

**Datasheet for 200-901-MH2****C9orf72 Antibody****Overview**

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|----------------------|-----------------------------------------------|
| <b>Description:</b>  | Anti-C9orf72 (CHICKEN) Antibody - 200-901-MH2 |
| <b>Item No.:</b>     | 200-901-MH2                                   |
| <b>Size:</b>         | 100 µg                                        |
| <b>Applications:</b> | FC, IF, IHC                                   |
| <b>Reactivity:</b>   | Human                                         |
| <b>Host Species:</b> | Chicken                                       |

**Product Details****Background:**

C9orf72 (Chromosome 9 Open Reading Frame 72) plays an important role in the regulation of endosomal trafficking and has been shown to interact with Rab proteins that are involved in autophagy and endocytic transport. It is a component of the C9orf72-SMCR8 complex, a complex that has guanine nucleotide exchange factor (GEF) activity and regulates autophagy. In the complex, C9orf72 and SMCR8 probably constitute the catalytic subunits that promote the exchange of GDP to GTP, converting inactive GDP-bound RAB8A and RAB39B into their active GTP-bound form, thereby promoting autophagosome maturation. The C9orf72-SMCR8 complex also acts as a regulator of autophagy initiation by interacting with the ATG1/ULK1 kinase complex and modulating its protein kinase activity. It positively regulates initiation of autophagy by regulating the RAB1A-dependent trafficking of the ATG1/ULK1 kinase complex to the phagophore which leads to autophagosome formation. And acts as a regulator of mTORC1 signaling by promoting phosphorylation of mTORC1 substrates. C9orf72 may be involved in regulating the maturation of phagosomes to lysosomes. It regulates actin dynamics in motor neurons by inhibiting the GTP-binding activity of ARF6, leading to ARF6 inactivation. This reduces the activity of the LIMK1 and LIMK2 kinases which are responsible for phosphorylation and inactivation of cofilin, leading to cofilin activation. It positively regulates axon extension and axon growth cone size in spinal motor neurons. C9orf72 plays a role within the hematopoietic system in restricting inflammation and the development of autoimmunity. Studies suggest that hexanucleotide expansions could result in the selective stabilization of repeat-containing pre-mRNA, and the accumulation of insoluble dipeptide repeat protein aggregates that could be pathogenic in FTD-ALS patients. Diseases associated with C9orf72 include Frontotemporal Dementia And/Or Amyotrophic Lateral Sclerosis 1 and Huntington Disease-Like Syndrome Due To C9orf72 Expansions. Anti-C9orf72 Antibody is useful for researchers interested in Neuroscience Research.

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|----------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Synonyms:</b>     | Chicken Anti-Chromosome 9 Open Reading Frame 72 Antibody, Chicken Anti-C9orf72 Antibody, Guanine Nucleotide Exchange, Protein C9orf72, DENNL72, FTDALS1, ALSFTD, FTDALS |
| <b>Host Species:</b> | Chicken                                                                                                                                                                 |
| <b>Clonality:</b>    | Polyclonal                                                                                                                                                              |
| <b>Format:</b>       | IgY                                                                                                                                                                     |

## Target Details

|                            |                                                                                                                                                                                                                                                                                                                                                                                                                                                             |
|----------------------------|-------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Gene Name:</b>          | C9orf72                                                                                                                                                                                                                                                                                                                                                                                                                                                     |
| <b>Reactivity:</b>         | Human                                                                                                                                                                                                                                                                                                                                                                                                                                                       |
| <b>Immunogen Type:</b>     | Conjugated Peptide                                                                                                                                                                                                                                                                                                                                                                                                                                          |
| <b>Immunogen:</b>          | Anti-C9orf72 antibody was prepared from eggs of chickens laid after repeated immunizations with a synthetic peptide corresponding to a C-terminal portion of human C9orf72 surrounding aa 425-481 conjugated to Keyhole Limpet Hemocyanin (KLH).                                                                                                                                                                                                            |
| <b>Purity/Specificity:</b> | This affinity purified antibody is directed against human C9orf72. This product is an IgY fraction antibody purified from monospecific chicken egg yolks by a multi-step process which includes selective precipitation and salt fractionation followed by extensive dialysis against the buffer stated above. A BLAST analysis was used to suggest cross-reactivity with the antigen based on 100% homology with the immunizing sequence to mouse and rat. |
| <b>Relevant Links:</b>     | <ul style="list-style-type: none"><li>• <a href="#">UniProtKB - Q96LT7</a></li><li>• <a href="#">NCBI - NP_001242983.1</a></li><li>• <a href="#">GeneID - 203228</a></li></ul>                                                                                                                                                                                                                                                                              |

