Supplementary material 1

In this retrospective study, all case records were read and reported complaints were written down. Similar complaints were combined into one umbrella term based on similar meaning and/or cognitive/ behavioural domains and some symptoms were excluded based on authors consensus.

Symptom identification and classification (Based on patient/ caregiver history)

Cognitive	Language	Behavioural	Other	Excluded
Memory Problems; forgetfulness, poor memory, worsening memory, memory deficit, problems remembering dates/ places/ addresses/ events	**Word-finding difficulties;** trouble finding correct words during spontaneous speech, pay more effort to find correct word	**Disinhibition;** social awkwardness, inappropriate behaviour, loss of manners or decorum, agitation, impulsiveness, aggression, rash actions, childish puns and jokes, childishness	**Motor/mental slowness;** parkinsonism, rigidity, spasticity, pyramidal symptoms, extrapyramidal symptoms, mental slowness, mental rigidity, slowness, slow walking, falls	**Personality changes;** since this term covers several behavioural changes and it is not specific for a cognitive domain, we excluded this symptom
Prosopagnosia; poor memory for faces, face recognition deficit, difficulties remembering famous faces or familiar faces, difficulties recognising people on family photo album	**Single word comprehension difficulties;** difficulties understanding words, trouble understanding sentences due to lack of word meaning	**Compulsive behaviour;** obsessiveness, hoarding, storing materials, pacing, counting, touching, ritualistic behaviour, preoccupation, fixed-rigid ideas, repeating the same routine or stories, doing puzzles/jigsaws, obsessive gambling, playing same games, clock watching	**Hyper-religiosity;** increased religious ideas, hyper-spiritualism, excessive piety, changing beliefs with regard to religion, tending to read holy books, increased frequency of church visits	**Loss of insight, problems understanding, spontaneous speech alterations, grammatical mistakes;** since these symptoms are not core diagnostic symptoms of any diagnostic group, we did not display these symptoms
Executive dysfunction; poor judgement, impaired planning and	**Paraphasias;** literal, verbal, semantic	**Apathy or inertia;** Loss of interest or initiative, withdrawn, lack of energy or	**Depression;** sadness, decreased mood, melancholia, suicidal ideas	**Motor restless, spending more/less money, mania, tiredness, sleeping problems,**

organisation skills, disorganization	paraphasias, using wrong words, neologisms	effort, lack of desire to learn new things or meet new people or have new experiences, emotionlessness, loss of creativity, carelessness, becoming introverted, flat affect		**libido changes, utilization behaviour, perseveration, apraxia, concentration problems and concretism;** These symptoms were recorded in less than 20% in rtvFTD and none of them is a significant symptom for either rtvFTD (based on literature review) or one of the control groups. Therefore, they were not displayed.
Orientation problems; problems recognising time or place	**Naming difficulties;** poor memory for names, remembering difficulties the names of objects/ familiar countries/ cities/ streets	**Loss of empathy and egocentrism;** diminished response to other people feelings or needs, interrelatedness, diminished personal warmth, emotion recognition difficulties, selfishness, self-centeredness	**Delusions/ hallucinations;** False beliefs for things that did not happen, incorrect perceptions of objects or events involving the senses, visual or auditory hallucinations	
Getting lost; losing way, loss of knowledge about streets/ buildings/ neighbourhood, navigation problems		**Hyper-orality and dietary changes;** appetite changes, increase in appetite, decrease in appetite, seeks out food between meals, overeats at meal times, requests larger or second portion of food, reports hunger, reports being overfull, binge eating, needs to limit food intake, changing food preference (sweet, salt or obsessed with same food	**Somatic complaints and aches;** hypochondria, seeking medical help for headache/ muscle pain or other aches without physiological explanation	

		or developed other food fads), drinks more soft drinks/ tea/ coffee/ alcohol, obsessed with eating time/ schedule/ routine, loss of table manners, eating fast, eating with hands, increased smoking, loss of taste/ flavours, hyper-orality		
Visio-spatial problems; problems in discriminating form and colour, inability to perceive contrast, difficulties in visual spatial orientation and motion detection, difficulties in developing visual strategies, inability to process visual sensory information			**Feeling of anxiety/ panic;** feeling anxious, scared, insecure and panic	

rtvFTD: right temporal variant frontotemporal dementia

Supplementary material 2

Validation of the purposed diagnostic tree

To validate proposed diagnostic features, sensitivity and specificity analysis was conducted in the diagnostic groups for each step (Table 1, 2, 3, 4). Details of the cases and diagnostic symptoms were displayed in table 5.

