

INTRODUCTION

The right temporal variant of frontotemporal dementia (rtvFTD) does not perfectly fit neither the criteria for the behavioural variant of FTD nor for the semantic variant of primary progressive aphasia. The aim of this study was to assess functional connectivity (FC) patterns associated with this rare clinical variant in comparison to normal aging.

METHODS

Table 1. Clinical and neuropsychological features of patients and controls.

	Patients	HC	P values
Number	6	20	-
Age, years	60.6 ± 4.2	61.7 ± 4.6	0.43
Education, years	12.3 ± 5.5	12.2 ± 3.4	0.96
Sex	2 M/ 4 F	8 M/ 12 F	0.78
Disease duration, years	1.7 ± 0.5	-	-
CSF, Aβ42	755.2 ± 189.9	-	-
CSF, total Tau	269.2 ± 133.7	-	-
CSF, phosphorylated-tau	33.9 ± 12.8	-	-
Global cognition			
MMSE	26.6 ± 2.3	29.3 ± 0.9	0.01
Frontal Assessment Battery	11.6 ± 4.3	-	-
Memory			
Digit Span, forward	4.6 ± 1.0	6.7 ± 1.6	0.01
RAVLT, delayed Recall	3.5 ± 2.8	9.8 ± 1.9	<0.001
Benson Figure, Recall	3.6 ± 4.6	11 ± 2.7	0.01
Executive functions			
MCST, perseverative errors	10.8 ± 10.1	3.1 ± 4.0	0.01
Trial Making Test (B-A)	114.4 ± 49.3	58.7 ± 19.9	0.02
Phonemic fluency	17.4 ± 10.6	41.9 ± 11.5	0.003
Category fluency	18.6 ± 8.6	48.5 ± 9.3	<0.001
Language ('left side')			
CaGi, Naming	39 ± 5	-	-
CaGi, Comprehension	45 ± 4.1	-	-
Pyramids and Palm Trees test	36.3 ± 7.4	-	-
Language ('Right side')			
BLED, Total score	30.5 ± 4.9	-	-
BLED, Figurative metaphors	1.8 ± 1.9	-	-
BLED, Written metaphors	4.8 ± 3.9	-	-
BLED, Inferences	5.9 ± 0.8	-	-
BLED, Requests	7.6 ± 3.2	-	-
BLED, Irony	3.5 ± 2.1	-	-
BLED, Prosody	6 ± 2.1	-	-
Social cognition			
SET, Global Score	8.6 ± 2.4	-	-
SET, Intention Attribution	3 ± 1.3	-	-
SET, Causal Inference	3.8 ± 1.5	-	-
SET, Emotion Attribution	1.8 ± 0.7	-	-
Visuospatial abilities			
Clock Drawing Test	5.1 ± 4.5	-	-
Benson Figure, copy	15.2 ± 0.8	-	-
Prosopagnosia			
Benton, face recognition	31.5 ± 5.8	-	-
Behavioural disturbances			
FBI, Total score	18.2 ± 8.2	-	-
FBI-A, negative symptoms	12.4 ± 6.8	-	-
FBI-B, positive symptoms	5.8 ± 2.9	-	-
Autonomy			
ADL	6 ± 0.0	-	-
IADL	6.8 ± 1.2	-	-
Disease severity			
CDR, Sum of Boxes	3.4 ± 2.7	-	-
CDR, FTD	6.1 ± 3.4	-	-

Values denote mean ± standard deviations (or frequencies). Cognitive tests are reported as raw values. P values refer to t-test models. *Abbreviations:* ADL= activities of daily living; BLED= 'Batteria per il Linguaggio dell'Emisfero Destro'; CDR= Clinical Dementia Rating scale; CSF= cerebrospinal fluid; FBI= Frontal Behavioural Inventory; FTD= frontotemporal dementia; HC= healthy controls; IADL= instrumental activities of daily living; MCST=Modified Card Sorting Test; MMSE=Mini Mental State Examination; RAVLT= Rey Auditory Verbal Learning Test; SET= Story-based Empathy Task.

Images acquisition (3.0 T Ingenia CX, Philips) and processing:

All subjects underwent an MRI scan with 3D T1-weighted and resting-state (RS) fMRI sequences.

RS-fMRI networks were identified using an independent component analysis (GIFT toolbox, SPM12).

For each network of interest, comparisons between groups were performed using T-test models in SPM12.

