



Universiteit
Leiden
The Netherlands

Neuroimaging biomarkers in genetic frontotemporal dementia : towards a timely diagnosis

Feis, R.A.

Citation

Feis, R. A. (2020, October 14). *Neuroimaging biomarkers in genetic frontotemporal dementia : towards a timely diagnosis*. Retrieved from <https://hdl.handle.net/1887/137726>

Version: Publisher's Version

License: [Licence agreement concerning inclusion of doctoral thesis in the Institutional Repository of the University of Leiden](#)

Downloaded from: <https://hdl.handle.net/1887/137726>

Note: To cite this publication please use the final published version (if applicable).

Cover Page



Universiteit Leiden

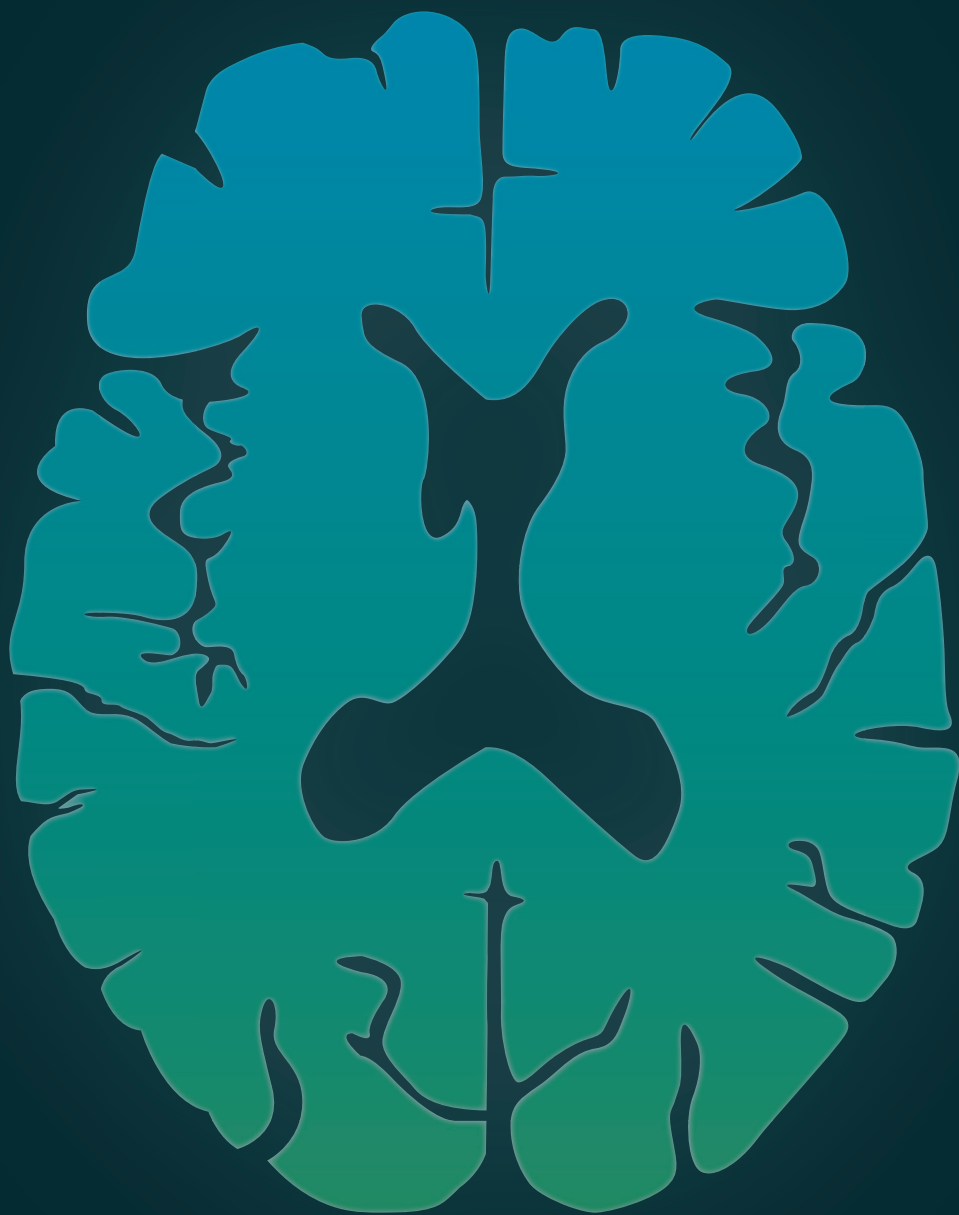


The handle <http://hdl.handle.net/1887/137726> holds various files of this Leiden University dissertation.

Author: Feis, R.A.

Title: Neuroimaging biomarkers in genetic frontotemporal dementia : towards a timely diagnosis

Issue Date: 2020-10-14



Appendix

Abbreviations

Bibliography

List of publications

Dankwoord

Curriculum vitae

Abbreviations

3DT _{1w}	3-dimensional T ₁ -weighted MRI
ABIDE	Autism Brain Imaging Data Exchange
AD	Alzheimer's disease
ALFF	amplitude of low frequency fluctuations
ALS	amyotrophic lateral sclerosis
<i>APOE4</i>	apolipoprotein E ϵ 4
ARTFL	Advancing Research and Treatment for FTLTD
ATR	anterior thalamic radiation
AxD	axial diffusivity
AUC	area under the ROC curve
BNT	Boston naming test
bvFTD	behavioural variant FTD
<i>C9orf72</i>	chromosome 9 open reading frame 72
CBS	corticobasal syndrome
CGC	cingulum in the cingulate gyrus area
CGH	cingulum in the hippocampal area
<i>CHMP2B</i>	charged multivesicular body protein 2b
CST	corticospinal tract
DNA	deoxyribonucleic acid
DTI	diffusion tensor imaging
DWI	diffusion-weighted imaging
FA	fractional anisotropy
FCor	full correlations between ICA components' time courses
FDA	United States Food and Drug Administration
FIX	FMRIB's ICA-based X-noiseifier
FMA	forceps major
FMI	forceps minor
fMRI	functional MRI
FMRIB	Functional Magnetic Resonance Imaging of the Brain Centre
FOV	field of view
FSL	FMRIB Software Library
FTD	frontotemporal dementia
FTD-RisC	FTD Risk Cohort
FTLD	frontotemporal lobar degeneration
FUS	fused in sarcoma protein
<i>FUS</i>	fused in sarcoma gene
FWE(RC)	family-wise error (rate corrected)
GENFI	Genetic Frontotemporal Dementia Initiative
GICA	group-level ICA
GMD	grey matter density
<i>GRN</i>	progranulin
IC(A)	independent component (analysis)
IFO	inferior fronto-occipital fasciculus
ILF	inferior longitudinal fasciculus

L	left
LDST	letter digit substitution test
LEFFTDS	Longitudinal Evaluation of Familial Frontotemporal Dementia Subjects
LUMC	Leiden University Medical Centre
lvPPA	logopenic variant PPA
<i>MAPT</i>	microtubule-associated protein tau
MD	mean diffusivity
MMSE	mini-mental state examination
MRI	magnetic resonance imaging
nvPPA	non-fluent variant PPA
OCMR	Oxford Centre for Clinical Magnetic Resonance Research
PCor	L1-regularised partial correlations between ICA components' time courses
PE	parameter estimate
PET	positron emission tomography
PPA	primary progressive aphasia
PSP	progressive supranuclear palsy
PVN	primary/medial visual network
R	right
RAVLT	Rey auditory verbal learning test
RD	radial diffusivity
RNA	ribonucleic acid
ROC	receiver operating characteristic
rs-fMRI	resting-state fMRI
RSN	resting-state network
SAT	semantic association test
SLF	superior longitudinal fasciculus
svPPA	semantic variant PPA
TBSS	tract-based spatial statistics
TDP-43	transactive response DNA binding protein 43 kDa
TE	echo time
TFCE	threshold-free cluster enhancement
TMT	trail making test
TNR	true-negative rate
TPR	true-positive rate
TR	repetition time
TSLF	temporal projection of the SLF
UF	uncinate fasciculus
VAT	visual association test
<i>VCP</i>	valosin-containing protein
WAIS	Wechsler adult intelligence scale
WCST	Wisconsin card sorting test
WMD	white matter density

Bibliography

- Adluru, N., Destiche, D.J., Lu, S.Y.-F., Doran, S.T., Birdsill, A.C., Melah, K.E., Okonkwo, O.C., Alexander, A.L., Dowling, N.M., Johnson, S.C., Sager, M.A., and Bendlin, B.B. (2014). White matter microstructure in late middle-age: Effects of apolipoprotein E4 and parental family history of Alzheimer's disease. *NeuroImage: Clinical*, 4:730–42.
- Agosta, F., Vessel, K.A., Miller, B.L., Migliaccio, R., Bonasera, S.J., Filippi, M., Boxer, A.L., Karydas, A., Possin, K.L., and Gorno-Tempini, M.L. (2009). Apolipoprotein E epsilon4 is associated with disease-specific effects on brain atrophy in Alzheimer's disease and frontotemporal dementia. *Proceedings of the National Academy of Sciences of the USA*, 106(6):2018–22.
- Agosta, F., Scola, E., Canu, E., Marcone, A., Magnani, G., Sarro, L., Copetti, M., Caso, F., Cerami, C., Comi, G., Cappa, S.F., Falini, A., and Filippi, M. (2012). White matter damage in frontotemporal lobar degeneration spectrum. *Cerebral Cortex*, 22(12):2705–14.
- Anderson, J.L.R., Jenkinson, M., and Smith, S.M. (2007). Non-linear registration aka Spatial normalisation. FMRIB Technical Report TR07JA2.
- Armstrong, M.J., Litvan, I., Lang, A.E., Bak, T.H., Bhatia, K.P., Borroni, B., Boxer, A.L., Dickson, D.W., Grossman, M., Hallett, M., Josephs, K.A., Kertesz, A., Lee, S.E., Miller, B.L., Reich, S.G., Riley, D.E., Tolosa, E., Tröster, A.I., Vidailhet, M., and Weiner, W.J. (2013). Criteria for the diagnosis of corticobasal degeneration. *Neurology*, 80(5):496–503.
- Army Individual Test Battery (1944). Manual of directions and scoring. *War Department, Adjunct General's Office, Washington, DC*.
- Bach, M., Laun, F.B., Leemans, A., Tax, C.M.W., Biessels, G.J., Stieltjes, B., and Maier-Hein, K.H. (2014). Methodological considerations on tract-based spatial statistics (TBSS). *NeuroImage*, 100:358–69.
- Basser, P.J., Mattiello, J., and LeBihan, D. (1994). MR diffusion tensor spectroscopy and imaging. *Biophysical Journal*, 66(1):259–67.
- Beckmann, C.F., and Smith, S.M. (2004). Probabilistic independent component analysis for functional magnetic resonance imaging. *IEEE Transactions on Medical Imaging*, 23(2):137–52.
- Beckmann, C.F., DeLuca, M., Devlin, J.T., and Smith, S.M. (2005). Investigations into resting-state connectivity using independent component analysis. *Philosophical Transactions of the Royal Society B, Biological Sciences*, 360(1457):1001–13.
- Beckmann, C.F., Mackay, C.E., Filippini, N., and Smith, S.M. (2009). Group comparison of resting-state fMRI data using multi-subject ICA and dual regression. *OHBM*.
- Benussi, A., Padovani, A., and Borroni, B. (2015). Phenotypic Heterogeneity of Monogenic Frontotemporal Dementia. *Frontiers in Aging Neuroscience*, 7:171.
- Bertrand, A., Wen, J., Rinaldi, D., Houot, M., Sayah, S., Camuzat, A., Fournier, C., Fontanella, S., Routier, A., Couratier, P., Pasquier, F., Habert, M.-O., Hannequin, D., Martinaud, O., Caroppo, P., Levy, R., Dubois, B., Brice, A., Durrleman, S., Colliot, O., and Le Ber, I. (2018). Early Cognitive, Structural, and Microstructural Changes in Presymptomatic C9orf72

