

Supplementary References

- S1. Zetterberg H, Blennow K. From cerebrospinal fluid to blood: The third wave of fluid biomarkers for Alzheimer's Disease. *J Alzheimer's Dis.* 2018;64(s1):S271-S279. doi:10.3233/JAD-179926
- S2. Lin J, Li J, Huang B, et al. Exosomes: Novel biomarkers for clinical diagnosis. *Sci World J.* 2015;657086. doi:10.1155/2015/657086
- S3. Saeedi S, Israel S, Nagy C, Turecki G. The emerging role of exosomes in mental disorders. *Transl Psychiatry.* 2019;9(1). doi:10.1038/s41398-019-0459-9
- S4. Jiang C, Hopfner F, Hopfner F, et al. Serum neuronal exosomes predict and differentiate Parkinson's disease from atypical parkinsonism. *J Neurol Neurosurg Psychiatry.* 2020;91(7):720-729. doi:10.1136/jnnp-2019-322588
- S5. Shephard SR, Wu J, Cardoso SM, et al. Urinary P75 ECD a prognostic, disease progression, and pharmacodynamic biomarker in ALS. *Neurology.* 2017;88(12):1137-1143. doi:10.1212/WNL.0000000000003741
- S6. González-Sánchez M, Bartolome F, Antequera D, et al. Decreased salivary lactoferrin levels are specific to Alzheimer's disease. *EBioMedicine.* 2020:102834. doi:10.1016/j.ebiom.2020.102834
- S7. Kan CW, Tobos CI, Rissin DM, et al. Digital enzyme-linked immunosorbent assays with sub-attomolar detection limits based on low numbers of capture beads combined with high efficiency bead analysis. *Lab Chip.* 2020;20(12):2122-2135. doi:10.1039/d0lc00267d
- S8. Yao NY, Braedersz CP, Lin YC, Kasza KE, MacKintosh FC, Weitz DA. Elasticity in ionically cross-linked neurofilament networks. *Biophys J.* 2010;98(10):2147-2153. doi:10.1016/j.bpj.2010.01.062
- S9. Scherling CS, Hall T, Berisha F, et al. Cerebrospinal fluid neurofilament concentration reflects disease severity in frontotemporal degeneration. *Ann Neurol.* 2014;75(1):116-126. doi:10.1002/ana.24052
- S10. Rohrer JD, Woollacott IOC, Dick KM, et al. Serum neurofilament light chain protein is a measure of disease intensity in frontotemporal dementia. *Neurology.* 2016;87(13):1329-1336. doi:10.1212/WNL.0000000000003154
- S11. Rojas JC, Bang J, Lobach I V., et al. CSF neurofilament light chain and phosphorylated tau 181 predict disease progression in PSP. *Neurology.* 2018;90(4):E273-E281. doi:10.1212/WNL.0000000000004859

- S12. Steinacker P, Anderl-Straub S, Diehl-Schmid J, et al. Serum neurofilament light chain in behavioral variant frontotemporal dementia. *Neurology*. 2018;91(15):E1390-E1401. doi:10.1212/WNL.0000000000006318
- S13. Meeter LH, Dopfer EG, Jiskoot LC, et al. Neurofilament light chain: a biomarker for genetic frontotemporal dementia. *Ann Clin Transl Neurol*. 2016;3(8):623-636. doi:10.1002/acn3.325
- S14. Donker Kaat L, Meeter LH, Chiu WZ, et al. Serum neurofilament light chain in progressive supranuclear palsy. *Park Relat Disord*. 2018;56:98-101. doi:10.1016/j.parkreldis.2018.06.018
- S15. Meeter LHH, Steketee RME, Salkovic D, et al. Clinical value of cerebrospinal fluid neurofilament light chain in semantic dementia. *J Neurol Neurosurg Psychiatry*. 2019;90(9):997-1004. doi:10.1136/jnnp-2018-319784
- S16. Steinacker P, Semler E, Anderl-Straub S, et al. Neurofilament as a blood marker for diagnosis and monitoring of primary progressive aphasia. *Neurology*. 2017;88(10):961-969. doi:10.1212/WNL.0000000000003688
- S17. Bridel C, Van Wieringen WN, Zetterberg H, et al. Diagnostic value of cerebrospinal fluid neurofilament light protein in neurology: A systematic review and meta-analysis. *JAMA Neurol*. 2019;76(9):1035-1048. doi:10.1001/jamaneurol.2019.1534
- S18. Katisko K, Cajanus A, Jääskeläinen O, et al. Serum neurofilament light chain is a discriminative biomarker between frontotemporal lobar degeneration and primary psychiatric disorders. *J Neurol*. 2020;267(1):162-167. doi:10.1007/s00415-019-09567-8
- S19. van der Ende EL, Meeter LH, Poos JM, et al. Serum neurofilament light chain in genetic frontotemporal dementia: a longitudinal, multicentre cohort study. *Lancet Neurol*. 2019;18(12):1103-1111. doi:10.1016/S1474-4422(19)30354-0
- S20. Winter B, Guenther R, Ludolph AC, Hermann A, Otto M, Wurster CD. Neurofilaments and tau in CSF in an infant with SMA type 1 treated with nusinersen. *J Neurol Neurosurg Psychiatry*. 2019;90(9):1068-1069. doi:10.1136/jnnp-2018-320033

