragility-woolly hair syndrome. Alternative splicing results in multiple transcript variants. [provided by RefSeq, Jan 2016],

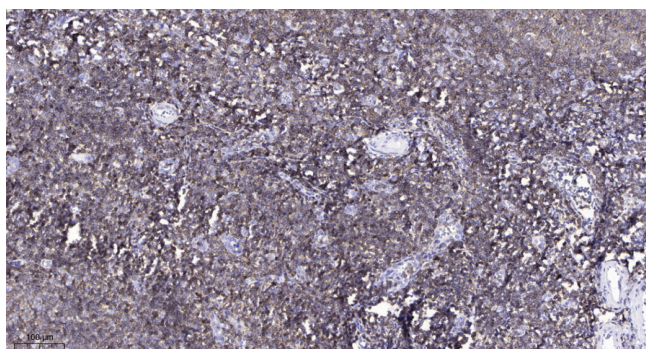
matters needing attention

Avoid repeated freezing and thawing!

Usage suggestions

This product can be used in immunological reaction related experiments. For more information, please consult technical personnel.

Products Images



Immunohistochemical analysis of paraffin-embedded human tonsil. 1, Antibody was diluted at 1:200 (4° overnight). 2, Tris-EDTA, pH 9.0 was used for antigen retrieval. 3, Secondary antibody was diluted at 1:200 (room temperature, 45min).