- Carriers Younger Than 40 Years. *JAMA Neurology*, 75(2):236–45.
- Bey, K., Montag, C., Reuter, M., Weber, B., and Markett, S. (2015). Susceptibility to everyday cognitive failure is reflected in functional network interactions in the resting brain. *NeuroImage*, 121:1–9.
- Biswal, B., Mennes, M., Zuo, X.-N., Gohel, S., Kelly, C., Smith, S.M., Beckmann, C.F., Adelstein, J.S., Buckner, R.L., Colcombe, S., Dogonowski, A., Ernst, M., Fair, D., Hampson, M., Hoptman, M.J., Hyde, J.S., Kiviniemi, V.J., Kötter, R., Li, S., Lin, C., Lowe, M.J., Mackay, C.E., Madden, D.J., Madsen, K.H., Margulies, D.S., Mayberg, H.S., McMahon, K., Monk, C.S., Mostofsky, S.H., Nagel, B.J., Pekar, J.J., Peltier, S.J., Petersen, S.E., Riedl, V., Rombouts, S.A.R.B., Rypma, B., Schlaggar, B.L., Schmidt, S., Seidler, R.D., Siegle, G.J., Sorg, C., Teng, G., Veijola, J., Villringer, A., Walter, M., Wang, L., Weng, X., Whitfield-Gabrieli, S., Williamson, P., Windischberger, C., Zang, Y.-F., Zhang, H., Castellanos, F.X., and Milham, M.P. (2010). Toward discovery science of human brain function. *Proceedings of the National Academy of Sciences of the USA*, 107(10):4734–9.
- Borroni, B., Alberici, A., Premi, E., Archetti, S., Garibotto, V., Agosti, C., Gasparotti, R., Di Luca, M., Perani, D., and Padovani, A. (2008). Brain magnetic resonance imaging structural changes in a pedigree of asymptomatic progranulin mutation carriers. *Rejuvenation Research*, 11(3):585–95.
- Borroni, B., Alberici, A., Cercignani, M., Premi, E., Serra, L., Cerini, C., Cosseddu, M., Pettenati, C., Turla, M., Archetti, S., Gasparotti, R., Caltagirone, C., Padovani, A., and Bozzali, M. (2012). Granulin mutation drives brain damage and reorganization from preclinical to symptomatic FTL. *Neurobiology of Aging*, 33(10):2506–20.
- Bouts, M.J.R.J., Möller, C., Hafkemeijer, A., van Swieten, J.C., Doppler, E.G.P., van der Flier, W.M., Vrenken, H., Wink, A.M., Pijnenburg, Y.A.L., Scheltens, P., Barkhof, F., Schouten, T.M., de Vos, F., Feis, R.A., van der Grond, J., de Rooij, M., and Rombouts, S.A.R.B. (2018). Single Subject Classification of Alzheimer's Disease and Behavioral Variant Frontotemporal Dementia Using Anatomical, Diffusion Tensor, and Resting-State Functional Magnetic Resonance Imaging. *Journal of Alzheimer's Disease*, 62(4):1827–39.
- Boxer, A.L., Yu, J.-T., Golbe, L.I., Litvan, I., Lang, A.E., and Höglinger, G.U. (2017). Advances in progressive supranuclear palsy: new diagnostic criteria, biomarkers, and therapeutic approaches. *The Lancet Neurology*, 16(7):552–63.
- Bron, E.E., Smits, M., Papma, J.M., Steketee, R.M.E., Meijboom, R., de Groot, M., van Swieten, J.C., Niessen, W.J., and Klein, S. (2017). Multiparametric computer-aided differential diagnosis of Alzheimer's disease and frontotemporal dementia using structural and advanced MRI. *European Radiology*, 27(8):3372–82.
- Brooks, B.R., Miller, R.G., Swash, M., Munsat, T.L., and World Federation of Neurology Research Group on Motor Neuron Diseases (2000). El Escorial revisited: revised criteria for the diagnosis of amyotrophic lateral sclerosis. *Amyotrophic Lateral Sclerosis and Other Motor Neuron Disorders*, 1(5):293–9.
- Cacciaglia, R., Molinuevo, J.L., Falcón, C., Brugalat-Serrat, A., Sánchez-Benavides, G., Gramunt, N., Esteller, M., Morán, S., Minguillón, C., Fauria, K., and Gispert, J.D. (2018). Effects of

- APOE- $\epsilon 4$ allele load on brain morphology in a cohort of middle-aged healthy individuals with enriched genetic risk for Alzheimer's disease. *Alzheimer's & Dementia*, 14(7):902–12.
- Canu, E., Agosta, F., Mandic-Stojmenovic, G., Stojković, T., Stefanova, E., Inuggi, A., Imperiale, F., Copetti, M., Kostic, V.S., and Filippi, M. (2017). Multiparametric MRI to distinguish early onset Alzheimer's disease and behavioural variant of frontotemporal dementia. *NeuroImage: Clinical*, 15:428–38.
- Casey, B.J., Cohen, J.D., O'Craven, K., Davidson, R.J., Irwin, W., Nelson, C., Noll, D.C., Hu, X., Lowe, M.J., Rosen, B.R., Truwitt, C.L., and Turski, P.A. (1998). Reproducibility of fMRI results across four institutions using a spatial working memory task. *NeuroImage*, 8(8):249–61.
- Cash, D.M., Bocchetta, M., Thomas, D.L., Dick, K.M., van Swieten, J.C., Borroni, B., Galimberti, D., Masellis, M., Tartaglia, M.C., Rowe, J.B., Graff, C., Tagliavini, F., Frisoni, G.B., Laforce, R., Finger, E., de Mendonça, A., Sorbi, S., Rossor, M.N., Ourselin, S., and Rohrer, J.D. (2018). Patterns of gray matter atrophy in genetic frontotemporal dementia: results from the GENFI study. *Neurobiology of Aging*, 62:191–6.
- Cavedo, E., Lista, S., Rojkova, K., Chiesa, P.A., Houot, M., Brueggen, K., Blautzik, J., Bokde, A.L.W., Dubois, B., Barkhof, F., Pouwels, P.J.W., Teipel, S., and Hampel, H. (2017). Disrupted white matter structural networks in healthy older adult APOE $\epsilon 4$ carriers – An international multicenter DTI study. *Neuroscience*, 357:119–33.
- Chao, L.L., Schuff, N., Clevenger, E.M., Mueller, S.G., Rosen, H.J., Gorno-Tempini, M.L., Kramer, J.H., Miller, B.L., and Weiner, M.W. (2007). Patterns of White Matter Atrophy in Frontotemporal Lobar Degeneration. *Archives of Neurology*, 64(11):1619–24.
- Chare, L., Hodges, J.R., Leyton, C.E., McGinley, C., Tan, R.H., Kril, J.J., and Halliday, G.M. (2014). New criteria for frontotemporal dementia syndromes: clinical and pathological diagnostic implications. *Journal of Neurology, Neurosurgery, and Psychiatry*, 85(8):865–70.
- Chen, G., Cox, R.W., Glen, D.R., Rajendra, J.K., Reynolds, R.C., and Taylor, P.A. (2019). A tail of two sides: Artificially doubled false positive rates in neuroimaging due to the sidedness choice with t-tests. *Human Brain Mapping*, 40(3):1037–43.
- Cherbuin, N., Anstey, K.J., Sachdev, P.S., Maller, J.J., Meslin, C., Mack, H.A., Wen, W., and Easteal, S. (2008). Total and regional gray matter volume is not related to APOE*E4 status in a community sample of middle-aged individuals. *The Journals of Gerontology: Series A*, 63(5):501–4.
- Cocozza, S., Saccà, F., Cervo, A., Marsili, A., Russo, C.V., Maria Delle Acque Giorgio, S., De Michele, G., Filla, A., Brunetti, A., and Quarantelli, M. (2015). Modifications of resting state networks in spinocerebellar ataxia type 2. *Movement Disorders*, 30(10):1382–90.
- Cohn-Hokke, P.E., van Swieten, J.C., Pijnenburg, Y.A.L., Tibben, A., Meijers-Heijboer, H., and Kievit, A. (2018). The Effect of Predictive Testing in Adult-Onset Neurodegenerative Diseases on Social and Personal Life. *Journal of Genetic Counseling*, 27(4):947–54.
- Costafreda, S.G., Brammer, M.J., Vêncio, R.Z.N., Mourão, M.L., Portela, L.A.P., de Castro, C.C., Giampietro, V.P., and Amaro, E. (2007). Multisite fMRI reproducibility of a motor task

- using identical MR systems. *Journal of Magnetic Resonance Imaging*, 26(4):1122–6.
- Cummings, J., Ritter, A., and Zhong, K. (2018). Clinical Trials for Disease-Modifying Therapies in Alzheimer's Disease: A Primer, Lessons Learned, and a Blueprint for the Future. *Journal of Alzheimer's Disease*, 64:3–22.
- Daianu, M., Mendez, M.F., Baboyan, V.G., Jin, Y., Melrose, R.J., Jimenez, E.E., and Thompson, P.M. (2016). An advanced white matter tract analysis in frontotemporal dementia and early-onset Alzheimer's disease. *Brain Imaging and Behavior*, 10(4):1038–53.
- Damoiseaux, J.S., Rombouts, S.A.R.B., Barkhof, F., Scheltens, P., Stam, C.J., Smith, S.M., and Beckmann, C.F. (2006). Consistent resting-state networks across healthy subjects. *Proceedings of the National Academy of Sciences of the USA*, 103(37):13848–53.
- Davatzikos, C., Resnick, S.M., Wu, X., Parnpi, P., and Clark, C.M. (2008). Individual patient diagnosis of AD and FTD via high-dimensional pattern classification of MRI. *NeuroImage*, 41(4):1220–7.
- De Luca, M., Beckmann, C.F., De Stefano, N., Matthews, P.M., and Smith, S.M. (2006). fMRI resting state networks define distinct modes of long-distance interactions in the human brain. *NeuroImage*, 29(4):1359–67.
- de Vos, F., Schouten, T.M., Hafkemeijer, A., Dopper, E.G.P., van Swieten, J.C., de Rooij, M., van der Grond, J., and Rombouts, S.A.R.B. (2016). Combining multiple anatomical MRI measures improves Alzheimer's disease classification. *Human Brain Mapping*, 37(5):1920–9.
- de Vos, F., Koini, M., Schouten, T.M., Seiler, S., van der Grond, J., Lechner, A., Schmidt, R., de Rooij, M., and Rombouts, S.A.R.B. (2018). A comprehensive analysis of resting state fMRI measures to classify individual patients with Alzheimer's disease. *NeuroImage*, 167:62–72.
- Dell'Acqua, F., Scifo, P., Rizzo, G., Catani, M., Simmons, A., Scotti, G., and Fazio, F. (2010). A modified damped Richardson-Lucy algorithm to reduce isotropic background effects in spherical deconvolution. *NeuroImage*, 49(2):1446–58.
- Dell'Acqua, F., and Tournier, J.-D. (2019). Modelling white matter with spherical deconvolution: How and why? *NMR in Biomedicine*, 32(4):e3945.
- Di Martino, A., Yan, C.-G., Li, Q., Denio, E., Castellanos, F.X., Alaerts, K., Anderson, J.S., Assaf, M., Bookheimer, S.Y., Dapretto, M., Deen, B., Delmonte, S., Dinstein, I., Ertl-Wagner, B., Fair, D.A., Gallagher, L., Kennedy, D.P., Keown, C.L., Keysers, C., Lainhart, J.E., Lord, C., Luna, B., Menon, V., Minshew, N.J., Monk, C.S., Mueller, S., Müller, R.-A., Nebel, M.B., Nigg, J.T., O'Hearn, K., Pelphrey, K.A., Peltier, S.J., Rudie, J.D., Sunaert, S., Thioux, M., Tyszka, J.M., Uddin, L.Q., Verhoeven, J.S., Wenderoth, N., Wiggins, J.L., Mostofsky, S.H., and Milham, M.P. (2014). The autism brain imaging data exchange: towards a large-scale evaluation of the intrinsic brain architecture in autism. *Molecular Psychiatry*, 19(6):659–67.
- Doesborgh, S.J.C., van de Sandt-Koenderman, W.M.E., Dippel, D.W.J., van Harskamp, F., Koudstaal, P.J., and Visch-Brink, E.G. (2003). Linguistic deficits in the acute phase of stroke. *Journal of Neurology*, 250(8):977–82.
- Dopper, E.G.P., Rombouts, S.A.R.B., Jiskoot, L.C., den Heijer, T., de Graaf, J.R.A., de Koning, I., Hammerschlag, A.R., Seelaar, H., Seeley, W.W., Veer, I.M., van Buchem, M.A., Rizzu, P., and