## Application Details

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|-----------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Tested Applications:</b> | FC, IF, IHC                                                                                                                                                                                                     |
| <b>Application Note:</b>    | Anti-C9orf72 Antibody has been tested in Immunohistochemistry, Immunofluorescence, and Flow Cytometry. Western blot not recommended. Positive control used: Human cerebellum in IHC, U2-OS in IF, MCF7 in FLOW. |
| <b>Assay Dilutions:</b>     | All assays should be optimized by the user. Recommended dilutions (if any) may be listed below.                                                                                                                 |
| <b>ELISA:</b>               | 1:10,000-1:50,000                                                                                                                                                                                               |
| <b>FC:</b>                  | 1:20                                                                                                                                                                                                            |
| <b>IF:</b>                  | 5µg/mL                                                                                                                                                                                                          |
| <b>IHC:</b>                 | 1:200                                                                                                                                                                                                           |

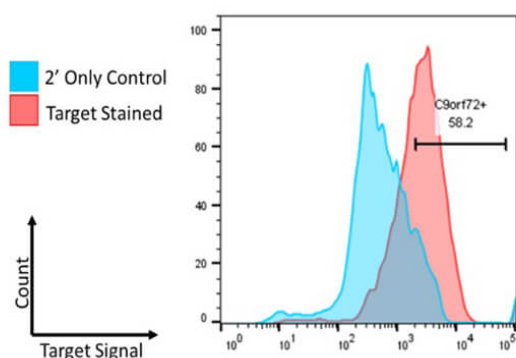
## Formulation

|                        |                                                          |
|------------------------|----------------------------------------------------------|
| <b>Physical State:</b> | Liquid (sterile filtered)                                |
| <b>Concentration:</b>  | 1.03 mg/ml by UV absorbance at 280 nm                    |
| <b>Buffer:</b>         | 0.002 M Sodium Phosphate, 0.15 M Sodium Chloride, pH 7.0 |
| <b>Preservative:</b>   | 0.01% (w/v) Sodium Azide                                 |
| <b>Stabilizer:</b>     | None                                                     |

## Shipping & Handling

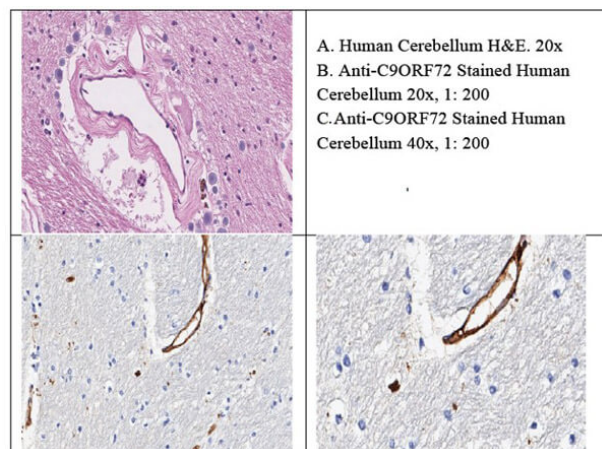
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|----------------------------|-----------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Shipping Condition:</b> | Dry Ice                                                                                                                                                                                                                                                                                                                                       |
| <b>Storage Condition:</b>  | Store vial at -20° C prior to opening. Aliquot contents and freeze at -20° C or below for extended storage. Avoid cycles of freezing and thawing. Centrifuge product if not completely clear after standing at room temperature. This product is stable for several weeks at 4° C as an undiluted liquid. Dilute only prior to immediate use. |
| <b>Expiration:</b>         | Expiration date is one (1) year from date of receipt.                                                                                                                                                                                                                                                                                         |

