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Connexin 26 Rabbit pAb

Catalog # AP96919

Product Information

Application	WB
Primary Accession	Q00977
Other Accession	Q00977
Reactivity	Mouse
Predicted	Rat
Host	Rabbit
Clonality	Polyclonal
Calculated MW	26411
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from mouse Connexin 26
Epitope Specificity	61-160/226
Isotype	IgG
Buffer	0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.
SIMILARITY	Belongs to the connexin family. Alpha-type (group II) subfamily.
Post-translational modifications	Belongs to the connexin family. Beta-type (group I) subfamily.
DISEASE	Defects in GJA1 are the cause of autosomal dominant oculodentodigital dysplasia (ODDD) [MIM:164200]; also known as oculodentoosseous dysplasia. ODDD is a highly penetrant syndrome presenting with craniofacial (ocular, nasal, dental) and limb dysmorphisms, spastic paraplegia, and neurodegeneration. Craniofacial anomalies tipically include a thin nose with hypoplastic alae nasi, small anteverted nares, prominent columnella, and microcephaly. Brittle nails and hair abnormalities of hypotrichosis and slow growth are present. Ocular defects include microphthalmia, microcornea, cataracts, glaucoma, and optic atrophy. Syndactyly type 3 and conductive deafness can occur in some cases. Cardiac abnormalities are observed in rare instances. Defects in GJA1 are the cause of autosomal recessive oculodentodigital dysplasia (ODDD autosomal recessive) [MIM:257850].
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	Gap junction channels consist of connexin protein subunits, which are encoded by a multigene family. GJBs (gap junction proteins or connexins) play crucial functional roles associated with these channels. Immunohistochemical staining of human cochlear cells demonstrated high levels of GJB2 expression. Mutations in GJB2 are associated with genetically derived hearing impairments, including autosomal recessive nonsyndromic deafness.

Additional Information

Gene ID	14619
Other Names	BAPS; CX26; DFNA3; DFNA3A; DFNB1; DFNB1A; HID; KID; NSRD1; PPK; Cnx26; Cxne; Gjb-2; CXN-26; CXB2_HUMAN; GJB2; Connexin-26 (Cx26); CXB2_MOUSE;

	CXB2_RAT; gap junction protein beta 2; gap junction protein, beta 2, 26kD (connexin 26); gap junction protein, beta 2, 26kDa (connexin 26); gap junction protein, beta 2, 26kDa; connexin 26
Dilution	WB=1:500-2000
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	Gjb2
Synonyms	Cxn-26
Function	Structural component of gap junctions (PubMed: 15692151). Gap junctions are dodecameric channels that connect the cytoplasm of adjoining cells. They are formed by the docking of two hexameric hemichannels, one from each cell membrane (By similarity). Small molecules and ions diffuse from one cell to a neighboring cell via the central pore (PubMed:15692151).
Cellular Location	Cell membrane; Multi-pass membrane protein {ECO:0000250|UniProtKB:P29033}. Cell junction, gap junction Note=Colocalizes with GJB4 at gap junction plaques in the cochlea
Tissue Location	Detected in cochlea (at protein level) (PubMed:15692151). Detected in cochlea (PubMed:15692151). Liver, kidney, intestine, lung, spleen, stomach, testis and brain, but not heart and adult skeletal muscle.

Please note: All products are 'FOR RESEARCH USE ONLY. NOT FOR USE IN DIAGNOSTIC OR THERAPEUTIC PROCEDURES'.