- van Swieten, J.C. (2014). Structural and functional brain connectivity in presymptomatic familial frontotemporal dementia. *Neurology*, 83(2):e19-26.
- Douaud, G., Smith, S.M., Jenkinson, M., Behrens, T., Johansen-Berg, H., Vickers, J., James, S., Voets, N., Watkins, K., Matthews, P.M., and James, A. (2007). Anatomically related grey and white matter abnormalities in adolescent-onset schizophrenia. *Brain*, 130(9):2375–86.
- Ekman, P., and Friesen, W.V. (1976). Pictures of facial affect. *Consulting Psychologists Press, Palo Alto*.
- Elahi, F.M., Marx, G., Cobigo, Y., Staffaroni, A.M., Kornak, J., Tosun, D., Boxer, A.L., Kramer, J.H., Miller, B.L., and Rosen, H.J. (2017). Longitudinal white matter change in frontotemporal dementia subtypes and sporadic late onset Alzheimer’s disease. *NeuroImage: Clinical*, 16:595–603.
- Farb, N.A.S., Grady, C.L., Strother, S., Tang-Wai, D.F., Masellis, M., Black, S., Freedman, M., Pollock, B.G., Campbell, K.L., Hasher, L., and Chow, T.W. (2013). Abnormal network connectivity in frontotemporal dementia: Evidence for prefrontal isolation. *Cortex*, 49(7):1856–73.
- Fawcett, T. (2006). An introduction to ROC analysis. *Pattern Recognition Letters*, 27(8):861–74.
- Feige, B., Scheffler, K., Esposito, F., Francesco, D.S., Hennig, J., and Seifritz, E. (2005). Cortical and subcortical correlates of electroencephalographic alpha rhythm modulation. *Journal of Neurophysiology*, 93:2864–72.
- Feinberg, D.A., Moeller, S., Smith, S.M., Auerbach, E., Ramanna, S., Glasser, M.F., Miller, K.L., Ugurbil, K., and Yacoub, E. (2010). Multiplexed Echo Planar Imaging for Sub-Second Whole Brain fMRI and Fast Diffusion Imaging. *PLOS One*, 5(12):e15710.
- Feis, R.A., Smith, S.M., Filippini, N., Douaud, G., Dopper, E.G.P., Heise, V., Trachtenberg, A.J., van Swieten, J.C., van Buchem, M.A., Rombouts, S.A.R.B., and Mackay, C.E. (2015). ICA-based artifact removal diminishes scan site differences in multi-center resting-state fMRI. *Frontiers in Neuroscience*, 9:395.
- Feis, R.A., Bouts, M.J.R.J., Panman, J.L., Jiskoot, L.C., Dopper, E.G.P., Schouten, T.M., de Vos, F., van der Grond, J., van Swieten, J.C., and Rombouts, S.A.R.B. (2019a). Single-subject classification of presymptomatic frontotemporal dementia mutation carriers using multimodal MRI. *NeuroImage: Clinical*, 22:101718.
- Feis, R.A., Bouts, M.J.R.J., de Vos, F., Schouten, T.M., Panman, J.L., Jiskoot, L.C., Dopper, E.G.P., van der Grond, J., van Swieten, J.C., and Rombouts, S.A.R.B. (2019b). A multimodal MRI-based classification signature emerges just prior to symptom onset in frontotemporal dementia mutation carriers. *Journal of Neurology, Neurosurgery, and Psychiatry*, 90(11):1207–14.
- Filippini, N., MacIntosh, B.J., Hough, M.G., Goodwin, G.M., Frisoni, G.B., Smith, S.M., Matthews, P.M., Beckmann, C.F., and Mackay, C.E. (2009). Distinct patterns of brain activity in young carriers of the APOE-epsilon4 allele. *Proceedings of the National Academy of Sciences of the USA*, 106(17):7209–14.
- Filippini, N., Ebmeier, K.P., MacIntosh, B.J., Trachtenberg, A.J., Frisoni, G.B., Wilcock, G.K., Beckmann, C.F., Smith, S.M., Matthews, P.M., and Mackay, C.E. (2011). Differential effects of the APOE genotype on brain function across the lifespan. *NeuroImage*, 54(1):602–10.

- Folstein, M.F., Folstein, S.E., and McHugh, P.R. (1975). "Mini-mental state". A practical method for grading the cognitive state of patients for the clinician. *Journal of Psychiatric Research*, 12(3):189–98.
- Fortin, J.-P., Parker, D., Tunç, B., Watanabe, T., Elliott, M.A., Ruparel, K., Roalf, D.R., Satterthwaite, T.D., Gur, R.C., Gur, R.E., Schultz, R.T., Verma, R., and Shinohara, R.T. (2017). Harmonization of multi-site diffusion tensor imaging data. *NeuroImage*, 161:149–70.
- Fortin, J.-P., Cullen, N., Sheline, Y.I., Taylor, W.D., Aselcioglu, I., Cook, P.A., Adams, P., Cooper, C., Fava, M., McGrath, P.J., McInnis, M., Phillips, M.L., Trivedi, M.H., Weissman, M.M., and Shinohara, R.T. (2018). Harmonization of cortical thickness measurements across scanners and sites. *NeuroImage*, 167:104–20.
- Fox, M.D., Snyder, A.Z., Vincent, J.L., Corbetta, M., Van Essen, D.C., and Raichle, M.E. (2005). The human brain is intrinsically organized into dynamic, anticorrelated functional networks. *Proceedings of the National Academy of Sciences of the USA*, 102(27):9673–8.
- Fox, M.D., and Raichle, M.E. (2007). Spontaneous fluctuations in brain activity observed with functional magnetic resonance imaging. *Nature Reviews Neuroscience*, 8(9):700–11.
- Friedman, J., Hastie, T., and Tibshirani, R. (2008). Sparse inverse covariance estimation with the graphical lasso. *Biostatistics*, 9(3):432–41.
- Friedman, J., Hastie, T., and Tibshirani, R. (2010). Regularization Paths for Generalized Linear Models via Coordinate Descent. *Journal of Statistical Software*, 33(1):1–22.
- Friedman, L., Stern, H., Brown, G.G., Mathalon, D.H., Turner, J., Glover, G.H., Gollub, R.L., Lauriello, J., Lim, K.O., Cannon, T., Greve, D.N., Bockholt, H.J., Belger, A., Mueller, B., Doty, M.J., He, J., Wells, W., Smyth, P., Pieper, S., Kim, S., Kubicki, M., Vangel, M., and Potkin, S.G. (2008). Test-retest and between-site reliability in a multicenter fMRI study. *Human Brain Mapping*, 29(8):958–72.
- Frings, L., Yew, B., Flanagan, E., Lam, B.Y.K., Hüll, M., Huppertz, H.-J., Hodges, J.R., and Hornberger, M. (2014). Longitudinal Grey and White Matter Changes in Frontotemporal Dementia and Alzheimer's Disease. *PLOS One*, 9(3):e90814.
- Friston, K.J., Ashburner, J., Kiebel, S., Nichols, T., and Penny, W.D. (2007). *Statistical parametric mapping: the analysis of functional brain images*. London: Elsevier/Academic Press.
- Galantucci, S., Tartaglia, M.C., Wilson, S.M., Henry, M.L., Filippi, M., Agosta, F., Dronkers, N.F., Henry, R.G., Ogar, J.M., Miller, B.L., and Gorno-Tempini, M.L. (2011). White matter damage in primary progressive aphasia: a diffusion tensor tractography study. *Brain*, 134(10):3011–29.
- Galimberti, D., and Scarpini, E. (2012). Clinical phenotypes and genetic biomarkers of FTL. *Journal of Neural Transmission*, 119(7):851–60.
- Galimberti, D., Fumagalli, G.G., Fenoglio, C., Cioffi, S.M.G., Arighi, A., Serpente, M., Borroni, B., Padovani, A., Tagliavini, F., Masellis, M., Tartaglia, M.C., van Swieten, J.C., Meeter, L.H.H., Graff, C., de Mendonça, A., Bocchetta, M., Rohrer, J.D., Scarpini, E., Andersson, C., Archetti, S., Arighi, A., Benussi, L., Binetti, G., Black, S.E., Cash, D.M., Cosseddu, M., Dick, K.M., Fallström, M., Ferreira, C., Finger, E., Fox, N.C., Freedman, M., Frisoni, G.B.,

- Gazzina, S., Ghidoni, R., Grisoli, M., Jelic, V., Jiskoot, L.C., Keren, R., Laforce, R., Lombardi, G., Maruta, C., Mead, S., van Minkelen, R., Nacmias, B., Öjjerstedt, L., Ourselin, S., Panman, J.L., Pievani, M., Polito, C., Prioni, S., Rademakers, R., Redaelli, V., Rogaeva, E., Rossi, G., Besta, C., Rossor, M.N., Rowe, J.B., Sorbi, S., Tang-Wai, D.F., Thomas, D.L., Thonberg, H., Tiraboschi, P., Verdelho, A., and Warren, J.D. (2018). Progranulin plasma levels predict the presence of GRN mutations in asymptomatic subjects and do not correlate with brain atrophy: results from the GENFI study. *Neurobiology of Aging*, 62:245.e9-12.
- Gaudio, S., Piervincenzi, C., Beomonte Zobel, B., Romana Montecchi, F., Riva, G., Carducci, F., and Quattrocchi, C.C. (2015). Altered resting state functional connectivity of anterior cingulate cortex in drug naïve adolescents at the earliest stages of anorexia nervosa. *Scientific Reports*, 5:10818.
- Genin, E., Hannequin, D., Wallon, D., Slegers, K., Hiltunen, M., Combarros, O., Bullido, M.J., Engelborghs, S., De Deyn, P., Berr, C., Pasquier, F., Dubois, B., Tognoni, G., Fiévet, N., Brouwers, N., Bettens, K., Arosio, B., Coto, E., Del Zompo, M., Mateo, I., Epelbaum, J., Frank-Garcia, A., Helisalmi, S., Porcellini, E., Pilotto, A., Forti, P., Ferri, R., Scarpini, E., Siciliano, G., Solfrizzi, V., Sorbi, S., Spalletta, G., Valdivieso, F., Vepsäläinen, S., Alvarez, V., Bosco, P., Mancuso, M., Panza, F., Nacmias, B., Bossù, P., Hanon, O., Piccardi, P., Annoni, G., Seripa, D., Galimberti, D., Licastro, F., Soininen, H., Dartigues, J-F, Kamboh, M.I., Van Broeckhoven, C., Lambert, J.C., Amouyel, P., and Campion, D. (2011). APOE and Alzheimer disease: a major gene with semi-dominant inheritance. *Molecular Psychiatry*, 16(9):903–7.
- Glover, G.H., Mueller, B.A., Turner, J.A., van Erp, T.G.M., Liu, T.T., Greve, D.N., Voyvodic, J.T., Rasmussen, J., Brown, G.G., Keator, D.B., Calhoun, V.D., Lee, H.J., Ford, J.M., Mathalon, D.H., Diaz, M., O’Leary, D.S., Gadde, S., Preda, A., Lim, K.O., Wible, C.G., Stern, H.S., Belger, A., McCarthy, G., Ozyurt, B., and Potkin, S.G. (2012). Function biomedical informatics research network recommendations for prospective multicenter functional MRI studies. *Journal of Magnetic Resonance Imaging*, 36(1):39–54.
- Gold, B.T., Powell, D.K., Andersen, A.H., and Smith, C.D. (2010). Alterations in multiple measures of white matter integrity in normal women at high risk for Alzheimer’s disease. *NeuroImage*, 52(4):1487–94.
- Good, C.D., Johnsrude, I.S., Ashburner, J., Henson, R.N.A., Friston, K.J., and Frackowiak, R.S.J. (2001). A Voxel-Based Morphometric Study of Ageing in 465 Normal Adult Human Brains. *NeuroImage*, 14(1):21–36.
- Gorgolewski, K.J., Varoquaux, G., Rivera, G., Schwarz, Y., Ghosh, S.S., Maumet, C., Sochat, V. V., Nichols, T.E., Poldrack, R.A., Poline, J-B., Yarkoni, T., and Margulies, D.S. (2015). NeuroVault.org: a web-based repository for collecting and sharing unthresholded statistical maps of the human brain. *Frontiers in Neuroinformatics*, 9:8.
- Gorno-Tempini, M.L., Hillis, A.E., Weintraub, S., Kertesz, A., Mendez, M., Cappa, S.F., Ogar, J.M., Rohrer, J.D., Black, S., Boeve, B.F., Manes, F., Dronkers, N.F., Vandenberghe, R., Rascovsky, K., Patterson, K., Miller, B.L., Knopman, D.S., Hodges, J.R., Mesulam, M.M., and Grossman, M. (2011). Classification of primary progressive aphasia and its variants. *Neurology*, 76(11):1006–14.
- Graham, A., Davies, R., Xuereb, J., Halliday, G., Kril, J., Creasey, H., Graham, K., and Hodges,

