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GJA8 Rabbit pAb

Catalog # AP99300

Product Information

Application	IHC-P, IHC-F, IF
Primary Accession	P48165
Other Accession	P48165
Reactivity	Mouse
Predicted	Rat, Human, Rabbit, Chicken, Dog, Sheep
Host	Rabbit
Clonality	Polyclonal
Calculated MW	48229
Physical State	Liquid
Immunogen	KLH conjugated synthetic peptide derived from human GJA8/Connexin 50
Epitope Specificity	151-250/433
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.
SUBUNIT	A connexon is composed of a hexamer of connexins. This particular connexin only forms junctional channels.
DISEASE	Defects in GJA8 are the cause of cataract zonular pulverulent type 1 (CZP1) [MIM:116200]. A form of zonular cataract. Zonular or lamellar cataracts are opacities, broad or narrow, usually consisting of powdery white dots affecting only certain layers or zones between the cortex and nucleus of an otherwise clear lens. The opacity may be so dense as to render the entire central region of the lens completely opaque, or so translucent that vision is hardly if at all impeded. Zonular cataracts generally do not involve the embryonic nucleus, though sometimes they involve the fetal nucleus. Usually sharply separated from a clear cortex outside them, they may have projections from their outer edges known as riders or spokes. Defects in GJA8 are the cause of cataract-microcornea syndrome (CAMIS) [MIM:116150]. Cataract-microcornea syndrome is characterized by the association of congenital cataract and microcornea without any other systemic anomaly or dysmorphism. Clinical findings include a corneal diameter inferior to 10 mm in both meridians in an otherwise normal eye, and an inherited cataract, which is most often bilateral posterior polar with opacification in the lens periphery. The cataract progresses to form a total cataract after visual maturity has been achieved, requiring cataract extraction in the first to third decade of life. Microcornea-cataract syndrome can be associated with other rare ocular manifestations, including myopia, iris coloboma, sclerocornea and Peters anomaly. Transmission is in most cases autosomal dominant, but cases of autosomal recessive transmission have recently been described.
Important Note	This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.
Background Descriptions	The connexin family of proteins form hexameric complexes called “connexons” that facilitate movement of low molecular weight proteins between cells via gap junctions (1). Connexin proteins share a common topology of 4 transmembrane alpha-helical domains, two extracellular loops,

a cytoplasmic loop, and cytoplasmic N- and C-termini (2). Many of the key functional differences arise from specific amino-acid substitutions in the most highly conserved domains, the transmembrane and extracellular regions (2). Each of the approximately 20 connexin isoforms produces channels with distinct permeabilities and electrical and chemical sensitivities; therefore, one connexin usually cannot fully substitute for another (2). Consequently, a wide variety of malignant phenotypes associate with decreased connexin expression and gap junction communication, dependent on the particular connexin that is effected (3, 2). For instance, deletion of the gene encoding connexin 50, normally expressed in the lens, produces cataracts, though not as severe as with deletion of connexin 46 (4).

Additional Information

Gene ID	2703
Other Names	CXA8_HUMAN; GJA8; Connexin-50 (Cx50); Lens fiber protein MP70; CXA8_MOUSE; CXA8_RAT; gap junction protein alpha 8; gap junction protein, alpha 8, 50kD (connexin 50); gap junction protein, alpha 8, 50kDa (connexin 50); gap junction protein, alpha 8, 50kDa; connexin 50
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	GJA8
Function	Structural component of eye lens gap junctions (PubMed: 18006672, PubMed:19756179). 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:18006672, PubMed:19756179).
Cellular Location	Cell membrane; Multi-pass membrane protein {ECO:0000250|UniProtKB:P55917}. Cell junction, gap junction
Tissue Location	Eye lens..

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