- S21. Wilke C, Pujol-Calderón F, Barro C, et al. Correlations between serum and CSF pNfH levels in ALS, FTD and controls: A comparison of three analytical approaches. *Clin Chem Lab Med*. 2019;57(10). doi:10.1515/cclm-2019-0015
- S22. Remnestål J, Öijerstedt L, Ullgren A, et al. Altered levels of CSF proteins in patients with FTD, presymptomatic mutation carriers and non-carriers. *Transl Neurodegener*. 2020;9(1). doi:10.1186/s40035-020-00198-y
- S23. Gendron TF, Daugherty LM, Heckman MG, et al. Phosphorylated neurofilament heavy chain: A biomarker of survival for C9ORF72-associated amyotrophic lateral sclerosis. *Ann Neurol*. 2017;82(1):139-146. doi:10.1002/ana.24980
- S24. Miller T, Cudkovicz M, Shaw PJ, et al. Phase 1–2 trial of antisense oligonucleotide tofersen for SOD1 ALS. *N Engl J Med*. 2020;383(2):109-119. doi:10.1056/NEJMoa2003715
- S25. Vatsavayai SC, Jin Yoon S, Gardner RC, et al. Timing and significance of pathological features in C9orf72 expansion-associated frontotemporal dementia. *Brain*. 2016;139(12):3202-3216. doi:10.1093/brain/aww250
- S26. Rascovsky K, Grossman M. Clinical diagnostic criteria and classification controversies in frontotemporal lobar degeneration. *Int Rev Psychiatry*. 2013;25(2):145. doi:10.3109/09540261.2013.763341
- S27. Ahmed RM, Latheef S, Bartley L, et al. Eating behavior in frontotemporal dementia. *Neurology*. 2015;85(15):1310-1317. doi:10.1212/WNL.0000000000002018
- S28. Ahmed RM, Irish M, Henning E, et al. Assessment of eating behavior disturbance and associated neural networks in frontotemporal dementia. *JAMA Neurol*. 2016;73(3):282. doi:10.1001/jamaneurol.2015.4478
- S29. Ikeda M, Brown J, Holland AJ, Fukuhara R, Hodges JR. Changes in appetite, food preference, and eating habits in frontotemporal dementia and Alzheimer's disease. *J Neurol Neurosurg Psychiatry*. 2002;73(4):371-376. doi:10.1136/jnnp.73.4.371
- S30. Ahmed RM, Irish M, Kam J, et al. Quantifying the eating abnormalities in frontotemporal dementia. *JAMA Neurol*. 2014;71(12):1540-1546. doi:10.1001/jamaneurol.2014.1931
- S31. Ahmed RM, Landin-Romero R, Collet TH, et al. Energy expenditure in frontotemporal dementia: A behavioural and imaging study. *Brain*. Published online 2017. doi:10.1093/brain/aww263

