

Supplementary material

Article title: Cognitive profiles discriminate between genetic variants of behavioral frontotemporal dementia

Journal name: Journal of Neurology

Author names and affiliation:

Poos, J.M. MSc¹², Jiskoot, L.C. PhD¹³, Leijdesdorff, S.M.J. MSc.⁴, Seelaar, H. MD PhD¹, Panman, J.L. MSc. ¹², van der Ende, E.L MD¹, Mol, M.O. MD¹, Meeter, L.H.H. MD PhD¹, Pijnenburg, Y.A.L. MD PhD⁵, Donker Kaat, L. MD PhD¹⁶, de Jong, F.J. MD PhD¹, van Swieten, J.C. MD PhD¹, Papma, J.M. PhD¹, van den Berg, E. PhD¹

¹Alzheimer Center and Department of Neurology, Erasmus MC University Medical Center, Rotterdam, the Netherlands

²Department of Radiology, Leiden University Medical Center, Leiden, the Netherlands

³Dementia Research Center, University College London, London, UK

⁴Department of Psychiatry and Psychology, Maastricht University, Maastricht, the Netherlands

⁵Alzheimer center, Department of Neurology, Amsterdam University Medical Center, location VU University Medical Center, Amsterdam Neuroscience, Amsterdam, the Netherlands

⁶ Department of Clinical Genetics, Erasmus MC University Medical Center, Rotterdam, The Netherlands

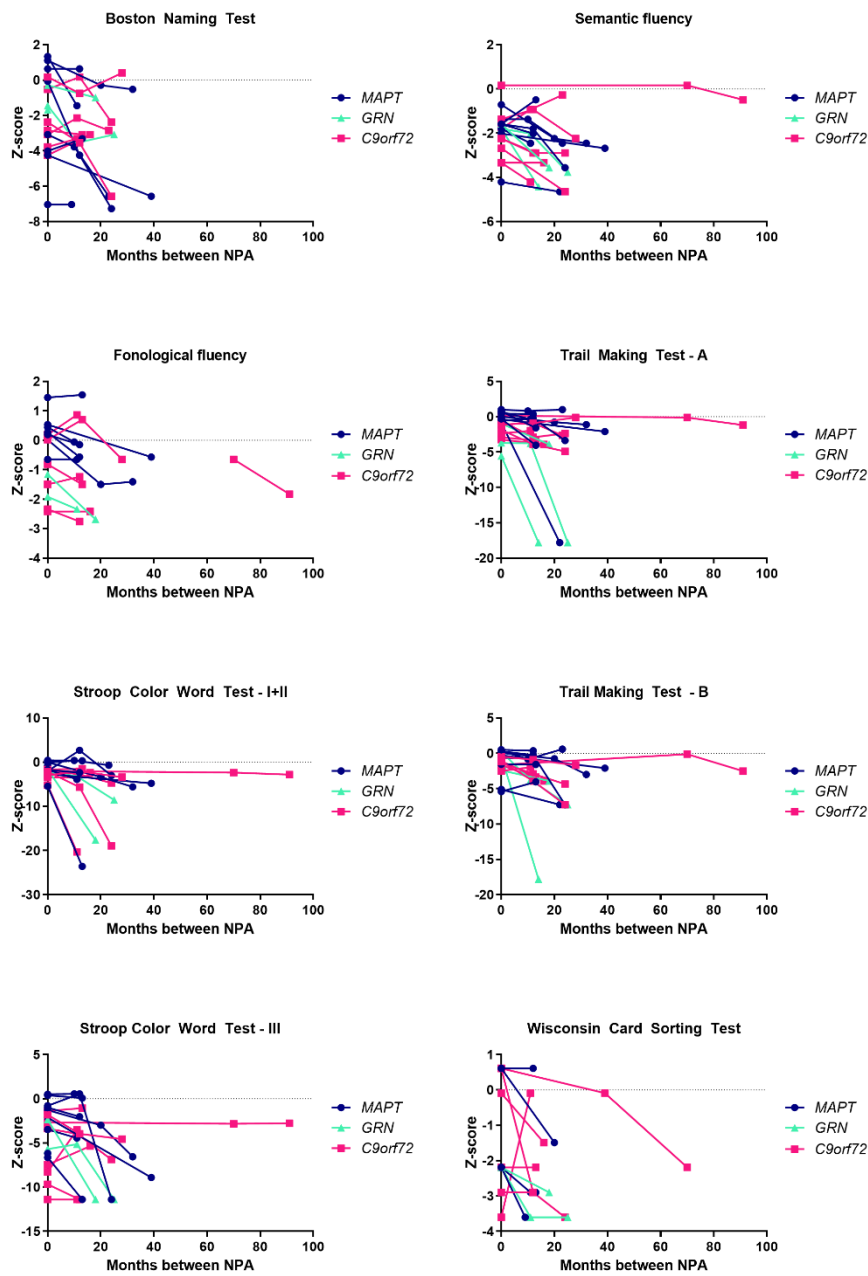
E-mail address corresponding author: j.m.poos@erasmusms.nl

