

# LRRC8A Rabbit pAb

Catalog # AP97798

## Product Information

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| <b>Application</b>             | WB, IHC-P, IHC-F, IF, E                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>Primary Accession</b>       | <a href="#">Q8IWT6</a>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
| <b>Other Accession</b>         | <a href="#">Q8IWT6</a>                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
| <b>Host</b>                    | Rabbit                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
| <b>Clonality</b>               | Polyclonal                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                            |
| <b>Calculated MW</b>           | 94199                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                 |
| <b>Physical State</b>          | Liquid                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
| <b>Immunogen</b>               | KLH conjugated synthetic peptide derived from human LRRC8A                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                            |
| <b>Epitope Specificity</b>     | 84-180/810                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                            |
| <b>Isotype</b>                 | IgG                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                   |
| <b>Purity</b>                  | affinity purified by Protein A                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                        |
| <b>Buffer</b>                  | 0.01M TBS (pH7.4) with 1% BSA, 0.02% Proclin300 and 50% Glycerol.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>SIMILARITY</b>              | Contains 17 LRR (leucine-rich) repeats.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                               |
| <b>DISEASE</b>                 | Defects in LRRC8A are the cause of agammaglobulinemia type 5 (AGM5) [MIM:613506]. It is a primary immunodeficiency characterized by profoundly low or absent serum antibodies and low or absent circulating B-cells due to an early block of B-cell development. Affected individuals develop severe infections in the first years of life. Note=A chromosomal aberration involving LRRC8 has been found in a patient with congenital agammaglobulinemia. Translocation t(9;20)(q33.2;q12). The translocation truncates the LRRC8 gene, resulting in deletion of the eighth, ninth, and half of the seventh LRR domains.                              |
| <b>Important Note</b>          | This product as supplied is intended for research use only, not for use in human, therapeutic or diagnostic applications.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                             |
| <b>Background Descriptions</b> | This gene encodes a protein belonging to the leucine-rich repeat family of proteins, which are involved in diverse biological processes, including cell adhesion, cellular trafficking, and hormone-receptor interactions. This family member is a putative four-pass transmembrane protein that plays a role in B cell development. Defects in this gene cause autosomal dominant non-Bruton type agammaglobulinemia, an immunodeficiency disease resulting from defects in B cell maturation. Multiple alternatively spliced transcript variants, which encode the same protein, have been identified for this gene. [provided by RefSeq, Jul 2008] |

## Additional Information

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| <b>Gene ID</b>     | 56262                                                                                                                                                                                                                                                                                                                                                                              |
| <b>Other Names</b> | LRC8A_HUMAN; LRRC8A; Leucine-rich repeat-containing protein 8A (HsLRRC8A); Swelling protein 1; KIAA1437; LRRC8; SWELL1; LRC8A_MOUSE; Leucine-rich repeat-containing protein 8A; Protein ebouriffe (ebo); LRC8A_RAT; Swelling protein 1 SWELL1; leucine rich repeat containing 8 VRAC subunit A; leucine rich repeat containing 8; leucine rich repeat containing 8 family member A |

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| <b>Dilution</b> | WB=1:500-2000,IHC-P=1:100-500,IHC-F=1:100-500,ICC/IF=1:100-500,IF=1:100-500,ELISA=1:5000-10000                                                                                                  |
| <b>Storage</b>  | 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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| <b>Name</b>              | LRRC8A {ECO:0000303 PubMed:22532330, ECO:0000312 HGNC:HGNC:19027}                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                 |
| <b>Function</b>          | <p>Essential component of the volume-regulated anion channel (VRAC, also named VSOAC channel), an anion channel required to maintain a constant cell volume in response to extracellular or intracellular osmotic changes (PubMed:<a href="#">24725410</a>, PubMed:<a href="#">24790029</a>, PubMed:<a href="#">26530471</a>, PubMed:<a href="#">26824658</a>, PubMed:<a href="#">28193731</a>, PubMed:<a href="#">29769723</a>). The VRAC channel conducts iodide better than chloride and can also conduct organic osmolytes like taurine (PubMed:<a href="#">24725410</a>, PubMed:<a href="#">24790029</a>, PubMed:<a href="#">26530471</a>, PubMed:<a href="#">26824658</a>, PubMed:<a href="#">28193731</a>, PubMed:<a href="#">30095067</a>). Mediates efflux of amino acids, such as aspartate and glutamate, in response to osmotic stress (PubMed:<a href="#">28193731</a>). LRRC8A and LRRC8D are required for the uptake of the drug cisplatin (PubMed:<a href="#">26530471</a>). In complex with LRRC8C or LRRC8E, acts as a transporter of immunoreactive cyclic dinucleotide GMP-AMP (2'-3'-cGAMP), an immune messenger produced in response to DNA virus in the cytosol: mediates both import and export of 2'-3'-cGAMP, thereby promoting transfer of 2'-3'-cGAMP to bystander cells (PubMed:<a href="#">33171122</a>). In contrast, complexes containing LRRC8D inhibit transport of 2'-3'-cGAMP (PubMed:<a href="#">33171122</a>). Required for in vivo channel activity, together with at least one other family member (LRRC8B, LRRC8C, LRRC8D or LRRC8E); channel characteristics depend on the precise subunit composition (PubMed:<a href="#">24790029</a>, PubMed:<a href="#">26824658</a>, PubMed:<a href="#">28193731</a>). Can form functional channels by itself (in vitro) (PubMed:<a href="#">26824658</a>). Involved in B-cell development: required for the pro-B cell to pre-B cell transition (PubMed:<a href="#">14660746</a>). Also required for T-cell development (By similarity). Required for myoblast differentiation: VRAC activity promotes membrane hyperpolarization and regulates insulin-stimulated glucose metabolism and oxygen consumption (By similarity). Also acts as a regulator of glucose-sensing in pancreatic beta cells: VRAC currents, generated in response to hypotonicity- or glucose-induced beta cell swelling, depolarize cells, thereby causing electrical excitation, leading to increase glucose sensitivity and insulin secretion (PubMed:<a href="#">29371604</a>). Also plays a role in lysosome homeostasis by forming functional lysosomal VRAC channels in response to low cytoplasmic ionic strength condition: lysosomal VRAC channels are necessary for the formation of large lysosome-derived vacuoles, which store and then expel excess water to maintain cytosolic water homeostasis (PubMed:<a href="#">31270356</a>, PubMed:<a href="#">33139539</a>). Acts as a key factor in NLRP3 inflammasome activation by modulating itaconate efflux and mitochondria function (PubMed:<a href="#">39909992</a>).</p> |
| <b>Cellular Location</b> | Cell membrane; Multi-pass membrane protein. Lysosome membrane; Multi-pass membrane protein. Note=Mainly localizes to the cell membrane, with some intracellular localization to lysosomes                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                         |
| <b>Tissue Location</b>   | Expressed in brain, kidney, ovary, lung, liver, heart, and fetal brain and liver. Found at high levels in bone marrow; lower levels are detected in peripheral blood cells. Expressed on T- cells as well as on B-lineage cells.                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                  |

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