Table 1. Core symptoms (At least 2 of 3 core symptoms**)**

	rtvFTD Positive	**rtvFTD Negative**
Criteria Positive	81% (n=57)	24% (n=51)
Criteria Negative	18% (n=13)	75% (n=159)
Statistic	**Value**	**95%Cl**
Sensitivity	81%	70.34% to 89.72%
Specificity	75%	69.34% to 81.35%
Positive predictive value	53%	46.19% to 59.27%
Negative predictive value	92%	88.16% to 95.26%

Table 2. Combination of core and supportive symptoms (At least 2 of 3 core symptoms and at least 1 of 3 supportive symptoms**)**

	rtvFTD Positive	**rtvFTD Negative**
Criteria Positive	70% (n=49)	12% (n=26)
Criteria Negative	30% (n=21)	88% (n=184)
Statistic	**Value**	**95%Cl**
Sensitivity	70%	60.02% to 78.76%
Specificity	88%	79.98% to 93.64%
Positive predictive value	65%	54.56% to 76.10%
Negative predictive value	89%	85.60% to 93.90%

Table 3. Combination of clinical and radiological features (At least 2 of 3 core symptoms, at least 1 of 3 supportive symptoms and at least 1 of 3 neuroimaging features**)**

	rtvFTD Positive	**rtvFTD Negative**
Criteria Positive	n=49	n=1
Criteria Negative	n=21	n=209
Statistic	**Value**	**95%Cl**
Sensitivity	70%	60.02% to 78.76%
Specificity	99%	97.38% to 99.99%
Positive predictive value	98%	87.33% to 99.71%
Negative predictive value	90%	87.43% to 93.44%

Table 4. Combination of amyloid status, clinical and radiological features (At least 2 of 3 core symptoms, at least 1 of 3 supportive symptoms, at least 1 of 3 neuroimaging features and negative amyloid status**)**

	rtvFTD Positive	**rtvFTD Negative**
Criteria Positive	n=49	n=0
Criteria Negative	n=21	n=210
Statistic	**Value**	**95%Cl**
Sensitivity	70%	60.02% to 78.76%
Specificity	100%	98.26% to 100.00%
Positive predictive value	100%	
Negative predictive value	90%	87.49% to 93.47%

rtvFTD: right temporal variant frontotemporal dementia

Table 5. Details of the subjects and diagnostic symptoms

Diagnostic group	Core clinical features			Supportive clinical features	Fulfil diagnostic criteria
	Prosopagnosia	Memory deficit	Behavioural Changes		
rtvFTD	1		4	3	+
rtvFTD		1	2		
rtvFTD	1	1	1		
rtvFTD	1	1	2	1	+
rtvFTD		1	4	1	+
rtvFTD	1		2	1	+
rtvFTD		1	2	1	+
rtvFTD		1	2	1	+
rtvFTD		1	2	1	+
rtvFTD		1	2		
rtvFTD			4		
rtvFTD	1		4	2	+
rtvFTD		1	2	1	+
rtvFTD		1	3	1	+
rtvFTD			4	1	
rtvFTD			2		
rtvFTD	1	1	3	1	+
rtvFTD	1		3	1	+
rtvFTD		1	2		
rtvFTD	1	1		1	+
rtvFTD			3		
rtvFTD			3		
rtvFTD		1	1		
rtvFTD		1	2	1	+
rtvFTD	1	1	1	1	+
rtvFTD	1	1	1	1	+
rtvFTD	1		4	1	+
rtvFTD	1	1	3	1	+
rtvFTD			3		
rtvFTD	1		3	1	+
rtvFTD	1		2	3	+
rtvFTD	1	1	3	1	+
rtvFTD	1	1	3	1	+
rtvFTD	1	1	3	1	+
rtvFTD	1	1	3	1	+
rtvFTD	1		2	1	+
rtvFTD	1		4	1	+
rtvFTD		1	2	1	+
rtvFTD	1	1	1	1	+
rtvFTD	1		3	1	+
rtvFTD	1		2	2	+
rtvFTD	1	1	2	1	+
rtvFTD		1	3	2	+
rtvFTD			3		
rtvFTD		1	2	1	+
rtvFTD	1	1		1	+
rtvFTD		1	1		