Table 1. Differences between *GRN* patients with predominant right-sided, left-sided or generalized atrophy on seven cognitive domains.

Domain	Left	<i>n</i>	Right	<i>n</i>	Generalized	<i>n</i>	<i>p</i> Value
Language	-3.3 ± 1.2	10	-1.0 ± 0.7	4	-2.1 ± 0.9	4	0.02
Attention and mental processing speed	-6.6 ± 4.2	9	-0.7 ± 0.7	4	-3.6 ± 3.8	3	0.2
Executive functioning	-6.2 ± 2.0	9	-2.2 ± 1.5	4	-5.1 ± 2.2	4	0.09
Memory - learning	-5.6 ± 6.5	9	-3.5 ± 5.2	4	-7.0 ± 9.3	3	0.8
Memory - recall	-1.1 ± 1.6	7	-0.3 ± 2.2	4	-1.0 ± 1.7	3	0.8
Working memory	-2.1 ± 1.0	4	-0.1 ± 2.7	4	-	0	0.5
Visuoconstruction	-1.2 ± 1.7	9	-0.3 ± 0.8	4	-0.9 ± 2.0	3	0.4

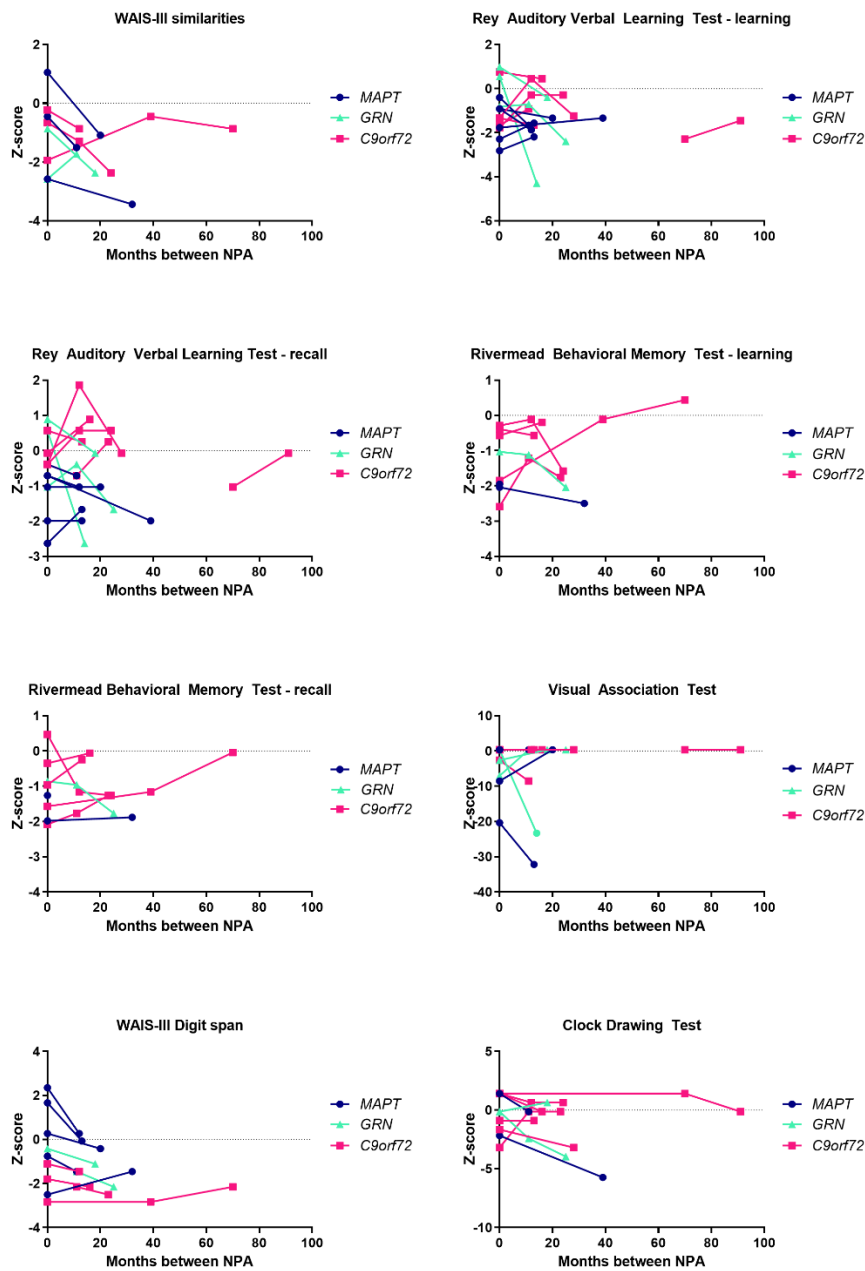
Values indicate mean ± SD. The *p* values constitute interaction terms of univariate analyses of covariance (corrected for age and education).