## Images



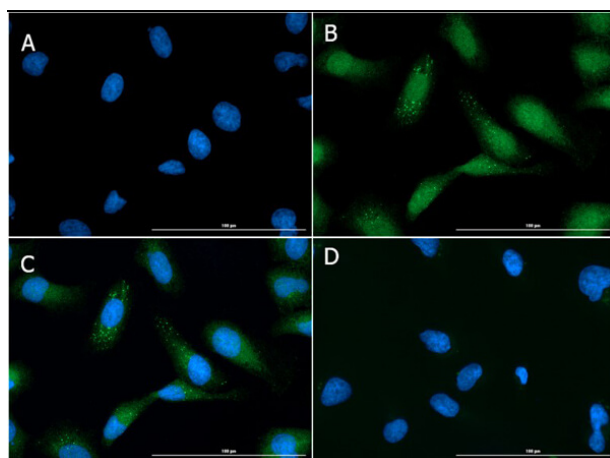
### Flow Cytometry

Flow Cytometry of Chicken Anti-C9orf72 Antibody. Cells: MCF-7 Cells. Primary Antibody: Anti-C9orf72 at 5µL in 100µL FACS buffer for 30 min at RT. Secondary Antibody: Goat Anti-Chicken IgG DyLight™488 (p/n 603-141-126) at 1:400 for 30 min at RT.



### Immunohistochemistry

**Immunohistochemistry of Chicken Anti-C9orf72 Antibody.**  
 Tissue: Human Cerebellum. Fixative: None. Antigen Retrieval: HIER using citrate buffer for 20 min. Primary Antibody: Anti-C9orf72 at 1:200 for 30 min at RT. Secondary Antibody: Alpha Anti Chicken HRP at 1:500 for 20 min at RT. Counterstain: Hematoxylin. Substrate: DAB. Analysis Results: C9ORF72 shows intense staining at a dilution of 1:100 and 1:200 of vascular endothelial cells, low intensity staining of neurophil and occasional more intense staining of glial cells. The specificity of this pattern of staining is consistent with C9ORF72 staining in the databases for cerebellum.



### Immunofluorescence Microscopy

**Immunofluorescence of Chicken Anti-C9orf72 Antibody.**  
 Cells: U-2OS Cells. Fixative: 4% PFA. Permeabilization: 0.3% Triton X-100. Primary Antibody: Anti-C9orf72 at 5µg/mL overnight at 2-8°C. Secondary Antibody: Goat Anti-Chicken IgG DyLight™488 (p/n 603-141-126) at 15µg/mL for 1hr at RT. Nuclear Counterstain: DAPI. Staining: (A) DAPI. (B) C9orf72 and secondary DyLight™488. (C) Merge A+B. (D) secondary only. Predicted localization: Nucleous, Endosome, Lysosome, Extracellular region or secreted. Image has been deconvoluted. Punctate staining may be indicative of endosomes in the cytoplasmic cell compartment.

## Disclaimer

This product is for research use only and is not intended for therapeutic or diagnostic applications. Please contact a technical service representative for more information. All products of animal origin manufactured by Rockland Immunochemicals are derived from starting materials of North American origin. Collection was performed in United States Department of Agriculture (USDA) inspected facilities and all materials have been inspected and certified to be free of disease and suitable for exportation. All properties listed are typical characteristics and are not specifications. All suggestions and data are offered in good faith but without guarantee as conditions and methods of use of our products are beyond our control. All claims must be made within 30 days following the date of delivery. The prospective user must determine the suitability of our materials before adopting them on a commercial scale. Suggested uses of our products are not recommendations to use our products in violation of any patent or as a license under any patent of Rockland Immunochemicals, Inc. If you require a commercial license to use this material and do not have one, then return this material, unopened to: Rockland Inc., P.O. BOX 5199, Limerick, Pennsylvania, USA.