RESULTS

Figure 1. Spatial maps of the RS networks are overlaid on the 3D Montreal Neurological Institute template in neurological convention (right is right): a) Default Mode; b) Salience; c) Sensorimotor; d) Anterior Temporal; e) Right Fronto-Parietal; f) Left Fronto-Parietal; g) Primary Visual; h) Associative Visual I; i) Associative Visual II; l) Cerebellar; m) Frontal. Coloured bars denote Z-values.

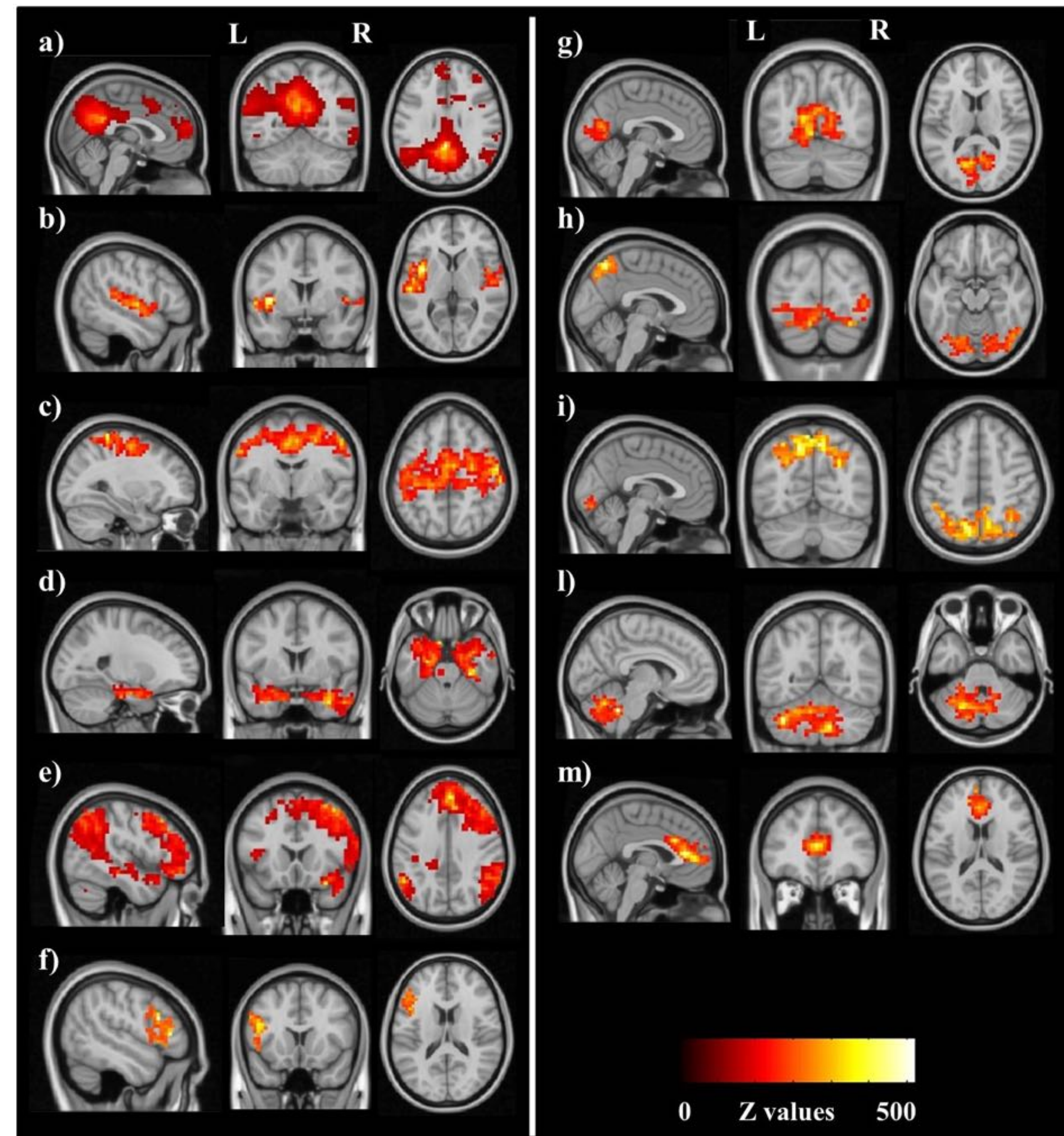
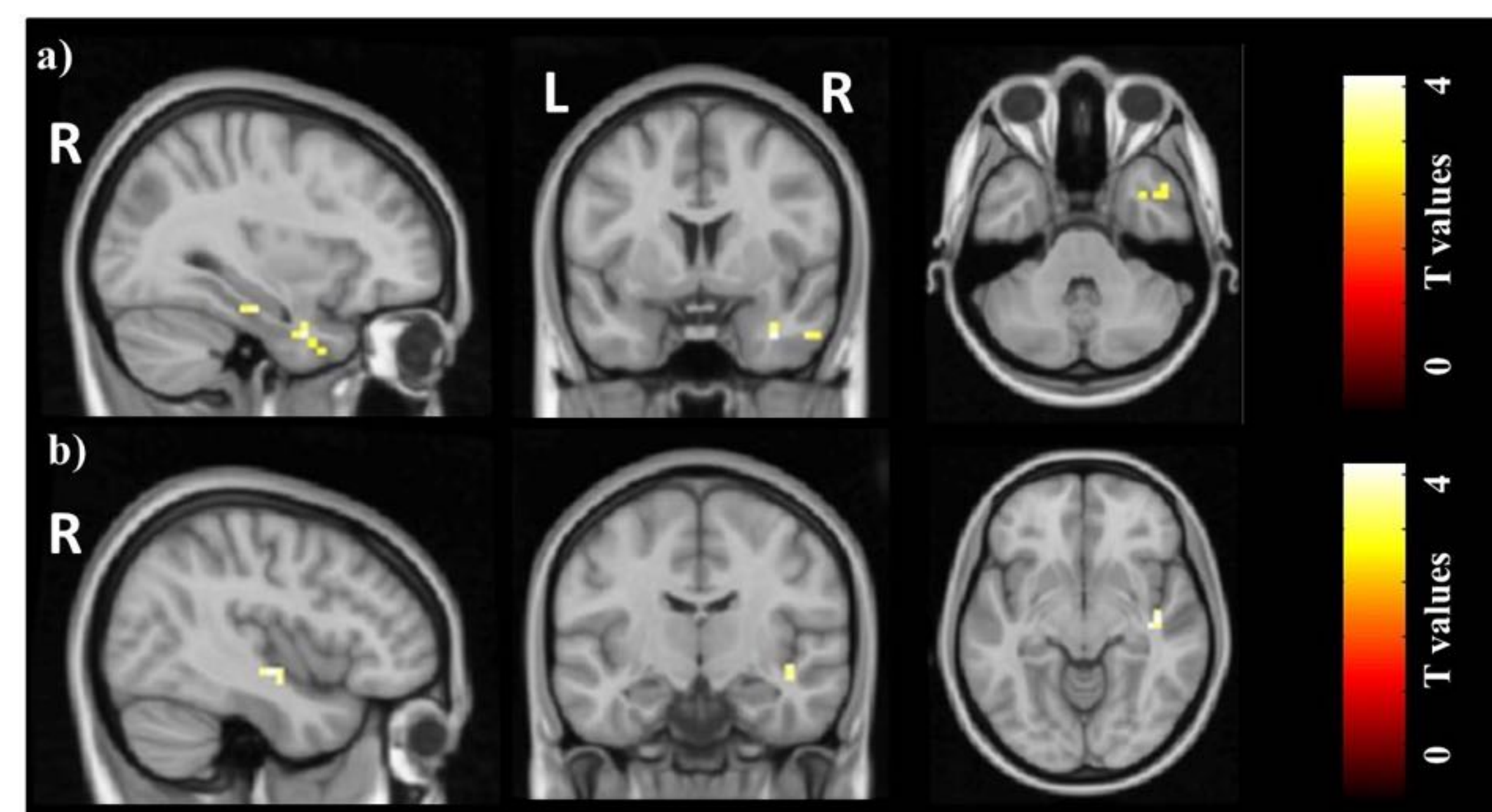


Figure 2. Significant increased FC in patients vs controls in the anterior temporal network (a) and in the right fronto-parietal network (b). Findings are overlaid on the 3D Montreal Neurological Institute template in neurological convention (right is right) and in warm colours. Coloured bars denote T-values.



CONCLUSIONS

- ✓ RtvFTD patients are characterized by altered FC in networks beyond the pure frontal and language circuits, mostly targeting pivotal regions involved in high-level visual processing.
- ✓ If the observed increased FC is a compensatory mechanism or rather reflects the underneath pathological process still needs to be determined.
- ✓ The RS-fMRI information might improve the distinction between this rare condition and other variants of FTD.

Supported by: Italian Ministry of Health (GR-2010-2303035 and GR-2011-02351217); European Research Council (StG-2016_714388_NeuroTRACK).