- J. (2005). Pathologically proven frontotemporal dementia presenting with severe amnesia. *Brain*, 128(3):597–605.
- Greicius, M.D., Krasnow, B., Reiss, A.L., and Menon, V. (2003). Functional connectivity in the resting brain: a network analysis of the default mode hypothesis. *Proceedings of the National Academy of Sciences of the USA*, 100(1):253–8.
- Greve, D.N., and Fischl, B. (2009). Accurate and Robust Brain Image Alignment using Boundary-based Registration. *NeuroImage*, 48(1):63–72.
- Griffanti, L., Salimi-Khorshidi, G., Beckmann, C.F., Auerbach, E.J., Douaud, G., Sexton, C.E., Zsoldos, E., Ebmeier, K.P., Filippini, N., Mackay, C.E., Moeller, S., Xu, J., Yacoub, E., Baselli, G., Ugurbil, K., Miller, K.L., and Smith, S.M. (2014). ICA-based artefact removal and accelerated fMRI acquisition for improved resting state network imaging. *NeuroImage*, 95:232–47.
- Hafkemeijer, A., Möller, C., Dopper, E.G.P., Jiskoot, L.C., Schouten, T.M., van Swieten, J.C., van der Flier, W.M., Vrenken, H., Pijnenburg, Y.A.L., Barkhof, F., Scheltens, P., van der Grond, J., and Rombouts, S.A.R.B. (2015). Resting state functional connectivity differences between behavioral variant frontotemporal dementia and Alzheimer’s disease. *Frontiers in Human Neuroscience*, 9:474.
- Hafkemeijer, A., Möller, C., Dopper, E.G.P., Jiskoot, L.C., van den Berg-Huysmans, A.A., van Swieten, J.C., van der Flier, W.M., Vrenken, H., Pijnenburg, Y.A.L., Barkhof, F., Scheltens, P., van der Grond, J., and Rombouts, S.A.R.B. (2016). Differences in structural covariance brain networks between behavioral variant frontotemporal dementia and Alzheimer’s disease. *Human Brain Mapping*, 37(3):978–88.
- Handels, R.L., Aalten, P., Wolfs, C.A., OldeRikkert, M., Scheltens, P., Visser, P.J., Joore, M.A., Severens, J.L., and Verhey, F.R. (2012). Diagnostic and economic evaluation of new biomarkers for Alzheimer’s disease: the research protocol of a prospective cohort study. *BMC Neurology*, 12(1):72.
- Happé, F., Brownell, H., and Winner, E. (1999). Acquired “theory of mind” impairments following stroke. *Cognition*, 70(3):211–40.
- Harvey, R.J., Skelton-Robinson, M., and Rossor, M.N. (2003). The prevalence and causes of dementia in people under the age of 65 years. *Journal of Neurology, Neurosurgery, and Psychiatry*, 74(9):1206–9.
- Heise, V., Filippini, N., Ebmeier, K.P., and Mackay, C.E. (2011). The APOE $\epsilon 4$ allele modulates brain white matter integrity in healthy adults. *Molecular Psychiatry*, 16(9):908–16.
- Hodges, J.R., Davies, R., Xuereb, J., Kril, J., and Halliday, G. (2003). Survival in frontotemporal dementia. *Neurology*, 61(3):349–54.
- Hoerl, A.E., and Kennard, R.W. (1970). Ridge Regression: Biased Estimation for Nonorthogonal Problems. *Technometrics*, 12(1):55–67.
- Hogan, D.B., Jetté, N., Fiest, K.M., Roberts, J.I., Pearson, D., Smith, E.E., Roach, P., Kirk, A., Pringsheim, T., and Maxwell, C.J. (2016). The Prevalence and Incidence of Frontotemporal Dementia: a Systematic Review. *Canadian Journal of Neurological Sciences / Journal Canadien*

- des Sciences Neurologiques*, 43 Suppl 1:S96-109.
- Honea, R.A., Vidoni, E., Harsha, A., and Burns, J.M. (2009). Impact of APOE on the Healthy Aging Brain: A Voxel-Based MRI and DTI Study. *Journal of Alzheimer's Disease*, 18:553–64.
- Huey, E.D., Armstrong, N., Momeni, P., and Grafman, J. (2008). Challenges and new opportunities in the investigation of new drug therapies to treat frontotemporal dementia. *Expert Opinion on Therapeutic Targets*, 12(11):1367–76.
- Hyvärinen, A. (1999). Fast and robust fixed-point algorithms for independent component analysis. *IEEE Transactions on Neural Networks*, 10(3):626–34.
- Jacova, C., Hsiung, G.-Y.R., Tawankanjanachot, I., Dinelle, K., McCormick, S., Gonzalez, M., Lee, H., Sengdy, P., Bouchard-Kerr, P., Baker, M., Rademakers, R., Sossi, V., Stoessl, A.J., Feldman, H.H., and Mackenzie, I.R. (2013). Anterior brain glucose hypometabolism predates dementia in progranulin mutation carriers. *Neurology*, 81(15):1322–31.
- Jao, T., Vértes, P.E., Alexander-Bloch, A.F., Tang, I.-N., Yu, Y.-C., Chen, J.-H., and Bullmore, E.T. (2013). Volitional eyes opening perturbs brain dynamics and functional connectivity regardless of light input. *NeuroImage*, 69:21–34.
- Jenkinson, M., and Smith, S.M. (2001). A global optimisation method for robust affine registration of brain images. *Medical Image Analysis*, 5(2):143–56.
- Jenkinson, M., Bannister, P., Brady, M., and Smith, S.M. (2002). Improved optimization for the robust and accurate linear registration and motion correction of brain images. *NeuroImage*, 17(2):825–41.
- Jenkinson, M., Beckmann, C.F., Behrens, T.E.J., Woolrich, M.W., and Smith, S.M. (2012). FSL. *NeuroImage*, 62(2):782–90.
- Jiskoot, L.C., Dopper, E.G.P., den Heijer, T., Timman, R., van Minkelen, R., van Swieten, J.C., and Papma, J.M. (2016). Presymptomatic cognitive decline in familial frontotemporal dementia: A longitudinal study. *Neurology*, 87(4):384–91.
- Jiskoot, L.C., Bocchetta, M., Nicholas, J.M., Cash, D.M., Thomas, D., Modat, M., Ourselin, S., Rombouts, S.A.R.B., Dopper, E.G.P., Meeter, L.H.H., Panman, J.L., van Minkelen, R., van der Ende, E.L., Donker Kaat, L., Pijnenburg, Y.A.L., Borroni, B., Galimberti, D., Masellis, M., Tartaglia, M.C., Rowe, J., Graff, C., Tagliavini, F., Frisoni, G.B., Laforce, R., Finger, E., de Mendonça, A., Sorbi, S., Papma, J.M., van Swieten, J.C., and Rohrer, J.D. (2018a). Presymptomatic white matter integrity loss in familial frontotemporal dementia in the GENFI cohort: A cross-sectional diffusion tensor imaging study. *Annals of Clinical and Translational Neurology*, 5(9):1025–36.
- Jiskoot, L.C., Panman, J.L., van Asseldonk, L., Franzen, S., Meeter, L.H.H., Donker Kaat, L., van der Ende, E.L., Dopper, E.G.P., Timman, R., van Minkelen, R., van Swieten, J.C., van den Berg, E., and Papma, J.M. (2018b). Longitudinal cognitive biomarkers predicting symptom onset in presymptomatic frontotemporal dementia. *Journal of Neurology*, 265(6):1381–92.
- Jiskoot, L.C., Panman, J.L., Meeter, L.H.H., Dopper, E.G.P., Donker Kaat, L., Franzen, S., van der Ende, E.L., van Minkelen, R., Rombouts, S.A.R.B., Papma, J.M., and van Swieten, J.C. (2019). Longitudinal multimodal MRI as prognostic and diagnostic biomarker in presymptomatic

- familial frontotemporal dementia. *Brain*, 142(1):193–208.
- Johnson, J.K., Head, E., Kim, R., Starr, A., and Cotman, C.W. (1999). Clinical and Pathological Evidence for a Frontal Variant of Alzheimer Disease. *Archives of Neurology*, 56(10):1233–9.
- Johnson, J.K., Diehl, J., Mendez, M.F., Neuhaus, J., Shapira, J.S., Forman, M., Chute, D.J., Roberson, E.D., Pace-Savitsky, C., Neumann, M., Chow, T.W., Rosen, H.J., Forstl, H., Kurz, A., and Miller, B.L. (2005). Frontotemporal Lobar Degeneration. *Archives of Neurology*, 62(6):925–30.
- Johnson, W.E., Li, C., and Rabinovic, A. (2007). Adjusting batch effects in microarray expression data using empirical Bayes methods. *Biostatistics*, 8(1):118–27.
- Jolles, J., Houx, P.J., van Boxtel, M.P.J., and Ponds, R. (1995). Maastricht Aging Study: determinants of cognitive aging. *Neuropsych Publishers, Maastricht*.
- Josephs, K.A., Petersen, R.C., Knopman, D.S., Boeve, B.F., Whitwell, J.L., Duffy, J.R., Parisi, J.E., and Dickson, D.W. (2006). Clinicopathologic analysis of frontotemporal and corticobasal degenerations and PSP. *Neurology*, 66(1):41–8.
- Josephs, K.A., Hodges, J.R., Snowden, J.S., Mackenzie, I.R., Neumann, M., Mann, D.M., and Dickson, D.W. (2011). Neuropathological background of phenotypical variability in frontotemporal dementia. *Acta Neuropathologica*, 122(2):137–53.
- Kansal, K., Mareddy, M., Sloane, K.L., Minc, A.A., Rabins, P. V., McGready, J.B., and Onyike, C.U. (2016). Survival in Frontotemporal Dementia Phenotypes: A Meta-Analysis. *Dementia and Geriatric Cognitive Disorders*, 41(1–2):109–22.
- Kaplan, E., Goodglass, H., and Weintraub, S. (1978). The Boston Naming Test. *Lea & Febiger, Philadelphia*.
- Kazemi, K., and Noorizadeh, N. (2014). Quantitative Comparison of SPM, FSL, and Brainsuite for Brain MR Image Segmentation. *Journal of Biomedical Physics & Engineering*, 4(1):13–26.
- Kertesz, A., McMonagle, P., Blair, M., Davidson, W., and Munoz, D.G. (2005). The evolution and pathology of frontotemporal dementia. *Brain*, 128(9):1996–2005.
- Kim, J.P., Kim, J., Park, Y.H., Park, S.B., Lee, J.S., Yoo, S., Kim, E.-J., Kim, H.J., Na, D.L., Brown, J.A., Lockhart, S.N., Seo, S.W., and Seong, J.-K. (2019). Machine learning based hierarchical classification of frontotemporal dementia and Alzheimer’s disease. *NeuroImage: Clinical*, 23:101811.
- Klöppel, S., Peter, J., Ludl, A., Pilatus, A., Maier, S., Mader, I., Heimbach, B., Frings, L., Egger, K., Dukart, J., Schroeter, M.L., Perneczky, R., Häussermann, P., Vach, W., Urbach, H., Teipel, S., Hüll, M., Abdulkadir, A., and Alzheimer’s Disease Neuroimaging Initiative (2015). Applying Automated MR-Based Diagnostic Methods to the Memory Clinic: A Prospective Study. *Journal of Alzheimer’s Disease*, 47(4):939–54.
- Koikkalainen, J., Rhodius-Meester, H., Tolonen, A., Barkhof, F., Tijms, B., Lemstra, A.W., Tong, T., Guerrero, R., Schuh, A., Ledig, C., Rueckert, D., Soininen, H., Remes, A.M., Waldemar, G., Hasselbalch, S., Mecocci, P., van der Flier, W., and Lötjönen, J. (2016). Differential diagnosis of neurodegenerative diseases using structural MRI data. *NeuroImage: Clinical*, 11:435–49.

