

# Frontotemporal dementia

Frontotemporal [dementia](#) is an umbrella term for a group of uncommon brain [disorders](#) that primarily affect the frontal and [temporal lobes](#) of the brain. These areas of the brain are generally associated with personality, behavior and language. In frontotemporal dementia, portions of these lobes shrink (atrophy).

## Diagnosis

Results indicate that the total levels of [TDP-43](#) in the serum are decreased especially in FTD patients with the [C9orf72](#) repeat expansion or FTD-MND phenotype, both subtypes strongly associated with TDP-43 type B brain pathology. Serum-based measurement of TDP-43 could represent a useful tool in indicating C9orf72 repeat expansion and FTD-MND-related TDP-43 neuropathology for future diagnostics and intervention studies <sup>1)</sup>.

TAR DNA-binding protein 43 ([TDP-43](#)) is a major protein component of pathological inclusions in [amyotrophic lateral sclerosis](#) and frontotemporal [dementia](#). Reducing aberrant aggregation of TDP-43 is a potential approach to prevent [cell death](#). To investigate whether DJ-1 might inhibit TDP-43 aggregation to exert a protective effect in oxidative stress-induced injury, Lei et al. tested the protein level and subcellular localization of TDP-43 and [DJ-1](#) in SH-SY5Y cells transfected with wild-type DJ-1, DJ-1 mutant (L166P) cDNA, or DJ-1 siRNA. They showed that [oxidative stress](#) induced by paraquat leads to the formation of cytosolic TDP-43 aggregation in SH-SY5Y cells. DJ-1 overexpression decreases paraquat-induced cytoplasmic accumulation of TDP-43 in SH-SY5Y cells and protects against paraquat-induced cell death. Transfection of DJ-1 L166P mutant or DJ-1 siRNA leads to increased cytosolic aggregation of TDP-43 in paraquat-treated SH-SY5Y cells and promotes cell death. These data suggest that [DJ-1](#) may protect against oxidative stress-induced cell death through the suppression of cytoplasmic TDP-43 aggregation <sup>2)</sup>.

## Genetic Frontotemporal dementia

[Genetic Frontotemporal dementia](#)

## Differential diagnosis

It tends to occur earlier than Alzheimer's, primarily between ages 35 and 75. Due to its earlier onset and prominent behavioral and personality symptoms, it may be mistaken for a primary psychiatric disorder.

<sup>1)</sup>

Katisko K, Huber N, Kokkola T, Hartikainen P, Krüger J, Heikkinen AL, Paananen V, Leinonen V, Korhonen VE, Helisalmi S, Herukka SK, Cantoni V, Gadola Y, Archetti S, Remes AM, Haapasalo A, Borroni B, Solje E. Serum total TDP-43 levels are decreased in frontotemporal dementia patients with

C9orf72 repeat expansion or concomitant motoneuron disease phenotype. *Alzheimers Res Ther.* 2022 Oct 11;14(1):151. doi: 10.1186/s13195-022-01091-8. PMID: 36217158.

<sup>2)</sup>

Lei Y, Zhang ZF, Lei RX, Wang S, Zhuang Y, Liu AC, Wu Y, Chen J, Tang JC, Pan MX, Liu R, Liao WJ, Feng YG, Wan Q, Zheng M. DJ-1 Suppresses Cytoplasmic TDP-43 Aggregation in Oxidative Stress-Induced Cell Injury. *J Alzheimers Dis.* 2018 Oct 22. doi: 10.3233/JAD-180460. [Epub ahead of print] PubMed PMID: 30372676.

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