





# Recombinant Human TAR DNA-binding protein 43 (TARDBP), partial

Product Code	CSB-YP023129HU
Relevance	DNA and RNA-binding protein which regulates transcription and splicing. Involved in the regulation of CFTR splicing. It promotes CFTR exon 9 skipping by binding to the UG repeated motifs in the polymorphic region near the 3'-splice site of this exon. The resulting aberrant splicing is associated with pathological features typical of cystic fibrosis. May also be involved in microRNA biogenesis, apoptosis and cell division. Can repress HIV-1 transcription by binding to the HIV-1 long terminal repeat. Stabilizes the low molecular weight neurofilament (NFL) mRNA through a direct interaction with the 3' UTR.
Abbreviation	Recombinant Human TARDBP protein, partial
Storage	The shelf life is related to many factors, storage state, buffer ingredients, storage temperature and the stability of the protein itself. Generally, the shelf life of liquid form is 6 months at -20°C/-80°C. The shelf life of lyophilized form is 12 months at -20°C/-80°C.
Uniprot No.	Q13148
Product Type	Recombinant Proteins
Immunogen Species	Homo sapiens (Human)
Purity	Greater than 95% as determined by SDS-PAGE. Greater than 95% as determined by SEC-HPLC.
Sequence	MSEYIRVTEDENDEPIEIPSEDDGTVLLSTVTAQFPGACGLRYRNPVSQCMRG VRLVEGILHAPDAGWGNLVYVVNYPKDNKRKMDETDASSAVKVKRAVQKTSD LIVLGLPWKTTEQDLKEYFSTFGEVLMVQVKKDLKTGHSKGFGFVRFTEYETQ VKVMSQRHMIDGRWCDCKLPNSKQSQDEPLRSRKVFVGRCTEDMTEDELRE FFSQYGDVMDVFIPKPFRAFAFVTFADDQIAQSLCGEDLIIKGISVHISNAEPKH NSNRQLERSGRFGGNPGGFGNQGGFGNSRGGGAGLGNNQGSNMGGGMN FGAFSINPAMMAAAQAALQSSWGMMGMLASQQNQSGPSGNNQNQGNMQR EPNQAFGSGNNSYSGSNSGAAIGWGSASNAGSGSG
Research Area	Microbiology
Source	Yeast
Target Names	TARDBP
Expression Region	1-396aa
Notes	Repeated freezing and thawing is not recommended. Store working aliquots at 4°C for up to one week.
Tag Info	N-terminal 6xHis-tagged
Mol. Weight	44.9kDa
Protein Length	Partial

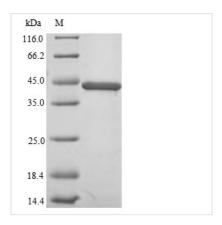
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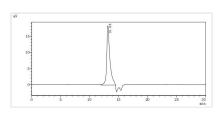




## **Image**



(Tris-Glycine gel) Discontinuous SDS-PAGE (reduced) with 5% enrichment gel and 15% separation gel.



The purity of TARDBP was greater than 95% as determined by SEC-HPLC

# **Description**

TAR DNA-binding protein 43 (TARDBP) is a crucial component in the pathogenesis of amyotrophic lateral sclerosis (ALS), a fatal motor neuron disorder. TARDBP encodes the transactive response DNA-binding protein 43 (TDP-43), which is frequently found in pathological inclusions in various forms of ALS [1]. Mutations in TARDBP have been identified in both familial and sporadic ALS cases, highlighting its significance in the disease [2][3][4]. TDP-43 is a 43 kDa nuclear protein involved in RNA metabolism [5]. It is the major protein found in the ubiquitinated inclusions in affected motor neurons in ALS cases [6]. Studies have shown that TARDBP mutations are prevalent in Italian ALS patients, further emphasizing its role in the disease [7].

TDP-43 is an RNA/DNA-binding protein that belongs to the heterogeneous nuclear ribonucleoprotein family [8]. It has been associated with various cellular functions, including transcriptional regulation and splicing [9]. The protein is essential for early embryonic development and is highly conserved across different species [10]. In ALS, TDP-43 forms aggregates in the motor cortex of aging brains, indicating its involvement in the disease progression [11].

The complexity of ALS pathophysiology involves interactions with other proteins and pathways. For instance, TDP-43 interacts with progranulin to reverse motor neuron defects, highlighting its neurotrophic effects [12]. Additionally, loss of TDP-43 in zebrafish models leads to muscle degeneration and reduced motor neuron axon outgrowth, further underlining its role in ALS-related degeneration [13].