- Kriegeskorte, N., Simmons, W.K., Bellgowan, P.S.F., and Baker, C.I. (2009). Circular analysis in systems neuroscience: the dangers of double dipping. *Nature Neuroscience*, 12(5):535–40.
- Krudop, W.A., Kerssens, C.J., Dols, A., Prins, N.D., Möller, C., Schouws, S., van der Flier, W.M., Scheltens, P., Sikkes, S., Stek, M.L., and Pijnenburg, Y.A.L. (2015). Identifying bvFTD Within the Wide Spectrum of Late Onset Frontal Lobe Syndrome: A Clinical Approach. *The American Journal of Geriatric Psychiatry*, 23(10):1056–66.
- Laird, N.M., and Ware, J.H. (1982). Random-Effects Models for Longitudinal Data. *Biometrics*, 38(4):963–74.
- Laukka, E.J., Lövdén, M., Kalpouzos, G., Papenberg, G., Keller, L., Graff, C., Li, T.-Q., Fratiglioni, L., and Bäckman, L. (2015). Microstructural White Matter Properties Mediate the Association between APOE and Perceptual Speed in Very Old Persons without Dementia. *PLOS One*, 10(8):e0134766.
- Lee, S.E., Khazenzon, A.M., Trujillo, A.J., Guo, C.C., Yokoyama, J.S., Sha, S.J., Takada, L.T., Karydas, A.M., Block, N.R., Coppola, G., Pribadi, M., Geschwind, D.H., Rademakers, R., Fong, J.C., Weiner, M.W., Boxer, A.L., Kramer, J.H., Rosen, H.J., Miller, B.L., and Seeley, W.W. (2014). Altered network connectivity in frontotemporal dementia with C9orf72 hexanucleotide repeat expansion. *Brain*, 137(11):3047–60.
- Lee, S.E., Sias, A.C., Mandelli, M.L., Brown, J.A., Brown, A.B., Khazenzon, A.M., Vidovszky, A.A., Zanto, T.P., Karydas, A.M., Pribadi, M., Dokuru, D., Coppola, G., Geschwind, D.H., Rademakers, R., Gorno-Tempini, M.L., Rosen, H.J., Miller, B.L., and Seeley, W.W. (2017). Network degeneration and dysfunction in presymptomatic C9orf72 expansion carriers. *NeuroImage: Clinical*, 14:286–97.
- Leemans, A., and Jones, D.K. (2009). The B-matrix must be rotated when correcting for subject motion in DTI data. *Magnetic Resonance in Medicine*, 61(6):1336–49.
- Lehmer, C., Oeckl, P., Weishaupt, J.H., Volk, A.E., Diehl-Schmid, J., Schroeter, M.L., Lauer, M., Kornhuber, J., Levin, J., Fassbender, K., Landwehrmeyer, B., Schludi, M.H., Arzberger, T., Kremmer, E., Flatley, A., Feederle, R., Steinacker, P., Weydt, P., Ludolph, A.C., Edbauer, D., and Otto, M. (2017). Poly-GP in cerebrospinal fluid links C9orf72-associated dipeptide repeat expression to the asymptomatic phase of ALS/FTD. *EMBO Molecular Medicine*, 9(7):859–68.
- Liang, B., Zhang, D., Wen, X., Xu, P., Peng, X., Huang, X., Liu, M., and Huang, R. (2014). Brain spontaneous fluctuations in sensorimotor regions were directly related to eyes open and eyes closed: evidences from a machine learning approach. *Frontiers in Human Neuroscience*, 8:645.
- Lindeboom, J., Schmand, B., Tulner, L., Walstra, G., and Jonker, C. (2002). Visual association test to detect early dementia of the Alzheimer type. *Journal of Neurology, Neurosurgery, and Psychiatry*, 73(2):126–33.
- Littow, H. (2010). Age-related differences in functional nodes of the brain cortex – a high model order group ICA study. *Frontiers in Systems Neuroscience*, 4:32.
- Liu, D.-Q., Dong, Z., Zuo, X.-N., Wang, J., and Zang, Y.-F. (2013). Eyes-open/eyes-closed dataset sharing for reproducibility evaluation of resting state fMRI data analysis methods.

- Neuroinformatics*, 11(4):469–76.
- Lobo, A., Launer, L.J., Fratiglioni, L., Andersen, K., Di Carlo, A., Breteler, M.M.B., Copeland, J.R.M., Dartigues, J.-F., Jagger, C., Martinez-Lage, J., Soininen, H., Hofman, A., and Group for the Neurologic Diseases in the Elderly Research (2000). Prevalence of dementia and major subtypes in Europe: A collaborative study of population-based cohorts. *Neurology*, 54(11 Suppl 5):S4-9.
- Logroschino, G., Imbimbo, B.P., Lozupone, M., Sardone, R., Capozzo, R., Battista, P., Zecca, C., Dibello, V., Giannelli, G., Bellomo, A., Greco, A., Daniele, A., Seripa, D., and Panza, F. (2019). Promising therapies for the treatment of frontotemporal dementia clinical phenotypes: from symptomatic to disease-modifying drugs. *Expert Opinion on Pharmacotherapy*, 20(9):1091–107.
- Lomen-Hoerth, C., Anderson, T., and Miller, B. (2002). The overlap of amyotrophic lateral sclerosis and frontotemporal dementia. *Neurology*, 59(7):1077–9.
- Long, X.-Y., Zuo, X.-N., Kiviniemi, V.J., Yang, Y., Zou, Q.-H., Zhu, C.-Z., Jiang, T.-Z., Yang, H., Gong, Q.-Y., Wang, L., Li, K.-C., Xie, S., and Zang, Y.-F. (2008). Default mode network as revealed with multiple methods for resting-state functional MRI analysis. *Journal of Neuroscience Methods*, 171(2):349–55.
- Ludolph, A., Drory, V., Hardiman, O., Nakano, I., Ravits, J., Robberecht, W., Shefner, J., and The WFN Research Group On ALS/MND (2015). A revision of the El Escorial criteria – 2015. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 16(5–6):291–2.
- Lyall, D.M., Harris, S.E., Bastin, M.E., Muñoz Maniega, S., Murray, C., Lutz, M.W., Saunders, A.M., Roses, A.D., del Carmen Valdés Hernández, M., Royle, N.A., Starr, J.M., Porteous, D.J., Wardlaw, J.M., and Deary, I.J. (2014). Alzheimer's disease susceptibility genes APOE and TOMM40, and brain white matter integrity in the Lothian Birth Cohort 1936. *Neurobiology of Aging*, 35(6):1513.e25-33.
- Machulda, M.M., Jones, D.T., Vemuri, P., McDade, E., Avula, R., Przybelski, S., Boeve, B.F., Knopman, D.S., Petersen, R.C., and Jack, C.R. (2011). Effect of APOE ϵ 4 Status on Intrinsic Network Connectivity in Cognitively Normal Elderly Subjects. *Archives of Neurology*, 68(9):1131–6.
- Mahoney, C.J., Ridgway, G.R., Malone, I.B., Downey, L.E., Beck, J., Kinnunen, K.M., Schmitz, N., Golden, H.L., Rohrer, J.D., Schott, J.M., Rossor, M.N., Ourselin, S., Mead, S., Fox, N.C., and Warren, J.D. (2014). Profiles of white matter tract pathology in frontotemporal dementia. *Human Brain Mapping*, 35(8):4163–79.
- Mann, D.M.A., and Snowden, J.S. (2017). Frontotemporal lobar degeneration: Pathogenesis, pathology and pathways to phenotype. *Brain Pathology*, 27(6):723–36.
- Matura, S., Prvulovic, D., Jurcoane, A., Hartmann, D., Miller, J., Scheibe, M., O'Dwyer, L., Oertel-Knöchel, V., Knöchel, C., Reinke, B., Karakaya, T., Fußer, F., and Pantel, J. (2014). Differential effects of the ApoE4 genotype on brain structure and function. *NeuroImage*, 89:81–91.
- McAvoy, M., Larson-Prior, L., Nolan, T.S., Vaishnavi, S.N., Raichle, M.E., and D'Avossa, G. (2008). Resting states affect spontaneous BOLD oscillations in sensory and paralimbic cortex.

- Journal of Neurophysiology*, 100(2):922–31.
- McKhann, G.M., Knopman, D.S., Chertkow, H., Hyman, B.T., Jack, C.R., Kawas, C.H., Klunk, W.E., Koroshetz, W.J., Manly, J.J., Mayeux, R., Mohs, R.C., Morris, J.C., Rossor, M.N., Scheltens, P., Carrillo, M.C., Thies, B., Weintraub, S., and Phelps, C.H. (2011). The diagnosis of dementia due to Alzheimer's disease: Recommendations from the National Institute on Aging-Alzheimer's Association workgroups on diagnostic guidelines for Alzheimer's disease. *Alzheimer's & Dementia*, 7(3):263–9.
- McMillan, C.T., Brun, C., Siddiqui, S., Churgin, M., Libon, D., Yushkevich, P., Zhang, H., Boller, A., Gee, J., and Grossman, M. (2012). White matter imaging contributes to the multimodal diagnosis of frontotemporal lobar degeneration. *Neurology*, 78(22):1761–8.
- McMillan, C.T., Avants, B.B., Cook, P., Ungar, L., Trojanowski, J.Q., and Grossman, M. (2014). The power of neuroimaging biomarkers for screening frontotemporal dementia. *Human Brain Mapping*, 35(9):4827–40.
- Meeter, L.H.H., Dopfer, E.G.P., Jiskoot, L.C., Sanchez-Valle, R., Graff, C., Benussi, L., Ghidoni, R., Pijnenburg, Y.A.L., Borroni, B., Galimberti, D., Laforce, R., Masellis, M., Vandenberghe, R., Le Ber, I., Otto, M., van Minkelen, R., Papma, J.M., Rombouts, S.A.R.B., Balasa, M., Öijerstedt, L., Jelic, V., Dick, K.M., Cash, D.M., Harding, S.R., Jorge Cardoso, M., Ourselin, S., Rossor, M.N., Padovani, A., Scarpini, E., Fenoglio, C., Tartaglia, M.C., Lamari, F., Barro, C., Kuhle, J., Rohrer, J.D., Teunissen, C.E., and van Swieten, J.C. (2016a). Neurofilament light chain: a biomarker for genetic frontotemporal dementia. *Annals of Clinical and Translational Neurology*, 3(8):623–36.
- Meeter, L.H.H., Patzke, H., Loewen, G., Dopfer, E.G.P., Pijnenburg, Y.A.L., van Minkelen, R., and van Swieten, J.C. (2016b). Progranulin Levels in Plasma and Cerebrospinal Fluid in Granulin Mutation Carriers. *Dementia and Geriatric Cognitive Disorders Extra*, 6(2):330–40.
- Meeter, L.H.H., Gendron, T.F., Sias, A.C., Jiskoot, L.C., Russo, S.P., Donker Kaat, L., Papma, J.M., Panman, J.L., van der Ende, E.L., Dopfer, E.G.P., Franzen, S., Graff, C., Boxer, A.L., Rosen, H.J., Sanchez-Valle, R., Galimberti, D., Pijnenburg, Y.A.L., Benussi, L., Ghidoni, R., Borroni, B., Laforce, R., Del Campo, M., Teunissen, C.E., van Minkelen, R., Rojas, J.C., Coppola, G., Geschwind, D.H., Rademakers, R., Karydas, A.M., Öijerstedt, L., Scarpini, E., Binetti, G., Padovani, A., Cash, D.M., Dick, K.M., Bocchetta, M., Miller, B.L., Rohrer, J.D., Petrucelli, L., van Swieten, J.C., and Lee, S.E. (2018a). Poly(GP), neurofilament and grey matter deficits in C9orf72 expansion carriers. *Annals of Clinical and Translational Neurology*, 5(5):583–97.
- Meeter, L.H.H., Vijverberg, E.G., Del Campo, M., Rozemuller, A.J.M., Donker Kaat, L., de Jong, F.J., van der Flier, W.M., Teunissen, C.E., van Swieten, J.C., and Pijnenburg, Y.A.L. (2018b). Clinical value of neurofilament and phospho-tau/tau ratio in the frontotemporal dementia spectrum. *Neurology*, 90(14):e1231-9.
- Mendez, M.F., Shapira, J.S., McMurtray, A., Licht, E., Miller, B.L., and Trojanowski, J.Q. (2007). Accuracy of the Clinical Evaluation for Frontotemporal Dementia. *Archives of Neurology*, 64(6):830–5.
- Mendez, M.F. (2009). 'Frontotemporal Dementia: Therapeutic Interventions', in: Giannakopoulos, P., and Hof, P.R. (Eds.), *Dementia in Clinical Practice*. KARGER, Basel, pp. 168–78.

