

Connexin 26 Rabbit pAb

Catalog # AP97301

Product Information

Application	WB, IHC-P, IHC-F, IF, E
Primary Accession	P29033
Other Accession	P29033
Host	Rabbit
Clonality	Polyclonal
Calculated MW	26215
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human Connexin-26
Epitope Specificity	81-180/226
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. 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 typically 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	2706
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,IHC-P=1:100-500,IHC-F=1:100-500,IF=1:100-500,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 GJB2

Function Structural component of gap junctions (PubMed: [16849369](#), PubMed:[17551008](#), PubMed:[19340074](#), PubMed:[19384972](#), PubMed:[21094651](#), PubMed:[26753910](#)). 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 (PubMed:[17551008](#), PubMed:[19340074](#), PubMed:[21094651](#), PubMed:[26753910](#)). Small molecules and ions diffuse from one cell to a neighboring cell via the central pore (PubMed:[16849369](#), PubMed:[19384972](#), PubMed:[21094651](#)).

Cellular Location Cell membrane; Multi-pass membrane protein. Cell junction, gap junction. Note=Colocalizes with GJB4 at gap junction plaques in the cochlea. {ECO:0000250|UniProtKB:Q00977}

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