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connexin 30 Recombinant Rabbit mAb

Catalog # AP99170

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	O95452
Other Accession	O95452
Reactivity	Human, Mouse
Host	Rabbit
Clonality	Recombinant
Calculated MW	30387
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].
Background Descriptions	Gap junctions allow the transport of ions and metabolites between the cytoplasm of adjacent cells. They are formed by two hemichannels, made up of six connexin proteins assembled in groups. Each connexin protein has four transmembrane segments, two extracellular loops, a cytoplasmic loop formed between the two inner transmembrane segments, and the N- and C-terminus both being in the cytoplasm. The specificity of the gap junction is determined by which connexin proteins comprise the hemichannel. In the past, connexin protein names were based on their molecular weight, however the new nomenclature uses sequential numbers based on which form (alpha or beta) of the gap junction is present. This gene encodes one of the connexin proteins. Mutations in this gene have been found in some forms of deafness and in some families with hidrotic ectodermal dysplasia. [provided by RefSeq, Jul 2008]

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; Gap junction beta-6 protein; Connexin-30
Dilution	IHC-P=1:50-200,IHC-F=1:50-200,WB=1:500-2000,IF=1:50-200,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	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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