- Meyer, S., Mueller, K., Stuke, K., Bisenius, S., Diehl-Schmid, J., Jessen, F., Kassubek, J., Kornhuber, J., Ludolph, A.C., Prudlo, J., Schneider, A., Schuemberg, K., Yakushev, I., Otto, M., and Schroeter, M.L. (2017). Predicting behavioral variant frontotemporal dementia with pattern classification in multi-center structural MRI data. *NeuroImage: Clinical*, 14:656–62.
- Mito, R., Raffelt, D., Dhollander, T., Vaughan, D.N., Tournier, J.-D., Salvado, O., Brodtmann, A., Rowe, C.C., Villemagne, V.L., and Connelly, A. (2018). Fibre-specific white matter reductions in Alzheimer's disease and mild cognitive impairment. *Brain*, 141(3):888–902.
- Miyoshi, M., Shinotoh, H., Wszolek, Z.K., Strongosky, A.J., Shimada, H., Arakawa, R., Higuchi, M., Ikoma, Y., Yasuno, F., Fukushi, K., Irie, T., Ito, H., and Suhara, T. (2010). In vivo detection of neuropathologic changes in presymptomatic MAPT mutation carriers: A PET and MRI study. *Parkinsonism & Related Disorders*, 16(6):404–8.
- Mohs, R.C., Doody, R.S., Morris, J.C., Ieni, J.R., Rogers, S.L., Perdomo, C.A., Pratt, R.D., and “312” Study Group (2001). A 1-year, placebo-controlled preservation of function survival study of donepezil in AD patients. *Neurology*, 57(3):481–8.
- Möller, C., Dieleman, N., van der Flier, W.M., Versteeg, A., Pijnenburg, Y.A.L., Scheltens, P., Barkhof, F., and Vrenken, H. (2015a). More Atrophy of Deep Gray Matter Structures in Frontotemporal Dementia Compared to Alzheimer's Disease. *Journal of Alzheimer's Disease*, 44(2):635–47.
- Möller, C., Hafkemeijer, A., Pijnenburg, Y.A.L., Rombouts, S.A.R.B., van der Grond, J., Dopper, E.G.P., van Swieten, J.C., Versteeg, A., Pouwels, P.J.W., Barkhof, F., Scheltens, P., Vrenken, H., and van Der Flier, W.M. (2015b). Joint assessment of white matter integrity, cortical and subcortical atrophy to distinguish AD from behavioral variant FTD: A two-center study. *NeuroImage: Clinical*, 9:418–29.
- Möller, C., Pijnenburg, Y.A.L., van der Flier, W.M., Versteeg, A., Tijms, B., de Munck, J.C., Hafkemeijer, A., Rombouts, S.A.R.B., van der Grond, J., van Swieten, J.C., Dopper, E.G.P., Scheltens, P., Barkhof, F., Vrenken, H., and Wink, A.M. (2016). Alzheimer Disease and Behavioral Variant Frontotemporal Dementia: Automatic Classification Based on Cortical Atrophy for Single-Subject Diagnosis. *Radiology*, 279(3):838–48.
- Neary, D., Snowden, J.S., Gustafson, L., Passant, U., Stuss, D., Black, S., Freedman, M., Kertesz, A., Robert, P.H., Albert, M., Boone, K., Miller, B.L., Cummings, J., and Benson, D.F. (1998). Frontotemporal lobar degeneration: a consensus on clinical diagnostic criteria. *Neurology*, 51(6):1546–54.
- Nelson, H.E. (1976). A modified card sorting test sensitive to frontal lobe defects. *Cortex*, 12(4):313–24.
- Nierenberg, J., Pomara, N., Hoptman, M.J., Sidtis, J.J., Ardekani, B.A., and Lim, K.O. (2005). Abnormal white matter integrity in healthy apolipoprotein E epsilon4 carriers. *Neuroreport*, 16(12):1369–72.
- Noirhomme, Q., Lesenfants, D., Gomez, F., Soddu, A., Schrouff, J., Garraux, G., Luxen, A., Phillips, C., and Laureys, S. (2014). Biased binomial assessment of cross-validated estimation of classification accuracies illustrated in diagnosis predictions. *NeuroImage: Clinical*, 4:687–94.

- Omer, T., Finegan, E., Hutchinson, S., Doherty, M., Vajda, A., McLaughlin, R.L., Pender, N., Hardiman, O., and Bede, P. (2017). Neuroimaging patterns along the ALS-FTD spectrum: a multiparametric imaging study. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*, 18(7–8):611–23.
- Operto, G., Cacciaglia, R., Grau-Rivera, O., Falcon, C., Brugulat-Serrat, A., Ródenas, P., Ramos, R., Morán, S., Esteller, M., Bargalló, N., Molinuevo, J.L., and Gispert, J.D. (2018). White matter microstructure is altered in cognitively normal middle-aged APOE- ϵ 4 homozygotes. *Alzheimer's Research & Therapy*, 10(1):48.
- Pan, P.L., Song, W., Yang, J., Huang, R., Chen, K., Gong, Q.-Y., Zhong, J.G., Shi, H.C., and Shang, H.F. (2012). Gray matter atrophy in behavioral variant frontotemporal dementia: a meta-analysis of voxel-based morphometry studies. *Dementia and Geriatric Cognitive Disorders*, 33(2–3):141–8.
- Panman, J.L., Jiskoot, L.C., Bouts, M.J.R.J., Meeter, L.H.H., van der Ende, E.L., Poos, J.M., Feis, R.A., Kievit, A.J.A., van Minkelen, R., Dopper, E.G.P., Rombouts, S.A.R.B., van Swieten, J.C., and Papma, J.M. (2019). Gray and white matter changes in presymptomatic genetic frontotemporal dementia: a longitudinal MRI study. *Neurobiology of Aging*, 76:115–24.
- Papma, J.M., Jiskoot, L.C., Panman, J.L., Dopper, E.G.P., den Heijer, T., Donker Kaat, L., Pijnenburg, Y.A.L., Meeter, L.H.H., van Minkelen, R., Rombouts, S.A.R.B., and van Swieten, J.C. (2017). Cognition and gray and white matter characteristics of presymptomatic C9orf72 repeat expansion. *Neurology*, 89(12):1256–64.
- Pasquier, F., Richard, F., and Lebert, F. (2004). Natural History of Frontotemporal Dementia: Comparison with Alzheimer's Disease. *Dementia and Geriatric Cognitive Disorders*, 17:253–7.
- Patenaude, B., Smith, S.M., Kennedy, D.N., and Jenkinson, M. (2011). A Bayesian model of shape and appearance for subcortical brain segmentation. *NeuroImage*, 56(3):907–22.
- Patriat, R., Molloy, E.K., Meier, T.B., Kirk, G.R., Nair, V.A., Meyerand, M.E., Prabhakaran, V., and Birn, R.M. (2013). The effect of resting condition on resting-state fMRI reliability and consistency: A comparison between resting with eyes open, closed, and fixated. *NeuroImage*, 78:463–73.
- Persson, J., Lind, J., Larsson, A., Ingvar, M., Cruts, M., Van Broeckhoven, C., Adolfsson, R., Nilsson, L.-G., and Nyberg, L. (2006). Altered brain white matter integrity in healthy carriers of the APOE epsilon4 allele: a risk for AD? *Neurology*, 66(7):1029–33.
- Pick, A. (1892). Über die Beziehungen der senilen Hirnatrophie zur Aphasie. *Prager Medicinische Wochenschrift*, 17:165–7.
- Pievani, M., Paternicò, D., Benussi, L., Binetti, G., Orlandini, A., Cobelli, M., Magnaldi, S., Ghidoni, R., and Frisoni, G.B. (2014). Pattern of structural and functional brain abnormalities in asymptomatic granulin mutation carriers. *Alzheimer's & Dementia*, 10(5 Suppl):S354–63.
- Pijnenburg, Y.A.L., Gillissen, F., Jonker, C., and Scheltens, P. (2004). Initial Complaints in Frontotemporal Lobar Degeneration. *Dementia and Geriatric Cognitive Disorders*, 17(4):302–6.

- Pinheiro, J., Bates, D., DebRoy S, Sarkar D, and R Core Team (2018). nlme: Linear and Nonlinear Mixed Effects Models [R package nlme version 3.1-137].
- Plassman, B.L., Langa, K.M., Fisher, G.G., Heeringa, S.G., Weir, D.R., Ofstedal, M.B., Burke, J.R., Hurd, M.D., Potter, G.G., Rodgers, W.L., Steffens, D.C., Willis, R.J., and Wallace, R.B. (2007). Prevalence of dementia in the United States: The aging, demographics, and memory study. *Neuroepidemiology*, 29(1–2):125–32.
- Premi, E., Cauda, F., Gasparotti, R., Diano, M., Archetti, S., Padovani, A., and Borroni, B. (2014). Multimodal fMRI Resting-State Functional Connectivity in Granulin Mutations: The Case of Fronto-Parietal Dementia. *PLOS One*, 9(9):e106500.
- Pressman, P.S., and Miller, B.L. (2014). Diagnosis and Management of Behavioral Variant Frontotemporal Dementia. *Biological Psychiatry*, 75(7):574–81.
- Pruim, R.H.R., Mennes, M., van Rooij, D., Llera, A., Buitelaar, J.K., and Beckmann, C.F. (2015). ICA-AROMA: A robust ICA-based strategy for removing motion artifacts from fMRI data. *NeuroImage*, 112:267–77.
- Raamana, P.R., Rosen, H.J., Miller, B., Weiner, M.W., Wang, L., and Beg, M.F. (2014). Three-Class Differential Diagnosis among Alzheimer Disease, Frontotemporal Dementia, and Controls. *Frontiers in Neurology*, 5:71.
- Rabinovici, G.D., Seeley, W.W., Kim, E.J., Gorno-Tempini, M.L., Rascovsky, K., Pagliaro, T.A., Allison, S.C., Halabi, C., Kramer, J.H., Johnson, J.K., Weiner, M.W., Forman, M.S., Trojanowski, J.Q., DeArmond, S.J., Miller, B.L., and Rosen, H.J. (2008). Distinct MRI atrophy patterns in autopsy-proven Alzheimer’s disease and frontotemporal lobar degeneration. *American Journal of Alzheimer’s Disease & Other Dementias*, 22(6):474–88.
- Rabinovici, G.D., and Miller, B.L. (2010). Frontotemporal Lobar Degeneration. *CNS Drugs*, 24(5):375–98.
- Raffelt, D.A., Smith, R.E., Ridgway, G.R., Tournier, J.-D., Vaughan, D.N., Rose, S., Henderson, R., and Connelly, A. (2015). Connectivity-based fixel enhancement: Whole-brain statistical analysis of diffusion MRI measures in the presence of crossing fibres. *NeuroImage*, 117:40–55.
- Raichle, M.E. (2015). The Brain’s Default Mode Network. *Annual Review of Neuroscience*, 38(1):433–47.
- Rascovsky, K., Hodges, J.R., Knopman, D., Mendez, M.F., Kramer, J.H., Neuhaus, J., van Swieten, J.C., Seeley, H., Dopper, E.G.P., Onyike, C.U., Hillis, A.E., Josephs, K.A., Boeve, B.F., Kertesz, A., Seeley, W.W., Rankin, K.P., Johnson, J.K., Gorno-Tempini, M.L., Rosen, H.J., Prigleau-Latham, C.E., Lee, A., Kipps, C.M., Lillo, P., Piguet, O., Rohrer, J.D., Rossor, M.N., Warren, J.D., Fox, N.C., Galasko, D., Salmon, D.P., Black, S.E., Mesulam, M., Weintraub, S., Dickerson, B.C., Diehl-Schmid, J., Pasquier, F., Deramecourt, V., Lebert, F., Pijnenburg, Y.A.L., Chow, T.W., Manes, F., Grafman, J., Cappa, S.F., Freedman, M., Grossman, M., and Miller, B.L. (2011). Sensitivity of revised diagnostic criteria for the behavioural variant of frontotemporal dementia. *Brain*, 134(9):2456–77.
- Ratnavalli, E., Brayne, C., Dawson, K., and Hodges, J.R. (2002). The prevalence of frontotemporal dementia. *Neurology*, 58(11):1615–21.

