

GJB3 Rabbit pAb

Catalog # AP97214

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	O75712
Other Accession	O75712
Host	Rabbit
Clonality	Polyclonal
Calculated MW	30818
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Connexin-31
Epitope Specificity	51-150/270
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 GJB3 are a cause of erythrokeratoderma variabilis (EKV) [MIM:133200]. EKV is a genodermatosis characterized by the appearance of two independent skin lesions: transient figurate erythematous patches and hyperkeratosis that is usually localized but occasionally occurs in its generalized form. Clinical presentation varies significantly within a family and from one family to another. Palmoplantar keratoderma is present in around 50% of cases. Defects in GJB3 are the cause of deafness autosomal dominant type 2B (DFNA2B) [MIM:612644]. DFNA2 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.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	This gene is a member of the connexin gene family. The encoded protein is a component of gap junctions, which are composed of arrays of intercellular channels that provide a route for the diffusion of low molecular weight materials from cell to cell. Mutations in this gene can cause non-syndromic deafness or erythrokeratoderma variabilis, a skin disorder. Alternative splicing results in multiple transcript variants encoding the same protein. [provided by RefSeq, Jul 2008].

Additional Information

Gene ID	2707
Other Names	CX31; DFNA2; DFNA2B; EKV; EKVP1; Cnx31; Cxnc; D4Wsu144e; Gjb-3; CXB3_HUMAN; GJB3; Connexin-31 (Cx31); CXB3_MOUSE; Cxn-31; CXB3_RAT; gap junction protein beta 3; gap junction protein, beta 3, 31kD (connexin 31); gap junction protein, beta 3, 31kDa (connexin 31); erythrokeratoderma

variabilis; gap junction protein, beta 3, 31kDa; connexin 31

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

GJB3

Synonyms

CX31

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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