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connexin 30 Rabbit pAb

Catalog # AP99063

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O95452
Other Accession	O95452
Reactivity	Rat
Predicted	Human, Mouse, Chicken, Dog, Horse, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	30387
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human connexin-30
Epitope Specificity	161-261/260
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 connexin family. Beta-type (group I) subfamily.
SUBUNIT	A connexon is composed of a hexamer of connexins. Interacts with CNST
DISEASE	Defects in GJB6 are the cause of ectodermal dysplasia type 2 (ED2) [MIM:129500]; also known as Clouston syndrome. Ectodermal dysplasia defines a heterogeneous group of disorders due to abnormal development of two or more ectodermal structures. ED2 is an autosomal dominant condition characterized by atrichosis, nail hypoplasia and deformities, hyperpigmentation of the skin, normal teeth, normal sweat and sebaceous gland function. Palmoplantar hyperkeratosis is a frequent features. Hearing impairment has been detected in few cases of ED2.Defects in GJB6 are the cause of deafness autosomal recessive type 1B (DFNB1B) [MIM:612645]. DFNB1B is a form of sensorineural hearing loss. Sensorineural deafness results from damage to the neural receptors of the inner ear, the nerve pathways to the brain, or the area of the brain that receives sound information. Defects in GJB6 are the cause of deafness autosomal dominant type 3B (DFNA3B) [MIM:612643].
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The connexin family of proteins form hexameric complexes called connexons that facilitate movement of low molecular weight proteins between cells via gap junctions. Connexin proteins share a common topology of four transmembrane alpha-helical domains, two extracellular loops, a cytoplasmic loop and cytoplasmic N- and C-termini. Many of the key functional differences between connexins arise from specific amino-acid substitutions in the most highly conserved domains: the transmembrane and extracellular regions. Connexin 30, also known as GJB6 (Gap junction beta 6), ED2, EDH, HED or DFNA3, is a 261 amino acid multi-pass membrane protein that localizes to the cell junction and belongs to the connexin family. Functioning as a hexamer with other connexin proteins, connexin 30 facilitates the diffusion of low molecular weight materials from one cell to another. Defects in the gene encoding connexin 30 are the cause of ectodermal dysplasia type 2 (ED2) and

non-syndromatic sensorineural deafness autosomal dominant type 3 (DFNA3), the former of which is characterized by abnormal development of ectodermal structures (such as skin and nails).

Additional Information

Gene ID	10804
Other Names	CX30; DFNA3; DFNA3B; DFNB1B; ECTD2; ED2; EDH; HED; HED2; Cxnf; D14Bwg0506e; CXB6_HUMAN; GJB6; Connexin-30 (Cx30); CXB6_MOUSE; Cxn-30; gap junction protein beta 6; ectodermal dysplasia 2, hidrotic (Clouston syndrome); gap junction protein, beta 6 (connexin 30); gap junction protein, beta 6; gap junction protein, beta 6, 30kDa; connexin 30
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	GJB6
Function	One gap junction consists of a cluster of closely packed pairs of transmembrane channels, the connexons, through which materials of low MW diffuse from one cell to a neighboring cell.
Cellular Location	Cell membrane; Multi-pass membrane protein. Cell junction, gap junction

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