间隙连接蛋白 26GJB2 抗体

产品货号： mlR21812

英文名称： Connexin 26

中文名称： 间隙连接蛋白 26/GJB2 抗体

别　　名： cx26; DFNA3; DFNB1; HID; KID; NSRD1; Connexin26; Connexin-26; GJB2; PPK; CXB2_HUMAN.

研究领域： 肿瘤　细胞生物　神经生物学　信号转导

抗体来源： Rabbit

克隆类型： Polyclonal

交叉反应　： Mouse, Rat,

产品应用： WB=1:500-2000 ELISA=1:500-1000

not yet tested in other applications.

optimal dilutions/concentrations should be determined by the end user.

分 子 量： 26kDa

细胞定位： 细胞膜

性　　状： Lyophilized or Liquid

浓　　度： 1mg/ml

免 疫 原： KLH conjugated synthetic peptide derived from mouse Connexin 26 :61-160/226 <Cytoplasmic>

亚　　型： IgG

纯化方法： affinity purified by Protein A

储 存 液： 0.01M TBS(pH7.4) with 1% BSA, 0.03% Proclin300 and 50% Glycerol.

保存条件： Store at -20 ° C for one year. Avoid repeated freeze/thaw cycles. The lyophilized antibody is stable at room temperature for at least one month and for greater than a year when kept at -20° C. 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.

PubMed： PubMed

产品介绍： 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.

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. May play a critical role in the physiology of hearing by participating in the recycling of potassium to the cochlear endolymph.

Subcellular Location:

Cell membrane. Cell junction, gap junction.

Tissue Specificity:

Expressed in the heart and fetal cochlea.

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].

Similarity:

Belongs to the connexin family. Alpha-type (group II) subfamily.

SWISS:

Q00977

Gene ID:

14619

Important Note:

This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.

间隙连接(Gap junction, GJ)是普遍存在于相邻细胞间的细胞连接方式，是相邻细胞膜上的一种膜蛋白通道结构。间隙连接允许邻近细胞之间的离子，核苷酸和小的调节分子以及药物或它们的代谢物进行转运，细胞通过它所介导的细胞间连接通讯,进行细胞间信息和能量的传递，调控细胞的生长、分化和内环境的稳定，对维持机体的功能发挥重要作用. 间隙连接蛋白 26 目前主要用于肿瘤方面的研究。

产品图片

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