- Rey, A. (1958). L'examen clinique en psychologie. *Presses Universitaires de France, Paris*.
- Riedijk, S.R., Niermeijer, M.F.N., Dooijes, D., and Tibben, A. (2009). A Decade of Genetic Counseling in Frontotemporal Dementia Affected Families: Few Counseling Requests and much Familial Opposition to Testing. *Journal of Genetic Counseling*, 18(4):350–6.
- Risacher, S., and Saykin, A. (2013). Neuroimaging Biomarkers of Neurodegenerative Diseases and Dementia. *Seminars in Neurology*, 33(4):386–416.
- Roberson, E.D., Hesse, J.H., Rose, K.D., Slama, H., Johnson, J.K., Yaffe, K., Forman, M.S., Miller, C.A., Trojanowski, J.Q., Kramer, J.H., and Miller, B.L. (2005). Frontotemporal dementia progresses to death faster than Alzheimer disease. *Neurology*, 65(5):719–25.
- Rohrer, J.D., Warren, J.D., Barnes, J., Mead, S., Beck, J., Pepple, T., Boyes, R., Omar, R., Collinge, J., Stevens, J.M., Warrington, E.K., Rossor, M.N., and Fox, N.C. (2008). Mapping the progression of progranulin-associated frontotemporal lobar degeneration. *Nature Clinical Practice Neurology*, 4(8):455–60.
- Rohrer, J.D., Guerreiro, R., Vandrovцова, J., Uphill, J., Reiman, D., Beck, J., Isaacs, A.M., Authier, A., Ferrari, R., Fox, N.C., Mackenzie, I.R.A., Warren, J.D., de Silva, R., Holton, J., Revesz, T., Hardy, J., Mead, S., and Rossor, M.N. (2009). The heritability and genetics of frontotemporal lobar degeneration. *Neurology*, 73(18):1451–6.
- Rohrer, J.D., and Rosen, H.J. (2013). Neuroimaging in frontotemporal dementia. *International Review of Psychiatry*, 25(2):221–9.
- Rohrer, J.D., Warren, J.D., Fox, N.C., and Rossor, M.N. (2013). Presymptomatic studies in genetic frontotemporal dementia. *Revue Neurologique*, 169(10):820–4.
- Rohrer, J.D., Nicholas, J.M., Cash, D.M., van Swieten, J.C., Dopper, E.G.P., Jiskoot, L.C., van Minkelen, R., Rombouts, S.A.R.B., Cardoso, M.J., Clegg, S., Espak, M., Mead, S., Thomas, D.L., De Vita, E., Masellis, M., Black, S.E., Freedman, M., Keren, R., MacIntosh, B.J., Rogava, E., Tang-Wai, D., Tartaglia, M.C., Laforce, R., Tagliavini, F., Tiraboschi, P., Redaelli, V., Prioni, S., Grisoli, M., Borroni, B., Padovani, A., Galimberti, D., Scarpini, E., Arighi, A., Fumagalli, G., Rowe, J.B., Coyle-Gilchrist, I., Graff, C., Fallström, M., Jelic, V., Ståhlbom, A.K., Andersson, C., Thonberg, H., Lilius, L., Frisoni, G.B., Binetti, G., Pievani, M., Bocchetta, M., Benussi, L., Ghidoni, R., Finger, E., Sorbi, S., Nacmias, B., Lombardi, G., Polito, C., Warren, J.D., Ourselin, S., Fox, N.C., and Rossor, M.N. (2015). Presymptomatic cognitive and neuroanatomical changes in genetic frontotemporal dementia in the Genetic Frontotemporal dementia Initiative (GENFI) study: a cross-sectional analysis. *The Lancet Neurology*, 14(3):253–62.
- Royall, D.R., Cordes, J.A., and Polk, M. (1998). CLOX: an executive clock drawing task. *Journal of Neurology, Neurosurgery, and Psychiatry*, 64(5):588–94.
- Rytty, R., Nikkinen, J., Paavola, L., Abou Elseoud, A., Moilanen, V., Visuri, A., Tervonen, O., Renton, A.E., Traynor, B.J., Kiviniemi, V., and Remes, A.M. (2013). GroupICA dual regression analysis of resting state networks in a behavioral variant of frontotemporal dementia. *Frontiers in Human Neuroscience*, 7:461.
- Salimi-Khorshidi, G., Douaud, G., Beckmann, C.F., Glasser, M.F., Griffanti, L., and Smith, S.M.

- (2014). Automatic denoising of functional MRI data: Combining independent component analysis and hierarchical fusion of classifiers. *NeuroImage*, 90:449–68.
- Schouten, T.M., Koini, M., de Vos, F., Seiler, S., van der Grond, J., Lechner, A., Hafkemeijer, A., Möller, C., Schmidt, R., de Rooij, M., and Rombouts, S.A.R.B. (2016). Combining anatomical, diffusion, and resting state functional magnetic resonance imaging for individual classification of mild and moderate Alzheimer’s disease. *NeuroImage: Clinical*, 11:46–51.
- Schouten, T.M., Koini, M., de Vos, F., Seiler, S., de Rooij, M., Lechner, A., Schmidt, R., van den Heuvel, M., van der Grond, J., and Rombouts, S.A.R.B. (2017). Individual classification of Alzheimer’s disease with diffusion magnetic resonance imaging. *NeuroImage*, 152:476–81.
- Seelaar, H., Kamphorst, W., Rosso, S.M., Azmani, A., Masdjedi, R., de Koning, I., Maat-Kievit, J.A., Anar, B., Donker Kaat, L., Breedveld, G.J., Dooijes, D., Rozemuller, J.M., Bronner, I.F., Rizzu, P., and van Swieten, J.C. (2008). Distinct genetic forms of frontotemporal dementia. *Neurology*, 71(16):1220–6.
- Seelaar, H., Rohrer, J.D., Pijnenburg, Y.A.L., Fox, N.C., and van Swieten, J.C. (2011). Clinical, genetic and pathological heterogeneity of frontotemporal dementia: a review. *Journal of Neurology, Neurosurgery, and Psychiatry*, 82(5):476–86.
- Seeley, W.W., Allman, J.M., Carlin, D.A., Crawford, R.K., Macedo, M.N., Greicius, M.D., DeArmond, S.J., and Miller, B.L. (2007). Divergent social functioning in behavioral variant frontotemporal dementia and Alzheimer disease: reciprocal networks and neuronal evolution. *Alzheimer Disease & Associated Disorders*, 21(4):S50-7.
- Seeley, W.W., Crawford, R.K., Zhou, J., Miller, B.L., and Greicius, M.D. (2009). Neurodegenerative diseases target large-scale human brain networks. *Neuron*, 62(1):42–52.
- Shmueli, G. (2010). To Explain or to Predict? *Statistical Science*, 25(3):289–310.
- Smith, C.D., Chebrolu, H., Andersen, A.H., Powell, D.A., Lovell, M.A., Xiong, S., and Gold, B.T. (2010). White matter diffusion alterations in normal women at risk of Alzheimer’s disease. *Neurobiology of Aging*, 31(7):1122–31.
- Smith, S.M. (2002). Fast robust automated brain extraction. *Human Brain Mapping*, 17(3):143–55.
- Smith, S.M., Jenkinson, M., Woolrich, M.W., Beckmann, C.F., Behrens, T.E.J., Johansen-Berg, H., Bannister, P.R., De Luca, M., Drobnjak, I., Flitney, D.E., Niazy, R.K., Saunders, J., Vickers, J., Zhang, Y., De Stefano, N., Brady, J.M., and Matthews, P.M. (2004). Advances in functional and structural MR image analysis and implementation as FSL. *NeuroImage*, 23 Suppl 1:S208-19.
- Smith, S.M., Jenkinson, M., Johansen-Berg, H., Rueckert, D., Nichols, T.E., Mackay, C.E., Watkins, K.E., Ciccarelli, O., Cader, M.Z., Matthews, P.M., and Behrens, T.E.J. (2006). Tract-based spatial statistics: Voxelwise analysis of multi-subject diffusion data. *NeuroImage*, 31(4):1487–505.
- Smith, S.M., Fox, P.T., Miller, K.L., Glahn, D.C., Fox, P.M., Mackay, C.E., Filippini, N., Watkins, K.E., Toro, R., Laird, A.R., and Beckmann, C.F. (2009). Correspondence of the brain’s functional architecture during activation and rest. *Proceedings of the National Academy of*

- Sciences of the USA*, 106(31):13040–5.
- Smith, S.M., and Nichols, T.E. (2009). Threshold-free cluster enhancement: addressing problems of smoothing, threshold dependence and localisation in cluster inference. *NeuroImage*, 44(1):83–98.
- Smith, S.M., Beckmann, C.F., Andersson, J., Auerbach, E.J., Bijsterbosch, J., Douaud, G., Duff, E., Feinberg, D.A., Griffanti, L., Harms, M.P., Kelly, M., Laumann, T., Miller, K.L., Moeller, S., Petersen, S., Power, J., Salimi-Khorshidi, G., Snyder, A.Z., Vu, A.T., Woolrich, M.W., Xu, J., Yacoub, E., Ugurbil, K., Van Essen, D.C., and Glasser, M.F. (2013). Resting-state fMRI in the Human Connectome Project. *NeuroImage*, 80:144–68.
- Sotiropoulos, S.N., Jbabdi, S., Xu, J., Andersson, J.L., Moeller, S., Auerbach, E.J., Glasser, M.F., Hernandez, M., Sapiro, G., Jenkinson, M., Feinberg, D.A., Yacoub, E., Lenglet, C., Van Essen, D.C., Ugurbil, K., and Behrens, T.E.J. (2013). Advances in diffusion MRI acquisition and processing in the Human Connectome Project. *NeuroImage*, 80:125–43.
- Spinelli, E.G., Mandelli, M.L., Miller, Z.A., Santos-Santos, M.A., Wilson, S.M., Agosta, F., Grinberg, L.T., Huang, E.J., Trojanowski, J.Q., Meyer, M., Henry, M.L., Comi, G., Rabinovici, G., Rosen, H.J., Filippi, M., Miller, B.L., Seeley, W.W., and Gorno-Tempini, M.L. (2017). Typical and atypical pathology in primary progressive aphasia variants. *Annals of Neurology*, 81(3):430–43.
- Staffaroni, A.M., Bajorek, L., Casaletto, K.B., Cobigo, Y., Goh, S.-Y.M., Wolf, A., Heuer, H.W., Elahi, F.M., Ljubenkov, P.A., Dever, R., Kornak, J., Appleby, B., Bove, J., Bordelon, Y., Brannelly, P., Brushaber, D., Caso, C., Coppola, G., Dheel, C., Dickerson, B.C., Dickinson, S., Dominguez, S., Domoto-Reilly, K., Faber, K., Ferrall, J., Fields, J.A., Fishman, A., Fong, J., Foroud, T., Forsberg, L.K., Gavrilova, R., Gearhart, D., Ghazanfari, B., Ghoshal, N., Goldman, J., Graff-Radford, J., Graff-Radford, N., Grant, I., Grossman, M., Haley, D., Hsiung, G.-Y., Huey, E.D., Irwin, D.J., Jones, D.T., Jones, L., Kantarci, K., Karydas, A., Kaufer, D.I., Kerwin, D.R., Knopman, D.S., Kraft, R., Kremers, W.K., Kukull, W.A., Litvan, I., Lucente, D., Lungu, C., Mackenzie, I.R., Maldonado, M., Manoochehri, M., McGinnis, S.M., McKinley, E., Mendez, M.F., Miller, B.L., Multani, N., Onyike, C.U., Padmanabhan, J., Pantelyat, A., Pearlman, R., Petrucelli, L., Potter, M., Rademakers, R., Ramos, E.M., Rankin, K.P., Rascovsky, K., Roberson, E.D., Rogalski, E., Sengdy, P., Shaw, L.M., Syrjanen, J., Tartaglia, M.C., Tatton, N., Taylor, J., Toga, A., Trojanowski, J.Q., Weintraub, S., Wang, P., Wong, B., Wszolek, Z., Boxer, A.L., Boeve, B.F., Kramer, J.H., and Rosen, H.J. (2019). Assessment of executive function declines in presymptomatic and mildly symptomatic familial frontotemporal dementia: NIH-EXAMINER as a potential clinical trial endpoint. *Alzheimer's & Dementia*, 16(1):11–21.
- Stroop, J.R. (1935). Studies of interference in serial verbal reactions. *Journal of Experimental Psychology*, 18:643–62.
- Sullivan, L.M., Dukes, K.A., and Losina, E. (1999). An introduction to hierarchical linear modelling. *Statistics in Medicine*, 18(7):855–88.
- Suri, S., Topiwala, A., Mackay, C.E., Ebmeier, K.P., and Filippini, N. (2014). Using Structural and Diffusion Magnetic Resonance Imaging To Differentiate the Dementias. *Current Neurology and Neuroscience Reports*, 14(9):475.