- S32. Piguet O, Petersén Å, Yin Ka Lam B, et al. Eating and hypothalamus changes in behavioral-variant frontotemporal dementia. *Ann Neurol*. 2011;69(2):312-319. doi:10.1002/ana.22244
- S33. Bocchetta M, Gordon E, Manning E, et al. Detailed volumetric analysis of the hypothalamus in behavioral variant frontotemporal dementia. *J Neurol*. 2015;262(12):2635-2642. doi:10.1007/s00415-015-7885-2
- S34. Vercruyse P, Vieau D, Blum D, Petersén Å, Dupuis L. Hypothalamic alterations in neurodegenerative diseases and their relation to abnormal energy metabolism. *Front Mol Neurosci*. 2018;11. doi:10.3389/fnmol.2018.00002
- S35. Woolley JD, Khan BK, Natesan A, et al. Satiety-related hormonal dysregulation in behavioral variant frontotemporal dementia. *Neurology*. 2014;82(6):512-520. doi:10.1212/WNL.000000000000106
- S36. Ahmed RM, Phan K, Highton-Williamson E, et al. Eating peptides: biomarkers of neurodegeneration in amyotrophic lateral sclerosis and frontotemporal dementia. *Ann Clin Transl Neurol*. 2019;6(3):486-495. doi:10.1002/acn3.721
- S37. Zanardini R, Benussi L, Fostinelli S, et al. Serum C-peptide, visfatin, resistin, and ghrelin are altered in sporadic and grn-associated frontotemporal lobar degeneration. *J Alzheimer's Dis*. 2018;61(3):1053-1060. doi:10.3233/JAD-170747
- S38. Liguori C, Romigi A, Mercuri NB, et al. Cerebrospinal-fluid orexin levels and daytime somnolence in frontotemporal dementia. *J Neurol*. 2014;261(9):1832-1836. doi:10.1007/s00415-014-7455-z
- S39. Çoban A, Bilgiç B, Lohmann E, et al. Reduced orexin-a levels in frontotemporal dementia: Possible association with sleep disturbance. *Am J Alzheimers Dis Other Demen*. 2013;28(6):606-611. doi:10.1177/1533317513494453
- S40. Vatsavayai SC, Yoon SJ, Gardner RC, Gendron TF, Vargas JN, Trujillo A, Pribadi M, Phillips JJ, Gaus SE, Hixson JD, Garcia PA, Rabinovici GD, Coppola G, Geschwind DH, Petrucelli L, Miller BL, Seeley WW. [Timing and significance of pathological features in C9orf72 expansion-associated frontotemporal dementia](#). *Brain*. 2016 Dec;139(Pt 12):3202-3216. doi: 10.1093/brain/aww250.
- S41. Rohrer JD, Nicholas JM, Cash DM, et al. Presymptomatic cognitive and neuroanatomical changes in genetic frontotemporal dementia in the Genetic Frontotemporal dementia Initiative (GENFI) study: A cross-sectional analysis. *Lancet Neurol*. 2015;14(3):253-262. doi:10.1016/S1474-4422(14)70324-2

- S42. Boeve B, Bove J, Brannelly P, et al. The longitudinal evaluation of familial frontotemporal dementia subjects protocol: Framework and methodology. In: *Alzheimer's and Dementia*. Vol 16. John Wiley and Sons Inc.; 2020:22-36. doi:10.1016/j.jalz.2019.06.4947
- S43. Rosen HJ, Boeve BF, Boxer AL. Tracking disease progression in familial and sporadic frontotemporal lobar degeneration: Recent findings from ARTFL and LEFFTDS. *Alzheimer's Dement*. 2020;16(1):71-78. doi:10.1002/alz.12004
- S44. Moore KM, Nicholas J, Grossman M, et al. Age at symptom onset and death and disease duration in genetic frontotemporal dementia: an international retrospective cohort study. *Lancet Neurol*. 2020;19(2):145-156. doi:10.1016/S1474-4422(19)30394-1
- S45. Otto M, Ludolph AC, Landwehrmeyer B, et al. Konsortium zur Erforschung der frontotemporalen Lobärdegeneration. *Nervenarzt*. 2011;82(8):1002-1005. doi:10.1007/s00115-011-3261-3
- S46. DZNE DESCRIBE (DZNE - Clinical Register Study of neurodegenerative Disorders) > DESCRIBE-FTD (DZNE - Clinical Registry Study on Frontotemporal Dementia (FTD)). Accessed July 29, 2020. <https://www.dzne.de/en/research/studies/clinical-studies/describe/describe-ftd>
- S47. Jabbari E, Holland N, Chelban V, et al. Diagnosis across the spectrum of progressive supranuclear palsy and corticobasal syndrome. *JAMA Neurol*. 2020;77(3):377-387. doi:10.1001/jamaneurol.2019.4347
- S48. 4 repeat tauopathy neuroimaging initiative. Accessed July 29, 2020. <http://4rtni-ftldni.ini.usc.edu/>
- S49. Galimberti D, Fumagalli GG, Fenoglio C, et al. Progranulin plasma levels predict the presence of GRN mutations in asymptomatic subjects and do not correlate with brain atrophy: results from the GENFI study. *Neurobiol Aging*. 2018;62:245.e9-245.e12. doi:10.1016/j.neurobiolaging.2017.10.016
- S50. Meeter LHH, Gendron TF, Sias AC, et al. Poly(GP), neurofilament and grey matter deficits in C9orf72 expansion carriers. *Ann Clin Transl Neurol*. 2018;5(5):583-597. doi:10.1002/acn3.559