



COSMO BIO Co., LTD.

Inspiration for Life Science

**POLYCLONAL ANTIBODY**

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**Catalog No. CAC-TIP-C9-P02**

## Anti-C9orf72 poly(GR)

### BACKGROUND

Hexanucleotide expansions in C9orf72 gene was identified in patients with frontotemporal lobar degeneration (FTLD) and Amyotrophic Lateral Sclerosis (ALS) in 2011. GGGGCC expansions are characterised pathologically by the presence of TDP-43 negative and p62 positive inclusions in granule cells of cerebellum and in cells of dentate gyrus and CA4 area of the hippocampus. It was reported that these inclusions included dipeptide repeat proteins, poly-GA, poly-GR and poly GP, arising from a putative non-ATG initiated sense translation of the GGGGCC expansion. These antibodies are powerful tools for IHC analysis of neurodegenerative diseases.

|                              |                                                                                                                                                                |
|------------------------------|----------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>Product type</b>          | Primary antibody                                                                                                                                               |
| <b>Immunogen</b>             | poly (GR) <sup>8</sup>                                                                                                                                         |
| <b>Raised in</b>             | Rabbit (New Zealand White)                                                                                                                                     |
| <b>Myeloma</b>               | -                                                                                                                                                              |
| <b>Clone number</b>          | -                                                                                                                                                              |
| <b>Isotype</b>               | -                                                                                                                                                              |
| <b>Source</b>                | Anti-serum                                                                                                                                                     |
| <b>Purification</b>          | -                                                                                                                                                              |
| <b>Form</b>                  | Liquid. Anti-serum with 0.1% NaN <sub>3</sub> as a preservative                                                                                                |
| <b>Concentration</b>         | -                                                                                                                                                              |
| <b>Volume</b>                | 50 µL                                                                                                                                                          |
| <b>Label</b>                 | Unlabeled                                                                                                                                                      |
| <b>Specificity</b>           | poly (GR) <sup>8</sup>                                                                                                                                         |
| <b>Cross reactivity</b>      | Human                                                                                                                                                          |
| <b>Storage</b>               | Store below -20°C. (below -70°C for prolonged storage).<br>Aliquot to avoid cycles of freeze/thaw.                                                             |
| <b>Application notes</b>     | ELISA                                                                                                                                                          |
| <b>Recommended dilutions</b> | Immunohistochemistry: 1/500-1/2000 (Ref.1)<br>Other applications have not been tested. Optimal dilutions/ concentrations should be determined by the end user. |

|                   |                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                                |
|-------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|
| <b>References</b> | 1) David MA Mann, et al. Dipeptide repeat proteins are present in the p62 positive inclusions in patients with frontotemporal lobar degeneration and motor neurone disease associated with expansions in C9ORF72. <i>Acta Neuropathologica Communications</i> (2013) 1:68. PMID 24252525<br><br>2) Tan RH, <i>et al.</i> Cerebellar neuronal loss in als cases with ATXN2 intermediate repeat expansions. <i>Ann Neurol.</i> 2015 Nov 24. doi: 10.1002/ana.24565. PMID:26599997<br><br>3) Davidson Y, <i>et al.</i> Neurodegeneration in Frontotemporal Lobar Degeneration and |
|-------------------|--------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------------|



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TOYO EKIMAE BLDG. 2-20, TOYO 2-CHOME, KOTO-KU, TOKYO 135-0016 JAPAN

TEL: (81)3-5632-9617 / FAX: (81)3-5632-9618 / e-mail: export@cosmobio.co.jp / URL:www.cosmobio.com



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Motor Neurone Disease associated with expansions in C9orf72 is linked to TDP-43 pathology and not associated with aggregated forms of dipeptide repeat proteins. Neuropathol Appl Neurobiol. 2015 Nov 5. doi: 10.1111/nan.12292. PMID: 26538301

4) Baborie A, *et al.* Accumulation of dipeptide repeat proteins predates that of TDP-43 in frontotemporal lobar degeneration associated with hexanucleotide repeat expansions in C9ORF72 gene. Neuropathol Appl Neurobiol. 2015 Aug;41(5):601-12. doi: 10.1111/nan.12178. Epub 2015 Apr 30. PMID: 25185840

5) Davidson YS, *et al.* Brain distribution of dipeptide repeat proteins in frontotemporal lobar degeneration and motor neurone disease associated with expansions in C9ORF72. Acta Neuropathol Commun. 2014 Jun 20;2:70. doi: 10.1186/2051-5960-2-70. PMID: 24950788

6) Konno T, *et al.* C9ORF72 repeat-associated non-ATG-translated polypeptides are distributed independently of TDP-43 in a Japanese patient with c9ALS. Neuropathol Appl Neurobiol. 2014 Oct;40(6):783-8. doi: 10.1111/nan.12157. No abstract available. PMID: 24861677

**RELATED PRODUCTS:**

| Product Name          | Quantity | Cat#           |
|-----------------------|----------|----------------|
| Anti-C9orf72 poly(GA) | 50 uL    | CAC-TIP-C9-P01 |
| Anti-C9orf72 poly(GP) | 50 uL    | CAC-TIP-C9-P03 |

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