## References:

[1] V. Soldatov, M. Kukharsky, ?. ?????, A. Sobolev, & A. Deykin, "Retinal damage in amyotrophic lateral sclerosis: underlying mechanisms", Eye and Brain, vol. Volume 13, p. 131-146, 2021. https://doi.org/10.2147/eb.s299423 [2] J. Sreedharan, I. Blair, V. Tripathi, X. Hu, C. Vance, B. Rogeljet al., "Tdp-43

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mutations in familial and sporadic amyotrophic lateral sclerosis", Science, vol. 319, no. 5870, p. 1668-1672, 2008. https://doi.org/10.1126/science.1154584 [3] D. Bosco, N. Lemay, H. Ko, H. Zhou, C. Burke, T. Kwiatkowskiet al., "Mutant fus proteins that cause amyotrophic lateral sclerosis incorporate into stress granules", Human Molecular Genetics, vol. 19, no. 21, p. 4160-4175, 2010. https://doi.org/10.1093/hmg/ddq335

- [4] S. Lattante, G. Rouleau, & E. Kabashi, "tardbpandfusmutations associated with amyotrophic lateral sclerosis: summary and update", Human Mutation, vol. 34, no. 6, p. 812-826, 2013. https://doi.org/10.1002/humu.22319
- [5] S. Dzieciolowska, P. Drapeau, & G. Armstrong, "Augmented quantal release of acetylcholine at the vertebrate neuromuscular junction following tdp-43 depletion", Plos One, vol. 12, no. 5, p. e0177005, 2017. https://doi.org/10.1371/journal.pone.0177005
- [6] P. Bose, G. Armstrong, & P. Drapeau, "Neuromuscular junction abnormalities in a zebrafish loss-of-function model of tdp-43", Journal of Neurophysiology, vol. 121, no. 1, p. 285-297, 2019. https://doi.org/10.1152/jn.00265.2018
- [7] L. Corrado, A. Ratti, C. Gellera, E. Buratti, B. Castellotti, Y. Carlomagnoet al., "High frequency oftardbpgene mutations in italian patients with amyotrophic lateral sclerosis", Human Mutation, vol. 30, no. 4, p. 688-694, 2009. https://doi.org/10.1002/humu.20950
- [8] J. Dou, "Tdp-43: a key of neurodegenerative disease", Asian Journal of Complementary and Alternative Medicine, vol. 10, no. 3, 2022. https://doi.org/10.53043/2347-3894.acam90039
- [9] C. Hewamadduma, A. Grierson, P. Taylur, L. Pan, C. Moens, P. Inghamet al., "Tardbpl splicing rescues motor neuron and axonal development in a mutant tardbp zebrafish", Human Molecular Genetics, vol. 22, no. 12, p. 2376-2386, 2013. https://doi.org/10.1093/hmg/ddt082
- [10] C. Sephton, S. Good, S. Atkin, C. Dewey, P. Mayer, J. Herzet al., "Tdp-43 is a developmentally regulated protein essential for early embryonic development", Journal of Biological Chemistry, vol. 285, no. 9, p. 6826-6834, 2010. https://doi.org/10.1074/jbc.m109.061846
- [11] Y. Koike, A. Sugai, N. Hara, J. Ito, A. Yokoseki, T. Ishiharaet al., "Agerelated demethylation of the tdp-43 autoregulatory region in the human motor cortex",, 2021. https://doi.org/10.1101/2021.01.13.426599
- [12] B. Chitramuthu, D. Kay, A. Bateman, & H. Bennett, "Neurotrophic effects of progranulin in vivo in reversing motor neuron defects caused by over or under expression of tdp-43 or fus", Plos One, vol. 12, no. 3, p. e0174784, 2017. https://doi.org/10.1371/journal.pone.0174784
- [13] B. Schmid, A. Hruscha, S. Hogl, J. Banzhaf-Strathmann, K. Strecker, J. Zeeet al., "Loss of als-associated tdp-43 in zebrafish causes muscle degeneration, vascular dysfunction, and reduced motor neuron axon outgrowth", Proceedings of the National Academy of Sciences, vol. 110, no. 13, p. 4986-4991, 2013. https://doi.org/10.1073/pnas.1218311110

### Shelf Life

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