rtvFTD		1	2	2	+
rtvFTD	1		4	1	+
rtvFTD	1	1	3	1	+
rtvFTD		1			
rtvFTD	1		2	1	+
rtvFTD	1		3	1	+
rtvFTD	1		3	1	+
rtvFTD		1	2		
rtvFTD		1	1		
rtvFTD		1	2		
rtvFTD	1	1	2	1	+
rtvFTD	1	1	3	1	+
rtvFTD	1		2	1	+
rtvFTD		1	3	2	+
rtvFTD			1		
rtvFTD	1		3	2	+
rtvFTD			2		
rtvFTD		1	4	1	+
rtvFTD	1		3	1	+
rtvFTD	1		2		
rtvFTD	1	1	2	2	+
rtvFTD	1	1	2	1	+
rtvFTD		1	2		
svPPA	1		1	1	
svPPA	1			2	
svPPA				2	
svPPA				2	
svPPA	1			1	
svPPA			3	2	
svPPA				2	
svPPA				1	
svPPA				2	
svPPA	1		2	1	+
svPPA			2	1	
svPPA			2	1	
svPPA			1	2	
svPPA				2	
svPPA				1	
svPPA		1	1	2	
svPPA			1	2	
svPPA			3	2	
svPPA			1	1	
svPPA	1		1	1	
svPPA	1		1	1	
svPPA				1	
svPPA		1		1	
svPPA		1	2	2	
svPPA			2	2	
svPPA		1	1	2	
svPPA		1	3	1	

svPPA				1	
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svPPA				2	
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svPPA	1			1	
svPPA				2	
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svPPA		1		2	
svPPA		1	1	2	
svPPA		1	3	2	+
svPPA		1	2	1	+
svPPA			3	2	
svPPA				2	
svPPA				2	
svPPA			2	2	
svPPA			2	2	
svPPA		1		2	
svPPA	1	1	3	2	+
svPPA	1	1	3	2	+
svPPA	1		4	2	+
svPPA	1	1		1	+
svPPA			1	2	
svPPA			3	2	
svPPA			3	2	
svPPA			4	2	
svPPA	1	1	2	2	+
svPPA		1	2	1	+
svPPA				2	
svPPA				2	
bvFTD		1	2		
bvFTD			1	2	
bvFTD		1	1		
bvFTD			2	1	
bvFTD			4	1	
bvFTD		1	2		
bvFTD			2		

bvFTD			1		
bvFTD		1	4		
bvFTD		1	1	1	
bvFTD			3	1	
bvFTD			4	2	
bvFTD		1	2		
bvFTD			1		
bvFTD		1	3		
bvFTD			1	1	
bvFTD			1	1	
bvFTD		1	2		
bvFTD		1	2		
bvFTD			2	2	
bvFTD	1		2		
bvFTD		1	1	1	
bvFTD		1	3		
bvFTD		1	1		
bvFTD		1	1	2	
bvFTD		1	2		
bvFTD			3		
bvFTD			1		
bvFTD			3		
bvFTD			4		
bvFTD		1	3	1	+
bvFTD		1	1		
bvFTD		1	2		
bvFTD	1		3		
bvFTD		1	3	1	+
bvFTD		1	4		
bvFTD		1	2		
bvFTD			4		
bvFTD			2		
bvFTD			2		
bvFTD		1	2		
bvFTD			1	1	
bvFTD			2		
bvFTD		1	1	2	
bvFTD			2	2	
bvFTD		1	3	2	+
bvFTD		1	4	1	+
bvFTD			2		
bvFTD		1	3	2	+
bvFTD			2		
bvFTD			2		
bvFTD			3		
bvFTD			3	1	
bvFTD			2	1	
bvFTD		1	2		
bvFTD			2	1	
bvFTD		1	3		

bvFTD			4	2	
bvFTD	1		3	2	+
bvFTD			2		
bvFTD		1	3	1	+
bvFTD		1	3		
bvFTD			3	1	
bvFTD			3	2	
bvFTD		1	3		
bvFTD		1	3	1	+
bvFTD			3	1	
bvFTD		1	2	1	+
bvFTD		1	2		
bvFTD		1	2		
AD		1	1	3	
AD		1	1	3	
AD		1		2	
AD		1	2	2	+
AD		1	1	1	
AD		1			
AD		1			
AD			1	2	
AD		1	2	2	+
AD		1	1		
AD		1			
AD		1	1	1	
AD		1		1	
AD		1	1	2	
AD		1	1		
AD		1		2	
AD		1	1	3	
AD		1	1	2	
AD		1	2	2	+
AD		1		1	
AD		1		1	
AD		1	1	2	
AD		1	2		
AD		1		1	
AD		1		2	
AD		1	2		
AD		1		1	
AD		1		2	
AD		1	2	2	+
AD		1		3	
AD		1	1	3	
AD		1	2	2	+
AD		1	1	1	
AD		1		1	
AD		1			
AD		1	2	1	+
AD		1		1	