Fig. 1 Within-individual trajectories on neuropsychological tests



Abbreviations: NPA = neuropsychological assessment; *MAPT* = microtubule-associated protein tau; *GRN* = progranulin; *C9orf72* = chromosome 9 open reading frame 72. Raw data for each neuropsychological test were first converted to z-scores by standardization to the baseline data of healthy controls. Composite cognitive domain scores were calculated. Each subplot presents the trajectory on a specific cognitive domain. Data are available in: Boston Naming Test (*MAPT* n=8; *GRN* n=2; *C9orf72* n=7); semantic fluency (*MAPT* n=8; *GRN* n=3; *C9orf72* n=8); letter fluency (*MAPT* n=7; *GRN* n=2; *C9orf72* n=6); Trail Making Test - A (*MAPT* n=9; *GRN* n=3; *C9orf72* n=8); Stroop Color Word Test I+II (*MAPT* n=8; *GRN* n=2; *C9orf72* n=8); Trail Making Test - B (*MAPT* n=8; *GRN* n=3; *C9orf72* n=9); Stroop Color Word Test III (*MAPT* n=8; *GRN* n=2; *C9orf72* n=8); Wisconsin Card Sorting Test (*MAPT* n=4; *GRN* n=2; *C9orf72* n=6).

Fig. 2 Within-individual trajectories on neuropsychological tests.



Abbreviations: NPA = neuropsychological assessment; *MAPT* = microtubule-associated protein tau; *GRN* = progranulin; *C9orf72* = chromosome 9 open reading frame 72. Raw data for each neuropsychological test were first converted to z-scores by standardization to the baseline data of healthy controls. Composite cognitive domain scores were calculated. Each subplot presents the trajectory on a specific cognitive domain. Data are available in: WAIS-III Similarities (*MAPT* n= 3; *GRN* n=2; *C9orf72* n= 3); Rey Auditory Verbal Learning Test – learning (*MAPT* n= 6; *GRN* n=3; *C9orf72* n=6); Rey Auditory Verbal Learning – recall (*MAPT* n=6; *GRN* n=3; *C9orf72* n=6); Rivermead Behavioral Memory Test - learning (*MAPT* n=1; *GRN* n=1; *C9orf72* n=6); Rivermead Behavioral Memory Test - recall (*MAPT* n=5; *GRN* n=1; *C9orf72* n= 6); Visual Association Test (*MAPT* n= 3; *GRN* n=3; *C9orf72* n=3); WAIS-III Digit Span (*MAPT* n= 5; *GRN* n=2; *C9orf72* n=4); Clock Drawing Test (*MAPT* n=2; *GRN* n=2; *C9orf72* n=6).