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# Plakophilin 2 Rabbit pAb

Catalog # AP98816

## Product Information

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| **Application** | IHC-P, IHC-F, IF |
| **Primary Accession** | Q99959 |
| **Other Accession** | Q99959 |
| **Reactivity** | Human |
| **Predicted** | Rat, Pig, Mouse, Rabbit, Dog, Horse, Sheep |
| **Host** | Rabbit |
| **Clonality** | Polyclonal |
| **Calculated MW** | 97415 |
| **Physical State** | Liquid |
| **Immunogen** | KLH conjugated synthetic peptide derived from human Plakophilin 2 |
| **Epitope Specificity** | 801-881/881 |
| **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 beta-catenin family.Contains 8 ARM repeats. |
| **SUBUNIT** | Interacts with DSC2. |
| **Post-translational modifications** | Phosphorylated upon DNA damage, probably by ATM or ATR. |
| **DISEASE** | Defects in PKP2 are the cause of familial arrhythmogenic right ventricular dysplasia type 9 (ARVD9) [MIM:609040]; also known as arrhythmogenic right ventricular cardiomyopathy 9 (ARVC9). ARVD is an autosomal dominant disease characterized by partial degeneration of the myocardium of the right ventricle, electrical instability, and sudden death. It is clinically defined by electrocardiographic and angiographic criteria; pathologic findings, replacement of ventricular myocardium with fatty and fibrous elements, preferentially involve the right ventricular free wall. |
| **Important Note** | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications. |
| **Background Descriptions** | Plakophilins 1, 2, 3 and 4 (PKP1-4) influence development and participate in linking cadherins to cytoskeletal intermediate filaments. Plakophilins 1-4 contain arm-repeat (armadillo) domains, and localize to nuclei and cell desmosomes (cell-cell junctions found in suprabasal layers of stratifying epithelia that undergo mechanical stress). Plakophilin-1 mediates increases in desmosomal protein content, desmosome assembly, and regulation of cell migration. Plakophilin-2 is important for desmosome assembly and is an essential morphogenic factor and architectural component of the heart. Plakophilin-3 plays a role in both desmosome-dependent adhesion and signaling pathways. Plakophilin-4 is a component of desmosomal adhesion plaques that regulates junctional plaque organization and cadherin function. |

## Additional Information

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| **Gene ID** | 5318 |
| **Other Names** | ARVD9; 1200008D14Rik; 1200012P04Rik; Pkp2l; PKP2_HUMAN; PKP2; plakophilin 2 |
| **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

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| **Name** | PKP2 ( HGNC:9024) |
| **Function** | A component of desmosome cell-cell junctions which are required for positive regulation of cellular adhesion (PubMed:25208567). Regulates focal adhesion turnover resulting in changes in focal adhesion size, cell adhesion and cell spreading, potentially via transcriptional modulation of beta-integrins (PubMed:23884246). Required to maintain gingival epithelial barrier function (PubMed:34368962). Important component of the desmosome that is also required for localization of desmosome component proteins such as DSC2, DSG2 and JUP to the desmosome cell-cell junction (PubMed:22781308, PubMed:25208567). Required for the formation of desmosome cell junctions in cardiomyocytes, thereby required for the correct formation of the heart, specifically trabeculation and formation of the atria walls (By similarity). Loss of desmosome cell junctions leads to mis-localization of DSP and DSG2 resulting in disruption of cell-cell adhesion and disordered intermediate filaments (By similarity). Modulates profibrotic gene expression in cardiomyocytes via regulation of DSP expression and subsequent activation of downstream TGFB1 and MAPK14/p38 MAPK signaling (By similarity). Required for cardiac sodium current propagation and electrical synchrony in cardiac myocytes, via ANK3 stabilization and modulation of SCN5A/Nav1.5 localization to cell-cell junctions (By similarity). Required for mitochondrial function, nuclear envelope integrity and positive regulation of SIRT3 transcription via maintaining DES localization at its nuclear envelope and cell tip anchoring points, and thereby preserving regulation of the transcriptional program (PubMed:35959657). Maintenance of nuclear envelope integrity protects against DNA damage and transcriptional dysregulation of genes, especially those involved in the electron transport chain, thereby preserving mitochondrial function and protecting against superoxide radical anion generation (PubMed:35959657). Binds single-stranded DNA (ssDNA) (PubMed:20613778). May regulate the localization of GJA1 to gap junctions in intercalated disks of the heart (PubMed:18662195). Involved in the inhibition of viral infection by influenza A viruses (IAV) (PubMed:28169297). Acts as a host restriction factor for IAV viral propagation, potentially via disrupting the interaction of IAV polymerase complex proteins (PubMed:28169297). |
| **Cellular Location** | Nucleus. Cell junction, desmosome. Cell junction. Cytoplasm Note=Colocalizes with CTNNA3 and SCN5A/Nav1.5 at intercalated disks in the heart. {ECO:0000250\|UniProtKB:Q9CQ73} |
| **Tissue Location** | Expressed at myocyte intercalated disks in the heart (at protein level) (PubMed:18662195, PubMed:31845994). Expressed in gingival epithelial, endothelial and fibroblast cells (at protein level) (PubMed:34368962). Faintly expressed in tracheal epithelial cells (at protein level) (PubMed:28169297). Widely expressed. Found at desmosomal plaques in simple and stratified epithelia and in non- epithelial tissues such as myocardium and lymph node |

follicles. In most stratified epithelia found in the desmosomes of the basal cell layer and seems to be absent from suprabasal strata

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