AD		1		2	
AD		1	2	1	+
AD		1	1		
AD		1		2	
AD		1		1	
AD		1	1		
AD		1		1	
AD		1		1	
AD		1		2	
AD		1		1	
AD		1		1	
AD		1		1	
AD		1	1	1	
AD		1	1	2	
AD		1			
AD		1			
AD		1		1	
AD		1		3	
AD		1			
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AD		1		1	
AD		1		1	
AD		1		2	
AD		1	1	1	
AD		1		1	
AD		1		2	
AD		1		3	
AD		1	1	1	
AD		1	1	2	
AD		1		2	
AD		1		2	
AD		1		1	
AD		1		2	

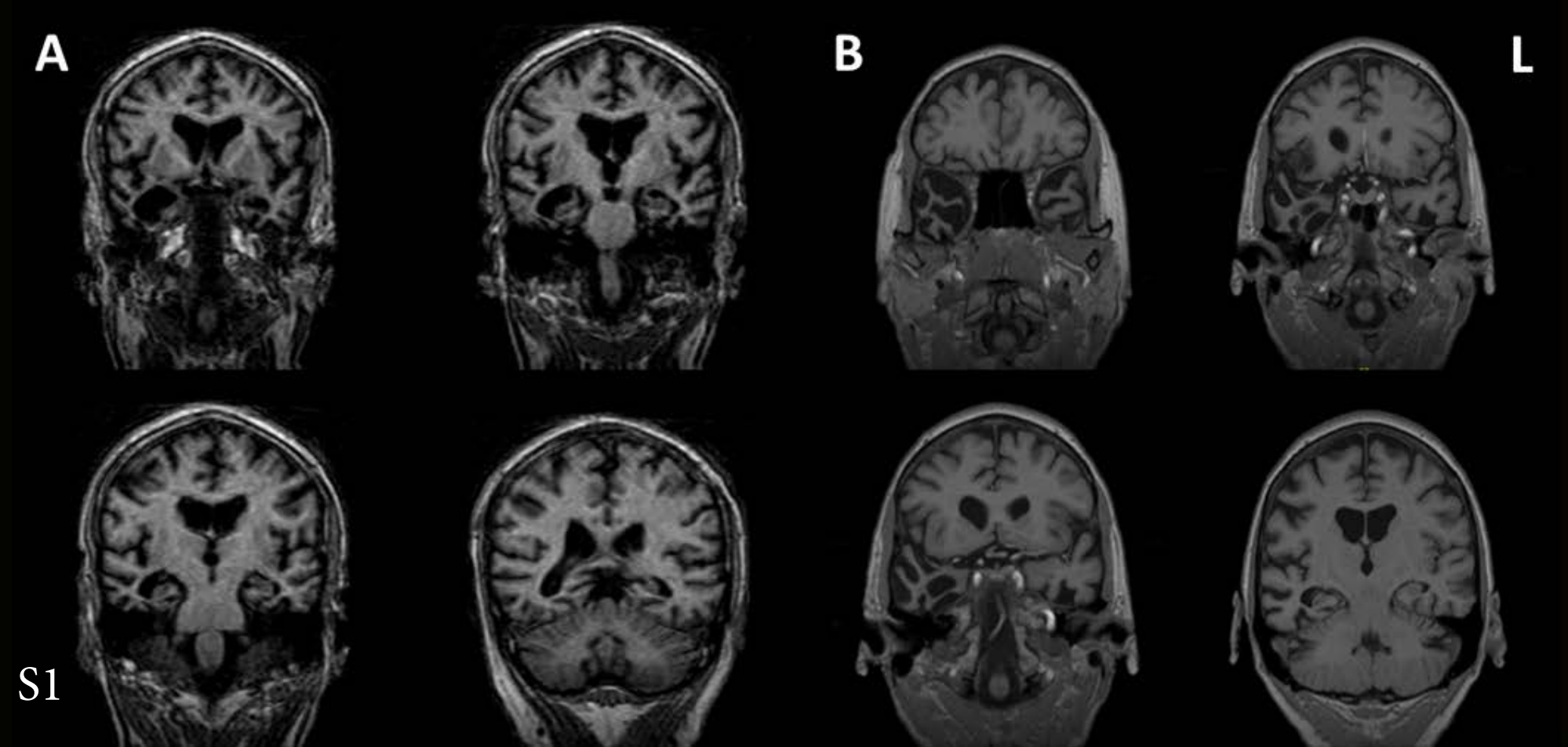
AD: Alzheimer’s disease; bvFTD: behavioural variant frontotemporal dementia; rtvFTD: right temporal variant frontotemporal dementia; svPPA: semantic variant frontotemporal dementia