- ten Kate, M., Sanz-Arigita, E.J., Tijms, B.M., Wink, A.M., Clerigue, M., Garcia-Sebastian, M., Izagirre, A., Ecay-Torres, M., Estanga, A., Villanua, J., Vrenken, H., Visser, P.J., Martinez-Lage, P., and Barkhof, F. (2016). Impact of APOE- ϵ 4 and family history of dementia on gray matter atrophy in cognitively healthy middle-aged adults. *Neurobiology of Aging*, 38:14–20.
- The ADHD-200 Consortium (2012). The ADHD-200 Consortium: A Model to Advance the Translational Potential of Neuroimaging in Clinical Neuroscience. *Frontiers in Systems Neuroscience*, 6:62.
- Thurstone, L.L.T., and Thurstone, T.G. (1962). Primary mental abilities. *Science Research Associates, Chicago*.
- Tian, L., Kong, Y., Ren, J., Varoquaux, G., Zang, Y.-F., and Smith, S.M. (2013). Spatial vs. Temporal Features in ICA of Resting-State fMRI – A Quantitative and Qualitative Investigation in the Context of Response Inhibition. *PLOS One*, 8(6):e66572.
- Tibshirani, R. (1996). Regression Shrinkage and Selection via the Lasso. *Journal of the Royal Statistical Society*, 58(1):267–88.
- Tournier, J.-D., Calamante, F., and Connelly, A. (2007). Robust determination of the fibre orientation distribution in diffusion MRI: Non-negativity constrained super-resolved spherical deconvolution. *NeuroImage*, 35(4):1459–72.
- Trachtenberg, A.J., Filippini, N., Cheeseman, J., Duff, E.P., Neville, M.J., Ebmeier, K.P., Karpe, F., and Mackay, C.E. (2012a). The effects of APOE on brain activity do not simply reflect the risk of Alzheimer’s disease. *Neurobiology of Aging*, 33(3):618.e1–13.
- Trachtenberg, A.J., Filippini, N., Ebmeier, K.P., Smith, S.M., Karpe, F., and Mackay, C.E. (2012b). The effects of APOE on the functional architecture of the resting brain. *NeuroImage*, 59(1):565–72.
- Tsai, R.M., and Boxer, A.L. (2016). Therapy and clinical trials in frontotemporal dementia: past, present, and future. *Journal of Neurochemistry*, 138:211–21.
- Tu, S., Leyton, C.E., Hodges, J.R., Piguet, O., and Hornberger, M. (2015). Divergent Longitudinal Propagation of White Matter Degradation in Logopenic and Semantic Variants of Primary Progressive Aphasia. *Journal of Alzheimer’s Disease*, 49(3):853–61.
- Tuovinen, T., Rytty, R., Moilanen, V., Abou Elseoud, A., Veijola, J., Remes, A.M., and Kiviniemi, V.J. (2017). The Effect of Gray Matter ICA and Coefficient of Variation Mapping of BOLD Data on the Detection of Functional Connectivity Changes in Alzheimer’s Disease and bvFTD. *Frontiers in Human Neuroscience*, 10:680.
- Tustison, N.J., Avants, B.B., Cook, P.A., Yuanjie Zheng, Egan, A., Yushkevich, P.A., and Gee, J.C. (2010). N4ITK: Improved N3 Bias Correction. *IEEE Transactions on Medical Imaging*, 29(6):1310–20.
- van Swieten, J.C., Rosso, S.M., and Heutink, P. (2000). ‘MAPT-Related Disorders’, in: Adam, M.P., Ardinger, H.H., Pagon, R.A., Wallace, S.E., Bean, L.J.H., Stephens, K., and Amemiya, A. (Eds.), GeneReviews®.
- van Swieten, J.C., and Heutink, P. (2008). Mutations in progranulin (GRN) within the spectrum

- of clinical and pathological phenotypes of frontotemporal dementia. *The Lancet Neurology*, 7(10):965–74.
- Varma, S., and Simon, R. (2006). Bias in error estimation when using cross-validation for model selection. *BMC Bioinformatics*, 7:91.
- Varoquaux, G. (2018). Cross-validation failure: Small sample sizes lead to large error bars. *NeuroImage*, 180:68–77.
- Veer, I.M., Beckmann, C.F., van Tol, M.-J., Ferrarini, L., Milles, J., Veltman, D.J., Aleman, A., van Buchem, M.A., van der Wee, N.J., and Rombouts, S.A.R.B. (2010). Whole brain resting-state analysis reveals decreased functional connectivity in major depression. *Frontiers in Systems Neuroscience*, 4:41.
- Vieira, R.T., Caixeta, L., Machado, S., Silva, A.C., Nardi, A.E., Arias-Carrión, O., and Carta, M.G. (2013). Epidemiology of early-onset dementia: a review of the literature. *Clinical Practice & Epidemiology in Mental Health*, 9:88–95.
- Visch-Brink, E., Stronks, D., and Denes, G. (2005). SAT: Semantische Associatie Test. *Swets & Zeitlinger, Lisse*.
- Wang, J., Redmond, S.J., Bertoux, M., Hodges, J.R., and Hornberger, M. (2016). A Comparison of Magnetic Resonance Imaging and Neuropsychological Examination in the Diagnostic Distinction of Alzheimer’s Disease and Behavioral Variant Frontotemporal Dementia. *Frontiers in Aging Neuroscience*, 8:119.
- Wechsler, D. (2005). WAIS-III Nederlandse bewerking. Technische Handleiding. *Harcourt Test Publishers, Lisse*.
- Wegner, C., Filippi, M., Korteweg, T., Beckmann, C.F., Ciccarelli, O., De Stefano, N., Enzinger, C., Fazekas, F., Agosta, F., Gass, A., Hirsch, J., Johansen-Berg, H., Kappos, L., Barkhof, F., Polman, C., Mancini, L., Manfredonia, F., Marino, S., Miller, D.H., Montalban, X., Palace, J., Rocca, M., Ropele, S., Rovira, A., Smith, S.M., Thompson, A., Thornton, J., Yousry, T., and Matthews, P.M. (2008). Relating functional changes during hand movement to clinical parameters in patients with multiple sclerosis in a multi-centre fMRI study. *European Journal of Neurology*, 15(2):113–22.
- Wheeler-Kingshott, C.A.M., and Cercignani, M. (2009). About “axial” and “radial” diffusivities. *Magnetic Resonance in Medicine*, 61(5):1255–60.
- Whitwell, J.L., and Jack, C.R. (2005). Comparisons Between Alzheimer Disease, Frontotemporal Lobar Degeneration, and Normal Aging With Brain Mapping. *Topics in Magnetic Resonance Imaging*, 16(6):409–25.
- Whitwell, J.L., Avula, R., Senjem, M.L., Kantarci, K., Weigand, S.D., Samikoglu, A., Edmonson, H.A., Vemuri, P., Knopman, D.S., Boeve, B.F., Petersen, R.C., Josephs, K.A., and Jack, C.R. (2010). Gray and white matter water diffusion in the syndromic variants of frontotemporal dementia. *Neurology*, 74(16):1279–87.
- Whitwell, J.L., Josephs, K.A., Avula, R., Tosakulwong, N., Weigand, S.D., Senjem, M.L., Vemuri, P., Jones, D.T., Gunter, J.L., Baker, M., Wszolek, Z.K., Knopman, D.S., Rademakers, R., Petersen, R.C., Boeve, B.F., and Jack, C.R. (2011a). Altered functional connectivity in asymptomatic

- MAPT subjects: a comparison to bvFTD. *Neurology*, 77(9):866–74.
- Whitwell, J.L., Weigand, S.D., Gunter, J.L., Boeve, B.F., Rademakers, R., Baker, M., Knopman, D.S., Wszolek, Z.K., Petersen, R.C., Jack, C.R., and Josephs, K.A. (2011b). Trajectories of brain and hippocampal atrophy in FTD with mutations in MAPT or GRN. *Neurology*, 77(4):393–8.
- Whitwell, J.L., Weigand, S.D., Boeve, B.F., Senjem, M.L., Gunter, J.L., DeJesus-Hernandez, M., Rutherford, N.J., Baker, M., Knopman, D.S., Wszolek, Z.K., Parisi, J.E., Dickson, D.W., Petersen, R.C., Rademakers, R., Jack, C.R., and Josephs, K.A. (2012). Neuroimaging signatures of frontotemporal dementia genetics: C9orf72, tau, progranulin and sporadics. *Brain*, 135(3):794–806.
- Whitwell, J.L., Boeve, B.F., Weigand, S.D., Senjem, M.L., Gunter, J.L., Baker, M.C., DeJesus-Hernandez, M., Knopman, D.S., Wszolek, Z.K., Petersen, R.C., Rademakers, R., Jack, C.R., and Josephs, K.A. (2015). Brain atrophy over time in genetic and sporadic frontotemporal dementia: A study of 198 serial magnetic resonance images. *European Journal of Neurology*, 22(5):745–52.
- Winkler, A.M., Ridgway, G.R., Webster, M.A., Smith, S.M., and Nichols, T.E. (2014). Permutation inference for the general linear model. *NeuroImage*, 92:381–97.
- Woolrich, M.W., Jbabdi, S., Patenaude, B., Chappell, M., Makni, S., Behrens, T., Beckmann, C.F., Jenkinson, M., and Smith, S.M. (2009). Bayesian analysis of neuroimaging data in FSL. *NeuroImage*, 45(1 Suppl 1):S173–86.
- Yan, C.-G., Liu, D.-Q., He, Y., Zou, Q.-H., Zhu, C.-Z., Zuo, X.-N., Long, X.-Y., and Zang, Y.-F. (2009). Spontaneous brain activity in the default mode network is sensitive to different resting-state conditions with limited cognitive load. *PLOS One*, 4(5):e5743.
- Yang, H., Long, X.-Y., Yang, Y., Yan, H., Zhu, C.-Z., Zhou, X.-P., Zang, Y.-F., and Gong, Q.-Y. (2007). Amplitude of low frequency fluctuation within visual areas revealed by resting-state functional MRI. *NeuroImage*, 36(1):144–52.
- Yu, M., Linn, K.A., Cook, P.A., Phillips, M.L., McInnis, M., Fava, M., Trivedi, M.H., Weissman, M.M., Shinohara, R.T., and Sheline, Y.I. (2018). Statistical harmonization corrects site effects in functional connectivity measurements from multi-site fMRI data. *Human Brain Mapping*, 39(11):4213–27.
- Yuan, B.-K., Wang, J., Zang, Y.-F., and Liu, D.-Q. (2014). Amplitude differences in high-frequency fMRI signals between eyes open and eyes closed resting states. *Frontiers in Human Neuroscience*, 8:503.
- Zhang, H., Schneider, T., Wheeler-Kingshott, C.A., and Alexander, D.C. (2012). NODDI: Practical in vivo neurite orientation dispersion and density imaging of the human brain. *NeuroImage*, 61(4):1000–16.
- Zhang, Y., Schuff, N., Du, A.-T., Rosen, H.J., Kramer, J.H., Gorno-Tempini, M.L., Miller, B.L., and Weiner, M.W. (2009). White matter damage in frontotemporal dementia and Alzheimer's disease measured by diffusion MRI. *Brain*, 132(9):2579–92.
- Zhang, Y., Schuff, N., Ching, C., Tosun, D., Zhan, W., Nezamzadeh, M., Rosen, H.J., Kramer, J.H.,

- Gorno-Tempini, M.L., Miller, B.L., and Weiner, M.W. (2011). Joint assessment of structural, perfusion, and diffusion MRI in Alzheimer's disease and frontotemporal dementia. *International Journal of Alzheimer's Disease*, 546871.
- Zhang, Y., Tartaglia, M.C., Schuff, N., Chiang, G.C., Ching, C., Rosen, H.J., Gorno-Tempini, M.L., Miller, B.L., and Weiner, M.W. (2013). MRI signatures of brain macrostructural atrophy and microstructural degradation in frontotemporal lobar degeneration subtypes. *Journal of Alzheimer's Disease*, 33(2):431–44.
- Zhou, J., Greicius, M.D., Gennatas, E.D., Growdon, M.E., Jang, J.Y., Rabinovici, G.D., Kramer, J.H., Weiner, M., Miller, B.L., and Seeley, W.W. (2010). Divergent network connectivity changes in behavioural variant frontotemporal dementia and Alzheimer's disease. *Brain*, 133(5):1352–67.
- Zhou, J., and Seeley, W.W. (2014). Network Dysfunction in Alzheimer's Disease and Frontotemporal Dementia: Implications for Psychiatry. *Biological Psychiatry*, 75(7):565–73.
- Zivadinov, R., and Cox, J.L. (2008). Is functional MRI feasible for multi-center studies on multiple sclerosis? *European Journal of Neurology*, 15(2):109–10.
- Zou, H., and Hastie, T. (2005). Regularization and variable selection via the elastic net. *Journal of the Royal Statistical Society: Series B (Statistical Methodology)*, 67(2):301–20.
- Zou, K.H., Greve, D.N., Wang, M., Pieper, S.D., Warfield, S.K., White, N.S., Manandhar, S., Brown, G.G., Vangel, M.G., Kikinis, R., and Wells, W.M. (2005). Reproducibility of functional MR imaging: preliminary results of prospective multi-institutional study performed by Biomedical Informatics Research Network. *Radiology*, 237(3):781–9.