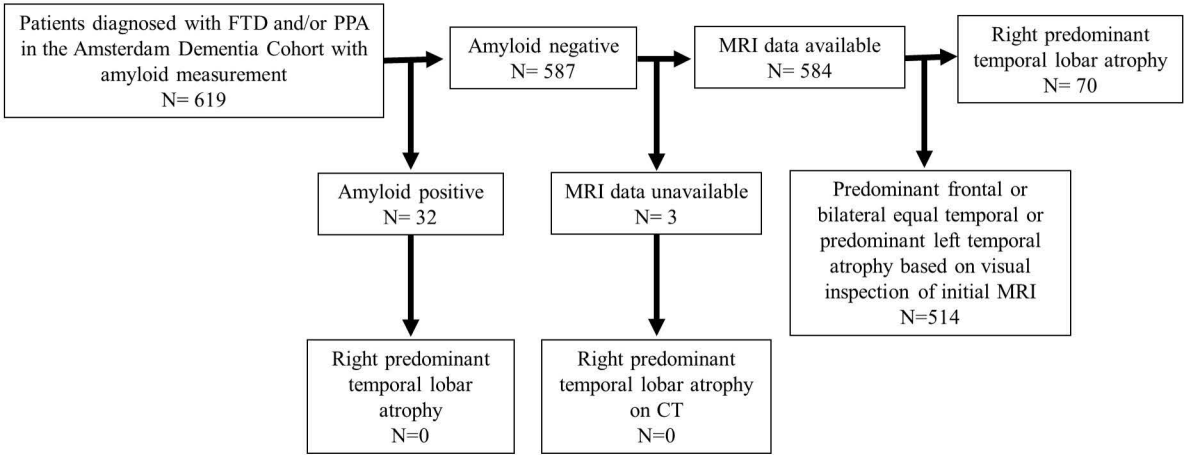
Supplementary figures:

Supplementary Figure 1: Right predominant temporal lobe atrophy. Not only mesial temporal atrophy (A) but also cortical temporal atrophy (B) was considered at visual inspection. L; Left

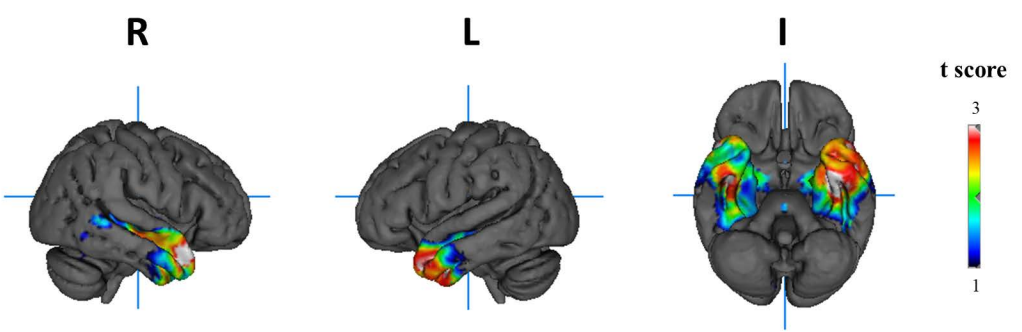
Supplementary Figure 2: Patient selection scheme. FTD: frontotemporal dementia; PPA: primary progressive aphasia

Supplementary Figure 3: 3D T-Maps of the radiological correlation of prosopagnosia in rtvFTD. R: right; L: left; I: inferior


A
B
L
S1


Patients diagnosed with FTD and/or PPA in the Amsterdam Dementia Cohort with amyloid measurement
N= 619
Amyloid negative
N= 587
MRI data available
N= 584
Right predominant temporal lobar atrophy
N= 70
Amyloid positive
N= 32
MRI data unavailable
N= 3
Predominant frontal or bilateral equal temporal or predominant left temporal atrophy based on visual inspection of initial MRI
N=514
Right predominant temporal lobar atrophy
N=0
Right predominant temporal lobar atrophy on CT
N=0

S2



S3